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Clinical Cases in Internal Medicine: Learning Through Practice (McMaster International Review Course in Internal Medicine, May 11–June 17, 2021, virtual course)

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ABSTRACT PROCEEDINGS OF THE BEST CASE REPORT CONTEST 2021

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AWARD-WINNING CASES

1ST PLACE: TANUKA MANDAL

Unusual neurological manifestations of scrub typhus

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INTRODUCTION Opsoclonus is a rare neurological manifestation of scrub typhus, usually occurring in association with myoclonus, cerebellar dysfunction, or extrapyramidal symptoms.

CASE REPORT A 55-year-old female patient from Burdwan presented with high fever lasting 12 days, vomiting lasting 10 days, and progressive, gradual deterioration of the level of consciousness lasting 7 days. On examination, the patient was drowsy, with a Glasgow Coma Score of 12 out of 15 points and meningeal signs present. Cranial nerve examination was unremarkable. Eye examination showed multiaxial, involuntary, saccadic movements of both eyes, suggestive of opsoclonus. There was no limitation of voluntary eye movements, a full range of both the uniocular and conjugate movements was possible. The patient was also unable to maintain the tongue in a protruded state; the tongue moved in and out, which was suggestive of tongue dyskinesia.

Respiratory examination revealed bilateral diffuse crepitations throughout both lung fields.

Cerebrospinal fluid (CSF) analysis and magnetic resonance imaging of the brain were performed. Empirical treatment with antibiotics and antivirals targeting meningitis was started. Analysis of the CSF showed a cell count of 10/mm^3, of which 80% were lymphocytes and 20% were polymorphonuclear neutrophils. Concentrations of protein and glucose in the CSF were 48 mg/dl and 53 mg/dl, respectively. The adenosine deaminase test, polymerase chain reaction–based assays for tuberculosis and *Herpes simplex* virus as well as the Japanese encephalitis virus IgM test came out to be negative. Magnetic resonance imaging did not reveal any abnormalities.

Other examinations revealed neutrophilic leukocytosis and conjugated hyperbilirubinemia with alanine transaminase levels higher than levels of aspartate aminotransferase. Chest X-ray showed bilateral diffuse areas of reticulonodular opacities, and abdominal ultrasonography revealed hepatosplenomegaly. High-resolution computed tomography of the thorax showed bilateral ground-glass opacity. The typhidot IgM, malarial parasite as well as dengue IgM and NS1 antigen tests were all negative.

Further diagnostic workup was performed, ultimately revealing a positive result of the scrub typhus IgM test.

The patient was diagnosed with scrub typhus with meningoencephalitis and pulmonary (alveolar) involvement. She was started on doxycycline 100 mg orally twice daily for 15 days. After the initiation of treatment, the patient gradually recovered and was discharged on the 26th day of admission.

CONCLUSIONS Opsoclonus appears to occur during the febrile phase of scrub typhus, with neurological manifestations completely resolving on completion of treatment.

Key words

meningoencephalitis, opsoclonus, pulmonary involvement, scrub typhus, tongue dyskinesia

2ND PLACE: KHALID EL KHOLY (*PRESENTED BY ELEANOR CRONIN*)

Systemic capillary leak syndrome: a rare, intriguing disease entity in the midst of the mass COVID-19 vaccination campaigns

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INTRODUCTION Systemic capillary leak syndrome (SCLS) is exceedingly rare, with less than 500 cases reported worldwide. It can be fatal in the initial episode; thus, it necessitates early recognition. It is characterized by acute profound disruption of the vascular endothelium, resulting in the classic triad of hypotension, hypoalbuminemia, and hemoconcentration.

CASE REPORT A previously healthy, 49-year-old woman with a 4-day history of malaise, worsening painful, bilateral, massive edema of the extremities, and syncope was diagnosed with SCLS upon presentation to our Emergency Department (ED). An initial dose of the Oxford-AstraZeneca COVID-19 vaccine was administered 4 days prior to her presentation.

In retrospect, the patient described symptoms of SCLS, such as episodes of cyclical edema and dehydration for which no cause had been identified. These episodes were often precipitated by an upper respiratory tract infection and associated with mild cyclical leg edema. They had been occurring for years prior to admission and had never been investigated.

During the acute phase, the patient complained of severe painful bilateral extremity edema, dizziness, and abdominal pain and had a syncopal episode which necessitated attendance to the ED. She quickly became hypotensive and developed tachycardia. Intravenous access was extremely difficult to maintain given the degree of diffuse edema. On examination, she had mottling of distal extremities, cool peripheries, and extensive peripheral pitting edema. Capillary refill time was prolonged but pulses were palpable.

Initial blood workup showed evidence of significant hemoconcentration, hypoalbuminemia, lactate acidosis, rhabdomyolysis, and acute kidney injury. Later investigations revealed an IgG lambda paraproteinemia. Additional extensive panel of blood workup was largely unremarkable.

A computed tomography angiography and venography of the aorta, thorax, abdomen, pelvis, and lower limbs were performed. There was no arterial or venous occlusion, but extensive anasarca with edema in the intramuscular fat planes and perivascular spaces was found, particularly in both lower limbs. Vascular specialist consultations were sought throughout her stay; reassuringly, no signs of limb ischemia were found.

The patient was initially treated with intravenous fluid boluses and broad-spectrum antibiotics. When the initial investigations pointed towards SCLS, high-dose intravenous methylprednisolone 1 g for 3 days was started together with intravenous immunoglobulin (IVIG) at a dose of 0.4 mg/kg for 5 days. There was a rapid clinical improvement following these treatments. The patient was then immediately transferred to the intensive care unit for close monitoring.

Following discharge from hospital 3 weeks after this major leak episode, the patient remains in good clinical condition and is scheduled for receiving IVIG 1 g/kg every 4 weeks.

DISCUSSION Medical therapy primarily targets prevention of recurrent episodes. Currently, IVIG is favored; other options include aminophylline, theophylline, and terbutaline. The differential diagnosis of SCLS includes septic shock, toxic shock syndrome, and anaphylaxis. Given the rarity of SCLS, patients are often initially treated empirically for these conditions.

CONCLUSION One should consider SCLS when severe hypoalbuminemia and hemoconcentration are also present, as it requires a unique approach to the management of shock. To our knowledge, this is the first case to describe this rare syndrome in temporal relation to a COVID-19 vaccination. However, while the vaccine may imply a direct relationship, there is insufficient evidence to infer causality.

Key words
COVID-19 vaccination, hypoalbuminemia, intravenous immunoglobulin, systemic capillary leak syndrome

3RD PLACE AND THE EUROPEAN JOURNAL OF CASE REPORTS IN INTERNAL MEDICINE AWARD: PAULINA GORZELAK-PABIŚ

Severe hypodysfibrinogenemia in a young patient with pulmonary thromboembolism: a case report

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INTRODUCTION Congenital fibrinogen disorders are the third most frequent rare coagulation defects. Mutations in fibrinogen genes can lead to quantitative (afibrinogenemia and hypofibrinogenemia) or qualitative (dysfibrinogenemia and hypodysfibrinogenemia) fibrinogen abnormalities. Hypodysfibrinogenemia, which is the least frequently reported congenital fibrinogen disorder, shares the features with both hypo- and dysfibrinogenemia.

CASE REPORT This study presents a case of a 44-year-old woman admitted to the hospital in April 2020 on account of nonspecific chest pain, exacerbating cough, and hemoptysis. The results of laboratory tests showed elevated levels of inflammatory markers, a thyroid-stimulating hormone level below 0.01 μU/ml, free thyroxine of up to 47.5 pmol/ml, fibrinogen concentration of 48 mg/dl, thrombin time of 25.6s, and D-dimer level of 1743 μg. Hyperthyroidism was diagnosed and thiamazole therapy was implemented. Computed tomography (CT) of the chest revealed pneumonia. Antibiotic therapy and a prophylactic dose of enoxaparin were administered.

During hospitalization, the patient's condition deteriorated; she presented persistent dyspnea and hemoptysis. The Wells and Revised Geneva scores indicated intermediate-risk pulmonary embolism, and due to an elevated D-dimer level, CT of the pulmonary arteries was performed. It revealed a massive embolic material in the right pulmonary artery and its branches.

The antibiotic treatment was modified and therapeutic doses of heparin were included. Due to the low level of fibrinogen, cryoprecipitate and fibrinogen concentrate (RiaSTAP) were administered. Additionally, screening for thrombophilia and antiphospholipid syndrome was performed, with negative results. The patient was discharged from hospital in good condition and prescribed with rivaroxaban 20 mg daily.

After 6 weeks, a hemostasis workup showed decreased functional (50 mg/dl based on the Clauss method) and antigenic (130 mg/dl) fibrinogen levels with a functional / antigenic fibrinogen ratio of 0.38 and prolonged thrombin times, which indicated hypodysfibrinogenemia. Next generation sequencing of the fibrinogen gene *FGG* revealed a heterozygous missense mutation in exon 8: p.1055 G>C; p.Cys352Ser. Severe hypodysfibrinogenemia was diagnosed.

DISCUSSION The prevalence of hypodysfibrinogenemia is unknown as the disease is often asymptomatic. The most extensive literature review published so far reports 51 patients diagnosed with this disorder. The cutoff value of 0.7 for the ratio of functional concentration of fibrinogen to its antigen level has an 86% diagnostic sensitivity for hypodysfibrinogenemia.

In the presented case, hypodysfibrinogenemia was diagnosed in a 44-year-old patient with massive pulmonary thromboembolism based on a functional/antigenic fibrinogen ratio of 0.38 and a heterozygous missense mutation in the fibrinogen gene *FGG* in exon 8: p.1055 G>C; p.Cys352Ser (p.Cys326Ser in the mature chain), resulting in an amino acid substitution of serine for cysteine. This mutation affects the same amino acid as fibrinogen Cordoba which, unlike in our patient, was associated with severe cerebral hemorrhage. So far, 2 cases with a γCys326Ser mutation coexisting with a thrombotic disease have been reported, and our study is the third to describe the abovementioned mutation in association with thrombotic complications.

It is clinically important to detect coagulation abnormalities like hypodysfibrinogenemia, which may cause thrombosis, and establish a proper anticoagulant therapy. In the presented case, a lifelong therapy with rivaroxaban 20 mg daily was recommended.

CONCLUSIONS When a young patient presents with unusual thrombosis, fibrinogen concentration should be determined and if it is low, a congenital fibrinogen disorder should be considered as a possible cause of the problem.

Key words
genetic mutation, hypodysfibrinogenemia, p.Cys352Ser, pulmonary thromboembolism, rivaroxaban

SPECIAL AWARD: LUCRECIA DI ROCCO

From shivers to mercury poisoning

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INTRODUCTION Tremor is defined as an involuntary, rhythmic, oscillatory movement. It is the most common of all movement disorders. Clinical classification of tremor is based on medical history, characteristics of movement disorders, and the associated neurological and systemic signs. Therefore, a comprehensive diagnostic evaluation is essential to identify the etiology.

Mercury poisoning, also known as hydrargyria or mercurialism, is a disease caused by exposure to mercury or mercury compounds. Its characteristics vary depending on the oxidation state of the metal and the route of entry to the organism. The manifestation of toxicity depends on the type and duration of exposure. The main targets of mercury absorption in the body are the kidneys, nervous system, skin, digestive system, blood, and lungs.

CASE REPORT We present a case of a 38-year-old woman with a history of viral pericarditis, surgical resection of acoustic neuroma, and previous hospitalization due to delirium accompanied by hyperglycemia and ketosis requiring insulin therapy. She also had a history of severe acute kidney failure with hyperchloremic acidosis, anemia, psychotic break, and behavioral disorders with paranoid features and kinesthetic hallucinations that improved after treatment with atypical antipsychotic drugs. *Klebsiella pneumoniae carbapenemase* infection complicated the clinical picture and was treated with imipenem and colistin. After discharge, the patient started to experience lower limb weakness due to peripheral neuropathy with mild-to-moderate axonomyelinic involvement confirmed by electromyography. In the course of the following 5 months, the clinical picture evolved into progressive and disabling shivering, together with decreased appetite, loss of weight and hair, and anemia. Upon presentation for consultation for all the above symptoms, the patient was admitted for further studies. Physical examination showed generalized disabling cerebellar, cephalic, and limb tremor exacerbated by change of position, as well as constant nystagmus, ataxia, and white corneal arcus. Nuclear magnetic resonance, magnetic resonance angiography of the brain, cerebrospinal fluid testing, thyroid panel, collagenogram, and viral serology were performed to rule out structural, vascular, demyelinating, pharmacological, infectious, and autoimmune diseases. Apart from clinical evaluation and laboratory test results, there was an epidemiological element to consider—the patient lived in a river basin highly contaminated by industrial activity and near a waste-processing plant, with no access to drinking water. Blood and urine toxicology tests for heavy metals showed high mercury plasma levels, indicating mercury poisoning.

DISCUSSION A broad approach in differential diagnosis of central tremor is a challenge due to a variety of clinical presentations in terms of the accompanying signs and symptomatology.

According to the World Health Organization, mercury is among the 10 chemical products or group of products that represent special public health threats, being present naturally in the air, water, and soil. Exposure to such metal, even in small doses, can cause severe health problems. The type and severity of the symptoms depend on individual susceptibility to toxicity, dose as well as the method and duration of exposure.

CONCLUSIONS When faced with a neurological disease presenting with central tremor, a deep and complete investigation of the causes must be carried out, including a toxicology screening. Appropriate diagnostic workup can lead to initiation of specific treatment, which can considerably reduce the risk of sequelae.

Key words

environmental pollution, mercury, poisoning, tremor

ORAL PRESENATIONS

AINAN ARSHAD

Lead encephalopathy in an adult opioid abuser

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CASE REPORT A 38-year-old man presented to the emergency department with abdominal pain, vomiting, general weakness, and altered level of consciousness. He had been ingesting opioids for more than 5 years and had a history of multiple hospital admissions for abdominal pain. His laboratory results showed deranged liver function tests, anemia, and basophilic stippling on the peripheral blood smear. Further investigation revealed markedly raised serum lead levels. We started chelation with oral penicillamine 250 mg every 6 hours for 2 days, which was subsequently switched to intramuscular dimercaprol 4 mg/kg twice daily and intravenous calcium disodium ethylenediamine tetraacetic acid 50 mg/kg in 2 divided doses for the next 5 days. By that time, the patient became clinically stable and his serum lead levels decreased. He was discharged and followed up at the outpatient clinic. This case report highlights the importance of consideration of lead toxicity in opioid abusers, and brings to attention new ways of lead chelation in resource-limited settings.

Key words
chelation, lead encephalopathy, opioid abuse

ANNAMÁRIA KÖVESDI

Parathyroid adenoma in Li-Fraumeni syndrome: a novel manifestation of the disease

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INTRODUCTION Carriers of germline mutations of the p53-encoding *TP53* gene are predisposed to multiple types of tumors at early age. The main manifestations of Li-Fraumeni syndrome (LFS) are soft tissue and bone sarcomas, breast cancers, brain tumors, adrenocortical carcinomas, and leukemias. Family history of patients with this autosomal, mainly inherited tumor syndrome usually includes multiple, even synchronous neoplasms among the relatives. We report a case of a patient with a genetically confirmed diagnosis of LFS, presenting with an unusual manifestation and a novel mutation of the disease.

CASE REPORT A 41-year-old woman with a history of bilateral ovarian dermoid cysts and no other comorbidities was admitted to the hospital due to severe pneumonia. The relatives seemed to neglect the positive family history of tumors (including 4 brain tumors and 2 lung cancers). Computed tomography of the chest revealed a lesion in segment S2 of the right lung and a tumor on the fifth rib, potentially indicative of metastasis. Fluorodeoxyglucose positron emission tomography / computed tomography showed an increased uptake behind the right thyroid lobe, in the left thyroid lobe, and in multiple lymph nodes in the chest and abdomen as well as a mass in the right breast. The ovary and bones were also involved. Further examination of the breast tumor revealed an invasive breast cancer. Cytology of the thyroid nodule showed a benign thyroid lesion. Hyperparathyroidism and hypercalcemia resulted from primary hyperparathyroidism. Bone scintigraphy did not confirm any malignancies; however, it was suggestive of osteitis fibrosa cystica caused by metabolic bone disease secondary to primary hyperparathyroidism. According to the recommendation of the tumor board, a synchronous resection of the thyroid gland and breast cancer was performed. Histological examination of these lesions revealed a benign parathyroid adenoma and a ductal adenocarcinoma of the breast. Core needle biopsy of the lesion in the right psoas region revealed a leiomyosarcoma, which was subsequently removed surgically. The diagnosis was histologically confirmed. The ovarian mass removed later was confirmed to be a metastasis of leiomyosarcoma. The patient refused any further surgical or medical treatment. Genetic analysis led to identification of a germline missense mutation of the *TP53* gene. Despite the unfortunate course of the disease in this patient, we managed to genetically test 4 of her 5 children; 2 of them also carried the mutation.

DISCUSSION The importance of early recognition of hereditary tumor syndromes and their surveillance are crucial. Parathyroid involvement in LFS has not yet been described in the literature. Our patient had severe primary hyperparathyroidism with high levels of parathyroid hormone and calcium resulting in metabolic bone disorder. Family history of the patient included at least 8 tumors among the relatives, reflecting a hereditary condition. Based on these findings and the subsequently confirmed genetic diagnosis of LFS, parathyroid malignancy was rather suspected. However, histological examination confirmed a benign parathyroid adenoma. To our best knowledge, this is the first report of a 1722G>C missense mutation in the literature.

Key words
Li-Fraumeni syndrome, *TP53*, mutation, parathyroid adenoma

ARTHUR RENAUD

Catastrophic multifocal sarcomatous hemorrhages triggered by acquired factor XIII deficiency

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INTRODUCTION Factor XIII (FXIII) deficiency is a very rare bleeding disorder that can be acquired and is associated with various pathological conditions in its nonimmune form. It can be responsible for a potentially severe deep hemorrhagic syndrome despite perfectly normal results of first-line hemostasis tests, which makes the diagnosis challenging.

CASE REPORT A 75-year-old man was referred to the internal medicine department due to acute, nontraumatic pain in the left knee, with swelling and inability to walk. This episode appeared in the context of longstanding fatigue and fluctuating fever. Blood samples showed a significant inflammatory syndrome with microcytic anemia and thrombocytosis. Puncture of the left knee revealed hemarthrosis attributed to treatment with vitamin K antagonists (international normalized ratio, 2.2), which was therefore interrupted. Analysis of the synovial fluid showed neither bacteria nor microcrystals. The initial suspicion of infectious arthritis of the left knee was ruled out and early intravenous antibiotics were discontinued. Also, an extensive infectious workup was not positive for any atypical bacteria. A whole-body computed tomography showed multiple bilateral pulmonary parenchymal nodules. Positron emission tomography / computed tomography revealed intense hypermetabolism of the micronodules as well as mediastinal and retroperitoneal lymph nodes, and intense and heterogeneous hypermetabolism of the anterior compartment of the left lower limb invading the lateral-external femoral canal and the patellofemoral joint. Subsequently, the patient developed a spontaneous large hematoma of the left thigh associated with 2 episodes of hemoptysis, which resulted in a massive drop of the hemoglobin level that required 7 red blood cell transfusions within a few days. This severe hemorrhagic syndrome led to extensive hemostasis investigations including clot factor (II,

V, VII, VIII, IX, X, XI) and von Willebrand factor activities, fibrinogen level, and platelet function assay that turned out to be subnormal. The patient was eventually transferred to the intensive care unit due to acute respiratory distress related to bilateral hemothorax and left lung atelectasis cause by a bronchial blood clot. At that time, the hemostasis department was contacted. A moderate FXIII deficiency with an activity level of 18% was finally diagnosed. No anti-FXIII autoantibodies were found. Administration of high-dose FXIII concentrate was not effective. The situation evolved towards multivisceral failure and death despite salvage therapy including corticosteroids, octreotide, and tranexamic acid. The autopsy resulted in the diagnosis of a metastatic angiosarcoma associated with nonimmune-mediated acquired FXIII deficiency responsible for a catastrophic hemorrhagic syndrome localized exclusively at the tumor sites. A hyperconsumption mechanism was assumed to be the cause of the deficit.

DISCUSSION Acquired FXIII deficiency is poorly known by internal medicine practitioners. Nonetheless, this rare bleeding disorder can be responsible for potentially life-threatening hemorrhages, as demonstrated here. Interestingly, in our case, hemorrhages were localized specifically at the tumor sites. This observation allows us to hypothesize that the vascular invasiveness of angiosarcoma could have played a role as a local weakening element. Factor XIII deficiency should be considered when standard hemostasis investigations remain normal. A close cooperation with hemostasis specialists is crucial in this setting to conduct an appropriate biological workup.

Key words
acquired factor XIII deficiency, angiosarcoma, hemorrhagic syndrome

BRAM VERDONCK

Hemifacial flushing and hyperhidrosis after general anesthesia in a patient with a metastatic recurrence of cervical carcinoma

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INTRODUCTION Transient hemifacial flushing and hyperhidrosis with contralateral pallor and anhidrosis are the cardinal symptoms of Harlequin syndrome resulting from a disruption to the sudo- and vasomotor innervation of the face (the oculosympathetic pathway). The term *Harlequin sign* is typically used when the condition is associated with other autonomic syndromes, such as Horner syndrome. It is a rare condition, often benign and idiopathic (in two-thirds of cases). In the remainder of cases, it is mostly secondary to compression by a mass along the course of the oculosympathetic chain.

As such, extensive work-up of patients with this syndrome is necessary to exclude serious underlying pathology. We report a case of a Harlequin sign after general anesthesia in a patient with an underlying metastatic recurrence of cervical carcinoma.

CASE REPORT A 75-year-old female patient was recently diagnosed with a metastatic recurrence of cervical carcinoma at the level of the left sulcus superior due to a new-onset left-sided Horner syndrome. She had a history of hypertension and type 2 diabetes mellitus. A right-sided Port-a-Cath device was surgically placed under general anesthesia with propofol. There was no abnormal body position during surgery. No thoracic epidural anesthesia nor paravertebral block was used. In the immediate postoperative period, the patient developed right-sided hemifacial flushing and hyperhidrosis with contralateral pallor and anhidrosis. There was no orthostatic hypotension and neurologic examination was normal except for the known Horner syndrome. A presumptive diagnosis of left-sided Harlequin sign was made as the hemifacial flushing and sweating diminished over the following hours. The patient received 3 cycles of palliative chemotherapy with paclitaxel-carboplatin; unfortunately, without tumor response. She died 5 months later.

DISCUSSION To our best knowledge, no case of Harlequin syndrome / sign after general anesthesia has been reported in the literature to date. The pathophysiologic mechanism of the condition in this case is not completely clear but is probably the same as in Harlequin syndrome / sign that is triggered by exercise. There was no abnormal body position during surgery. No thoracic epidural anesthesia was administered. No additional damage to the oculosympathetic chain could have occurred during surgery as the Port-a-Cath device was placed on the right side (which would have then produced a right-sided Harlequin sign). It is evident that there was previous damage to the oculosympathetic chain in this patient, mainly neuropraxia caused by the tumor and possibly also due to diabetes mellitus and hypertension. Recovery from surgery and anesthesia is associated with a stress response, mainly sympathetically mediated, which is quite similar to exercise. In this case, previous damage to the oculosympathetic chain on the left side would prevent a sympathetic response of the left-sided cutaneous blood vessels and sweat glands, resulting in an (over)compensatory reaction on the patient's right side.

CONCLUSIONS Harlequin syndrome is a striking clinical picture which should be recognized easily despite its rarity. Surgeons, anesthesiologists, and oncologists should be familiar with this syndrome.

It is extremely important to perform thorough investigations in patients with Harlequin syndrome, mainly to exclude underlying malignancy. In this case, the underlying malignancy was already known.

Key words
cervical carcinoma, general anesthesia, Harlequin syndrome, hemifacial flushing

CATALINA MOSNA

Fabry disease: a case report

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INTRODUCTION Fabry disease is a lysosomal storage disorder caused by a deficiency of alpha-galactosidase (α-Gal A). It is considered a rare condition due to its low frequency (incidence of approximately 1:117 000 live male births); in Bahía Blanca, Argentina there have been no records of patients affected in the last 20 years. We report a case of a 28-year-old patient with a late diagnosis of the disease.

CASE REPORT A 28-year-old man presented to hospital in July 2019 with skin angiokeratomas. He also reported other symptoms such as polyarthralgia and paresthesia in the upper and lower limbs, anhidrosis, exhaustion, and nonspecific digestive symptoms since the age of 9 years. His previous medical consultations were inconclusive. Due to the suspicion of lysosomal storage disease (Fabry disease), diagnostic studies were performed, which revealed deficient levels of α-Gal A. Further evaluation showed cardiac and renal involvement. In November 2019, systemic enzyme replacement therapy with agalsidase alfa (0.2 mg/kg every 15 days) was initiated.

DISCUSSION Fabry disease is an X-linked lysosomal storage disorder caused by a deficiency of α-Gal A leading to an accumulation of glycosphingolipids (mostly globotriaosylceramide) in the lysosomes of the cells of different organs (such as the skin, heart, or kidneys). The classic phenotype is associated with early onset of symptoms

(during childhood or adolescence), followed by a progressive multiorgan failure. Clinical manifestations include neuropathic pain and episodic severe pain crises, hypohidrosis, skin abnormalities (angiokeratomas), gastrointestinal disturbances, and a characteristic asymptomatic corneal opacity. Kidney injury findings are generally albuminuria and glomerulosclerosis, and cardiac involvement is frequent due to the accumulation of globotriaosylceramide in cardiac tissues. In male patients, the α-Gal A activity testing is a standard diagnostic tool; however, confirmation of the disease-causing *GLA* gene mutation is important to establish the phenotype of the disease and rule out benign polymorphisms. It also allows the testing of at-risk family members. The management of Fabry disease includes enzyme replacement therapy by intravenous infusion; it is available in the form of agalsidase alfa and agalsidase beta. Adjunctive therapies aimed to treat symptoms of tissue injury and its progression are also important. The assessment of patients with Fabry disease should involve a multidisciplinary care team to assist in the management of organ-specific complications.

CONCLUSIONS Fabry disease is a rare metabolic disorder. It is important to know about this condition since the diagnosis is usually delayed because the symptoms of the disease may not be spontaneously reported by patients or they may be reluctant to talk about them if they have experienced multiple failures in relation to the diagnosis.

Key words
alpha-galactosidase, case report, Fabry disease, lysosomal storage disorder

CSABA SUMÁNSZKI

Secondary hypertension due to middle aortic syndrome in a young adult

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INTRODUCTION Middle aortic syndrome (MAS) is a rare cause of secondary hypertension in children and adolescents. It constitutes 0.5% to 2% of all cases of aortic stenosis. The stenosis involves the abdominal aorta and/or distal descending thoracic aorta and can include renal and visceral branches. Although the disease was first described in 1963, its etiology remains controversial. The majority of MAS cases (64%) are idiopathic, 17% are caused by acquired inflammatory diseases such as Takayasu's arteritis or intrauterine rubella infection, while the remaining 15% are associated with Mendelian disorders such as neurofibromatosis type I (von Recklinghausen disease) and Williams syndrome.

CASE REPORT In February 2016, a 21-year-old female patient was referred to our tertiary referral endocrine center with resistant hypertension diagnosed at the age of 18 years. Her blood pressure exceeded normal ranges (max, 190–200/100–110 mm Hg) despite a triple combination of antihypertensive drugs (angiotensin-converting enzyme inhibitor, calcium channel blocker, and diuretics).

Physical examination on her first visit revealed equally elevated blood pressure on both arms (180/100 mm Hg) and significantly lower blood pressure values on both legs (150/80 mm Hg). Electrocardiography showed no signs of abnormal heart rhythm or cardiopathy. According to the 24-hour blood pressure monitoring, median blood pressure was 155/80 mm Hg (daytime median, 137/67 mm Hg with maximum systolic blood pressure, 220 mm Hg; night-time median, 164/86 mm Hg with maximum systolic blood pressure, 175 mm Hg). Routine laboratory examinations revealed normal potassium and sodium levels, with no signs of hyperlipidemia or diabetes mellitus. Echocardiography revealed normal ventricular function.

Hormonal investigations were performed after optimizing antihypertensive drugs not affecting the renin-angiotensin-aldosterone system. The laboratory results confirmed hyperreninemic hyperaldosteronism—supine aldosterone level was 17.2 ng/dl (reference range, 0.7–15.0 ng/dl), plasma renin activity was 4.41 ng/ml/h (reference range, 0.2–2.8 ng/ml/h). The patient was euthyroid and eucortisolemic, and urinary metanephrines were within the normal range.

Diagnostic imaging workup was performed to investigate the cause of secondary aldosteronism. Both kidneys showed normal parenchymal function and ultrasound morphology. Abdominal and consecutive thoracic computed tomography–angiography revealed significant stenosis of thoracic aorta at the segment of the X–XI thoracic vertebrae.

Percutaneous transluminal stent-graft implantation and postdilatation with balloon catheter was successfully performed and the patient's blood pressure normalized after the intervention.

Long-term blood pressure control has been achieved with low-dose doxazosin monotherapy.

DISCUSSION We report a case of MAS diagnosed in a young adult patient. Middle aortic syndrome is associated with high morbidity and mortality due to the increased risk of cardiovascular disorders. Conservative drug treatment is rarely effective, and endovascular or surgical management is required to normalize blood pressure. Residual hypertension should be accounted for as it can require medication or reintervention.

CONCLUSIONS Our case highlights that secondary hypertension should always be considered in young patients with hypertension. Management of MAS calls for a multidisciplinary strategy, which involves thorough investigations and medical management coupled with catheter-based and surgical interventions, in order to achieve blood pressure control and prevention of end-organ impairment.

Key words
endovascular grafting, middle aortic syndrome, secondary aldosteronism, secondary hypertension, thoracic aorta

JOOYEON LEE

Correct diagnosis of X-linked hypophosphatemia after 46 years of misdiagnosis: seeing the unseen

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INTRODUCTION Diagnosis of X-linked hypophosphatemia (XLH), an inherited metabolic bone disorder caused by excessive production of fibroblast growth factor 23 (FGF-23), is often delayed in cases without specific family history due to the rarity of the disease and diversity of clinical manifestations.

CASE REPORT A 60-year-old woman presented to the outpatient clinic due to aggravated musculoskeletal pain at multiple sites. She was of short stature and underweight (128 cm, 16 kg), and had a bow-shaped deformity of the extremities and severe kyphoscoliosis developed since childhood. She wore dentures since the age of 14 years due to spontaneous teeth loosening without any specific family history. Her initial phosphate level was 1.8 mg/dl with normocalcemia, vitamin D deficiency (12.14 ng/ml), elevated parathyroid hormone levels (99.3 pg/ml), and marginally reduced renal tubular reabsorption of phosphate (TmP/GFR, 84.3%). Her bone mass was very low (T-score: lumbar spine, 5.3; femur neck, 6.7; total hip, 6.1). She underwent osteotomy 40 years ago with delayed wound healing that lead to the diagnosis of osteogenesis imperfecta. She was started on calcium, vitamin D_3 (1000 IU daily), and calcitriol (0.5 mcg daily) supplementation. However, her genetic evaluation

for osteogenesis imperfecta (*COL1A1*) was negative. Nevertheless, treatment for osteoporosis, along with the use of ibandronate and zoledronic acid, showed minimal improvement in her bone mass and pain. However, hypophosphatemia and low borderline calcium level persisted despite vitamin D supplementation. A re-evaluation of chronic hypophosphatemia was performed, which showed lower borderline TmP/GFR (85%), a highly elevated level of FGF23 (119.46 pg/ml), and normal level of 25-hydroxyvitamin D (52.19 ng/ml). Next generation sequencing panel showed a *PHEX* mutation from the genomic DNA. The patient was diagnosed with XLH.

Currently, conservative treatment of XLH consists of persistent supply of phosphorus and vitamin D, and after appropriate supplementation of phosphate (1500 mg daily), the patient's laboratory and radiologic findings showed significant improvement. Meanwhile, with the development of burosumab, an anti-FGF23 human antibody, a new era of XLH treatment will begin.

Key words
fibroblast growth factor 23, osteogenesis imperfecta, *PHEX*, phosphate regulating endopeptidase homologue, X-linked hypophosphatemia

JUNGMIN CHOI

Recurrent pancreatitis in a pregnant woman with severe hypertriglyceridemia successfully managed by multiple plasmapheresis

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INTRODUCTION Hypertriglyceridemia is a state of increased serum triglyceride levels triggered by multigenetic and multifactorial causes. The serum triglyceride concentration can be markedly elevated in the presence of precipitating factors such as a high level of estrogen or pregnancy. We report a case of a patient with recurrent pancreatitis during pregnancy conceived with in vitro fertilization-embryo transfer (IVF-ET), in whom severe hypertriglyceridemia was successfully controlled by multiple sessions of plasmapheresis.

CASE REPORT A 24-year-old pregnant woman with newly diagnosed severe hypertriglyceridemia was admitted to the hospital due to a sudden onset of severe abdominal pain at 26 weeks of gestation conceived using IVF-ET. She had been experiencing recurrent pancreatitis despite using the dyslipidemia medications that were allowed in pregnancy. On admission, serum levels of amylase and lipase were elevated to 347 U/l and 627 U/l, respectively, along with fasting triglyceride levels elevated to 4809 mg/dl. A clinical diagnosis of hypertriglyceridemia-induced acute pancreatitis was made and plasmapheresis was administered. After the procedure, serum levels of triglycerides, amylase, and lipase decreased to 556 mg/dl, 60 U/l, and 69 U/l, respectively, along with subsequent pain relief. The patient underwent a total of 9 sessions of plasmapheresis to retain serum triglyceride levels below 1000 mg/dl during pregnancy, with no further recurrence of acute pancreatitis. After delivery, the serum triglyceride level was maintained below 500 mg/dl with a combination treatment including fenofibrate, statin, and ezetimibe.

DISCUSSION Although severe hypertriglyceridemia is usually asymptomatic, it can cause fatal complications, such as acute pancreatitis, when precipitating factors are present. Early administration of plasmapheresis can be a useful option in hypertriglyceridemia-induced acute pancreatitis resistant to medical treatment.

Key words
acute pancreatitis, IVF-ET, plasmapheresis, pregnancy, severe hypertriglyceridemia

KARIMULLA MONDAL

A rare presentation of scrub typhus: myocarditis, acute liver failure, and leukemoid reaction

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CASE REPORT Scrub typhus is a mite-borne zoonosis, caused by Orientia tsutsugamushi, a Gram-negative intracellular organism. This infection has a high prevalence in rural areas of East Asia and Western Pacific islands. It usually presents with fever, chills, myalgia, headache, skin rashes, and pathognomonic skin lesions (eschar) which can be found in around 10% cases in the Indian subcontinent. Occasionally, it can lead to life-threatening complications.

Simultaneous presentation of more than 2 complications is uncommon and rarely reported in the literature. Here we report a case of a 37-year-old woman with acute febrile illness complicated with myocarditis, acute liver failure, and leukemoid reaction. She was diagnosed promptly and successfully treated with doxycycline, with full recovery.

Key words
acute liver failure, leukemoid reaction, myocarditis, scrub typhus

LAURI HEIN

Idiopathic systemic capillary leak syndrome: a rare cause of shock

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INTRODUCTION Idiopathic systemic capillary leak syndrome (ISCLS) is a rare disorder characterized by episodes of severe hypotension, hemoconcentration, and hypoalbuminemia caused by paroxysmal capillary hyperpermeability. This case report describes a patient with recurrent episodes of ISCLS to enhance awareness of the disease, as early recognition and preventive treatment reduce morbidity.

CASE REPORT A 63-year-old woman presented with hypovolemic shock, hemoconcentration, and hypoalbuminemia. Thorough radiological studies did not reveal any relevant abnormalities. She required hemodynamic resuscitation with intravenous fluids and vasopressors in the intensive care unit, following pulmonary edema and bilateral pleural effusions due to volume overload. Thoracocentesis was performed and after a few days of treatment with intravenous diuretics, the patient was discharged in good condition. Several causes of distributive, cardiogenic, hypovolemic, and obstructive shock were considered and excluded.

Fourteen months later the patient was admitted again, in a clinical condition similar to the one she was in during the first hospitalization. Considering several episodes of rapidly developed shock syndrome, hemoconcentration, and hypoalbuminemia, the diagnosis of ISCLS was made and preventive treatment with terbutaline and theophylline was initiated.

Another episode of ISCLS occurred 6 months later; therefore, prophylactic treatment was changed to intravenous immunoglobulin (IVIG) 2 g/kg/month. Hereafter, prolonging the administration interval or reducing the dose of IVIG led to new episodes of ISCLS. During these attacks, the patient presented with prodromal complaints and hemoconcentration, which resolved quickly after commencing treatment with intravenous fluids and IVIG (1 g/kg for 2 consecutive days). To date, the patient has been in remission while receiving regular IVIG infusions at a dose of 2 g/kg/month.

DISCUSSION In the presented case, the patient experienced recurrent episodes of hypotension, hemoconcentration, and hypoalbumin-

emia, following complications from volume overload in the recovery phase. These findings are suggestive of ISCLS and the diagnosis was made after exclusion of other alternative causes. Although less than 500 cases of ISCLS have been reported in the literature to date, the condition is probably underdiagnosed because of the lack of awareness and high mortality without treatment.

The cornerstone of acute treatment is supportive care with intense hemodynamic stabilization and meticulous monitoring of the fluid status in the recovery phase. Preventive treatment with IVIG (2 g/kg/month) seems to be effective; our case supports this treatment regimen. A few case reports previously described successful treatment with IVIG during attacks. In our patient, early infusion with IVIG in the prodromal phase appeared to be beneficial in terms of preventing hypotension, hypoalbuminemia, and the possible complications arising from hypoperfusion or subsequent volume overload.

CONCLUSIONS Idiopathic systemic capillary leak syndrome should be suspected in the presence of the triad of hypotension, hemoconcentration, and hypoalbuminemia. Recognition of this syndrome is important as early diagnosis and preventive treatment reduce morbidity. This report suggests that administration of IVIG in the prodromal phase might prevent subsequent capillary leakage.

Key words
hemoconcentration, hypoalbuminemia, idiopathic systemic capillary leak syndrome, intravenous immunoglobulin, shock

MICHELLE BRENNAN

A novel technique for the diagnosis of a rare presentation of renal cell carcinoma

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INTRODUCTION Metastatic renal cell carcinoma (RCC) is common, with 20% to 30% of patients presenting with metastatic disease at the time of diagnosis. Inferior vena caval (IVC) involvement occurs in up to 15% of individuals; however, cardiac metastasis without IVC involvement is rare. Left atrial (LA) involvement is extremely uncommon, and LA involvement without right heart involvement occurs even less often.

Vascular metastasis is a recognized but uncommon cause of acquired pulmonary vein stenosis (PVS). Clinical symptoms include exertional dyspnea, cough, hemoptysis, or chest pain. It can be a fatal condition due to the risk of systemic embolism, cardiac arrhythmias, or circulatory collapse.

Metastatic spread of RCC occurs via several mechanisms—hematogenous spread occurs via IVC with or without direct renal vein invasion and lymphatic spread, via the intrathoracic lymphatic system without IVC involvement or direct extension from adjacent metastatic lesions.

CASE REPORT A 48-year-old man presented with hemoptysis, cough, and dyspnea that occurred 10 months after radical nephrectomy for Fuhrman grade 3 undifferentiated RCC. Computed tomography pulmonary angiography (CTPA) revealed left lower zone consolidation and small pleural effusion. Bronchoscopy was normal. The patient clinically improved after antibiotics for community acquired pneumonia and was discharged home. Two months later, he re-presented with worsening symptoms.

COURSE OF EVENTS On second presentation, CTPA showed a large filling defect in the left inferior pulmonary vein with extension to the LA and progressive left lower zone consolidation. The differential diagnosis for this acquired PVS included myocardial metastasis from RCC or another primary tumor or thrombosis secondary to a paraneoplastic phenomenon. Transthoracic echocardiogram confirmed a moderately-sized mobile mass in the LA, adjacent to the pulmonary vein. Further CT imaging did not show evidence of malignancy elsewhere. Following a multidisciplinary discussion, the patient was anticoagulated and an endobronchial ultrasound (EBUS) was arranged. The primary vascular mass was identified using EBUS and it was subsequently removed using intravascular aspiration.

CLINICAL RESOLUTION Histopathology of the specimens obtained by EBUS confirmed metastatic RCC. The patient was referred to the oncology department and first-line treatment with multireceptor tyrosine kinase inhibitor (pazopanib) was initiated.

DISCUSSION This case describes an extremely rare presentation of pulmonary vein metastasis arising from RCC, most likely occurring via hematogenous spread through the systemic circulation. Pulmonary vein and left atrial metastatic disease from RCC, in the absence of IVC involvement, is extremely rare and a review of the literature revealed only 10 other previously published cases. Notable findings are the absence of IVC involvement, lack of disseminated metastasis, and the atypical route of metastatic spread.

CONCLUSIONS

- Any patient with a history of RCC presenting with dyspnea, hemoptysis, and progressive consolidation should raise suspicion for pulmonary vein involvement.
- Acquired PVS is uncommon and is associated with mortality due to systemic thromboembolism and circulatory collapse.
- New developments in targeted therapies require identification of tumor-specific biomarkers to predict the response, and molecular analysis is key to identifying patients who may benefit from immunotherapy treatments.
- EBUS-guided intravascular aspiration is a novel technique and may facilitate a diagnosis when radiological or surgical approach is not feasible.

Key words
cardiac metastasis, endobronchial ultrasound, intravascular aspiration pulmonary vein stenosis, renal cell carcinoma

MIN SEOK CHANG

Erdheim-Chester disease mimicking IgG4-related disease

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INTRODUCTION Erdheim-Chester disease (ECD) is a rare form of non–Langerhans cell histiocytosis with multiorgan involvement. Although awareness and diagnostic acumen regarding ECD are increasing, the condition is still challenging to diagnose because of its rarity. Here, we report a case of a patient who had been treated for central diabetes insipidus (DI) for 2 years and immunoglobulin G4–related disease (IgG4RD) for 1 year before diagnosis of ECD.

CASE REPORT A 47-year-old woman was admitted to the emergency department due to persistent chest pain and dyspnea, pain in both lower extremities, blurred vision, and headache. She had a history of steroid treatment for IgG4RD and had also been taking desmopressin for the treatment of central DI.

Chest computed tomography (CT) for chest pain evaluation showed a moderate pericardial effusion. A pericardial window surgery was performed for symptom relief and a biopsy of the pericardium revealed mild chronic inflammation with diffuse fibrosis.

The diagnostic workup was continued to find out other possible causes of the symptoms. Abdominal CT showed perinephric infiltration with retroperitoneal soft tissue ("hairy kidney") and multiple sclerotic lesions in the spine and pelvic bone. Bone

scintigraphy revealed localized increased technetium ^{99}m-methylene diphosphonate accumulations in the bilateral zygomaticomaxillary bones, bilateral distal humerus, and bilateral distal femur and tibia. Positron emission tomography-computed tomography (PET-CT) with ^{18}F-labeled fluorodeoxyglucose showed multiple hypermetabolic bone lesions, especially in bilateral distal femurs and tibia and bilateral distal humerus.

Based on these findings ECD was suspected. A bone biopsy was performed at the distal femur but the result was not diagnostic. Bone marrow biopsy showed an increased number of histiocytes with hemophagocytosis, which was suggestive of ECD.

Since *BRAF* mutation was not detected, a high dose of pegylated interferon-α (PEG-INFα) was initiated, as it seems to be the best initial treatment for ECD. After taking PEG-INFα, the patient showed confusion, delusion, and violent behavior. However, these symptoms disappeared after a month and the original symptoms, including chest pain, dyspnea, headache, blurred vision, and pain in both lower extremities, gradually improved. Furthermore, a follow-up PET-CT showed a decreased fluorodeoxyglucose uptake in the lower extremities and a complete resolution of pericardial effusion.

DISCUSSION This case can be an excellent example of diagnosing ECD based on careful history-taking and symptom evaluation. Biopsies in several sites did not help diagnose the disease; however, the symptoms and patient's history were particularly important and prompted imaging studies. Thus, when facing an unexplained inflammatory organ disease, careful history-taking should be performed and ECD should be considered as a differential diagnosis.

Key words
bone scan, Erdhemin-Chester disease, IgG4-related disease, interferon-α

MONIKA RZEŹNIK

Mild elevation of liver function tests associated with renal cell carcinoma hidden in a plethora of symptoms: a case of Stauffer's syndrome

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INTRODUCTION Renal cell carcinoma (RCC) is one of the most common cancers in adults. It is frequently asymptomatic and the diagnosis is often made incidentally during a radiological examination. Paraneoplastic syndromes occur in 20% of patients diagnosed with RCC, and 10% to 40% of them develop a paraneoplastic syndrome after the diagnosis in the course of the disease. One of these syndromes, Stauffer's syndrome, occurs in 3% to 20% of RCC cases, but its exact epidemiology is unknown.

CASE REPORT A 74-year-old patient was admitted to the Department of Internal Medicine and Geriatrics, University Hospital, Kraków, Poland, due to nonspecific general symptoms ("type-B" symptoms) that had lasted for approximately 2 weeks prior to admission. The symptoms included recurrent fever of up to 38.5 °C, nocturnal sweating, weight loss of 4 kg, and noncharacteristic pain in diverse localizations. The pain affected the chest, right side of the neck, middle and left sides of the epigastrium, and the shoulder girdle. The episodes of pain were not related to physical exertion, time of the day, or injuries; they appeared abruptly in different locations. The symptoms were accompanied by abnormal laboratory findings, including elevated inflammatory markers and moderate elevation of liver function tests. The broad diagnostic workup covered the fields of various subspecialties such as pulmonary medicine, cardiology, rheumatology / immunology, oncology, and infectious diseases. During hospitalization, computed tomography revealed a mass in the right kidney and a small, nonspecific nodule in the left lung. Fluorodeoxyglucose positron emission tomography excluded the possibility of neoplastic activity in the lung; no elevated tumor markers were observed. The initial suggestion of the consultant urologist, based on the tumor size and radiographic morphology, was to perform a follow-up scan after 6 months. Due to the fact that the findings were compatible with symptoms of Stauffer's syndrome, the decision was changed to expedient surgery. The patient successfully underwent laparoscopic heminephrectomy. Histological examination revealed clear-cell carcinoma of the right kidney (cT1a). The abnormalities in laboratory tests and most clinical symptoms (particularly fever) abated 2 weeks post surgery.

DISCUSSION Stauffer's syndrome is a rare paraneoplastic syndrome that occurs most commonly in patients with RCC. It is characterized by hepatic dysfunction and elevated levels of the liver enzyme with the absence of metastatic lesions. The main laboratory abnormalities include hypoalbuminemia, hypergammaglobulinemia, high level of alkaline phosphatase, prolonged prothrombin time, and elevated levels of gamma-glutamyl transpeptidase, alfa 2 globulin, interleukin 6, and platelets—all which return to normal values after the removal of the tumor. In the majority of cases documented to date, the liver function tests were 2 to 3-fold higher than the upper normal limit.

CONCLUSIONS The presented case underscores the need to involve a team of multiple specialists in the diagnostic process to reach the appropriate diagnosis. It also points to the possibility that many cases of Stauffer's syndrome may be overlooked. Although our finding is casuistic, we hypothesize that inclusion of elements of mildly expressed Stauffer's syndrome in the diagnostic workup may help in deciding about which radiographically detected renal masses should be promptly removed.

Key words
paraneoplastic syndrome, renal cell carcinoma, Stauffer's syndrome

ROMAN KRÁLIK

Generalized lymphadenopathy in Rosai–Dorfman disease

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INTRODUCTION Rosai–Dorfman disease, also known as sinus histiocytosis with massive lymphadenopathy, belongs to a group of rare hematological diseases with a prevalence of 1:200 000. It was first described in the second half of the 20th century by Juan Rosai and Ronald Dorfman. Children and young adults, especially those around the age of 20 years, are the most frequently affected age groups. In the clinical aspect of the disease, painless lymphadenopathy of the neck dominates in a large spectrum of clinical signs. Diagnosis is based on histological verification. Trademark of the disease are histiocytes positive for proteins S100, CD68, and CD163.

CASE REPORT We present a case of a 64-year-old patient with generalized lymphadenopathy, fever, malaise, and headache. The diagnostic process was arduous from the start, with findings leading to a differential diagnosis of various conditions including neural infection, pneumonia, and lymphoma. Several inconclusive test results, with numerous biopsies among them, caused a prolonged diagnostic workup and hospital stay. After careful evaluation of the obtained results, frequent consultations with specialists, and repeated assessment of histological findings, we were able to diagnose our patient with a rare clinical entity—Rosai–Dormfan disease.

Key words
histiocytosis, lymphadenopathy, Rosai–Dorfman disease

SHAHROZ AZHAR

Warm autoimmune hemolytic anemia and COVID-19

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INTRODUCTION Autoimmune hemolytic anemia (AIHA) has been associated with many conditions, such as autoimmune diseases (systemic lupus erythematosus), lymphoproliferative disorder, and certain infectious diseases as well as the use of various medications. Studies found that severe cases of COVID-19 may be associated with coagulopathies; however, the potential association with AIHA is not clear.

CASE REPORT A patient with no known risk factors or underlying predisposing factors for AIHA presented to hospital with unusual symptoms and profound anemia with complicated blood bank evaluation. He was found to have COVID-19 and AIHA for which extensive laboratory testing was performed, including direct antiglobulin tests and cold agglutinin titers, to identify the causative antibody. A steroid therapy was necessary for clinical stabilization.

DISCUSSION Autoimmune hemolytic anemia is a complex disease with a spectrum of presentations and varying degrees of clinical severity. Many diseases have been identified as predisposing factors for AIHA; however, there are few published reports of patients with COVID-19 and AIHA, most of them involving individuals with other underlying conditions that are known to be associated with AIHA.

CONCLUSIONS There are few reports of patients with concurrent COVID-19 and AIHA and the association between these 2 conditions is not clear. Although COVID-19 has been shown to be linked with coagulopathies, more research is required to determine whether AIHA may also be a potential complication of the disease.

Key words

anemia, autoimmune hemolytic anemia, COVID-19, infectious disease, SARS-CoV-2

TANIA TOFAIL

Hypokalemic paralysis and renal tubular acidosis: initial presentation of Sjogren syndrome

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CASE REPORT Sjogren syndrome is a rare autoimmune disease affecting multiple systems, with a variety of clinical manifestations. We report a case of a 37-year-old woman who presented with recurrent episodes of quadriparesis which was attributable to hypokalemia and was initially diagnosed with hypokalemic periodic paralysis. Later on, she was found to have metabolic acidosis rather than alkalosis, which pointed towards the diagnosis of renal tubular acidosis (RTA) in the absence of apparent gastrointestinal tract loss. Once the diagnosis of RTA was established, an attempt to determine the etiology revealed that the patient had primary Sjogren syndrome, although she did not show any symptoms at the time of diagnosis. She was found positive for anti-SSA antibodies. Lip biopsy revealed lymphocytic infiltration in the periductal and parenchymal regions. Schirmer's test confirmed severe dry eye disease. Of note, concomitant autoimmune hypothyroidism was also found. The patient responded well to potassium supplementation and symptomatic treatment. Presentation of this case highlights the importance of a careful management of recurrent hypokalemia as it might be a rare manifestation of primary Sjogren syndrome.

Key words

distal renal tubular acidosis, hypokalemic paralysis, primary Sjogren syndrome

VALDIS ĢĪBIETIS

Renal and intestinal AA amyloidosis after breast augmentation with silicone gel implants

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INTRODUCTION The report describes a case of renal and intestinal AA amyloidosis of uncertain etiology in a patient with a history of breast augmentation with silicone gel implants and multiple reoperations due to inflammation. Management of AA amyloidosis is based on treatment of the primary cause—chronic infection, inflammation, hereditary disorders, or neoplastic diseases. The primary cause is not identified in 20% of cases.

CASE REPORT A 61-year-old female patient was admitted to a tertiary care center with complaints of constipation, lower left abdominal tenderness, and progressive fatigue. Abdominal computed tomography demonstrated fecal impaction and abdominal lymphadenopathy. Blood tests revealed hypoproteinemia of 44 g/l (reference range, 65–85 g/l), hypoalbuminemia of 21 g/l (reference range, 34–50 g/l), and microcytic anemia (hemoglobin, 79 g/l; mean corpuscular volume, 61 fl; mean corpuscular hemoglobin, 20 pg). The level of C-reactive protein (CRP) was 81.5 mg/l. The patient did not have arthralgia or any signs of synovitis. The serum level of amyloid A was 166 mg/l (reference range, 0–6 mg/l). Serum levels of free kappa and lambda light chains were mildly decreased. Screening for antinuclear antibodies, extractable nuclear antigen antibodies, antineutrophil cytoplasmic antibodies (p-ANCA, c-ANCA), and HIV was negative; the level of anti–double stranded DNA antibodies was normal.

Colonoscopy showed clusters of petechial patches in the mucosa of the rectum and sigmoid colon. Biopsy revealed Congo red–positive intestinal amyloidosis.

The patient had a 4-year history of periodic constipation, diarrhea, and fatigue with episodes of subfebrile body temperature. Biopsy of the enlarged inguinal lymph node had revealed nonspecific reactive lesions. Two bone marrow biopsies had shown signs of reactive lesions. The level of CRP had been persistently elevated to a mild extent. The patient had undergone a cosmetic breast augmentation surgery using silicone gel implants 20 years previously. The implants were reoperated twice because of recurrent inflammation and possible leakage. Presently, mammography, ultrasound, and magnetic resonance imaging of the breasts visualized no pathologies.

Two months after the initial presentation, the patient was hospitalized with nephrotic syndrome and subsegmental pulmonary embolism. Treatment included methylprednisolone, loop diuretics, and rivaroxaban. Kidney biopsy immunohistochemically demonstrated AA amyloid in the glomeruli. Laparotomic abdominal lymph node biopsy confirmed xanthogranulomatous cell infiltration and lipomatosis.

Due to malabsorption and hypoproteinemia, the patient was started on parenteral nutrition. She received a course of 12 mg of dexamethasone, which led to a reduction of the CRP level to reference values and decreased fatigue. The patient sustained a catheter-associated methicillin-resistant *Staphylococcus aureus* infection which was successfully treated with intravenous vancomycin. She was discharged on supplemental parenteral nutrition, low-molecular-weight heparin, methylprednisolone (8 mg daily), spironolactone (25 mg daily), ramipril (1.25 mg daily), and L-thyroxine (50 μg dialy) because of new-onset hypothyroidism.

DISCUSSION In the presented case, no primary disease with proven association with AA amyloidosis was found. Inflammatory reaction after breast augmentation with silicone gel implants and multiple reoperations were considered a probable initiation factor for AA amyloidosis. Absence of current pathology at the site of

breast implantation led to a decision of retaining the implants as their removal would not be beneficial for the patient. The treatment strategy in this patient focused on symptomatic approach. The prognosis remains poor. Systemic amyloidosis causes unspecific symptoms that lead to a prolonged diagnosis. Adverse reaction to silicone gel implants has been scarcely described as a cause of renal amyloidosis but this association remains unclear.

Key words
amyloidosis, breast implants; gastrointestinal, nephrotic syndrome, silicone

OTHER PRESENTATIONS

ADNANUL ALAM

Metastatic tuberculous abscess: a rare manifestation of cutaneous tuberculosis

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INTRODUCTION Cutaneous tuberculosis (CTB) is a rare manifestation of tuberculosis, accounting for 1% to 1.5% of all extrapulmonary tuberculosis manifestations and only 8.4% to 13.7% of all cases of tuberculosis. Metastatic tuberculous abscess, also known as tuberculous gumma, is a type of CTB characterized by the development of cold abscesses which form on the patient's extremities or trunk, without any underlying tissue involvement. We report a case of a patient diagnosed with metastatic tuberculous abscess on the background of immunosuppression caused by prolonged corticosteroid use, without any previous history of tuberculosis.

CASE REPORT A 56-year-old Bangladeshi man presented with high-grade fever and nodular skin lesions which had begun to show 3 months before. The skin lesions first appeared on the thigh, followed by involvement of the trunk. Some of the lesions became larger and ulcerated over time, releasing pus. The patient's comorbidities included hypertension, ischemic heart disease, chronic kidney disease, and undifferentiated spondyloarthritis. He took a number of nonsteroidal anti-inflammatory drugs, oral corticosteroids, and intra-articular steroid injections for pain relief. He had no history of tuberculosis and reported no contact with a person with tuberculosis.

On examination, his face was puffy and neither lymphadenopathy nor organomegaly was observed. Warm, tender, nodular lesions of variable size and shape were found on the abdomen, back, and thigh. Laboratory tests revealed normal blood count with high erythrocyte sedimentation rate and elevated levels of C-reactive protein. Aspiration of pus from a skin lesion revealed numerous *Acid-Fast Baccilli* (AFBs) on staining. Skin biopsy showed perivascular infiltration of chronic inflammatory cells. Myobacterium tuberculosis (MTB) polymerase chain reaction assay of skin biopsy samples detected MTB DNA which was susceptible to rifampicin. Culture of pus in mycobacteria growth indicator tube revealed growth of MTB. Sputum smear test for AFB and culture test for MTB were negative. High-resolution computed tomography of the chest and colonoscopy were performed in search of primary focus of tuberculosis, but neither of them revealed any abnormalities.

The skin lesions were finally diagnosed as metastatic tubercular abscesses. The patient received antitubercular therapy comprising isoniazid, rifampicin, and pyrazinamide in full dose as well as ethambutol in modified doses for renal impairment. There was a significant improvement of the lesions after 1 month of treatment.

CONCLUSIONS A thorough understanding of various presentations of TB is a must-have skill for all clinicians working in countries where the prevalence of this disease is very high. Tuberculous gumma, being a rare type of CTB, has always been challenging to diagnose because of the wide variety of differential diagnoses and difficulty in obtaining microbiological confirmation. The possibility of this condition should be kept in mind to avoid misdiagnosis and case underreporting in countries with a high burden of tuberculosis.

Key words
cutaneous tuberculosis, metastatic tuberculous abscess, tuberculous gumma

ALAN G. ALTAMIRANO

Third time lucky: 3 unusual manifestations of a common disease. A case report of miliary tuberculosis

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INTRODUCTION Tuberculosis is a serious global health problem. The variety of its manifestations, high frequency of atypical presentations, and difficulty in obtaining a microbiological confirmation make it a true diagnostic challenge. A simultaneous presentation of several atypical manifestations makes clinical reasoning not only extraordinarily difficult, but also fascinating.

CASE REPORT A 35-year-old man with a history of dilated cardiomyopathy with ejection fraction of 10% due to noncompaction cardiomyopathy, chronic atrial fibrillation, and previous hospitalizations for acute decompensated heart failure was initially admitted for oppressive chest pain, dyspnea, and fever, with echocardiographic evidence of constrictive pericarditis and pericardial effusion associated with nonerosive symmetrical polyarthritis. Laboratory tests revealed an elevated erythrocyte sedimentation rate; other results were normal. Pericardiocentesis was not performed. Empirical treatment with acetylsalicylic acid, colchicine, and prednisone at 25 mg per day was initiated. The patient was discharged after follow-up echocardiogram showing a decreased volume of pericardial effusion. He was readmitted 19 days after discharge due to dyspnea and persistent polyarthritis, now associated with morning stiffness. Physical examination did not show significant abnormalities. Blood cell count revealed severe isolated thrombocytopenia and chest X-ray showed miliary opacities. High-resolution computed tomography of the chest and bronchoalveolar lavage were performed and immune thrombocytopenic purpura secondary to lung infection with diffuse alveolar hemorrhage was suspected based on the results. Antibiotics (piperacillin / tazobactam and cotrimoxazole), tuberculostatic drugs, and intravenous immunoglobulin were administered. On the fifth day of readmission, the patient presented refractory epistaxis associated with progressive thrombocytopenia; thus, corticosteroid therapy with dexamethasone and a second infusion of intravenous immunoglobulin were initiated. Clinical improvement was observed based on complete remission of thrombocytopenia by day 15, leading to the patient's discharge on tuberculostatic treatment. The bronchoalveolar lavage mycobacterial culture tested positive for *Mycobacterium tuberculosis complex*. A diagnosis of miliary tuberculosis associated with tuberculous pericarditis, Poncet disease, and immune thrombocytopenic purpura was confirmed. On follow-up clinical evaluation after tuberculostatic treatment, remission of pericardial effusion, polyarthritis, and thrombocytopenia was observed.

Although pericarditis is a common manifestation of tuberculosis, there are only 200 reported cases of Poncet disease, and 50 reported cases of immune thrombocytopenic purpura secondary to tuberculosis. However, this is the first case report describing the association of these 3 manifestations in the same patient. Occasionally, unusual presentations of common diseases can lead to misdiagnosis; therefore, physicians must take them into consideration during the diagnostic process.

Key words
immune thrombocytopenic purpura, miliary tuberculosis, pericarditis, Poncet disease, tuberculosis

MARÍA BERNARDA ÁLVAREZ APARICIO

Paradoxical embolism secondary to patent foramen ovale in a 95-year-old woman

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INTRODUCTION Patent foramen ovale (PFO) is a subclinical anomaly present in 25% to 30% of the adult population. Although most individuals are asymptomatic, PFO can serve as a pathway for transit of emboli from the venous to the arterial circulation via right-to-left shunting, leading to paradoxical embolisms that can involve almost any artery of the body. This can result in significant morbidity and mortality, depending on the location of the embolism.

CASE REPORT We describe a case of a 95-year-old woman with a history of hyperthyroidism and ischemic stroke, who presented with sudden dyspnea at rest accompanied by precordial and left arm pain lasting for 3 hours. Physical examination showed normal blood pressure, tachycardia, tachypnea and use of accessory muscles, hypoxemia (saturation of 70% on room air), and an absent left radial pulse with conserved distal perfusion. Electrocardiography revealed sinus tachycardia and highlighted R wave from lead V1 to V2. Chest X-ray was unremarkable.

The ultrasensitive troponin assay was positive (728 pg/ml). Due to the suspicion of pulmonary embolism (PE), anticoagulation with low-molecular-weight heparin was started, and the patient was admitted to the intensive care unit. Computed tomography angiography of the chest confirmed the clinical suspicion of acute bilateral PE with contrast passage through the interatrial septum and a thrombus in the left subclavian artery ostium. Doppler echocardiogram revealed abnormal movement of the interventricular septum and a dilated right ventricle with moderate deterioration of systolic function. The clinical case was interpreted as an intermediate–high risk (PE Severity Score score class III) PE with paradoxical embolism due to PFO.

Emergency arteriography of the left upper limb was performed, showing total occlusion of the left subclavian artery with the presence of a thrombus. Thromboaspiration and balloon angioplasty were performed. Control angiography revealed occlusion of the proximal segment of the humeral artery and thrombi in the radial and cubital arteries. The thrombotic material was surgically removed and oral anticoagulation with warfarin was started. The patient's clinical status was good, without sequelae, and she was discharged 10 days after the diagnosis.

DISCUSSION The most common cause of paradoxical embolism is PFO; therefore, its presence in all patients diagnosed with this type of embolism should be ruled out.

It is important to recognize the signs and symptoms of paradoxical embolism, allowing early diagnosis and, consequently, rapid intervention to minimize morbidity and mortality.

The present case describes a patient with arterial ischemia secondary to PE in relation to PFO. While this association is not frequent, it is a life-threatening complication and may cause severe sequelae. Due to the poor prognosis not only for limb viability but also for survival, further studies should be encouraged to clarify in which cases it would be beneficial to perform a percutaneous closure of PFO or proceed with a medical treatment.

Key words
paradoxical embolism, patent foramen ovale

LORIS AZOYAN

Fever in the French Amazon: the ecstasy of gold

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INTRODUCTION Disseminated histoplasmosis is considered the most common cause of AIDS-defining opportunistic infection in French Guiana. This fungal infection may cause secondary hemophagocytic lymphohistiocytosis.

CASE REPORT We describe a case of a newly diagnosed HIV infection at the AIDS stage revealed by disseminated histoplasmosis complicated by secondary hemophagocytic lymphohistiocytosis, pneumocystis pneumonia, cytomegalovirus retinitis, splenic artery thrombosis, and suspected pleural tuberculosis in a 32-year-old man working in a gold mine in the Amazon rainforest. The clinical outcome was favorable after treatment combining liposomal amphotericin B, hydrocortisone, trimethoprim/sulfamethoxazole, ganciclovir, and antituberculosis drugs followed by antiretroviral therapy with emtricitabine/tenofovir/dolutegravir.

We discuss the reasoning leading to the different diagnoses in a severely ill patient requiring a rapid diagnostic and therapeutic management. This case emphasizes the importance of a patient's environment and its microbial ecology in the diagnostic process in light of the preponderant role of disseminated histoplasmosis as a cause of secondary hemophagocytic lymphohistiocytosis among HIV-infected patients in French Guiana. The other important lesson from this case is that the principle of diagnostic parsimony is not always applicable, especially in immunocompromised patients.

Key words
disseminated histoplasmosis, hemophagocytic lymphohistiocytosis, HIV, splenic infarction

SOFIA BARRETO

From Hippocrates to today. Tetanus: the myocardium also suffers

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INTRODUCTION Tetanus is an acute and globally distributed disease caused by a toxin produced by the Gram-negative and anaerobic bacterium, *Clostridium tetani*. There are 3 clinical presentations of tetanus: generalized, localized, and cephalic. A large number of people are annually affected worldwide: in 2015, this disease killed 50 000 to 80 000 individuals, including approximately 34 000 newborns. In Spain, a mean of 10 yearly cases was reported between the years 2009 and 2015; 69.1% of the diseased individuals were people aged 64 years and over, mostly unvaccinated.

CASE REPORT A 73-year-old woman complained of dyspnea and dysphagia with muscular pain and weakness in the lower extremities (which progressively spread in an ascending pattern to the upper extremities and the jaw), proximal rigidity, and cervical contracture. She had a medical history of chronic venous ulcers in the left lower extremity which she had healed herself with a mixture of clay and banana peels. In 2018, she had been diagnosed with right-sided breast cancer and received surgical treatment accordingly. On admission, the patient was febrile and had stimulation-triggered generalized spasms as well as trismus, facial grimacing (*risus sardonicus*), generalized rigidity, and diminished sensitivity in the proximal region of all extremities, a grade 3 systolic murmur at the mitral focus, and an ulcer (15 × 8 cm) of the distal one-third of the left lower extremity, with foul-smelling, purulent discharge. Laboratory

tests revealed anemia, leukocytosis, an erythrocyte sedimentation rate of 75 mm, a creatine phosphokinase level of 713 U/l, creatine kinase–myocardial band level of 13 U/l, and lactate dehydrogenase level of 860 U/l. To rule out the stiff-person syndrome, computed tomography scans of the brain, neck, and chest were performed as well as a glutamic acid decarboxylase test, which yielded a negative result. Clinical suspicion prompted the treatment of tetanus with human tetanus immune globulin and tetanus toxoid. A medical intervention was performed during hospitalization and an episode of generalized rigidity with respiratory difficulty occurred, requiring an electrocardiogram which showed changes in leads V1, V2, and V3 with ST-segment elevation consistent with anteroseptal acute myocardial infarction. Coronary angiography was performed and no stenotic lesions were identified. The patient was admitted to the intensive care unit, where she received ventilatory support. She developed cardiogenic shock and renal failure requiring hemodialysis. The patient was in the intensive care unit for 25 days with poor treatment outcome and eventually died.

DISCUSSION This case report is clinically significant as there are no previous descriptions of the relationship between acute myocardial infarction and increased sympathetic nervous system activity and autonomic dysfunction. This case also highlights the possibility of re-emergence of preventable diseases in antivaccine communities and incomplete vaccination schemes in people aged 65 years and older.

Key words
Clostridium tetani, stiff-person syndrome, tetanus

ONUR BAS

Nivolumab-induced demyelinating polyneuropathy in a patient with lung cancer

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INTRODUCTION Immunotherapy is a novel treatment option for lung carcinoma which is widely used in clinical practice. However, it is associated with various immune-related adverse events (IRAEs) as a consequence of T-cell activation. Herein, we report a rare case of acute demyelinating polyneuropathy secondary to nivolumab in a lung cancer patient.

CASE REPORT A 66-year-old Caucasian man diagnosed with metastatic squamous cell lung cancer complained of severe cough and weight loss. The patient started first-line gemcitabine and cisplatin and nivolumab treatment due to high (90%) levels of tumoral programmed cell death ligand 1 expression. A week later after the first dose of nivolumab, he was admitted to our clinic with weakness in lower extremities and difficulty in walking. Deep tendon reflexes were hypoactive. The results of laboratory and radiological examinations including cranial magnetic resonance imaging, spinal magnetic resonance imaging, and lumbar puncture were all normal. Electromyography was suggestive of demyelinating polyneuropathy. We started treatment with pulse steroid course of 1 g/d for 5 days. We continued a daily dose of 1 mg/kg methylprednisolone as maintenance therapy. Due to protracted recovery, we started intravenous immunoglobulin (IVIG) for 5 days. One week after IVIG therapy, he started to regain strength in his lower extremity. Upon writing of this text, he was still on a tapering scheme of steroids with continuous physiotherapy.

DISCUSSION Neurological adverse events can be classified in 2 main groups: IRAEs associated with the peripheral nervous system (PNS) and with the central nervous system. Adverse events related to the PNS are more common and symptoms are more specific. Ptosis as well as limb weakness and numbness are the main symptoms. Most well-known IRAEs associated with the PNS are myasthenia and myositis; however, demyelinating neuropathies can occur, as in our patient. Although they are rare, neurological IRAEs are an important problem during treatment and should always be kept in mind for patients who are on immunotherapy.

CONCLUSIONS

- Neurological IRAEs are one of the rarest side effects of immunotherapy.
- Prompt recognition of neurological IRAEs are important in daily clinical practice to prevent potential morbid and fatal outcomes.
- Steroids and IVIG are main treatment options for neurological IRAEs.

Key words
demyelinating polyneuropathy, immunotherapy, neurological immune-related adverse events, nivolumab

MARTA BIANCUCCI

At the crossroads

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INTRODUCTION Duplication of the inferior vena cava (DIVC) is an anatomical variation with a prevalence of 0.1% to 3.5%. It is associated with almost 5% of all deep vein thromboses. Sometimes, DIVC is a contraindication to caval filter placement. Optimal management of patients not eligible for anticoagulation is still being debated.

CASE REPORT An 81-year-old woman with dyslipidemia was admitted to the hospital due to left-sided hyposthenia; brain computed tomography (CT) scan showed a right temporal hyperdense lesion surrounded by edema, compatible with ischemia. Neurosurgery prescribed intravenous corticosteroids and brain magnetic resonance imaging. A nasopharyngeal swab for SARS-CoV-2 tested positive and the patient was admitted to a COVID-19 designated ward. After a sudden worsening in her neurological state, new CT scan was performed. It showed an extension of the temporo-occipital hypodensity, cortical-subcortical laminar necrosis, and focal parietal subarachnoid hemorrhage. Brain magnetic resonance imaging confirmed complete occlusion of the internal carotid artery and showed gyriform hematic leaking. Moreover, atrial fibrillation was newly diagnosed. A neurologist prescribed antiepileptic and antiplatelet therapy; prophylactic low-molecular-weight heparin (LMWH) was introduced. Initially stable, a new hemorrhagic foci developed within the ischemic lesion—antiplatelet therapy was discontinued and LMWH was carried on. The patient's hospital stay was finally complicated by a deep venous thrombosis of the right femoral vein. Although there was a clinical indication for caval filter placement, the abdominal angio-CT scan revealed DIVC. Prophylactic LMWH was continued and, after initial signs of hematic reabsorption, the therapy was administered twice daily but still, full anticoagulation was not achieved.

DISCUSSION DIVC results from the persistence of the left and right supracardinal veins. It can be classified into 4 types depending on the size of the veins or as complete and incomplete depending on the drainage of the left renal vein.

The diagnosis is incidental; a meta-analysis revealed that anatomic studies yield a significantly higher prevalence than imaging and surgery. Literature search revealed numerous cases of missed DIVC with noneffective deployment of caval filters. Generally, nonselective venograms are performed before their positioning, unless the patient has already undergone other forms of imaging. Some studies suggest selective venograms in patients with no clear

visualization of the iliac veins confluence or of the origin of the contralateral common iliac vein.

Nonetheless, cases of successful caval filter placing have been reported despite the absence of established guidelines. There are 4 therapeutic options: bilateral placement of inferior vena cava (IVC) filters, which is the most frequently used; insertion of a single filter with contralateral steel coil embolization; insertion of a single filter with catheter-directed thrombolysis; placement of a single suprarenal IVC filter. The latter option does not protect the renal veins. Dual filter placement seems to be the safest and most effective long term.

CONCLUSIONS DIVC is associated with an increased risk of deep venous thrombosis and should be suspected in cases of recurrent pulmonary embolism. Correct diagnosis of this malformation is crucial for the management and treatment of its complications.

Key words
caval filter, deep vein thrombosis, duplication of the inferior vena cava

PURBASHA BISWAS

An unusual case of monoarthritis and rash

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INTRODUCTION Syphilis is an infectious disease caused by the bacterium Treponema pallidum that is most often transmitted between individuals by sexual contact. The number of syphilis cases have been increasing in various parts of the world, but numerous clinicians have limited experience in the diagnosis and treatment of this disease. If an early diagnosis is not made and treated promptly with antibiotics, the disease may remain latent until a more serious manifestation of tertiary syphilis develops.

CASE REPORT A 48-year-old man presented with a 4-week history of right-sided wrist joint pain with gradual swelling without any other small joint involvement or without any morning stiffness. There was no preceding trauma. His serology for rheumatoid factor was weakly positive and he was prescribed methotrexate (weekly dose) and daily sulfasalazine. Accidentally, our patient took methotrexate continuously for 7 days and again presented to us with multiple genital ulcers and vesicopustular lesions on bilateral palms and soles along with oral mucosa. He was initially evaluated for an overdose of methotrexate. Over the ensuing 4 days, wrist pain and swelling reduced in severity and active motion improved with analgesics without any resolution of rashes. During that time, he was assessed by dermatology and genitourinary specialists and was referred for a VDRL slide test (rapid plasma regain card test). This test came out to be reactive with both diluted and undiluted serum along with a positive modified *Treponema pallidum* hemagglutination assay test.

CLINICAL RESOLUTION The patient was given an injection of benzathine penicillin and his skin lesions started resolving. He was discharged and referred to an antiretroviral clinic for the management of his ICTC positive status.

DISCUSSION Syphilis has an early infectious phase and a late noninfectious phase. Stages of the disease have been described as primary, secondary, and tertiary, but the natural history of the disease is highly variable. Primary syphilis usually starts with a small genital or perianal ulcer, known as chancre. It develops 2 to 3 weeks following contact and may take up to 3 months to appear. Secondary syphilis most commonly presents with a prodrome with fever in addition to rash which involves hands, feet, and also the mucous membrane. Polyarthritis with synovitis associated with early syphilis have been described usually 3 to 12 weeks into the secondary stage. The case we report is unusual because monoarthritis rather than polyarthritis was the presenting feature. Diagnosing syphilis in HIV-positive patients remains a clinical challenge since in this population, it frequently manifests with atypical features and chronology. Our patient had an acute history of skin lesions and orthopedic involvement, but absence of primary syphilis was a diagnostic difficulty. This case is noteworthy in that to our knowledge, it is one of the first reports of syphilis in HIV-positive patients in whom the skin manifestation did occur in the secondary stage of syphilis along with monoarthritis instead of polyarthritis.

Key words
HIV, monoarthritis, syphilis

PAULA BORT

Hemorrhagic stroke in a young patient with infective endocarditis: a case report

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INTRODUCTION Infective endocarditis is a major clinical problem with mortality rates reaching 20% to 40%, with a similar percentage of neurological complications that are associated with poorer outcomes. Deciding whether to perform cardiac surgery and when to do it proves challenging, as it is important to determine the influence of neurological complications on the surgical intervention, and vice versa.

CASE REPORT A 25-year-old woman with a history of mitral valve prolapse with associated mild insufficiency presented to the emergency department due to atypical behavior and expressive aphasia that were detected on the morning of the consultation. On physical examination, she was somnolent, with right faciobrachiocrural hemiparesis, and an already known mitral regurgitation murmur. Magnetic resonance imaging showed a left frontal intraparenchymal hematoma with a 8-mm shift in the midline. It was interpreted as hemorrhagic stroke of unknown cause. Brain angiography showed no abnormalities. The condition of the patient deteriorated with impaired sensorium requiring mechanical ventilation and vasoactive drugs. Wide craniectomy and hematoma evacuation was performed with resection of what appeared to be a cavernoma; intracranial pressure catheter was placed. Biopsy obtained during surgery showed fibrinohematic material, which ruled out cavernoma.

The patient had a history of fever and reported having isolated episodes of fever for 4 months. *Streptococcus mitis* was isolated in 2 blood cultures and antibiotic treatment was initiated. Transesophageal echocardiogram showed a vegetation measuring 10×5 mm, with moderate insufficiency associated. New brain magnetic resonance imaging showed pinpoint ischemic foci probably related to septic impacts. The vascular surgery service was consulted and the surgical option was dismissed due to a high risk of new intraparenchymal bleeding.

DISCUSSION Infective endocarditis is associated with a broad array of systemic complications. Neurological complications are more common than previously believed, prevalence rates reaching between 23% to 50% in the literature. Factors that were more associated with neurological complications were the isolation of *S. aureus* in the cultures, involvement of the mitral valve, and other non-neurological embolic events, while the variable that was mainly associated with mortality to 3 months was sensory impairment on admission, defined as Glasgow Coma Scale score of less than 10. Within the group of hemorrhagic complications, mortality at 1 year was higher in patients with bleeding of unknown etiology compared with those with bleeding due to hemorrhagic transformation of ischemic strokes or aneurysm rupture.

Key words
hemorrhagic stroke, infective endocarditis, mitral insufficiency, surgery, young

DANIELA CABRAL

Perseverance and extrapulmonary sarcoidosis: a case report

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INTRODUCTION Sarcoidosis is a multisystemic granulomatous disease of unknown cause. It can present at any age but is more common in adults younger than 50 years.

CASE REPORT A 26-year-old male patient presented with irritating cough, oppressive chest pain 7/10, and night sweats that started 4 months ago. The symptoms worsened to World Health Organization functional class (WHO FC) II/III dyspnea, 10/10 chest pain, for which he presented again at a tertiary hospital where echocardiogram was performed. It showed severe pericardial effusion with compression of right cavities without cardiac tamponade. He was admitted to the coronary care unit where pericardiocentesis and thoracentesis were performed. Tuberculosis was suspected but purified protein derivative, BAAR cultures of pleural fluid, and adenosine deaminase were negative. Blood, urine, and mycological cultures were also negative. Immunological profile FAN 1/80 and the rest were negative. The diagnosis of polyserositis of immunological cause was assumed and empiric therapy with prednisone at a dose of 1 mg/kg of body weight for 3 days was started with improvement of the patient, reduction of lymphadenopathy, and resolution of serosal involvement. He was discharged and referred for a follow-up visit.

He was readmitted 45 days later due to pleuritic pain and a 7-day history of fever. Chest computed tomography was performed and showed lobulated pleural effusion probably related to a major fissure in the right pulmonary branch. The histological examination of fragments of pleural and lung tissue showed chronic granulomatous inflammatory appearance consisting of tuberculoid-type granulomas with areas of central necrosis and multinucleated giant cells of the Langhans type. Ziehl–Neelsen stains were negative. Pleural peel culture for BAAR was also negative. A diagnosis of extrapulmonary sarcoidosis was made and treatment with prednisone 1 mg/kg of body weight was started with good response.

DISCUSSION Since sarcoidosis is the great imitator with no specific standard test for its diagnosis, it can be confused with other collagenopathies, such as systemic lupus erythematosus. It is also a common manifestation of granulomatous infections such as tuberculosis and even systemic mycoses—that is why the start of treatment with corticosteroids was delayed in our case.

Key words
chest pain, polyserositis, sarcoidosis, tuberculosis

LORENZO MARIA CANZIANI

From pulmonary embolism to spontaneous iliopsoas hematoma: a COVID-19 tale

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INTRODUCTION In 2020, numerous countries faced the COVID-19 pandemic, which hit at varying times and with varying strength all over the world. As physicians soon learned, the most common problems related to COVID-19, rarer complications, need to be highlighted to improve current clinical care.

CASE REPORT During the second COVID-19 pandemic wave in northern Italy, a man in his 80's with a history of hypertension and ischemic cardiopathy with preserved ejection fraction presented to the emergency department complaining of fever, cough, and asthenia for some days. His wife was recently admitted for COVID-19.

The patient was eupneic with O_2 saturation of 93% on ambient air and mildly febrile. Physical examination showed mildly reduced vesicular murmur in the lungs. Blood exams revealed leukocyte count within the reference range, a hemoglobin level of 12.5 g/dl (reference range, 13–18 g/dl), a C-reactive protein level of 13.01 mg/dl (reference range <0.50 mg/dl), a procalcitonin level of 0.2 ng/ml (reference range <0.5 ng/l), a D-dimer level of 1330 ng/ml (reference range of 22–350 ng/ml), and an estimated glomerular filtration rate of 51 ml/min/1.73 m^2. Computed tomography (CT) pulmonary angiography showed bilateral ground-glass areas, without evidence of pulmonary embolism. A nasopharyngeal swab confirmed SARS-CoV-2 infection.

On admission, intravenous dexamethasone at a dose of 6 mg daily and subcutaneous nadroparin at a dose of 3800 IU daily were prescribed, along with usual therapy. Mild respiratory failure resolved in a few days; the C-reactive protein level decreased to 3.2 mg/dl and D-dimer, to 1273 ng/ml.

On day 7, the need for oxygen rose to 4 l/min and the D-dimer level increased (4741 ng/ml) without signs of deep venous thrombosis or hemodynamic instability. Subsequent CT pulmonary angiography revealed left lobar pulmonary embolism. Subcutaneous nadroparin dose was increased to 5700 IU twice daily. Echocardiogram did not show indirect signs of pulmonary hypertension.

On day 12, the patient reported lower back pain and new onset weakness of the right quadriceps femoris (grade 3/5) with associated hypotension (95/55 mm Hg). Blood tests showed acute kidney injury (estimated glomerular filtration rate, 14 ml/min/1.73 m^2), and a decrease in the level of hemoglobin (7.5 g/dl). Brain CT excluded acute neurological event. Total body CT showed right iliopsoas hematoma (16 cm in diameter) with a minor active bleeding. Hemotransfusion was performed with volume support, and nadroparin was discontinued. As the patient rapidly reached hemodynamic stability, no endovascular treatment was indicated.

Serial CT scans confirmed the progressive volumetric reduction of the hematoma. The level of hemoglobin remained stable, and acute kidney injury resolved. Nadroparin was gradually reintroduced with antifactor Xa activity monitoring. After 30 days, the patient was transferred to rehabilitation for reduced mobility due to prolonged bed rest.

DISCUSSION Pulmonary embolism is a common complication in COVID-19 that happens despite weight-adjusted prophylactic dose of nadroparin. Spontaneous hematomas are reported in COVID-19 with estimated incidence of 7.6 cases per 1000 hospitalizations, especially in patients receiving full-dose heparin and those critically ill. Our case illustrates the variability of coagulopathy complications that can arise from COVID-19. Besides thromboembolic complications, also bleeding risk is increased. This case was challenging due to the occurrence of both complications. We encourage clinicians to consider both risks in this patient population.

Key words
bleeding, COVID-19, pulmonary embolism

MARIO ARIEL CASTILLO

Leprosy, a condition with past and present: a case report

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INTRODUCTION Leprosy or Hansen disease is a bacterial infection caused by the *Mycobacterium leprae* complex. Although it remains a major global public health concern, highly effective therapies are

available. Its prevalence varies widely: on a yearly basis, 300 to 400 cases are identified in Argentina with the endemic areas in the northwestern, northeastern, and central provinces. The precise pathogenic mechanisms of the disease are not fully understood; however, a number of factors contribute to its development, including the immune status and genetic influences. The clinical classification of leprosy differentiates the following forms: tuberculoid leprosy, borderline tuberculoid leprosy, mid-borderline leprosy, borderline lepromatous leprosy, lepromatous leprosy, and indeterminate leprosy. Patients with high levels of cellular immunity and delayed sensitivity reaction are likely to have tuberculoid leprosy with relatively few well-defined lesions. They are otherwise likely to have lepromatous leprosy with a number of ill-defined lesions. The pathophysiology of the disease is characterized by an initial increased blood level of bacilli which can lead to recurrent episodes or progress to sustained bacteremia. Multiple organs are exposed to the bacilli during these episodes. The abundance of surrounding bacteria allows their identification in peripheral blood smears.

CASE REPORT A 36-year-old man with past medical history of severe chronic alcoholic intoxication and a 2-year history of hyperchromic lesions in the lower extremities reported interscapular burning pain in the dorsal region, subjective fevers, night sweats, and a 3-week history of 10-kg weight loss. On presentation, the patient had multiple skin lesions, madarosis, widespread nodular subcutaneous infiltrates, bullous lesions, and hypoesthetic hypopigmented macules with predominance in the lower extremities. Computed tomography scan showed multiple swollen glands, bilateral pleural effusion, and heterogeneous splenomegaly with multiple focal images. RMI revealed hyperintense areas in several lumbar and dorsal vertebral bodies with compromised spinal canal diameters. Bacilloscopy was 3+ with morphological index denoting high infectivity in specimens from the earlobe and nasal mucosa; biopsy specimens from the lymph nodes and dorsal vertebrae revealed findings consistent with lepromatous leprosy. Specific treatment was started with partial improvement and multiple intercurrent infectious diseases. Conditions such as HIV infection, multiple myeloma, myeloproliferative and lymphoproliferative syndromes were ruled out as possible causes of immunosuppression. The patient did well and was discharged from the hospital. Seven days later the patient was readmitted and diagnosed with septic syndrome. He died in the emergency room.

DISCUSSION We report an unusual case of organ involvement in Hansen disease. Spondylodiscitis and spleen involvement in this condition have not been previously reported in the literature. It is remarkable that basic test did not reveal a specific cause of immunological compromise, which is a prerequisite for the development of lepromatous leprosy; however, crowding, alcohol abuse, and poor nutrition were likely contributors to the development of an aggressive form of the disease. Our report is clinically significant as it highlights the rarity of the disease as well as lack of knowledge and attitudes of prejudice surrounding it. Moreover, it is usually neglected in the differential diagnosis in the daily clinical practice.

Key words
Mycobacterium leprae, lepromatous leprosy, leprosy, spondylodiscitis

UDDALAK CHAKRABORTY

An unusual case of recurrent ataxia and dysarthria

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INTRODUCTION The spectrum of the demyelinating diseases of the central nervous system is vast, heterogenous, and often with overlapping clinical presentations. Misdiagnosis might occur in up to 10% of cases with serious therapeutic repercussions. However, the introduction of several new biomarkers such as aquaporin 4 immunoglobulin G (IgG) and myelin oligodendrocyte glycoprotein IgG has made distinction between multiple sclerosis (MS) and other primary demyelination diseases of the central nervous system easier.

CASE REPORT A 15-year-old man presented with an acute onset unsteadiness and a nonprogressive weakness on his left side for 2 days. He also experienced clumsiness of fine movements and difficulty in feeding himself along with slurring of speech for 1 month. His mother noticed a high stepping gait on his right side. During this episode, neurological examination revealed an upper motor neuron weakness with an extensor plantar response on his left side, accompanied by cerebellar signs and impaired posterior column sensations on his right side. The patient had recurrent episodes of vomiting 3 months back which was treated conservatively with symptomatic improvement. Five years back, he had an episode of acute onset dysarthria, vertical misalignment of eyes, and unsteadiness. Brain magnetic resonance imaging (MRI) revealed hyperintensities in the midbrain, superior cerebellar peduncles, and right occipital periventricular region. The patient was initially diagnosed with acute demyelinating encephalomyelitis and treated with intravenous methylprednisolone during that episode with symptomatic improvement over 1 month. Follow-up brain MRI revealed no abnormalities. None of the above-mentioned episodes had any previous antecedent illness. During the current episode, brain MRI revealed hyperintense lesions in the right midbrain, right cerebellar hemisphere, and both occipital periventricular regions in T2 fluid-attenuated inversion recovery sequence; without any restriction in diffusion-weighted imaging. Screening MRI of both orbits and whole spine did not reveal any abnormality. Cerebrospinal fluid study revealed mild lymphocytic pleocytosis with mild elevated protein and absence of oligoclonal bands with an IgG index of 0.56. Visual evoked potential showed bilaterally prolonged P100 latency. A secondary demyelinating etiology was ruled out by supportive investigations. A test for serum IgG antibodies to aquaporin 4, specific for neuromyelitis optica (NMO), and for serum IgG antibodies to myelin oligodendrocyte glycoprotein (MOG) was performed and was positive for IgG antibodies to MOG. The patient was started on pulse methylprednisolone therapy followed by oral corticosteroids, with prompt recovery. He was finally diagnosed with MOG antibody disease (MOGAD), and is currently maintained on mycophenolate mofetil and oral prednisolone. MOGAD is distinct from MS and aquaporin 4 antibody NMO spectrum disorders (NMOSD) with varied phenotypes ranging from classic NMO to acute demyelinating encephalomyelitis and cortical encephalitis. Clinical and imaging features of MOGAD may overlap with NMO, but can be usually distinguished from MS. The disease is highly responsive to corticosteroid therapy and immunosuppressants and the course of the disease may be monophasic or relapsing despite good response.

Keywords
ataxia, dysarthria, acute demyelinating encephalomyelitis, myelin oligodendrocyte glycoprotein antibody

MANALI CHANDRA

Isolated third cranial nerve palsy as a presenting manifestation of protein S deficiency: a report of an exceedingly rare clinical entity

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CASE REPORT Cranial mononeuropathy is a commonly encountered neurological problem in day to day practice. Isolated third cranial

palsy is commonly caused by trauma, microvascular obstruction, compression from aneurysm or neoplasm, stroke, and Tolosa-Hunt syndrome. We report a case of a 30-year-old woman who was admitted to our facility with a sudden onset right of sided ptosis and ophthalmoplegia. Detailed clinical features, neuroimaging, and laboratory evaluation clinched the diagnosis of ischemic stroke in the midbrain from inherited thrombophilia due to protein S deficiency. She was treated with aspirin, atorvastatin, and physiotherapy.

Key words
cranial mononeuropathy, ischemic stroke, protein S deficiency

BELÉN CONDE SERRA

Mediastinal lymph nodes: a challenge between benign and malignant pathology

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INTRODUCTION Mediastinal lymphadenopathy can be caused by multiple factors and requires extensive differential diagnosis. Tuberculosis is a systemic infection caused by *Mycobacterium tuberculosis*, and although the lungs are the major site of infection, it can reach other organs by hematogenous or lymphatic dissemination.
CASE REPORT A previously healthy 28 year-old woman presented to the emergency department with a 2-week history of fever, non-productive cough, and night sweats. In terms of epidemiological background, she lived in Buenos Aires City, Argentina, worked as a surgical technologist in a private hospital, and had not travelled in the previous 6 months. The physical examination did not reveal any abnormalities. She was admitted to the general ward for further assessment. Blood tests showed increased acute phase reactants; blood and urine cultures, nasopharyngeal and oropharyngeal swabs for COVID-19, and viral serologies were negative. Chest, abdomen, and pelvic tomography showed multiple enlarged mediastinal lymph nodes. Due to a suspicion of lymphoproliferative disorder, a positron emission tomography scan was performed which revealed hypermetabolic supradiaphragmatic lymph nodes and new bilateral pulmonary opacities. A Mantoux tuberculin skin test was positive; therefore, treatment with isoniazid was started for latent tuberculosis. The patient ultimately underwent a left videothoracoscopy with a lymph node biopsy. Histopathological analysis revealed necrotizing granulomatous adenitis and the biopsy culture was positive for *M. tuberculosis* complex. The condition was then interpreted as tuberculosis and first-line treatment with 4 drugs was started.
CONCLUSIONS The clinical presentation of tuberculosis is heterogeneous and there is an increased risk of acquisition amongst health workers from endemic regions due to higher exposure rates. This case emphasizes the importance of a high level of pretest clinical suspicion based on the epidemiology, history, and physical examination to establish an adequate diagnostic work-up resulting in early treatment.

Key words
health workers, lymphadenopathies, malignancies, tuberculosis

VANESA DADON

When you don't know why the patients bleed... think of scurvy

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CASE REPORT Scurvy results from dietary deficiency of vitamin C, which is mostly found in citruses and fresh vegetables. It was described more than 3 centuries ago, when access to these foods was limited on ships, especially during long sea voyages. The main symptoms of scurvy are fatigue, muscle or bone pain, and spontaneous hemorrhages. Risk factors are poor socioeconomic conditions, alcoholism, or psychiatric disorders leading to malnutrition.

We present a 52-year-old man with a history of alcoholism admitted for weakness, sacrum pain, and multiple spontaneous hematomas in the lower extremities. We found gingival hypertrophy with severe periodontal inflammation and skin lesions characterized by perifollicular hyperkeratotic papules and purpura. He had pancytopenia and prolonged prothrombin time that improved with vitamin K supplementation. Abdomen and pelvis computed tomography scan was normal, without evidence of cirrhosis or portal hypertension. He developed episodes of hematochezia. We suspected scurvy. A great clinical improvement was achieved with treatment with intravenous vitamin C (1 g/d).

In the event of spontaneous hemorrhage associated with normal coagulation tests, the possibility of scurvy should be considered.

Key words
alcoholism, hematoma, scurvy, vitamin C

LEILA DUARTE

Fever and giant liver hemangioma: when Occam's razor is the answer

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INTRODUCTION Giant liver hemangioma (GLH) is mostly asymptomatic, and therefore, it is usually identified as an incidental finding on imaging. Cases of fever associated with GLH are rare and seldom reported in the literature.
CASE REPORT We describe a case of fever in a 54-year-old woman justified by GLH with signs of acute internal bleeding and infection, the latter resulting from a dental procedure performed weeks before. Hepatic ultrasound, computed tomography, and magnetic resonance imaging showed characteristics typical of hemangioma, although the findings, apart from recent hemorrhage, could not be used to clearly exclude infection. In the meantime, an extensive diagnostic workup was performed, and the patient received antibiotic empiric therapy with ceftriaxone, metronidazole, and doxycycline.
DISCUSSION In spite of being a rare association, GLH and fever could be presumably explained by immunological mediation due to pyrogens produced by Kupffer and endothelial cells. Surgery is an option for GLH complications, notwithstanding its morbidity and mortality. However, it apparently solves the inflammatory syndrome. In this clinical case, the option pursued by the medical team consisted of conservative management with empiric antibiotics. This proved to be a positive decision as the fever was eradicated.

After 2 years of follow-up, the patient did not report any relapsed fever, size of the hemangioma remained the same, and there was no further development of any malignancy nor autoimmune or infectious disease. Careful assessment excluded all other sources of inflammation except for GLH, proving to be a case of Occam's razor.

Key words
fever, giant liver hemangioma

HASAN ERUZUN

Late-onset common variable immunodeficiency: a case report

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INTRODUCTION Common variable immunodeficiency is a condition characterized by inadequate immunoglobulin synthesis due to impaired B cell differentiation. It is the most common primary immune deficiency in adults.

CASE REPORT A 60-year-old man was admitted with symptoms of decreased vision, weight loss, and diarrhea that had been ongoing for the last 8 months. The patient, who had a history of diabetes and ischemic heart disease, had sinusitis for the past 1 year. Perianal abscess drainage was performed 4 months ago. Vision loss started as blurred vision and progressed. The diarrhea was watery, occurring 4 or 5 times a day. On examination, bowel sounds were increased. The liver edge and splenic edge were 2 cm and 4 cm below the costal margin, respectively. Cardiac, respiratory, and neurological examinations were unremarkable. Anti-HIV antibodies were negative and there was no proteinuria. Hemogram was normal, except for mild anemia and lymphopenia. No parasite and parasite eggs were seen and feces cultures were negative. The C-reactive protein level and sedimentation rate were normal. The Brucella and Salmonella tube agglutination test was negative, but the direct Coombs test was positive. Antinuclear antibody, anti–double stranded DNA, and tissue transglutaminase immunoglobulin (Ig) A were negative. Immunoglobulin analyses revealed the IgA level of 24 mg/dl (reference range, 70–400 mg/dl), IgG level of 135 mg/dl (reference range, 700–1600 mg/dl), and IgM level of 15.3 mg/dl (reference range, 40–230 mg/dl). All of the subgroups of IgG were low. An increase in focal reticulin fiber, intertrabecular micronodules, rare atypical B lymphoid cells, and reactive T cells was seen in the examination of the bone marrow. Visual evoked potentials were observed, which were compatible with toxic / paraneoplastic optic neuropathy. There were no findings of uveitis in the fundus examination. Positron emission tomography was performed. The spleen was enlarged, fluorodeoxyglucose involvement was diffuse and slightly above the liver. It was decided to perform a splenectomy as a step towards diagnosis and treatment. An enlarged spleen and B-cell proliferation with an IgM kappa-type zonal pattern in the spleen suggested that there may be a minimal zonal involvement of low-grade B-cell lymphoproliferative disease. In addition, it was suggested to evaluate the patient in terms of immune deficiency syndromes due to absence of germinal center organization and no evidence of IgG and IgA synthesis. When all the findings were evaluated, the case was considered as splenic B-cell lymphoma secondary to common variable immune deficiency. Intravenous immunoglobulin treatment was started and performed every 3 weeks. The patient was followed for 4 years with subcutaneous IG and no new infection episodes. There are no new findings in favor of malignancy.

Key words
common variable immunodeficiency, hypogammaglobulinemia, immune deficiency

SEDAT IRMAK

A case of amyloidosis presenting with neurological symptoms

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INTRODUCTION Amyloidosis is the accumulation of insoluble fibril proteins in tissues in an amount sufficient to disrupt normal function. It is a rare condition that can present as a systemic or local disease and can be diagnosed by histopathological confirmation. Patients with amyloidosis present with a wide array of signs and symptoms which depend on the site of amyloid accumulation.

CASE REPORT A 58-year old Caucasian man was admitted to the hospital with a 10-month history of dysphagia, fatigue, and muscle weakness. He reported a weight loss of 30 kg over 10 months and has not been able to walk for the past 3 months. The past medical history of the patient included pulmonary tuberculosis, which had been treated successfully 20 years ago.

Numerous physicians examined him in different hospitals during the past year. A battery of tests was performed during the diagnostic workup, including gastroscopy, positron emission tomography, and thoracentesis. However, the diagnosis remained unclear and the patient referred to our clinic for further investigation.

Physical examination revealed a blood pressure of 80/40 mm Hg, regular pulse of 80/min, hepatomegaly, mild mydriasis in the left eye, diminished breath sounds in lung bases, and grade 2 pitting edema in lower legs. He had generalized muscle weakness of the body.

On admission, hemoglobin, platelets, neutrophils, and lymphocytes were within reference ranges. Laboratory values on admission were as follows: creatinine, 1.36 mg/dl (reference range, 0.67–1.17 mg/dl), urea, 97 mg/dl (reference range, 19–55 mg/dl), aspartate aminotransferase, 72 U/l (reference range, 5–34 U/l), alanine aminotransferase, 80 U/l (reference range, 0–41 U/l), alkaline phosphatase, 709 U/l (reference range, 40–20 U/l), and γ-glutamyltransferase, 687 U/l (reference range, 12–64 U/l). Lactate dehydrogenase was elevated at 271 U/l (reference range, 125–220 U/l). Albumin level on admission was 2.93 g/dl (reference range, 3.5–5.2 g/dl). Tests for hepatitis B and C were negative. Urinalysis on admission showed 2+ protein and microhematuria.

We performed the barium swallow test to determine the underlying cause of dysphagia. The test showed no sign of esophageal obstruction but gave us a clue that the patient might have a motor neuron disease. The patient subsequently was evaluated by a neurologist who performed additional testing, including cranial magnetic resonance imaging with normal results and electromyography which showed nothing but mixed-type polyneuropathy.

Abdomen ultrasonography revealed an enlarged liver and enlarged left kidney. The antimitochondrial antibody, antismooth muscle antibody, and liver-kidney microsomal autoantibody type 1 were negative. The protein level measured in a 24-hour urine sample was 5.5 g. These findings made us think of dense deposit diseases, especially amyloidosis. Kidney biopsy confirmed the diagnosis of amyloid A (AA) amyloidosis.

We speculated that tuberculosis, which the patient had 20 years ago, caused AA amyloidosis despite successful treatment. Once established, there is no specific treatment for amyloidosis due to tuberculosis. Unfortunately, the patient died on the day after the diagnosis was established due to ventricular arrhythmia.

DISCUSSION In our case, findings such as dysphagia, muscle atrophy, and neurologic abnormalities misled the physicians who

the patient saw in the past year and us as well. Proteinuria was a key diagnostic clue in this case and led us to the diagnosis.

Key words
amyloidosis, autonomic neuropathy, dysphagia

BAS R. P. JONKERS

It takes 2 to tango; disseminated cutaneous *Mycobacterium chelonae* infection in Cushing syndrome

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CASE REPORT A 56-year old woman was referred to the infectious diseases department for further evaluation and treatment of a cutaneous infection with nontuberculous mycobacteria species. She was recently treated by a surgeon for a number of skin abscesses by means of drainage and antibiotics. An additional skin biopsy demonstrated purulent and lipogranulomatous inflammation and the acid fast staining confirmed mycobacteria. Cultures grew *Mycobacterium chelonae*, usually found in immunocompromised hosts.

Previous medical history was significant for hypertension and in the year prior to presentation she was diagnosed with difficult-to-manage type 2 diabetes and polyneuropathy of uncertain origin for which she had been using a walking aid. Her weight increased slightly over the past months and she denied fever or night sweats. On physical examination, multiple erythematous maculopapular lesions were noted, some of which contained vesicles. Apart from the skin lesions, physical examination showed a cushingoid phenotype with moon facies, central obesity, proximal muscle atrophy, and easy bruising.

After ruling out HIV infection, we suspected an immunosuppressed state due to hypercortisolism. Biochemical hypercortisolism was proven by an abnormal dexamethasone suppression test, elevated urinary free and midnight salivary cortisol. Adrenocorticotrophic hormone was suppressed and computed tomography of the abdomen revealed a nodular lesion in the left adrenal gland, suspect for adrenal adenoma. Surgical resection was performed and pathology confirmed an adrenal adenoma. Treatment was started with clarithromycin, imipenem, and clofazimine based on susceptibility results. After a month of triple therapy, the imipenem was discontinued and dual therapy was continued with good clinical response.

In conclusion, we report a disseminated cutaneous *M. chelonae* infection in an immunocompromised patient due to underlying Cushing syndrome caused by adrenal adenoma.

Key words
Cushing syndrome, *Mycobacterium chelonae*, nontuberculous mycobacteria, opportunistic infection

KATRIN KEERMA

A case report of adrenal pheocromocytoma

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INTRODUCTION Pheochromocytoma is a rare catecholamine-secreting tumor usually originating from the adrenal medulla and producing signs and symptoms caused by excessive catecholamine secretion from tumor with a prevalence of approximately 0.1% in patients with hypertension and 4% in patients with incidentally discovered adrenal mass.

CASE REPORT A 42-year-old man presenting with nausea and dyspnea was admitted to a county hospital with hypertensive urgency, diabetes decompensation, and pneumonia of the left lung. Two days later, the patient presented with left-sided chest pain. Echocardiography and electrocardiography were performed and blood samples were obtained. Based on the new contractile dysfunction on echocardiography, T-wave inversion on electrocardiography, and increased cardiac troponin T level (>300 ng/l), the patient was admitted to a regional hospital with non–ST-segment elevation myocardial infarction. Coronary angiography was performed, no indication for therapeutic intervention was needed. The patient was then tested for pheochromocytoma. Based on the 24-hour urinary catecholamine level, serum catecholamine levels, and previous abdominal computed tomography scan, the patient was diagnosed with pheochromocytoma with a mass in the left adrenal gland. Previously prescribed extended-release β-blocker was discontinued, and perioperative doxazocin therapy was initiated. Left adrenalectomy was performed approximately 5 weeks after admission to the hospital. In the postoperative period, the patient was evaluated by an endocrinologists for residual tumor tissue or malignancy, which were not confirmed. The patient is currently being followed by an endocrinologist every 1 year. Pheochromocytoma is a rare, but potentially fatal, disease if left undiagnosed and untreated. Pheochromocytoma, when discovered in time, is a potentially curable cause of hypertension with an excellent overall prognosis.

Key words
adrenalectomy, adrenal glands, pheochromocytoma

CATALINA KLUG

Hypovolemic shock due to spontaneous rupture of hepatocarcinoma as the initial manifestation of the disease: a case report

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CASE REPORT Hepatocellular carcinoma is the leading primary liver cancer and it is one of the main causes of neoplasm-related deaths worldwide. Both its incidence and prevalence are on the rise. Spontaneous rupture is not a common complication (approximately 10%) but it is extremely serious. Clinical manifestations and therapeutic approach contribute to a better management of these cases. Currently, in instances of emergency, the chosen therapeutic approach is the selective hepatic transarterial embolization. We report a case of a patient who presented with hypovolemic shock and was found to have spontaneous rupture of hepatocellular carcinoma.

Key words
cirrhosis, hepatocellular carcinoma, hemoperitoneum, transarterial hepatic embolization

TAYYEBA KOMAL

Cerebral venous sinus thrombosis in a pregnant woman presenting with hemiplegia and aphasia

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CASE REPORT Cerebral venous sinus thrombosis is a rare and potentially deadly condition during pregnancy due to hypercoagubility. It can result in a wide range of neurological symptoms and very rarely, hemiplegia. We report a case of a young pregnant Pakistani

woman presenting with right hemiplegia and aphasia after undergoing lower-segment caesarean section for intrauterine fetal death due to eclampsia. On examination, her blood pressure was 150/100 mm Hg and manual muscle testing showed 0/5 grade in right upper and lower limbs and an upward response of the right plantar reflex. Brain computed tomography showed brain edema, and magnetic resonance venogram confirmed cerebral venous sinus thrombosis and the area of infarction. The patient was treated in the intensive care unit with anticoagulants, antihypertensives, anticonvulsants, and brain edema–lowering drugs. She was discharged with full recovery after 29 days of admission. Cerebral venous sinus thrombosis with a cerebrovascular event should always be considered in the differential diagnosis in a patient presenting with neurological symptoms during pregnancy.

Key words
aphasia, cerebral venous sinus thrombosis, hemiplegia, pregnancy

HINA LATIF

Extranodal manifestation of Kikuchi disease: a rare presentation

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CASE REPORT Kikuchi-Fujimoto disease is a self-limiting disease which usually presents with lymphadenopathy and fever. We report a case of a man who presented with complaints of cervical lymphadenopathy and fever. A lymph node biopsy was performed which was suggestive of Kikuchi disease. Awareness and knowledge of this disorder and its extranodal presentation are essential for physicians and healthcare workers to ensure appropriate care and treatment of such patients.

Key words
histiocytic necrotizing lymphadenitis, Kikuchi disease, lymphadenopathy, systemic symptoms

HINA LATIF

Bardet-Biedl syndrome with megaloblastic anemia

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CASE REPORT Bardet-Biedl syndrome is a rare multisystem autosomal recessive disorder having variable symptoms ranging from peripheral obesity, retinal degeneration, polydactyly, hypogonadism, and renal impairment among numerous other features. We present a case of a 16-year-old woman exhibiting characteristic features of Bardet-Biedl syndrome.

Key words
Bardet-Biedl syndrome, ciliopathic genetic disorder, clinical diagnosis, multisystem involvement

MASSIMO LEO

Spur cell anemia and cold agglutinin disease in cirrhosis: resolution after liver transplantation

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INTRODUCTION Spur cell anemia and cold agglutinin disease (CAD) are 2 rare conditions causing hemolytic anemia that may arise in cirrhotic patients. Spur cell anemia is an independent negative prognostic factor that warrants prioritization of liver transplantation (LT). Spur cells are aberrant red blood cells (RBCs) with irregular spike-like projections of the membrane morphologically similar to acanthocytes but chemically distinct since spur cells are characterized by an increased level of membrane free cholesterol. This acquired deformation reduces their deformability and makes them more prone to entrapment and destruction by the spleen. Cold agglutinin disease is characterized by autoantibodies that react with surface antigens on RBCs at cold temperatures; hypergammaglobulinemia, hallmark of cirrhosis, may represent a substrate for its development. In this clinical case, we highlight the impact of LT on the regression of these entities and on the biomechanical properties of erythrocytes.

CASE REPORT A 68-year-old man with alcohol-related liver failure (Model for End-Stage Liver Disease Score, 28; Child-Pugh-Turcotte score, C11) was referred to our liver transplantation unit to evaluate his eligibility for LT. Home therapy included antacid, diuretics, and laxatives. On physical examination, cutaneous jaundice was noted. No macroscopic loss of blood was observed. Pretransplant blood tests revealed severe macrocytic hyperchromic anemia refractory to transfusions associated with marked reticulocytosis, mixed hyperbilirubinemia, reduced levels of haptoglobin, and increased levels of lactate dehydrogenase. Hypolipoproteinemia and low levels of apolipoprotein A1 were also observed. Serum protein electrophoresis showed hypergammaglobulinemia. Endoscopic tests were negative for bleeding. Computed tomography revealed splenomegaly and mild ascites. Hemolytic anemia was suspected. Peripheral blood smear showed a heterogeneous population made up mainly of acanthocytes. Agglutinated RBCs were also present and since they had low titer and low thermal amplitude, nonpharmacologic management was adopted. The patient, after completing the full diagnostic protocol, was considered eligible for LT: a month later, he underwent a transplant with no intraoperative complications.

Laboratory testing 3 months after the LT showed progressive normalization of the complete blood counts, serum chemistry values, and serum protein electrophoresis, attesting restored liver function and complete absence of hemolysis. In parallel, peripheral blood smear showed predominance of discocytes, with just a small amount of acanthocytes without agglutinated RBCs. Moreover, before LT, the biomechanical properties of RBCs assessed with an atomic force microscopy included an abrupt stiffening when progressive stress was applied. On the contrary, post LT, RBCs showed a more physiological response, confirming a significant recovery of their functionality.

DISCUSSION Peripheral blood smear is an easily accessible, inexpensive and highly informative exam that represents a diagnostic keystone when hemolysis is suspected. Spur cell anemia and CAD are 2 peculiar conditions associated with advanced liver failure that have to be considered when laboratory findings indicate hemolysis, hypocholesterolemia, and hypergammaglobulinemia. Spur cell anemia has a negative prognostic value and is an indication for clinicians to prioritize these patients for LT, which, thanks to the restoration of optimal hepatic function including physiological metabolism of cholesterol and immunological status, represents the only etiological treatment for both conditions.

Keywords
biomechanics, cirrhosis, cold agglutinin disease, liver transplantation, spur cell anemia

CELESTINE LIM

Telbivudine-induced myopathy: an interesting presentation of swallowing impairment and proximal weakness in an elderly patient

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INTRODUCTION Dysphagia is common in older adults and it becomes increasingly so as people age. This is due to normal aging and aging-related multimorbidity, and increases the risk of aspiration pneumonia. Telbivudine is a potent antiviral against hepatitis B virus, which has not been widely studied in the geriatric population. While creatine kinase elevations were found to be common with telbivudine therapy, myopathy was rare. Patients who do develop myopathy usually do so within 2 years of treatment and present with muscle aches and limb weakness. We report a case of telbivudine-induced myopathy in an elderly man who presented atypically with predominant dysphagia and aspiration pneumonia.

CASE REPORT A 90-year-old Chinese man with chronic hepatitis B infection received telbivudine 600 mg daily for 4 years. He presented to hospital with pneumonia and was subsequently found to have severe pharyngeal dysphagia. Physical examination revealed generalized fasiculations, bulbar speech, and proximal weakness. Muscle enzyme levels were normal; however, nerve conduction studies and electromyography revealed irritable myopathy. Telbivudine and atorvastatin—the only 2 possibly myotoxic drugs that the patient was on—were stopped. Subsequent muscle biopsy revealed findings suggestive of mitochondrial toxicity. Telbivudine-induced myopathy was suspected and the offending drug was switched to entecavir with subsequent improvement of the patient's muscle weakness.

CONCLUSIONS To promptly detect this reversible adverse event, it is important to monitor side effects of telbivudine, even after long-term use. This is especially so in the elderly in whom presentation may be atypical. Further studies should be carried out to investigate the mechanism and risk factors of telbivudine-induced myopathy in the geriatric population.

Key words
adverse effects, hepatitis B, myopathy, telbivudine

HELENA LUÍS

A typical case of multiple myeloma revealed by cardiac amyloidosis

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INTRODUCTION Amyloidosis is characterized by clinically significant extracellular amyloid infiltration in different tissues and organs. Cardiac involvement represents the most important prognostic factor especially in amyloid light-chain. This type of amyloidosis occurs in about 15% of patients with multiple myeloma.

CASE REPORT We present a case of a 56-year-old man without a specific medical history who presented with constitutional symptoms and anemia, and who was ultimately diagnosed with light chain myeloma with cardiac amyloidosis.

CONCLUSIONS Primary amyloidosis associated with light chain myeloma is a rare, and often late, diagnosis with a dismal prognosis. The present case report illustrates the diagnostic challenge.

Key words
cardiac amyloidosis, light chain myeloma

GIOVANNA MACELLO

Congenital anomalies of the inferior vena cava: a factor to consider in the algorithm of deep vein thrombosis with therapeutic implications

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CASE REPORT Inferior vena cava (IVC) anomalies are rare defects resulting from aberrant development during embryogenesis and constitute a risk factor for thromboembolic disease. They are usually observed in asymptomatic patients as an incidental finding or together with other cardiac and / or abdominal anomalies.

With advances in diagnostic imaging, IVC anomalies have emerged as a new factor to be considered in the etiologic diagnostic algorithm of deep vein thrombosis (DVT) and as a thrombotic risk factor.

Inferior vena cava anomaly should be suspected in young patients with lower limb DVT when one or both iliac veins are involved and in patients with recurrent thrombosis. Whenever a dilated azygos or Hemi-azygos vein is found on chest computed tomography, additional images should be obtained to identify IVC abnormalities. The coexistence of other risk factors for thrombosis should always be considered.

We present a patient with intrahepatic IVC hypoplasia who consulted for DVT of the left external iliac vein and in whom we could not demonstrate the presence of other prothrombotic risk factors. We emphasize the importance of the role of computed tomography angiography in the diagnosis and the therapeutic decision of permanent anticoagulation.

Key words
anticoagulation, deep vein thrombosis, inferior vena cava anomalies

TANUKA MANDAL

Subcutaneous edema: an unusual presentation

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INTRODUCTION Polymyositis is a type of inflammatory myopathy characterized by proximal muscle weakness due to mononuclear infiltration of the endomysial layer of the skeletal muscle. It may also weaken the axial muscles. The hip extensors are most severely affected, leading to difficulty in climbing stairs and rising from a seated position. It can be associated with low-grade fever, enlarged lymph nodes or even dysphagia, and foot drop in advanced cases. Other systems involvement may occur in the form of interstitial lung disease and heart failure.

Subcutaneous edema has been reported in very few cases which was the presenting symptom in our case.

CASE REPORT A 28-year-old woman presented with a history of swelling of the right upper limb for last 1 month which gradually progressed to involve face, neck, left upper limb, and upper part of the chest within few days of onset. On examination, palpable cervical and axillary lymph nodes were found. During her stay in the hospital, the swelling gradually progressed to involve the bilateral lower limbs and feet. She complained of difficulty in getting up from a sitting position. She also developed nasal intonation, nasal regurgitation, and difficulty in swallowing of food.

Basic investigations revealed high erythrocyte sedimentation rate, C-reactive protein, and lactate dehydrogenase levels. The level of creatine kinase was raised and electromyography revealed a

myopathic pattern. The nerve conduction velocity and electrolyte report was normal. The anti-nuclear antibody profile was negative. The extractable nuclear antigen profile with U1RNP was negative. The myositis profile was negative. Serum protein electrophoresis with immunofixation and urinary protein electrophoresis was within normal limits.

The carcinoembryonic antigen and cancer antigens 125 and 19.9 were normal. The histopathological examination of the axillary lymph nodes revealed reactive hyperplasia.

Magnetic resonance imaging of the proximal muscles revealed multiple areas of edema at different muscles of the upper thigh with significant soft tissue edema.

On muscle biopsy, muscle fibers showed infiltration of endomysial cells by mononuclear lymphocytes. Myositis with focal myonecrosis with invasion of lymphocytes and macrophages was noted. Some muscle fibers showed loss of striations and signs of active phagocytosis.

The patient was finally diagnosed with polymyositis mimicking superior vena cava syndrome initially with generalized subcutaneous edema and lymphadenopathy as the unusual and rare presenting feature.

The patient was treated with pulse methyl prednisolone followed by oral steroids. Her clinical condition improved dramatically and subcutaneous edema subsided. Nasal intonation and dysphagia improved significantly but still persisted.

CONCLUSIONS Although very few studies stated subcutaneous edema to be the presenting symptom in polymyositis, this case has clearly established that it cannot be disregarded as a cause.

Key words
lymphadenopathy, polymyositis, subcutaneous edema, superior vena cava syndrome

MEHREEN MEHMOOD

Rosai-Dorfman syndrome: a case report

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INTRODUCTION Rosai-Dorfman syndrome (RDS) is a rare entity that has been reported across the world over years. Recently, it was included as a different category in the R group of histiocytosis classification. It has expressed itself in variable forms giving rise to variety of symptoms and widespread extranodal involvement. This case reports widespread collusion of lymph nodes and extranodal tissues by disease over some time with an underlying autoimmune disease process.

CASE REPORT A 40-year-old woman with known history of ulcerative colitis for last 4 years presented with a gradual protrusion of the right eye ball, axillary and breast lumps located bilaterally, and shortness of breath. Examination revealed proptosis of the right eye with large sized lumps in bilateral axilla and breasts. We performed routine baselines, thyroid function tests, fine needle aspiration of the axillary lump, and magnetic resonance imaging of the head. Histopathology of the tissue supported the diagnosis of RDS. The patient was started on steroids with partial relief of symptoms but later developed an enlarged subglottic mass that was compromising her airway. Tracheostomy was done. She partially healed later with treatment and tracheostomy was reversed.

CONCLUSIONS Rosai-Dorfman syndrome is a chronic ailment that impairs the normal lifestyle of a patient with remission and recurrences. Bilateral lymphadenopathy with unilateral proptosis of the eye with subjacent autoimmune disease should raise suspicion of RDS.

MARET MOISA

Pheochromocytoma complicated with multiple organ failure and posterior reversible encephalopathy syndrome

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INTRODUCTION Clinical presentation of pheochromocytoma can be highly variable and it may rarely present acutely with multiple organ failure. Acute hypertension may lead to posterior reversible encephalopathy syndrome. Therefore, a high index of suspicion resulting in timely and accurate diagnosis and treatment of pheochromocytoma is paramount to avoid possible severe complications.

CASE REPORT A 47-year-old previously healthy Caucasian woman was admitted to the intensive care unit with high blood pressure, multiple organ failure, and a decreased level of consciousness. Contrast-enhanced computed tomography showed an enlarged right adrenal gland with a heterogeneous structure. Due to high clinical suspicion of pheochromocytoma, preoperative treatment with an alpha-adrenergic receptor blocker, doxazosin, was started. The diagnosis was later confirmed by highly elevated free metanephrine and normetanephrine levels in plasma (34 100 ng/l and 8050 ng/l respectively, reference range $<$90 ng/l and $<$180 ng/l) and elective adrenalectomy was planned. The patient had acute heart, kidney, and liver failure, but with treatment her condition improved. A couple of days later her level of consciousness once again started to decrease, delirium developed and visual impairment was noticed. Diagnosis of posterior reversible encephalopathy syndrome (PRES) was confirmed by magnetic resonance imaging of the brain showing bilateral areas of edema in cortical and subcortical white matter of the occipital lobes consistent with PRES. Adequate management of elevated blood pressure remained challenging and a few days before scheduled adrenalectomy the patient had a generalized epileptic seizure resulting in clinical death. Cardiopulmonary resuscitation was performed and return of spontaneous circulation was achieved. Computed tomography scan of the brain revealed hemorrhage in the previously described occipital edematous areas and electroencephalography indicated persistent epileptic activity likely caused by diffuse hypoxic damage of the brain. The patient did not regain consciousness and following repeated electroencephalography studies were continuously indicating diffuse post-anoxic damage of the brain. Considering the patient's condition, the results of previous investigations, and poor prognosis, it was decided not to escalate the extent of treatment any further and she eventually died.

DISCUSSION This case report demonstrates a severe and complicated clinical presentation and course of pheochromocytoma, its association with posterior reversible encephalopathy syndrome, and the importance of timely and accurate diagnosis and management. In our case, the suspicion of pheochromocytoma arose quickly but confirming the diagnosis and scheduling adrenalectomy took several weeks. According to the literature, emergency adrenalectomy may be an option in extreme conditions. In this case, the patient's condition was initially stabilized but was later complicated by PRES and its complications resulting in poor outcome. Considering that in the management of PRES, treatment and, if possible, elimination of triggering condition is of major importance, it is possible that the patient may have benefited from better-timed adrenalectomy.

Key words
adrenalectomy, multiple organ failure, pheochromocytoma, posterior reversible encephalopathy syndrome

MARCO MORETTI

Cirrhosis and partial portal thrombosis leading to severe variceal bleeding, an unusual presentation of sarcoidosis

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INTRODUCTION Sarcoidosis is a systemic granulomatous disease, which is characterized by the formation of non-necrotizing granulomas. Although 50% to 80% of affected patients have granulomas on liver biopsy, related symptoms occur in only 5% to 20% of those patients. Sarcoidosis rarely leads to portal hypertension and cirrhosis (6%–8%). We present a case report to increase the knowledge on portal hypertension in hepatic sarcoidosis.

CASE REPORT A 62-year-old woman with medical history of type 2 diabetes mellitus was diagnosed with variceal bleeding for which elastic banding was performed. The patient was admitted to the intensive care unit as the bleeding persisted. She became hemodynamically unstable due to an ongoing hemorrhagic shock which required vasopressors and eventually intubation. Liver ultrasound detected splenomegaly and nodular hepatomegaly. Doppler ultrasound found partial portal thrombosis. Chest computed tomography (CT) showed diffuse hilar adenopathies and interstitial micronodular lesion. Finally, positron emission tomography with CT detected metabolic active liver, bone marrow, and upper and lower diaphragmatic adenopathies.

COURSE OF EVENTS A hematologist advised adenopathy and bone marrow biopsy. The latter ruled out lymphoma and revealed profound erythropoiesis reactive to anemia. A systemic disease specialist requested liver biopsy and blood tests for autoimmune and microbiological causes.

However, the condition of the patient deteriorated due to a residual post-banding bleeding for which a hemostatic cardia stent was placed.

CLINICAL RESOLUTION The diagnosis of sarcoidosis was supported by biopsies of the liver and lymph node, which yielded noncaseating granulomas infiltration. Chest CT and PET-CT were also consistent with this diagnosis. The complementary analysis excluded tuberculosis, invasive mycosis, primary biliary cholangitis, and primary sclerosing cholangitis. The patient was treated with high-dose methylprednisolone with notable clinical improvements and discharge from the intensive care unit. Nevertheless, cirrhosis was confirmed 6 weeks later by liver fibroscan.

CONCLUSIONS Hepatic sarcoidosis can present as life-threatening bleeding due to variceal bleeding caused by portal hypertension. It can also evolve in cirrhosis. Differentials diagnosis is broad when hepatic sarcoidosis is suspected. Therefore, multidisciplinary discussion is warranted. Anatomopathological examination of 2 potentially involved organs should be considered to make the appropriate diagnosis to promptly begin appropriate treatment. Further studies are requested to shed more light on the pathophysiological mechanism of portal hypertension.

Key words
hepatic sarcoidosis, portal hypertension, portal thrombosis, systemic sarcoidosis, variceal bleeding

KOUSHIK MUKHERJEE

Isolated right lateral rectus palsy: a rare and initial presentation in systemic lupus erythematosus

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INTRODUCTION Systemic lupus erythematosus (SLE) is an autoimmune disease primarily affecting women of childbearing age with complex pathogenesis and multiple organ involvement which mainly presents with arthritis and malar rash. The initial presentation as an isolated cranial nerve palsy is a rare and unusual manifestation of SLE which accounts for only 0.5% to 1% among other neuropsychiatric manifestations.

CASE REPORT A 16-year-old adolescent girl presented to the emergency department in a hemodynamically unstable condition and bleeding from mouth for the last 3 days. Her family gave history of progressive malar rash for last 1 week and difficulty in looking to the right side along with double vision for last 2 weeks for which she had visited multiple doctors. She had no history of fever, headache, nausea, vomiting, hearing loss, facial weakness, and decreased facial sensation. All cranial nerve functions were unremarkable apart from difficulty in lateral gaze of the right side which indicated right sixth cranial nerve palsy. On auscultation, she had coarse crackles over both lung fields. Her blood tests revealed the ANA-HEP2 titer of 1:240 and were positive for anti-dsDNA which led to the diagnosis of SLE.

The girl was treated with pulse therapy of intravenous methylprednisolone (1 mg/kg body weight) for 3 days and oral prednisolone (40 mg/d) for the next 4 weeks and then it was tapered to maintenance therapy along with hydroxychloroquine and immunosuppression.

DISCUSSION Isolated right lateral rectus palsy in a patient with SLE is an extremely rare initial presentation based on the literature review. Vasculitis is often suspected as a pathogenic mechanism of cranial neuropathies. Systemic lupus erythematosus is a disease that may eventually affect numerous organ systems. Although most patients demonstrate the classic diagnostic signs of skin rash and arthritis, the case reported here of an isolated sixth cranial nerve palsy suggests that such uncharacteristic presentation of SLE is possible.

Key words
anti-dsDNA, cranial nerve palsy, methylprednisolone, systemic lupus erythematosus

SYED OSAMA HUSAIN

Pancytopenia: a rare presentation of syphilis

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CASE REPORT Syphilis is a common public health concern which is caused by spirochete Treponema pallidum. It is one of the 4 curable sexually transmitted infections caused by bacteria. A common mode of transmission is via sexual contact and mainly targeting high-risk populations such as those with HIV and men who have sex with men. It has a wide range of presentations based on the 4 main overlapping clinical stages. Due to lack of vaccines to prevent syphilis, prompt diagnosis and management of infected persons are warranted to reduce disease burden and its clinical effects. Of note, not every pancytopenia in the elderly is due to malignancy, bone marrow aplasia, or acute viral/parasitic insult; other common causes coexist. Here, we present an unorthodox case of pancytopenia in a 55-year-old male patient who developed chronic progressive weakness, generalized body aches, and intermittent fever.

Key words
pancytopenia, sexually transmitted infections, syphilis

PRANABANANDA PAL

A case of rectal malignancy presenting with paraneoplastic myopathy and hyperpigmentation of skin

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INTRODUCTION Paraneoplastic syndromes are frequently associated with colorectal carcinoma, rarely even in absence of classic symptoms of cancer itself. Dermatomyositis is an inflammatory myopathy usually presenting with proximal muscle weakness and pathognomonic dermatological manifestations. Dermatomyositis is known to be associated with different types of malignancy, including colorectal carcinoma. However, rarely, the paraneoplastic manifestations precede the gastrointestinal symptoms and signs of carcinoma. **CASE REPORT** We present a case of a 60-year-old woman who presented generalized hyperpigmentation of skin for 3 months and proximal muscle weakness for 1 month. On clinical examination, she had pallor and significant left supraclavicular lymphadenopathy but classic skin manifestations of dermatomyositis were absent. On investigation, she had raised erythrocyte sedimentation rate and lactate dehydrogenase. Electromyography and magnetic resonance imaging of the thighs showed evidence of proximal myopathy which was subsequently confirmed on muscle biopsy. Contrast enhanced computed tomography of the abdomen to rule out primary malignancy revealed diffuse circumferential thickening of the distal and mid-rectum which was confirmed as adenocarcinoma of the rectum in histopathology. Dermatomyositis developed as a paraneoplastic syndrome of rectal carcinoma in absence of gastrointestinal symptoms. **CONCLUSIONS** The diagnosis of dermatomyositis in the elderly population should raise suspicion of an underlying malignancy. Early diagnosis of cancer by paraneoplastic manifestations will improve the outcome of the malignancy. Physicians should be aware of paraneoplastic syndromes which may be the first presentation of their etiological malignancies.

Key words
dermatomyositis, colorectal carcinoma, paraneoplastic

ANKAN PATHAK

Fever: an internist's nightmare

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CASE REPORT A 64-year-old man, a resident of Kolkata, India with a history of type 2 diabetes mellitus, hypertension, primary hypothyroidism, and chronic kidney disease VD was admitted with fever of 5 months' duration. Fever was marked by temporary relief from time to time by the use of empirical oral antibiotics followed by a subsequent relapse of fever. On physical examination, the patient was alert and conscious with normal vitals as well as no meningeal sign, no icterus, mild pallor, no rash with no organomegaly, and no palpable lymph nodes. The dialysis site was clean. There were no obvious potential diagnostic clues. Ear, nose, and throat assessment as well as dental examination were unremarkable. Ophthalmology revealed bilateral mild nonproliferative diabetic retinopathy.

On admission at our tertiary teaching hospital, pan culture with procalcitonin and routine blood tests were performed. The patient was initially started on piperacillin and Tazobactum which led to some improvement. However, all cultures were negative and procalcitonin was repeatedly elevated. Abdominal and chest imaging was inconclusive, and so was transthoracic echocardiogram. The patient continued to be febrile after temporary improvement. At this point, positron emission tomography (PET) with computed tomography (CT) was performed. Bone marrow examination and culture were unremarkable. PET CT revealed lymphoproliferative disorder with marrow infiltration, hypermetabolic bilateral cervical, rt supraclavicular, mediastinal, periportal, and left inguinal pelvic lymphadenopathy. An abscess in the right obturator internus was the occult source of infection. Unfortunately, the patient was lost to follow-up after PET CT.

CONCLUSIONS

1 A sufficient number of diagnoses should be proposed for a patient with pyrexia of unknown origin.

2 Clinical suspicion is always of paramount importance even if an early basic investigation may negate a possibility. For example, as in our case, contrast-enhanced CT of the thorax and the abdomen was normal and did not show any lymph nodes; however, PET CT showed multiple lymph nodes.

3 Fever in special populations, particularly those with chronic kidney disease VD and other immunocompromised patients (HIV, primary immunodeficiency disorders, and cancer chemotherapy) should not be treated empirically.

4 Since these cases often lead to frustrating results, the family should always be kept in confidence regarding the adverse outcomes.

Key words
fever

WAFA QAISAR

Chest pain in a patient with aortic aneurysm: a diagnosis missed!

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CASE REPORT Central chest pain in a patient presenting to the emergency department may indicate simpler disease processes, such as acid reflux or muscular spasm, or more serious conditions, such as myocardial infarction or rarely, aortic dissection that is a vascular emergency with potentially fatal outcomes if not treated in time and appropriately with a mortality of up to 50% in the first 48 hours. To make things more challenging, the common diagnostic tool, electrocardiography, used to diagnose heart conditions, shows an overlap of findings between myocardial infarction and dissecting aortic aneurysm while the management of the 2 conditions is poles apart. This leads to both a diagnostic dilemma as well as a treatment predicament and the window to take action remains narrow. We report a case of acute aortic dissection in which a patient with chest pain had ST-segment elevations on electrocardiography and no clinical findings to suggest a dissecting aortic aneurysm were present. Consequently, a misdiagnosis of acute myocardial infarction was made. The patient was given thrombolytic therapy that led to catastrophic results.

Key words
aortic aneurysm, dissection, electrocardiography, myocardial infarction

ANIL RENGAN

Plasma cell dyscrasia 8 years later: evolution of 3 forms of presentation in the same patient

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INTRODUCTION Plasma cell dyscrasias are rare diseases characterized by clonal proliferation of plasma cells and include monoclonal

gammopathy of uncertain significance, smoldering myeloma (SM), multiple myeloma (MM), solitary plasmacytoma, Waldenström macroglobulinemia, and plasma cell leukemia (PCL). MM and PCL have the highest and lowest frequency of reported cases (10% vs 2%).

CASE REPORT A 67-year-old man diagnosed with SM in September 2012 in the context of anemia, presence of monoclonal component (MC) IgG kappa (3 g/dl) and 10% of plasma cells in bone marrow biopsy (BMB) without organ damage White. He kept watchful waiting until September 2016 when he progressed to MM due to an increase in MC (5 g/dl), with 60% of plasma cells in BMB, as well as kidney (creatinine, 2 mg/dl) and bone involvement (lytic lesions in the femur and pelvis). He underwent 4 treatment cycles with bortezomib 1.3 mg/m^2 weekly, thalidomide 100 mg/d and dexamethasone 20 mg/wk with good response, subsequently receiving treatment with melphalan 200 mg/m^2, bone marrow transplantation in 2017 and maintenance with thalidomide 100 mg/d until 2018. In June 2019, he presented relapse with an increase in MC (2.85 g/dl), proteinuria (2.5 g in 24 hours), lytic lesions on the right scapula, and 40% infiltration of plasma cells in BMB. He received 6 cycles of treatment interrupted with carfilzomib 27 mg/m^2 twice weekly, lenalidomide 25 mg/d, and dexamethasone 40 mg/wk for multiple infections and cardiological toxicity to carfilzomib in the last cycle. In September 2020, he presented general deterioration, an increase in MC (5.5 g/dl), kidney failure, and bicytopenia (hemoglobin, 10 g/dl and platelets, 34 000/mm^3). The blood smear showed 60% of plasma cells and the BMB reported 80% of clonal plasma cells. With a diagnosis of PCL, he underwent treatment with VDT-PACE (days 1 to 4: cisplatin, 10 mg/m^2; doxorubicin, 10 mg/m^2; cyclophosphamide, 400 mg/m^2; etoposide; 40 mg/m^2; and dexamethasone, 40 mg; days 1, 4, 8, and 11: bortezomib, 1 mg/m^2; and thalidomide 200 mg per day) presenting poor evolution. He died 60 days later.

DISCUSSION The present case made it possible to evaluate the 8-year evolution of 3 forms of presentation associated with the group of plasma cell dyscrasias (SM, MM, PCL) in the context of relapse and subsequent refractoriness to first-line treatments including bone marrow transplantation. The last stage (PCL) is an extremely rare variant, the diagnosis of which is established based on the presence of >20% or >2000 plasma cells/mm^3 in the peripheral blood smear and its short-term prognosis is unfavorable.

CONCLUSIONS Although plasma cell dyscrasias are rare, their comprehensive knowledge is important for an early diagnosis and reasonable therapeutic decisions.

Key words
case report, dyscrasia, leukemia, myeloma, plasma cells

ELIANA RENSNER

Neurosyphilis: a case of ocular involvement

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INTRODUCTION Known as the great imitator due to its various manifestations, syphilis is a multisystemic chronic infection by Treponema pallidum. Neurosyphilis, the infection of the central nervous system (CNS) can happen any time after the initial onset of the disease. While any CNS structure can be affected, ocular involvement is considered a rare manifestation. We herein report a case of a patient with ocular syphilis.

CASE REPORT A 37-year-old woman presented to the emergency department with retroocular headache, fever, blurry vision, and scotomas. She had no history of headaches, migraines or recent head trauma. Physical examination showed bilateral hyperemia, sluggish pupillary reaction, and painful but unrestricted eye movement, diminished visual acuity, impaired color vision, and a superior arcuate defect in both eyes. Additionally, both wrists and the acromioclavicular joint appeared tender and stiff upon examination.

Ophthalmoscopy revealed bilateral papilledema with no signs of increased intracranial pressure. A routine laboratory exam showed elevated levels of erythrocyte sedimentation rate (96 m/h) and C-reactive protein (3.9 mg/dl). Imaging showed no abnormalities except for ultrasound imaging of the affected joints which demonstrated bilateral synovitis of the wrists, first metacarpal and acromioclavicular joints.

Lumbar puncture was performed with normal findings and negative cultures. Serologic workup revealed a highly positive venereal disease research laboratory test (titer 1/512) and a positive treponemal enzyme and chemiluminescence immunoassays. Nonetheless, the cerebrospinal fluid–venereal disease research laboratory test was negative.

Based on our clinical and serologic findings, a diagnosis of ocular syphilis was established. The patient was started on penicillin G 24 million units daily for 3 weeks and corticosteroid pulses after 2 weeks of treatment due to an initial exacerbation of ocular symptoms while on penicillin alone. The patient described an improvement in visual acuity, corroborated by an ophthalmologic evaluation. Headaches subsided as well and patient was able to complete treatment in an outpatient setting.

DISCUSSION Neurosyphilis develops with the infection of the CNS by the spirochete Treponema pallidum and any part of the CNS can be affected by the disease, including the eyes. While posterior uveitis and panuveitis are the most common and should be highly suggestive of syphilis, any eye structure can be affected, like in this case, the optic nerve.

Due to its ability to mimic different ocular disorders, there can be delays in reaching a proper diagnosis and subsequent treatment which can lead to long-term complications such as loss of visual acuity, glaucoma, or even blindness.

Therefore, it seems imperative that internists should be on the lookout for this form of the disease, and should consider it as a differential diagnosis whenever ocular structures are affected with no other clear etiology.

Key words
bilateral papillitis, neurosyphilis, ocular syphilis, optic neuritis, syphilis

SHAMBO SAMRAT SAMAJDAR

Human cytomegalovirus disease during treatment with tocilizumab for severe COVID-19: a case report

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CASE REPORT An adult patient with underlying type 2 diabetes mellitus and severe COVID-19 disease presented to our hospital with severe acute respiratory distress syndrome. He was treated with corticosteroids, tocilizumab, broad spectrum antibiotics, hydroxychloroquine, and mechanical ventilation. He deteriorated and went into multiple organ dysfunction syndrome. Eventually, a diagnosis of severe human cytomegalovirus disease was established. The patient was started on ganciclovir, but he succumbed to his illness. This case highlights that CMV reactivation, which is an opportunistic infection, remains a possible complication even with short-term use of tocilizumab in severe COVID-19 disease.

Key words
COVID-19, cytomegalovirus, interleukin 6, tocilizumab

SUBHRA SANKAR SEN

Neuroborreliosis: the great mimicker

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INTRODUCTION Neuroborreliosis is a manifestation of Lyme disease, which necessitates treatment with antibiotics. As the incidence of neuroborreliosis is quite infrequent in India and is known to mimic numerous neurological conditions, it is an important differential to bear in mind and should be investigated when necessary.

CASE REPORT A 17-year-old man presented to the inpatient department of medicine with progressive ascending weakness of both lower limbs for the last 3 days and pain in his lower back. He also complained of drooling and inability to completely close his right eye. On examination, he was areflexic at the knees, ankles, and plantars were non responsive. Over the next 2 days, the weakness started to ascend and affected both of his upper limbs. Examination revealed hyporeflexia in his upper limb jerks as well, and a right sided lower motor neuron (LMN) facial nerve palsy. He also became completely bedbound during the same period. A clinical diagnosis of Guillain-Barré syndrome with isolated right-sided LMN facial nerve palsy was established and he was consequently started on intravenous immunoglobulins. Nerve conduction velocity showed acute axonal motor polyneuropathy in addition to S1 radiculopathy, which presumably confirmed our diagnosis of Guillain-Barré syndrome. Owing to poor initial response to the immunoglobulin and the fact that the patient had right-sided LMN 7th nerve palsy and S1 radiculopathy, and despite no travel history or history of a possible tick bite, serology for antibodies against Borrelia burgdorferi was performed. The IgM for antibodies against *B. burgdorferi* were strongly positive and led us to the diagnosis of Lyme disease presenting as neuroborreliosis.

The patient was started on intravenous ceftriaxone and over a few days, his weakness improved and with inpatient neuro rehabilitation, he made quite a good recovery and was able to mobilize with the support of a walking stick after being completely bedbound 4 weeks after the completion of intravenous antibiotics.

DISCUSSION Neuroborreliosis is the most common complication of Lyme disease. Most commonly, it can present with acute meningoencephalomyelitis, or acute cranial neuropathy and radiculopathy, and rarely can manifest as a Guillain-Barré–like presentation. Lyme disease should be considered in anyone who presents with symptoms and signs, as above, suggestive of the early disseminated phase of the infection, and these patients should be questioned about possible tick bites in the preceding few months. Blood and cerebrospinal fluid samples from these patients should be sent for B. burgdorferi–specific antibodies and testing by polymerase chain reaction. Treatment of early localized Lyme disease can be carried out in the community with a course of oral doxycycline. However, severe early disseminated or late Lyme disease warrants intravenous antibiotics, such as ceftriaxone, for 2 to 3 weeks.

Key words
ceftriaxone, Guillain-Barré syndrome, neuroborreliosis

MARTA SKOCZYŃSKA

Cryoglobulinemic vasculitis as the first manifestation of type C hepatitis

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INTRODUCTION Extrahepatic lesions in the course of hepatitis C virus (HCV) are observed in 38% to 40% of patients with hepatitis C and, sporadically, may be the first clinical manifestation of previously undiagnosed HCV infection.

CASE REPORT A 31-year-old man was admitted to Wroclaw University Hospital, Poland, on May 10, 2017. On admission he reported dyspnea, cough, high fever, high blood pressure, and general weakness. Over the course of hospitalization, he developed fine skin lesions, myalgia, arthralgia, and abdominal pain, and was transferred to the rheumatology department due to a suspicion of systemic vasculitis.

COURSE OF EVENTS Physical examination revealed fine skin rash, high blood pressure, and lower limbs edema. Results of blood tests showed normocytic anemia, renal function impairment, features of nephrotic syndrome, lowered levels of C3 and C4, elevated levels of aspartate transaminase, and presence of anti-HCV. Fecal occult blood test was positive. Abdominal ultrasound revealed peritoneal and bilateral pleural effusion. Based on clinical signs and laboratory test results, the consulting nephrologist diagnosed rapidly progressing glomerulonephritis. The patient tested positive for HCV (3.75×107 copies/ml, genotype 1B) and cryoglobulins (70% after 24 hours). The diagnosis of cryoglobulinemic vasculitis with gastrointestinal and renal involvement (secondary to chronic HCV infection) was made. The patient was treated with pulses of methylprednisolone, followed by oral steroids and plasmapheresis.

CLINICAL RESOLUTION After 16 weeks of therapy with elbasvir/grazoprevir, the patient reported resolution of all symptoms of the diagnosed condition. HCV-RNA test performed at that time was negative. Cryoglobulins were still present (5%).

DISCUSSION HCV stimulates B cells to produce cryoglobulins, which are detected in serum in approximately 50% of patients infected with HCV, but most patients are asympotomatic. Clinical cryoglobulinemia occurs in 25% of patients with hepatitis C and concomitant cryoglobulinemia, while cryoglobulinemic vasculitis develops in approximately 5% to 10% of those cases. It is a potentially life-threatening condition as it may lead to multi-organ failure. Aside from antiviral drugs, treatment of cryoglobulinemic vasculitis secondary to HCV includes symptomatic treatment: nonsteroidal anti-inflammatory drugs in cases without organ function impairment, oral steroids in patients with renal function deterioration, and pulses of steroids and plasmapheresis in patients with progressive organ damage.

CONCLUSIONS Vasculitis in the course of mixed cryoglobulinemia secondary to HCV infection may be responsible for a wide spectrum of extrahepatic symptoms. These unusual symptoms, autoimmune in nature, may be the first and potentially life-threatening manifestation of hepatitis. In such cases, timely diagnosis as well as treatment of both secondary vasculitis and underlying infection are essential.

Key words
cryoglobulinemia, hepatitis C, vasculitis

ANTONIO SOUDO VENTURA

Hepatic failure due to AA amyloidosis

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CASE REPORT We describe a case of a 75-year-old man presenting with abdominal pain and constitutional syndrome for the preceding month. The physical examination revealed slight jaundice. The initial laboratory tests revealed mild cholestasis and abdominal ultrasound / computed tomography only showed cholelithiasis. Serologic markers of hepatitis viruses and autoimmune serology profile were negative. A drug-induced liver injury was suspected at first. Afterwards, the patient started with clinical signs of heart failure and deterioration of liver function. Ultrasound-guided percutaneous liver biopsy was performed showing amyloid deposition in sinusoidal spaces with positive Congo red stain. The patient had a fatal clinical outcome with progressive liver and heart failure. We present a rare case of AA amyloidosis with severe intrahepatic cholestasis and review the literature regarding varied presentations of hepatic involvement in amyloidosis.

Key words
AA amyloidosis, hepatic amyloidosis, intrahepatic cholestasis, liver failure

ANNA SUSKA

Burkitt lymphoma secondary to multiple myeloma

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CASE REPORT A 65-year-old woman with a history of multiple myeloma (MM) IgG kappa, International Staging System stage III, Durie and Salmon stage III, diagnosed 3 years earlier, after tandem autologous hematopoietic stem cell transplant (auto-HSCT) with complete remission, was admitted to the hematology department with signs of thrombocytopenic purpura accompanied by significant malaise.

COURSE OF EVENTS The primary diagnosis of multiple myeloma was based on bone marrow trephine biopsy which revealed 85% to 90 % diffuse infiltration of atypical plasma cells, confirmed by CD138+ immunohistochemical staining. Cytogenetic abnormalities included *TP53* gene deletion in 7% of studied cells. Electrophoresis revealed increased levels of α1-globulin, α2-globulin, and γ-globulin. Immunofixation confirmed the presence of clonal IgG kappa protein. The patient was treated with combination chemotherapy of 4 cycles of CTD (Morgan protocol) without response, and later with 4 cycles of VTD with partial remission. Then, she underwent auto-HSCT with complete hematological remission. After 23 months of remission, the patient presented with thrombocytopenic purpura.

CLINICAL RESOLUTION Additional laboratory tests showed pancytopenia with agranulocytosis and profound anemia, high ferritin, and lactate dehydrogenase. Contrast-enhanced abdominal computed tomography revealed no irregular mass or lymph node enlargement. Trephine biopsy of bone marrow revealed 95% pattern of infiltration with Burkitt lymphoma cells ("starry sky pattern"), which refers to stage IV in the Ann Arbor System. Immunophenotyping of the neoplastic cells showed high level of c-MYC and CD38+ expression. The R-CODOX-M/R-IVAC chemotherapy was initiated immediately, with complete remission for one year. Due to a persistent pancytopenia, she was transferred to the hematology department from the local hospital where she was admitted because of pneumonia (*P. aeruginosa*) and urinary tract infection (*E. faecium*). Bone marrow aspirate smears showed profound hypocellularity. Trephine biopsy presented with signs of myelodysplastic syndrome. The patient required continuation of an antibiotic and antifungal therapy, and also an implementation of oseltamivir due to influenza A(H1N1). During the hospitalization, she required numerous blood transfusions. Because of poor general health status, poor performance status (Eastern Cooperative Oncology Group status, 4), transfusion-dependence, and mobility-impairment, the patient was referred only for symptomatic treatment.

DISCUSSION Patients with MM are at higher risk of developing secondary malignancies. Variant Burkitt–type translocation (8;22) (q24;q11) in plasma cell myeloma has been also described in the literature. Nonetheless, none of patients with MM and with that translocation developed Burkitt lymphoma afterwards.

CONCLUSIONS Although the patient was treated with highly effective bortezomib-based protocol, followed by tandem auto-HSCT, it cannot be excluded that in the setting of high-risk deletion in the *TP53* gene, the new aggressive neoplastic cells with Burkitt lymphoma morphology derive from residual clonal plasma cells, even after achieving complete remission after induction. Therefore, such occurrence of MM and Burkitt lymphoma needs further investigation.

Key words
Burkitt lymphoma, multiple myeloma, *TP53* deletion

JOANNA SZAFRAN-DOBROWOLSKA

Fever in a patient with end-stage renal disease in the course of hepatitis C virus infection

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CASE REPORT There are currently 3.9 million people infected with hepatitis C virus (HCV) in the European Union and most of them are not aware of the infection. The most common clinical manifestation of HCV infection is chronic viral hepatitis, which, if left untreated, can lead to cirrhosis in some patients, as well as the development of hepatocellular carcinoma. Therefore, early diagnosis is particularly important especially in the era of modern therapy with direct-acting antiviral (DAA) drugs allowing complete recovery.

We present the case of a 55-year-old patient who was diagnosed with atypical HCV infection, end-stage renal disease (stage G5A3), and liver cirrhosis upon admission to our clinic due to an elevated creatinine level and fever. The patient did not manifest symptoms of liver cirrhosis, only fevers up to 39 °C were observed, which had a self-limiting tendency after the introduction of direct-acting antiviral treatment; as well as symptoms of end-stage renal disease in the course of nephrotic syndrome and its complications. During the differential diagnosis, no other recurrent cause of fever was found and the whole picture suggested that renal failure arose against the background of HCV infection. The extrahepatic manifestations of HCV infection affect up to 74% of patients and are associated with an approximately 20% increase in death risk among untreated patients with HCV infection. Considering the above, it is important to draw clinicians' attention to extrahepatic manifestations of HCV infection, especially in the context of the implementation of modern DAA therapy before complications.

Key words
antiviral agents, chronic hepatitis C, end-stage renal disease, fever

JUULI-ANN TÄHISTE

An elusive diagnosis of alcoholic hepatitis presenting with leukemoid reaction: a case report

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INTRODUCTION Establishing the diagnosis of alcoholic hepatitis (AH) can be challenging but is of great importance, since severe AH has a mortality up to 50% and patients could benefit from glucocorticosteroid treatment. This case report demonstrates a rare presentation of alcoholic hepatitis with leukemoid reaction and the winding road to the diagnosis.

CASE REPORT A 66-year-old man presented with weakness, anorexia, and weight loss in recent months. He had no drug history and reported consuming 200 g of vodka a few times a month. Physical findings were cachexia with muscle wasting, icteric sclerae, palmar erythema, enlarged painless stomach, hypotension 87/66 mm Hg, pulse 108 bpm. There was no mental confusion or fever.

Blood analysis revealed neutrophilic leukocytosis with the leucocyte count of 43×10^9/l, C-reactive protein, 152 mg/l, aspartate aminotransferase level of 78 U/l, alanine aminotransferase level of 37 U/l, bilirubin level of 97 μmol/l, international normalized ratio of 1.8, and albumin level of 17 g/l.

He received empiric antibiotics, but no definite infection was diagnosed on full-body computed tomography scan, echocardiography, or after repeated microbiological cultures.

A workup for liver disease showed ascites (transudate, with low PMN count), diffuse liver damage on magnetic resonance imaging, no portal thrombosis, steatosis, or cirrhosis, no focal findings (abscess or neoplasm). Tests were negative for hepatotropic viruses, autoimmune and metabolic liver diseases. The patient consistently denied heavier alcohol use; however, his daughter-in-law reported him an alcohol abuser, which resulted in diagnosis of severe alcoholic hepatitis 12 days after admission. His Maddrey score was 61, so treatment with prednisolone was started, then tapered down from a full dose after 10 days because of a suspected infection. Moreover, his Lille score was 0.59, meaning nonresponse to treatment. The patient's leukocytosis was constantly above 30×10^9/l and leukemoid reaction was diagnosed after bone marrow biopsy.

The patient remained clinically stable, and after 30 days, he was discharged with still elevated markers of cholestasis and white blood cell count of 48×10^9/l. He continued glucocorticosteroids in diminishing doses under close surveillance by his family physician. Two months later, having abstained from alcohol, his white blood cell count was 11×10^9/l and liver tests were normal.

DISCUSSION This patient fulfilled the criteria for clinical diagnosis of alcoholic hepatitis, but diagnosis was delayed by misleading information on the use of alcohol—AH occurs after heavy and long-standing drinking, which the patient denied. Secondly, although AH is an inflammatory condition often accompanied by features of systemic inflammatory response syndrome, infections are also common in these patients and need to be excluded or treated, since uncontrolled infection is a contraindication to glucocorticosteroid treatment. Lastly, a moderate neutrophilic leukocytosis is frequent in AH, but leukemoid reaction is rare and was somewhat confounding at first.

CONCLUSIONS Alcoholic hepatitis is mostly a clinical diagnosis that relies greatly on patient history—accurate anamnesis is crucial. A throughout workup with focus on excluding infection and search for other liver diseases needs to be performed. Rarely, a leukemoid reaction is seen in AH.

Key words
alcoholic hepatitis, alcohol-associated liver disease, leukemoid reaction

ANASTASIOS TENTOLOURIS

A young medical attendant presenting with non-palpable purpura and fever: discovering the unexpected

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INTRODUCTION Abdominal tuberculosis is indeed a rare entity and the sixth most frequent form of extrapulmonary tuberculosis. Granulomatous peritonitis due to tuberculosis is equally rare and difficult to diagnose primarily due to its insidious onset, its variability in symptoms and lack of easily applicable diagnostic biomarkers. It is characterized by epithelioid cell granulomas with central caseous necrosis and should be suspected in patients who present with abdominal tenderness / pain and fever with progressive malaise. Scurvy, on the other hand, is characterized by deficiency of vitamin C, and is still potentially lethal, although rare, in the developed world. It occurs most commonly in people with alcoholism, mental disorders, anorexia nervosa, Crohn disease, as well as abdominal tuberculosis. Herein, we present a very interesting case of scurvy due to abdominal tuberculosis presenting with fever, abdominal distention and rash.

CASE REPORT A 29-year-old Greek medical attendant, with unremarkable prior history, was admitted to our Clinic due to diffuse abdominal distention and pain, fever up to 38.5 °C unresponsive to paracetamol, NSAIDs and antibiotics for the last 5 days (cefuroxime po) and lower extremity rash for the last month. Her diet was restricted, excluding any fresh fruit and vegetables, which were thought to deteriorate her abdominal discomfort. The patient was febrile, her physical examination revealed diffuse abdominal tenderness and nonpalpable perifollicular purpura of both her legs. Blood chemistry showed elevated inflammation markers, normocytic anemia and lymphopenia, while no pathogen was isolated in multiple blood and urine cultures. Her vitamin C levels were measured <0.5 mg/l (max normal, 6–20) and vitamin C was immediately administered po (lack of IV solution). She underwent endoscopies with no findings and full body computed tomographies, which revealed ascites and edema of the mesentery. The tuberculin skin test was performed and measured 23 mm. The ascetic fluid culture was negative for *Mycobacterium tuberculosis*, ADA was measured 31 U/l (max normal, 40 U/l). She finally underwent exploratory laparotomy, where multiple extensive adhesions between intestinal loops and the right ovary were detected. The histopathological analysis showed epithelioid cell granulomas with central caseous necrosis. No acid-fast bacilli were detected with Ziehl-Neelsen staining of the tissue samples, PCR was also negative. However, the culture for *M. tuberculosis* was positive after 8 weeks of incubation. The patient was already treated with rifampicin / isoniazid, ethambutol, and pyrazinamide. She showed rapid clinical response and was soon discharged with follow-up in our Infectious Diseases Outpatient Clinic.

DISCUSSION Scurvy is rare compared to other nutritional deficiencies. It occurs more often in the developing world in association with malnutrition and among refugees, as well in patients with unusual nutrition habits or intestinal inflammation. Abdominal tuberculosis is indeed difficult to diagnose, as it mimics many abdominal diseases. Complete recovery from scurvy is expected after 3 months of supplementation, from tuberculous peritonitis may take though some more.

CONCLUSIONS Although scurvy and tuberculosis peritonitis are rare nowadays, both should be considered in high risk patient groups, since cases may be easily missed due to non-specific initial symptoms, and if left untreated, they are potentially lethal.

Key words
abdominal tuberculosis, granulomatous peritonitis, scurvy, vitamin C

MUHAMMAD USMAN

Benign metastasizing leiomyoma: a rare phenomenon with even rarer radiology and disease course

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INTRODUCTION Benign metastasizing leiomyoma (BML) is a rare phenomenon where uterine leiomyomas exist in extra-uterine sites. It commonly involves the lungs and follows a steady course. Our case report aims to present a rare case of BML with atypical organ involvement and radiological appearance as well as rapid progression.
CASE REPORT Our patient had a total hysterectomy due to PV bleeding showing benign leiomyomas in her late 40s. Several years later, she presented with dyspnea and her Computed tomography (CT) scan of the chest, abdomen, and pelvis showed innumerable bilateral lung nodules, pleural effusions, and hilar and mediastinal lymphadenopathy. CT-guided biopsy showed spindle-shaped cells with limited atypia and mitosis, consistent with BML. Later, positron emission tomography (PET) showed increased cellular uptake in the areas identified by CT and additional involvement of the pleura as well. Unfortunately, she deteriorated rapidly and passed away.
DISCUSSION Our literature review showed that the lungs are the primary site for BML metastasis with occasional lymph node involvement. Our case is unique as the patient had simultaneous involvement of the lungs, lymph nodes, and pleura, which have not been reported in the literature before. CT is the imaging modality of choice but PET can provide additional information. Increased uptake of fluorodeoxyglucose during PET could mean a rapidly progressing disease. The expected prognosis is 94 months from the diagnosis. Our patient deteriorated rapidly and passed away within 4 months of the initial presentation despite receiving anastrozole.
CONCLUSIONS Benign metastasizing leiomyoma is a rare condition but should be considered in women with a prior history of uterine leiomyomas. PET is an additional imaging modality that can provide key information about disease course. Although most patients have a stable disease course, rapid deterioration can occur.
LESSONS TO BE LEARNED Benign metastasizing leiomyoma should be considered as a differential diagnosis in women with a history of uterine leiomyoma presenting with numerous nodules in the lungs or other sites. It tends to involve the lungs, but it is not uncommon for it to involve atypical sites, such as the pleura in our case. Although it follows a steady course, rapid deterioration can occur, thus, patient / family counselling and treatment planning should be carried out accordingly. Finally, PET can be used as an additional imaging modality that can give more clues as to the aggressive nature of the disease.

YANINA VALDEZ

Behçet disease with neurological involvement associated to HLA-B35 and HLA-B52

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INTRODUCTION Behçet syndrome is a multisystemic disease that affects several organs. Oral ulcerations are typically the first sign of the disease. Worldwide, the highest numbers of cases were registered in the areas surrounding the old silk road which span form the Mediterranean area to Japan. In American countries, the incidence of the disease is lower.
CASE REPORT A 20-year-old woman form Formosa city without European ascendants and with Creole parents presented with a fever of more than 38 ºC, paraesthesia in the left side of the body, and muscle and joint pain that subsided with nonsteroidal anti-inflammatory drugs. The patient had functional dyspnea and nonspecific thoracic pain on admission. The admission laboratory test results showed normal renal hepatic profile, normal muscle enzymes coagulation, viral serologic, negative HIV result, negative toxoplasmosis result, negative complete collagenogram included rheumatoid factor, antinuclear anti-body, anti DNA anti-body, anti-RO; anti-LA, anti-SM, anti-RNP, and anti-neutrophil cytoplasm antibodies (perinuclear and cytoplasmic). Echocardiogram showed mild pericardial effusion. Lumbar puncture was normal. Brain and cervical spinal magnetic resonance imaging was normal. We observed hyperintensity in sequence T2 and hypointensity in T1 with enhancement of homogenous contrast at C4 to C6. The case was assumed to be myelitis with myelopathic pain and serositis being studied. Daily 1-g pulses of methylprednisolone were started for 3 days, meprednisone was continued orally with gradually decreasing dose. Treatment with 75-g pregabalin was starter and 400-g carbamazepine was administered because of pain persistence.

After 11 months, the patient presented with inflammatory and mechanic lumbago, left sacroiliitis, mouth dryness, oral ulcers, hair loss, and diffuse edema in the left foot, and no arthritis signs. Pain in wrists and hands with bilateral stiffness of proximal and distal interphalangeal joints together with muscle and joint pain. A new collagenogram showed antinuclear antibodies titer of 1:80 with a thin speckled pattern and other results were all negative. Articular echography of the right hand showed in the 2nd articulation metacarpophalangeal, articular distension exudative grade II; negative power Doppler. It was considered as myelitis secondary to collagen disease. Seronegative spondyloarthropathy and probable Behçet disease. Treatment with 8 mg/d meprednisone was begun, AINES, 200 mg/d of hydroxychloroquine, and 15 mg methotrexate for week.

The patient showed a bad clinical evolution, with myoclonic movements in the right hand and recurrent genital and oral ulcers. A new brain RMI was performed, evoked potentials, electroencephalogram, all visual campus normal and AC negative anti aquaporin 4.

HLA B 52 is received and a positive 35. After that, we established the diagnosis of Behçet syndrome with neurological, articular, and mucosal involvement. The patient is currently treated with 100 mg/d of azathioprine plus 20 mg/d of meprednisone with clinical improvement.

Key words
neuro-Behçet disease, myelitis, HLA

SYLVAIN VERBANCK

Lupus mimicker and "full-house" glomerulonephritis

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INTRODUCTION The spectrum of renal insufficiency, either acute or chronic, related to HIV is varied and includes: iatrogenic complications induced by anti-retroviral treatments, pathologies induced by the virus itself, others that are linked to the systemic immune response to infection, or opportunistic infections. We present the case of a patient presenting with nephrotic syndrome. Renal biopsy was performed, highlighting an immunological "full house" pattern, highly suggestive of lupus nephropathy. However, as the blood test for antinuclear antibodies was negative and the HIV was serology positive, the diagnosis of HIV-associated immune complex kidney disease (HIVICK) was retained.
CASE REPORT A 48-year-old patient with a history of well-controlled type 1 diabetes mellitus under prandial–basal regimen, hypertension treated with perindopril, amlodipine, and bisoprolol, and hypercholesterolemia treated with simvastatin presented with edematous syndrome (morning swelling of the eyelids and limbs) since one week. Systematic anamnesis reported an influenza-like episode with sore throat and myalgia one month earlier, currently resolved. The vitals were normal and the rest of the clinical examination did not reveal any additional signs. Blood testing showed a hemoglobin level of 9 g/dl (mean corpuscular volume of 94 fl, low haptoglobin schistocytes at 16/1000). Platelet count was within the reference range, and Coombs test as well as irregular antibodies were nega-

tive. White blood cell count demonstrated total lymphocytosis of 7750/μl. Renal test showed stage 1 acute kidney injury according to the Acute Kidney Injury Network (urea, 89 mg/dl; creatinine, 1.36 mg/dl; DFG calculated according to the Chronic Kidney Disease-Epidemiology Collaboration equation, 62 ml/min/1.73 m^2). The level of albumin indicated albuminemia at 29 g/l. Immune testing demonstrated polyclonal hypergammaglobulinemia at 13.9 g/l, antinuclear antibodies were negative, C3 was normal (72 mg/dl), C4 was lowered (7 mg/dl), and C3d/C3 ratio was increased at 1.7. A slight inflammatory syndrome was present (C-reactive protein, 12 mg/l). 24-hour urine analysis showed microscopic hematuria and total proteinuria at 8 g/d (albuminuria A3 at 5 g/d).

Seen the table of "impure" nephrotic syndrome, corticotherapy was started (1 mg/kg) together with furosemide. As type 1 diabetes was well controlled (normal albuminuria one year earlier), renal biopsy was performed, which highlighted a "full house" immunological pattern, first suggestive of a diagnosis of lupus nephropathy. However, in the view of a HIV-positive serology, the diagnosis of HIV-associated immune complex kidney disease (HIVICK) was finally retained. There was no hepatitis B nor C virus coinfection. The patient was followed for over 18 months, corticosteroids were stopped, and he is currently receiving mycophenolate mofetil and dual antiretroviral therapy (dolutegravir plus emtricitabine) with resolution of proteinuria, but persistence of grade 2 chronic kidney disease.

DISCUSSION This case of HIV-associated nephropathy (HIVAN) highlights the importance of exhaustive assessment of adult nephrotic syndrome. The originality of this case lies in the kidney biopsy which is almost pathognomonic of lupus nephropathy. Two major subgroups of HIV-related nephropathies have been described: HIVAN, secondary to the direct action of the virus and HIVICK, resulting from the immune response associated with HIV systemic proliferation. Both groups differ in terms of diagnosis, histopathology, therapeutic management, and patient outcome (HIVAN has a poorer renal outcome).

In conclusion, the diagnosis of nephrotic syndrome must be exhaustive, including renal biopsy if indicated. Untreated HIV infection as well as antiretroviral therapy are associated with multiple kidney diseases.

Key words
complex kidney disease, full-house pattern, HIV-associated immune, lupus-like glomerulonephritis, lupus mimicker

ANNA ZUBKIEWICZ-ZARĘBSKA

Clinical picture suggesting Crohn disease as the manifestation of multiple myeloma

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CASE REPORT Multiple myeloma (MM) is a neoplastic proliferation of plasma cells producing a monoclonal immunoglobulin. Extramedullary plasmacytomas are rare, and seen in approximately 7% of patients with MM at the time of diagnosis. Symptomatic gastrointestinal involvement in MM has been rarely reported. We report a case of MM with gastrointestinal involvement suggesting Crohn disease at disease manifestation. Radiologic and endoscopic studies revealed colon lesions similar to those found in inflammatory bowel disease. Due to laboratory and radiological findings, proliferative disease was suspected and later confirmed on basis of trepanobiopsy which revealed infiltration of plasmacytes. The patient was treated according to appropriate guidelines and the disease remains in remission. Presented case demonstrates that although colon lesions in MM are rare, they should be considered during differential diagnosis of patients with chronic diarrhea, especially when they coexist with other signs of plasma cell neoplasm.

Key words
diarrhea, inflammatory bowel disease, multiple myeloma