POLISH ARCHIVES OF
Internal Medicine
POLSKIE ARCHIWUM MEDYCYNY WEWNĘTRZNEJ

ESTABLISHED 1923 VOL.133 (2023) Special Issue 1 WWW.MP.PL/PAIM

SPECIAL ISSUE

IMPACT FACTOR 2022
4.8

Abstract proceedings of the best case report contest 2023

Clinical Cases in Internal Medicine: Learning Through Practice
(8th McMaster International Review Course in Internal Medicine,
May 11–13, 2023, Kraków, Poland, hybrid course)

medycyna praktyczna

POLISH ARCHIVES OF
Internal Medicine
POLSKIE ARCHIWUM MEDYCYNY WEWNĘTRZNEJ

ADDRESS
Cholerzyn 445
32-060 Liszki, Poland
phone: +48 12 293 42 20
fax: +48 12 293 40 10
email: pamw@mp.pl
www.mp.pl/paim

PUBLISHER
Medycyna Praktyczna

INDEXED IN
Chemical Abstracts Service (CAS), Crossref, Database of Abstracts of Reviews of Effects (DARE), EBSCO, EMBASE, GENAMICS, Geneva Foundation Free Medical Journals, Index Copernicus (IC), J-Gate, Master Journal List, MEiN, National Library of Medicine (NLM), Polish Medical Library (GBL), PubMed/MEDLINE, Science Citation Index Expanded (SCI-Ex), SciFinder, Scopus, Web of Science Core Collection

Online ISSN: 1897-9483

Impact Factor 2022 = 4.8

Partially financed through state budget funds granted by the Republic of Poland's Minister of Education and Science under the program "Doskonała nauka II" ("Excellent Science II").

Minister of Education and Science
Republic of Poland

ABSTRACT PROCEEDINGS OF THE BEST CASE REPORT CONTEST 2023

Clinical Cases in Internal Medicine: Learning Through Practice
(8th McMaster International Review Course in Internal Medicine,
May 11–13, 2023, Kraków, Poland, hybrid course)

AWARD-WINNING CASES

1ST PLACE: JIRI MÜLLER

Alpha-gal syndrome: a case report of tick-borne anaphylactic shock

Jiri Müller*, Jaroslav Radej, Martin Matejovic

First Department of Internal Medicine, Pilsen University Hospital, Charles University in Prague, Pilsen, Czech Republic

*Corresponding author: mullerji@fnplzen.cz

INTRODUCTION The most common cause of vasoplegic shock in critical care is sepsis. However, although rarely and only in specifically-sensitized individuals previously bitten by a tick, red meat may provoke a delayed allergic reaction called an alpha-gal syndrome. We present a case of protracted, life-threatening manifestation of alpha-gal syndrome, which, due to an unusual absence of typical features of anaphylaxis can mask septic shock. Our case indicated that premature end of the diagnostic process can contribute to a diagnostic error.

CASE DESCRIPTION A previously healthy 68-year-old man collapsed and gasped while sitting in a chair 60 minutes after finishing his lunch of spicy pork kidneys. Telephone-assisted cardiopulmonary resuscitation was performed by his wife for 15 minutes with automated external defibrillator not recommending defibrillation. Upon arrival of an ambulance, spontaneous circulation had already been restored, but severe hypotension persisted requiring fluid and norepinephrine administration. On admission to an emergency department, the patient was awake yet drowsy, with no apparent source of infection on physical examination. A tick embedded in the skin of the left arm was found and extracted. Vasoplegic shock progressed rapidly, necessitating intubation, mechanical ventilation, and continuous hemodynamic support. The patient's wife denied any history of allergy or medication use. Due to an absence of apparent culprit allergen, no history of allergies, and laboratory and computed tomography findings pointing toward an infection, fulminant septic shock was considered a working diagnosis, and empiric broad-spectrum antibiotic treatment was initiated. However, after carefully reviewing the salient features of the case, including an abrupt pace of the life-threatening disease process, additional anamnestic information about itchy skin right after eating the lunch, and a tick removed during the admission, protracted anaphylaxis was reconsidered. Elevated serum tryptase confirmed mast cell involvement, and later specific immunoglobulin E against alpha-gal confirmed the diagnosis of alpha-gal syndrome as a causative factor of vasoplegic shock with rapid progression to cardiac arrest.

CONCLUSIONS Alpha-gal syndrome is a relatively new, but increasingly recognized health issue. Here, we present a case of life-threatening, protracted anaphylactic shock following innards consumption, which was initially mistaken for a septic shock. We propose that alpha-gal syndrome should be considered in the differential diagnosis of vasoplegic shock of unclear etiology even in the absence of typical allergic symptomatology and typical allergen exposure, as alpha-gal is present in a wide variety of carriers.

Key words
alpha-gal syndrome, anaphylaxis, meat allergy, tick allergy, sepsis mimics

2ND PLACE: DANIEL N. MARCO

Gastric mucormycosis presenting as fever of unknown origin in an immunocompetent host after heatstroke

Daniel N. Marco[1]*, Borja González[2], Pamela-Inés Doti[1]

1 Intensive Care Unit, Department of Internal Medicine, ICMID, Hospital Clínic Barcelona, Spain

2 Department of Internal Medicine, Hospital La Paz, Barcelona, Spain

*Corresponding author: dnmarco@clinic.cat

INTRODUCTION Mucormycosis is an aggressive fungal disease caused by fast-growing environmental molds. An infection usually occurs in immunocompromised hosts after inhalation of spores but can also occur via injured skin or ingestion of contaminated food. On rarer occasions, the infection manifests in immunocompetent hosts. Among different clinical subtypes, classified as per the site of occurrence, rhino-ocular-cerebral mucormycosis is the most frequent, followed by gastrointestinal location. The diagnosis of gastrointestinal mucormycosis is a complicated task due to its unspecific features, and the need for invasive collection of microbiological samples. The majority of cases are diagnosed postmortem. Among the cases diagnosed in alive patients, the most frequent scenario is sudden visceral perforation or gastrointestinal bleeding, leading to urgent surgery and delayed etiologic diagnosis on pathologic examination.

CASE DESCRIPTION We present a case of gastric mucormycosis affecting a young man without any classic risk factors. He was admitted to a hospital in the course of a severe heatstroke, with multiorgan failure. After initially favorable evolution, he started with persistent fever and elevation of acute-phase reactants. The origin of the fever was unknown until he presented abdominal retentions with coffee-grounds appearance. Gastroscopy and computed tomography scan showed an extensive necrotic ulcer in the gastric wall, whose cultures revealed invasive mucormycosis caused by *Rhizopus microsporus*. A combined therapy with antifungal agents was introduced and surgery was performed. Satisfactory control of the infection was achieved, without evidence of relapse after 6 weeks of treatment.

DISCUSSION The relevance of the case lies both in the diagnosis of the invasive fungal infection in an immunocompetent host with heatstroke (to date, this has been the second reported case in the literature), and in the fact that the infection could be diagnosed in vivo, via endoscopy and subsequent pathologic specimen examination, which allowed for treating the infection at a localized stage. Heatstroke may cause severe pathophysiologic multiorgan disturbances and promote severe infections, with sepsis due to translocation of gram-negative bacteria from the gastrointestinal (GI) tract being the most common. Analogous mechanisms could lead to fungal infections. Thus, apart from classic risk factors, a suspicion of mucormycosis may be raised in other clinical settings, such as heatstroke.

LESSONS TO BE LEARNED

- Mucormycosis is a life-threatening fungal infection affecting mostly immunocompromised hosts. The hallmark of its evolution is angioinvasion and progressive local tissue necrosis, with high mortality rates despite proper therapy.
- A combination of pharmacologic agents and surgery is the cornerstone of the treatment. However, even appropriate antifungal treatments at correct dosage may not penetrate into necroischemic lesions caused by angioinvasion.
- Apart from the classic risk factors, mucormycosis should be suspected in other clinical settings where the internal medium is intensely disrupted. These include, for example, heatstroke (similarly to increased awareness among intensive care unit caregivers following the emergence of COVID-19–associated mucormycosis in relation to glucocorticoid use).
- Heatstroke entails severe pathophysiologic multiorgan disturbances promoting severe infections, with sepsis caused by translocation of gram-negative bacteria from the GI tract being the most common. Analogous mechanisms could lead to fungal infections. To date, this has been the second reported case of invasive mucormycosis after heatstroke.

Key words
antifungal treatment, gastrointestinal mucormycosis, heatstroke

3RD PLACE: MONIKA GÓRSKA

Diffuse alveolar hemorrhage in a chronically hemodialyzed patient with progressive necrotic skin lesions as a manifestation of calciphylaxis

Monika Górska*

Department of Internal Medicine and Transplant Nephrology, Medical University of Lodz, Łódź, Poland

*Corresponding author: monika.gorska@umed.lodz.pl

INTRODUCTION Calciphylaxis is a rare but severe and life-threatening complication of end-stage kidney disease (ESKD) and the associated calcium-phosphate imbalance. It develops slowly with late clinical manifestation. Currently, there are no defined diagnostic standards or treatment regimens for this condition.

CASE REPORT A 49-year-old woman hemodialyzed (HD) for 9 years complained of leg pain (7/10), described as a burning, bursting sensation, exacerbated during HD sessions, with firm subcutaneous tumors on both thighs and livedo reticularis on both lower limbs. Her comorbidities included myocardial infarction treated with percutaneous coronary intervention (2016), paroxysmal atrial fibrillation treated with vitamin K antagonists (VKAs) for 5 months, tertiary hyperparathyroidism, Crohn disease, and recently diagnosed erythema nodosum. Laboratory workup showed anemia, elevated C-reactive protein, hyperphosphatemia with hypocalcemia, and elevated parathormone.

Due to suspected erythema nodosum, systemic methylprednisolone followed by oral prednisone were introduced. In the following days, the patient showed no response to the glucocorticoids, complained of progressive exacerbated pain with muscle weakness in lower extremities, with severely impaired gait, skin lesions progressed into areas of livedo racemosa and necrosis. Differential diagnoses excluded arterial / venous ischemia, steroid-induced myopathy, or dermatomyositis. Skin biopsy revealed leukocytoclastic vasculitis but the titers of antinuclear and antineutrophil cytoplasmic antibodies were negative. High-resolution computed tomography (HRCT) of the lungs revealed diffuse alveolar hemorrhage (DAH), confirmed by bronchoalveolar lavage, without hemoptysis, dyspnea, or auscultatory findings. Since all the findings strongly indicated seronegative vasculitis, intravenous cyclophosphamide (2 pulses of 600 mg) followed by rituximab (1 pulse of 600 mg) were administered, with no clinical response in the following 2 weeks.

Considering the overall clinical presentation with progressive skin necrosis located mainly in the proximal extremities with concomitant DAH, no response to immunosuppression, long history of severe hyperparathyroidism and VKA treatment, severe calciphylaxis was diagnosed. Immunosuppression was discontinued, HD was intensified with conversion to hemodiafiltration (HDF) with simultaneous calcium chelating treatment involving intravenous infusions of natrium thiosulphate during the last 30 minutes of each HDF session. Hyperparathyroidism treatment consisted of cinacalcet and sevelamer. Hyperbaric oxygen therapy was included to supplement wound healing. In a 3-month follow-up the patient showed areas of demarcation, with margins of healthy tissue surrounding the lesions, and reduced lung involvement.

DISCUSSION The case exemplifies difficulties in establishing the diagnosis of calciphylaxis. Skin necrosis and DAH in a patient with ESKD caused by glomerulonephritis, and with Crohn disease, indicated development of autoimmune vasculitis. Unsuccessful immunosuppressive treatment, developed clinical presentation, and thorough literature search allowed us to reach a proper diagnosis. Visceral manifestations of calciphylaxis are rare, however, a few cases of calciphylaxis misdiagnosed as vasculitis are available in the literature.

LESSONS TO BE LEARNED

- Conditions with different pathomechanism and treatment may share clinical presentation.
- In ESKD, calciphylaxis must be included in differential diagnosis of atypical skin and visceral lesions.
- Intensification of renal replacement therapy, calcium-phosphate disturbances management, chelating agents, and wound care are the basis of calciphylaxis treatment.

Key words
calcyphilaxis, diffuse alveolar hemorhage, hemodialysis

ORAL PRESENATIONS

MERVE ADIYAMAN

A case of Waldenstrom macroglobulinemia presenting with acute kidney injury

Merve Adıyaman*, Harun Akar

Health Sciences University, Tepecik Education and Research Hospital, Department of Internal Medicine, Hematology and Pathology, Izmir, Turkey

*Corresponding author: drmerveadymn@gmail.com

INTRODUCTION Waldenstrom macroglobulinemia (WM) is a lymphoplasmacytic hematologic malignancy that may present with infiltration of the bone marrow, spleen and lymph nodes, and immunoglobulin (Ig) M monoclonal gammopathy. WM rarely shows extranodal involvement, and the diagnosis is made by lymph node or bone marrow biopsy. The rates of hepatomegaly, splenomegaly, and lymphadenopathy in patients with WM are 20%, 15%, and 15%, respectively. Symptoms usually include dizziness, blurred vision, epistaxis, or exacerbation of congestive heart failure when the IgM level exceeds 3 g/l. High IgM level and hyperviscosity syndrome require treatment.
CASE DESCRIPTION In this case report, we discuss a diagnosis of WM and regression of renal damage following treatment in a 72-year-old woman admitted with dizziness and nausea, who was hospitalized with acute kidney injury.

Key words
acute kidney injury, Waldenstrom macroglobulinemia

SARFRAZ BEGG

A case of copper sulfate poisoning

Sarfraz Begg*

University of Cape Town, Grooteschuur Hospital, Cape Town, South Africa

*Corresponding author: sbegg01@gmail.com

INTRODUCTION Copper sulphate is an easily available compound widely used around the world as a pesticide and fungicide, and commonly used in the agricultural industry. It is a rare cause of poisoning in western countries, with the exception of the subcontinent, where cases are mainly suicidal in nature. Copper sulphate is an inorganic salt that is highly soluble in water, bright blue in color, and odorless. The bright blue color of the hydrated form of copper sulphate crystals is visually attractive to children and may be a reason for unintentional poisoning in kids.
CASE DESCRIPTION We report a rare case of a 24-year-old man, who is HIV-positive and on antiretroviral therapy, who presented to the emergency department with gastrointestinal upset, jaundice, and oliguria, complicated by multiorgan dysfunction. He reported ingesting a powder that he had received from a friend, a practicing traditional healer. The diagnosis of copper sulphate poisoning was suspected after reviewing the powder ingested and the unique color of the powder, which was bright blue.
DISCUSSION Common complications of copper sulphate poisoning include gastrointestinal tract hemorrhage, hemolysis, methemoglobinemia, rhabdomyolysis, liver impairment, cardiac involvement, and renal failure. Our patient received symptomatic treatment with fluids, multiple blood transfusions, proton pump inhibitors, methylene blue, and hemodialysis. He had a favorable outcome with his renal function recovering to near normal. Copper sulphate poisoning is unheard of in South Africa, however, up to 72% of traditional healers in a province in South Africa use copper sulphate in their daily treatments. Hopefully, this case raises awareness and highlights the importance of recognizing the complications and providing prompt treatment.

Key words
copper sulphate, dialysis, methemoglobinemia, renal failure, suicide

GEORGE CIULEI

Pembrolizumab-induced colitis: diagnosis and management

George Ciulei*, Mirela G. Perné, Olga H. Orășan

Iuliu Haţieganu University of Medicine and Pharmacy, Cluj-Napoca, Romania

*Corresponding author: geo.ciulei@yahoo.com

INTRODUCTION Immune checkpoint inhibitors (ICIs), such as pembrolizumab or ipilimumab, have improved the management and survival rate of patients with metastatic non–small-cell lung carcinoma (NSLC) or metastatic melanoma. Immune-related adverse effects can be located at the gastrointestinal tract. The case report describes the diagnosis and management of a patient with metastatic NSLC treated with pembrolizumab.
CASE DESCRIPTION A 50-year-old woman was diagnosed with stage 4 NSLC with cerebral, mediastinal, and left adrenal metastases. She was enrolled in a pembrolizumab trial and tolerated the treatment initially. After 4 months, she complained of diarrhea and abdominal pain. Colonoscopy showed changes typical of colitis and the diagnosis was corroborated by histopathologic examination. A differential diagnosis included new-onset ulcerative pancolitis, pembrolizumab-induced pancolitis, and pancolitis caused by Epstein-Barr virus, HIV, or cytomegalovirus infection. The patient was diagnosed with grade 2 pembrolizumab-induced colitis and treatment was started with prednisone and 5-aminosalycilic acid. Symptom remission was obtained in a few days but after a month the patient returned again with the symptoms of colitis.
CONCLUSIONS Colitis induced by ICIs that has not responded to prednisone should be treated with a biologic therapy.

Key words
colitis, immune checkpoint inhibitors, non–small-cell lung carcinoma

JOÃO Á. DE MELO E SOUSA

An unusual presentation of TAFRO syndrome

João Á. de Melo e Sousa[1]*, Isabelle Nubourgh[2], Daphné Papazoglou[2]

1 Hôpital Erasme, Brussels, Belgium
2 CHU Saint-Pierre, Brussels, Belgium

*Corresponding author: jdemeloe@ulb.be

INTRODUCTION TAFRO syndrome (TAFROs) is characterized by thrombocytopenia, anasarca, fever, reticulin myelofibrosis, and organomegaly. Due to similar histopathologic findings on lymph node biopsy, TAFROs is often considered a variant of Castleman disease (CD). CD is a rare lymphoproliferative disorder that can be unicentric or multicentric (MCD). According to its systemic manifestations and the HHV8 negativity, TAFROs is classified as a subtype of idiopathic MCD (iMCD). This classification is relevant because the 5-year prognosis in iMCD and TAFROs is, respectively, 100% and 66.5%. Sjögren syndrome (SjS) is a systemic autoimmune disease characterized by sicca syndrome, systemic manifestations, and lymphocytic infiltration of the exocrine glands (especially salivary and lacrimal glands). Despite their low prevalence, an association of TAFROs and SjS has already been reported 10 times in the literature.
CASE DESCRIPTION A 44-year-old woman was admitted for fever, diarrhea, bicytopenia (anemia and thrombocytopenia), edema, xerostomia, and asthenia. Her physical examination showed mild anasarca, hard cervical lymph nodes (approximately 1 cm in diameter), and hepatosplenomegaly. Bone marrow aspiration resulted in a "dry tap." Bone marrow biopsy revealed reticular myelofibrosis with hypercellular marrow and increased megakaryocytes. Cervical lymph node biopsy was consistent with CD, and salivary gland biopsy with SjS. Treatment with corticosteroids, rituximab, and siltuximab was administered from day 18, 20, and 27, respectively.

The patient showed a good clinical and biological response but developed autoimmune hemolytic anemia and a persistent low platelet count that was treated with intravenous immunoglobulin on day 39. Thrombocytopenia recovery occurred after 30 days of treatment. According to the 2019 TAFROs diagnosis criteria, our patient presented 3 major and 3 minor criteria and SjS, which is an exclusion criterion in the 2019 classification, but not in the 2015 classification. Our case shows, once more, the association between these 2 entities. TAFROs could be a severe manifestation of SjS. It is well known that SjS is associated with an increased risk of lymphoid malignancy. TAFROs could be a manifestation of a nonclonal secondary lymphoid proliferation of SjS. This association should be sought in the presence of one of these diseases.

DISCUSSION There are no clear guidelines for the treatment of TAFROs and the platelet count is often the last parameter to improve after initiation of the therapy. In the literature, corticosteroids are the first-line treatment. In a review of 38 cases, 9 were treated with corticosteroids alone, and 29 patients received other treatments. The median time of thrombocytopenia recovery was, respectively, 25 and 50 days. In our case, we chose an aggressive approach, as we know that the prognosis of TAFROs differs from iMCD in the first months.

CONCLUSIONS The association between TAFROs and SjS raises a question of a continuum between these 2 pathologies, and opens a research field on SjS and lymphoproliferative disorders. Finally, TAFROs prognosis is critical and requires further studies on the effectiveness of the aggressive and conservative treatment.

Key words
fever, Sjögren syndrome, TAFRO syndrome, thrombocytopenia

MARKÉTA DUDKOVÁ

Superior vena cava syndrome as a manifestation of antineutrophil cytoplasmic antibodies–associated aortitis

Markéta Dudková*, Martina Skacelová, Pavel Horak

Third Department of Internal Medicine – Nephrology, Rheumatology and Endocrinology, Faculty of Medicine and Dentistry, Palacky University Olomouc, Olomouc University Hospital, Olomouc, Czech Republic

*Corresponding author: marketa.dudkova@fnol.cz

INTRODUCTION Antineutrophil cytoplasmic antibodies (ANCA)-associated vasculitis is a necrotizing small vessel vasculitis associated with ANCA positivity. Patients with systemic form of ANCA-associated vasculitis (AAV) usually present with general symptoms of inflammation together with respiratory system and kidney impairment. Aortic vasculitis is rare and potentially life-threatening manifestation of AAV.

CASE DESCRIPTION We discuss a case of AAV aortitis in a 71-year-old man, presenting as superior vena cava syndrome (SVCS) caused by inflammation of the ascending aorta. The patient with a history of lumbar spinal stenosis, hearing loss, and thrombocytosis presented with face edema and neck swelling, intermittent face cyanosis, and acute decompensation of hypertension. Hypertensive medication was changed due to a suspected allergic reaction, and a small dose of peroral corticosteroids was administered by his family doctor. Despite a minor improvement, the patient was referred to an internist.

Laboratory workup showed only mild elevation of inflammatory parameters (C-reactive protein), mild thrombocytosis, and microscopic hematuria with normal kidney function. During the search for the cause of SVCS, abdominal ultrasound and computed tomography (CT) scan of the lungs and neck were performed. Several nodules sized under 10 mm and interstitial fibrosis were found on CT scan of the lungs. There was an enlargement of the ascending aortic wall that caused compression of the superior vena cava. No thrombosis or tumors were found. Cystoscopy was negative. High positivity of antimyeloperoxidase antibodies was detected.

Positron emission tomography-CT scan confirmed aortitis. Malignancy and infection were repeatedly excluded. The American College of Rheumatology / European Alliance of Associations for Rheumatology classification criteria for microscopic polyangiitis were met. We started treatment with high doses of corticosteroids and immunosuppressive drugs according to the guidelines with a good effect on SVCS symptoms.

DISCUSSION It is unusual for AAV to cause aortic inflammation. It is suggested that ANCA lead to aortitis by causing vasorum vasculitis in the adventitia, which later spreads to the intima and media. ANCA-associated aortitis may present as an aneurysmal disease, aortic dissection, rupture, aortic regurgitation, and even death. Other infectious and noninfectious causes must be excluded before immunosuppressive treatment is implemented.

The main purpose of this case report is to emphasize the possibility of aortic involvement in AAV as a rare and possibly life-threatening condition. Right diagnosis and treatment are necessary to avoid serious manifestation of AAV.

Key words
aortitis, ANCA-associated vasculitis, superior vena cava syndrome

ANTONELLA FONTANA

Platypnea-orthodeoxia syndrome: a rare clinical case

Antonella Fontana, Paolo Carducci, Clara Balsano*

School of Emergency and Urgency Medicine, MESVA Department, University of L'Aquila, Coppito, L'Aquila, Italy

*Corresponding author: clara.balsano@univaq.it

CASE DESCRIPTION A 32-year-old nonsmoking woman was admitted to the emergency department of the San Salvatore Hospital of L'Aquila experiencing worsening of dyspnea. Three months earlier the patient had undergone left pneumonectomy. Arterial blood gas analysis performed on air revealed pH 7.53, pCO_2 23.7 mm Hg, pO_2 38.7 mm Hg, and SpO_2 80%. Blood tests were within normal limits, except for a moderate increase in D-dimer.

Considering the diagnostic hypothesis of pulmonary embolism, computed tomography angiography of the chest was performed and showed no radiological characteristics suggesting parenchymal pathology and no evidence of thromboembolic or other vascular pathologies.

Based on the clinical evaluation of the patient, features of severe platypnea were evident: supine decubitus position was recommended. Together with the above position variations, a strong desaturation was observed, going from almost normal values of 94%–96% SpO_2 to 80%–82% SpO_2, when the patient was sitting up in bed.

Based on the data above, a right-left shunt was assumed, and transesophageal contrast echocardiogram was requested. The latter demonstrated patent foramen ovale (PFO).

DISCUSSION The first description of a condition very similar to platypnea-orthodeoxia syndrome (POS) following pneumonectomy dates to 1956, by Schnabel et al.

A recent Pubmed database-EMBASE systematic review identified 8 studies concerning 10 patients (6 men, 4 women) of mean age 62 years. All the patients underwent resection of the right lung. The time interval between the operation and POS was from 2 days to 3 years. PFO was present in 90% of cases.

Considering the data, our patient shows at least 2 peculiarities; the first and most relevant is her young age: in fact, there is only 1 description of POS in a patient of younger age.

The second peculiarity is the left localization of the previous pneumonectomy, much rarer than the contralateral in the known cases. Partial or total pulmonary resection can rarely be followed by the onset of POS. This syndrome can be severely disabling, so physicians must be prepared for its prompt recognition, maintaining a high degree of suspicion.

Key words
dyspnea, left pneumonectomy, patent foramen ovale, platypnea-orthodeoxia syndrome

AGNIESZKA JAROSIŃSKA

Atypical presentation of Lyme disease

Agnieszka Jarosińska[1]*, Aleksandra Pilśniak[1], Aleksandra Cieśla-Fuławka[2]

1 Department of Internal Medicine, Autoimmune and Metabolic Diseases, Faculty of Medical Sciences in Katowice, Medical University of Silesia, Katowice, Poland
2 Department of Neurology, Faculty of Medical Sciences in Katowice, Medical University of Silesia in Katowice, Katowice, Poland

*Corresponding author: ajarosinska@sum.edu.pl

CASE DESCRIPTION A 64-year-old man was admitted to the Department of Internal Medicine with symptoms of acute respiratory failure occurring in the supine position and oxygen saturation decreasing to 75%–80%. Physical examination revealed tachypnea and increased respiratory effort with the activation of accessory respiratory muscles, without any signs of an acute respiratory infection. Arterial blood gases indicated hypoxemic respiratory failure, but the levels of inflammatory markers were low and the D-dimer concentration was normal. Medical imaging, such as computed tomography (CT) pulmonary angiography, and chest CT excluded the diagnosis of pulmonary embolism or pneumonia. Echocardiography did not reveal any significant abnormalities. Bronchoscopy results were normal and brain magnetic resonance imaging (MRI) showed no pathologies. The initial diagnosis of left-sided phrenic nerve palsy was made. Chest fluoroscopy revealed bilateral phrenic nerve palsy. Due to the unknown etiology of his condition, the patient was transferred to the Department of Neurology for further evaluation.

Upon admission to the Department of Neurology, neurologic examination revealed no significant abnormalities. MRI of the cervical spine showed cervical spondylosis. Electromyography and electroneuronography did not reveal the cause of the phrenic nerve palsy. Serologic tests excluded autoimmune neuromyelitis, myasthenia gravis, or herpes simplex virus infection as possible causes of the symptoms. A positive result was obtained for both immunoglobulin (Ig) M and IgG antibodies in the blood serum, specific for *Borrelia burgdorferi* antigens. The analysis of the cerebrospinal fluid (CSF) obtained by a lumbar puncture revealed the presence of oligoclonal bands and positive enzyme-linked immunosorbent assay and Western Blot for IgM and IgG antibodies specific for *B. burgdorferi* antigens. Hence, the diagnosis of probable Lyme neuroborreliosis complicated by the diaphragm paralysis was made. Targeted antibiotic therapy against *B. burgdorferi* was introduced and partial clinical improvement was observed.

DISCUSSION Typical neurologic symptoms of Lyme neuroborreliosis include headaches and cranial nerve paralysis or paresis, and rarely radiculitis and peripheral neuritis, peripheral polyneuropathy, and chronic encephalomyelitis. Only 16 cases of Lyme neuroborreliosis complicated by paralysis of the diaphragm are available in the literature. Serologic diagnostics of Lyme disease should be considered in patients with diaphragmatic paralysis of undetermined etiology.

Key words
acute respiratory failure, diaphragm paralysis, Lyme disease, Lyme neuroborreliosis

HYEONGMIN KIM

A case report of spontaneous remission of acromegaly after pituitary apoplexy

Hyeongmin Kim[1]*, Eun Hee Koh[2]

1 Department of Internal Medicine, Asan Medical Center, University of Ulsan College of Medicine, Seoul, Republic of Korea
2 Department of Endocrinology, Asan Medical Center, University of Ulsan College of Medicine, Seoul, Republic of Korea

*Corresponding author: freewillboy01@gmail.com

INTRODUCTION Pituitary apoplexy is an endocrine emergency resulting from infarction or hemorrhage of the pituitary gland. Rarely, pituitary apoplexy can be the first manifestation of functioning adenomas, and lead to spontaneous remission of hormonal hypersecretion. We report a case of a man with acromegaly who showed signs of pituitary apoplexy but no growth hormone (GH) hypersecretion.

CASE DESCRIPTION A previously healthy 54-year-old man, complaining of ongoing headache, was diagnosed with a 3.5-cm pituitary mass on brain magnetic resonance imaging (MRI). He was admitted to a neurosurgery department for acromegaly-like features, which had progressed during the prior 7 years. He experienced severe general weakness and nausea. Laboratory workup revealed hyponatremia, normal level of insulin-like growth factor 1 (IGF-1), and panhypopituitarism. Initially, the patient was diagnosed with macroprolactinoma and panhypopituitarism. Glucocorticoid replacement therapy was implemented, and then he was discharged on cabergoline for macroprolactinoma. On 25th day of cabergoline administration, he was referred to the emergency room for severe headache, suggesting pituitary apoplexy after cabergoline, and consequent adrenal insufficiency. Lastly, the remnant pituitary mass was refractory to cabergoline and the patient had trans-sphenoidal surgery for increasing tumor and subsequent visual impairment.

DISCUSSION The patient should have been initially diagnosed with synchronous GH- and prolactin-secreting pituitary adenoma. Considering minimal intratumoral hemorrhage on initial brain MRI, there may be recurrent subclinical pituitary apoplexy including the initial episode, and then the GH-secreting tumor spontaneously resolved, presenting normal IGF-1.

This case describes a rare type of pituitary adenoma, synchronous GH- and prolactin-secreting pituitary adenoma, with spontaneously cured hormonal hypersecretion after pituitary apoplexy.

Key words
acromegaly, pituitary apoplexy, prolactinoma, spontaneous remission

CHUNG M. A. LIN

Giant cell arteritis: an interesting case

Chung M. A. Lin[1]*, Alice R. Lorenzi[2], Faye A. H. Cooles[1,3,4]

1 Translational and Clinical Research Institute, Newcastle University, Newcastle-upon-Tyne, United Kingdom
2 Department of Rheumatology, Newcastle upon Tyne Foundation Trust Hospitals, Newcastle-upon-Tyne, United Kingdom
3 Musculoskeletal Research Group and Arthritis Research UK Centre of Excellence in Rheumatoid Arthritis Pathogenesis, Newcastle University, Newcastle-upon-Tyne, United Kingdom
4 Newcastle NIHR Biomedical Research Centre, Newcastle upon Tyne Hospitals NHS Foundation Trust and Newcastle University, Newcastle-upon-Tyne, United Kingdom

*Corresponding author: ncmal@newcastle.ac.uk

INTRODUCTION Giant cell arteritis (GCA), otherwise known as temporal arteritis, is a form of large vessel vasculitis. Patients may present with a variety of symptoms but the most common include headaches, jaw claudication, fatigue, night sweats, and visual disturbance. Diagnosis is made using a combination of temporal artery ultrasound, clinical findings, and blood tests, and the diagnostic gold standard of a temporal artery biopsy (TAB) is sometimes needed for histopathologic confirmation.

CASE DESCRIPTION We report a case of a 60-year-old man who presented with a 4-month history of sudden onset, episodic, pulsating temporo-occipital headaches that lasted for 10 to 25 minutes

at a time and resolved spontaneously. No associated history of jaw claudication, visual symptoms, or polymyalgia rheumatic symptoms were reported. However, he did report drenching night sweats and fatigue. His medical history included cholecystectomy secondary to splenic infarct and appendicectomy, and he had no relevant family history. The only relevant examination finding was minimal tenderness over the right temporal artery and a slightly reduced pulse volume. Blood tests revealed erythrocyte sedimentation rate of 30 mm/h and a negative immune profile. The temporal artery ultrasound scan revealed increased vessel wall thickness of 0.04 cm, suggestive of GCA. In addition, a computed tomography (CT) scan of the chest, abdomen, and pelvis was performed to look for evidence of large vessel disease in view of atypical presentation, which revealed a new 24 mm fusiform aneurysm of the common hepatic artery.

A diagnosis of atypical GCA was made, and the patient was started on high-dose prednisolone (40 mg) for 1 week, followed by a tapering course. However, at 15 mg of prednisolone, he developed visual disturbances in the right eye, later diagnosed at ophthalmology review as the right cilioretinal artery occlusion, presumed secondary to GCA, and he was started on intravenous methylprednisolone. Due to the acute presentation, the patient was investigated and treated for a transient ischemic attack. Imaging demonstrated normal head CT and mildly calcified bilateral carotid bulbs with increased external carotid velocity but no atherosclerosis. The patient was additionally screened for syphilis and following strongly positive serology (serodia particle agglutination reactive at dilution >1:1280, treponemal antibody positive, rapid plasma reagin 1:16 [later dropped to 1:2 with treatment], and detectable immunoglobulin M), he was diagnosed with neurosyphilis and started on a 14-day course of intravenous ceftriaxone (2 g daily for 2 weeks). During a follow-up review by the ophthalmology team, examination findings were typical of syphilitic posterior placoid uveitis, which in this case had convincingly mimicked the cilioretinal artery occlusion. The patient has since recovered well, with no recurrence of the symptoms, however, his fusiform aneurysm of the common hepatic artery has since grown by 4 mm over a 7-month period.

CONCLUSIONS Syphilis is a sexually transmitted disease caused by the spirochaete *Treponema pallidum*. It can infect the central nervous system and cause neurosyphilis, which can mimic GCA. As shown in this interesting case study, it is useful to consider syphilis screening in patients with atypical presentations of GCA, as this can help determine correct and timely treatment but also initiate necessary screening of previous / current partners.

Key words
giant cell arteritis, large vessel vasculitis, syphilis, temporal arteritis

THOMAS J. MATTHEWS

Internal medusa: a large arteriovenous malformation as a cause of refractory seizures

Thomas J. Matthews*, Nikhil Jacobs, Niall S. Colwell

Department of Medicine, Tipperary University Hospital, Clonmel, County Tipperary, Ireland

*Corresponding author: matthetj@tcd.ie

INTRODUCTION Arteriovenous malformations (AVMs) are abnormal direct connections between arterial and venous systems without an interposed capillary bed. This permits high-flow arteriovenous shunting which precipitates structural changes in the afferent and efferent vessels, namely arterial smooth muscle hyperplasia and thinning of venous walls.

CASE REPORT We describe the case of a 43-year-old woman presenting to our emergency department in 2012 with a tonic-clonic seizure secondary to an 8-cm Martin-Spetzler grade V arteriovenous malformation in her left parietal lobe, which was deemed unsuitable for operative or radiotherapy-based intervention. She was managed observationally and relatively good control of her breakthrough seizures was achieved through the addition of brivaracetam in 2018 to her lamotrigine and carbamazepine-based therapy.

Key words
arteriovenous malformation, Martin-Spetzler grade

LUCÍA B. MOLINERO

Bilateral primary frosted branch angiitis

Lucía B. Molinero*, Andrés Serra, Roberto Parodi

Internal Medicine Division, Hospital Provincial del Centenario, National University of Rosario, Rosario, Santa Fe, Argentina

*Corresponding author: lucia.molinero94@gmail.com

INTRODUCTION Frosted branch angiitis represents an extremely rare type of retinal vasculitis that could be primary or secondary. The primary form can usually be generalized and bilateral, and presents as a translucent perivascular sheathing of both arterioles and venules with uveitis, retinal edema, and visual loss. The secondary form is characteristically focal, and it is associated with infectious retinitis, especially by cytomegalovirus, and to a lesser extent by herpes simplex virus, toxoplasmosis, or *Fusarium dimerum* or autoimmune diseases. Other less frequent causes of retinal vasculitis are sarcoidosis, multiple sclerosis, syphilis, lymphoma, and leukemia. Regarding treatment, most patients respond well to topical and / or systemic steroids with a low recurrence rate. We present a case with a primary frosted branch angiitis with bilateral compromise, in which improvement was due to regular therapies as well as the use of mycophenolate.

CASE DESCRIPTION A 23-year-old man, with a history of allergic rhinitis and marihuana consumption, was admitted for 15 days of headaches, retro-orbital pain, photophobia, vomiting, diarrhea, and fever. He had consulted another health care institution, where SARS-CoV-2 infection was ruled out, and was given antibiotics for 7 days. After 6 days he added blurred vision, and saw an ophthalmologist who indicated prednisone 40 mg/day. The patient persisted with visual alterations and consulted our hospital. On examination he was afebrile, with tachycardia, deteriorated visual acuity (counting fingers at 3 m). The laboratory workup showed leukocytosis (11 630 cells/mm^3) and negative antineutrophil cytoplasmic antibody titer. HIV, hepatitis virus B and C, and toxoplasmosis assays were negative, as was viral polymerase chain reaction (PCR) in the cerebrospinal fluid.

Aqueous humor was evaluated by puncture of the anterior chamber with a PCR negative sample for cytomegalovirus, herpes zoster virus, chicken pox, and tuberculosis. Anticardiolipin antibodies, anti-β-2-glycoprotein-I antibodies, Lupus anticoagulants, procalcitonin, and human leukocyte antigen (HLA) B-27 and HLA B-51 were also negative.

Ophthalmologic examination revealed retinal vasculitis, hemorrhages in the 4 quadrants with peripheral dominance, and macular oedema. The patient was diagnosed with bilateral frosted branch vasculitis and was given an intravitreal injection of bevacizumab and triamcinolone and started a high dose of intravenous steroids, with continuation of oral steroids. He was discharged with mild improvement, and after a year of outpatient controls, the acuity of his vision improved after panretinal photocoagulation and mycophenolate administration.

CONCLUSIONS Frosted branch angiitis stands out as a severe and extremely infrequent entity, with fewer than 60 cases reported in the global literature. Although most cases are idiopathic, we highlight the importance of the clinical approach to rule out infectious, autoimmune, or lymphoproliferative causes in a differential diagnosis. Early recognition and implementation of a timely treatment with steroids

allows, in most cases, for rapid recovery and reduces permanent visual impairment due to neovascularization, glaucoma, macular scarring, and retinal detachment. Although mycophenolate was used and showed favorable response in our case, further studies are needed to recommend it to other patients.

Key words
frosted branch, primary, retinal vasculitis

GEORGIOS PERIFANOS

Lower limb edema and cholestasis: it is not always the liver

Georgios Perifanos[1,2]*, Eirini I. Rigopoulou[1,2], George N. Dalekos[1,2]

1 Department of Medicine and Research Laboratory of Internal Medicine, National Expertise Center of Greece in Autoimmune Liver Diseases, Larissa, Greece
2 European Reference Network on Hepatological Diseases (ERN-Rare Liver), General University Hospital of Larissa, Larissa, Greece

*Corresponding author: geoproud1@gmail.com

INTRODUCTION Immunoglobulin (Ig) G4-related disease is a rare, relatively recently described disease, affecting almost any system, with a wide range of clinical presentations, mimicking many other common diseases. Therefore, its diagnosis remains a unique challenge for internists.

CASE DESCRIPTION A 74-year-old man with a history of diabetes mellitus was admitted to our department for further investigation of lower limb edema and increased cholestatic enzymes over the last 15 days. Clinical examination revealed periorbital and lower limb edema, hepatosplenomegaly, and jugular vein distention. Laboratory workup showed normocytic-normochromic anemia, thrombocytopenia, and increased international normalized ratio. A differential diagnosis included cirrhosis, heart failure, nephrotic syndrome, hypothyroidism, and malignancy.

Chronic hepatopathies, nephrotic syndrome, and hypothyroidism were ruled out by extensive evaluation. Immunologic studies exhibited increased IgG of 2190 mg/dl (upper limit of normal [ULN], 1690 mg/dl) and IgG4 of 425 mg/dl (ULN, 200 mg/dl). Computed tomography (CT) showed enlarged mediastinal and paraaortic lymph nodes and bilateral pleural effusions. Pleural effusion examination was indicative of exudate with low adenosine deaminase, negative cytology for malignancy, and negative culture for tuberculosis. Initial cardiac ultrasound was not suggestive of significant heart abnormality. Further investigation with gastroscopy, colonoscopy, magnetic resonance, cholangiopancreatography, and myocardial scintigraphy did not reveal any abnormal findings. A following CT scan demonstrated worsening of pleural effusions and punctate pericardial calcifications. Re-evaluation by an experienced cardiologist with ultrasound showed septal bounce, mildly affected right ventricular contractility, dilated inferior vena cava, and a small amount of pericardial fluid. These findings were suggestive of constrictive pericarditis and the diagnosis was confirmed by the right heart catheterization. The patient was referred to a thoracic surgeon for pericardiotomy. Pericardial tissue was histologically examined, pointing out plasmacytic infiltration. Immunohistochemistry revealed high concentration of IgG4+ plasmacytes, resulting in our final diagnosis. Therapy started with prednisolone (1 mg/kg of body weight); however, the follow-up ended after 2 weeks due to personal reasons.

CONCLUSIONS Constrictive pericarditis should be considered in cases of unexplained cholestasis. Diagnosis usually relies on careful combination of clinical, laboratory, and ultrasound findings, including a confirmatory right heart catheterization. IgG4-related disease should be included in the differential diagnosis of this type of pericarditis, while tuberculosis and cancer must be ruled out.

Key words
cholestasis, constrictive pericarditis, IgG4

ANNA SAMAKIDOU

Resistant anemia due to copper deficiency: think out of the box

Anna Samakidou[1,2]*, Nikolaos K. Gatselis[1,2], George N. Dalekos[1,2]

1 Department of Medicine and Research Laboratory of Internal Medicine, National Expertise Center of Greece in Autoimmune Liver Diseases, Larissa, Greece
2 European Reference Network on Hepatological Diseases (ERN-Rare Liver), General University Hospital of Larissa, Larissa, Greece

*Corresponding author: anna__sam@hotmail.com

INTRODUCTION Chronic malabsorption syndrome is a disorder than can affect almost any system, with numerous clinical and laboratory presentations. Resistant anemia is one of the most common manifestations. Iron, vitamin B_{12}, and folic acid are the hematopoietic factors primarily evaluated to define the type of deficiency in this situation. Are these enough?

CASE DESCRIPTION An 80-year-old man with a history of resection of the two-thirds of the jejunum and of the ileum due to colon ischemia was admitted to our department for further investigation of refractory anemia, lower limb edema, and fatigue. Clinical examination revealed lower limb edema and paleness. Laboratory workup showed normocytic-normochromic anemia with increased absolute reticulocyte count but with low reticulocyte production index (RPI), increased prothrombin time, hypocalcemia, low albumin level, and hypogammaglobulinemia. Considering that the patient has not so far responded to supplemental therapy with parenteral administration of iron and vitamin B_{12}, and oral folic acid, our differential diagnosis was expanded to investigate other causes or refractory anemia.

Bowel malignancy, celiac disease, pernicious anemia, and nephrotic syndrome were ruled out upon extensive evaluation. Ferritin, vitamin B_{12}, folic acid, and soluble transferrin receptors were within normal limits (for 10 months prior to the evaluation the patient had been receiving oral folic acid, intravenous iron, and intramuscular vitamin B_{12}). The peripheral blood smear did not reveal abnormalities, while bone marrow biopsy revealed reactive marrow with dyserythropoiesis, dysplastic megakaryocytes, and immature myeloid cells, findings suggestive of dysregulation of the bone marrow but not pathognomic of myelodysplastic syndrome. Therefore, during the differential diagnosis we investigated other factors, whose deficiency due to malabsorption could lead to dysregulation of the bone marrow and subsequently to refractory anemia, provided that all the other hematopoietic factors were replaced. Copper is one of such factors. Both serum ceruloplasmin (9 mg/dl; lower limit of normal [LLN], 20 mg/dl) and serum copper levels (30 µg/dl; LLN, 70 µg/dl in men) were low. Replacement therapy started with chelated copper at a daily dose of 5 mg orally. A month after the initiation of therapy, we noticed a significant increase in hemoglobulin levels with increased RPI, while no side effects were reported.

CONCLUSIONS Copper deficiency should be considered in cases of refractory anemia in patients with malabsorption syndrome, especially when all the other hematopoietic factors are within normal limits. The diagnosis relies on laboratory findings, while bone marrow biopsy could be helpful. Replacement therapy is effective, without side effects.

Key words
anemia, copper, malabsorption syndrome

RACHEL SMITH

Successful treatment of calciphylaxis in a renal transplant patient with previous total parathyroidectomy

Rachel Smith[1]*, Naomi Bulteel[2], Annette Alfonzo[2]

1 NHS Grampian, Aberdeen, United Kingdom
2 NHS Fife, Kirkcaldy, United Kingdom

*Corresponding author: rachel.smith4@nhs.scot

INTRODUCTION Calciphylaxis is a rare dermatologic condition strongly associated with chronic kidney disease (CKD). The clinical course can be devastating, with high associated morbidity and mortality. The pathophysiology and optimum treatment remain uncertain. Calciphylaxis is known mostly to affect dialysis patients but is less frequently reported in renal transplant recipients.
CASE DESCRIPTION We report the case of a renal transplant recipient who had undergone previous total parathyroidectomy. Diagnosis was made at the time of limb amputation, yet despite this relatively late diagnosis the patient made a good recovery. We believe this resulted from a multimodal approach including surgical debridement, sodium thiosulphate (STS), and vitamin K_2 therapy. The patient remained free from recurrence 2 years after completion of the treatment, despite ongoing vitamin D replacement. There is still much to ascertain regarding calciphylaxis treatment, including the optimal regimens and duration of therapy, particularly in the transplant recipient population, where data are limited.
LESSONS TO BE LEARNED

- Calciphylaxis is a rare complication of CKD, most often seen in dialysis patients. Its rarity in the transplant population makes diagnosis challenging and may delay treatment.
- Modifiable risk factors in both the dialysis and transplant populations include vitamin K antagonist use.
- Treatments are limited but include wound care, STS, and vitamin K.

Key words
calcific uremic arteriolopathy, parathyroidectomy, renal transplant, sodium thiosulphate, vitamin K_2

APARNA TEWARI

A curious case of hyponatremia

Aparna Tewari[1]*, Ankan Pathak[2]

1 Medical College Kolkata, Kolkata, India
2 Department of Cardiology, Apollo Multispecialty Hospitals, Kolkata, India

*Corresponding author: aparnatewari03@gmail.com

INTRODUCTION Panhypopituitarism is an underdiagnosed entity. As the physicians are often unaware of its existence, the condition remains underdiagnosed for a significant amount of time. The thyroid function often remains normal, which could be attributed to differential effects of various trophic hormones. Possibly the patients retain residual function of the trophic hormones, which is manifested during acute stress.
CASE DESCRIPTION A 50-year-old woman presented to the emergency room with obtundation, severe hypotension, and in acute confusion state. She had no history of a fall, seizure, or fever. The patient had a history of hyponatremia (3 episodes) in the last 6 months, which required hospital admission. She was recently discharged from another hospital with a diagnosis of hyponatremia with acute gastroenteritis. A family member reported 3 spontaneous abortions (the 1st and 2nd trimester). On examination, the patient was febrile and pale but no icterus, rash, palpable lymph nodes, ecchymotic spots, or dehydration were observed. On a systemic examination, no abdomen organomegaly was detected, chest X-ray was unremarkable, S1 and S2 heart tones were audible, bilateral plantar reflex was normal, Glasgow Coma Scale was E4V4M6, pulse 70 bpm, blood pressure 100/60 mm Hg, and capillary blood glucose was 70 mg/dl. Keeping in mind recurrent hyponatremia with a hypotensive state the appropriate laboratory workup was performed.

Keeping in mind the presentation of reduced cortisol with reduced luteinizing hormone, follicle-stimulating hormone, and central hypothyroidism, brain magnetic resonance imaging was ordered and revealed empty sella consistent with hypopituitarism.
CONCLUSIONS Panhypopituitarism is a disease characterized by total or partial deficiency of hormones secreted by the pituitary gland. The etiology may include cranial surgery, radiotherapy, tumors, hereditary causes, infiltrative lesions, infections, and head trauma. The clinical presentation may vary from person to person. It may range from asymptomatic to life-threatening features of adrenal insufficiency. Signs and symptoms of the disease may persist for several years without diagnosis. Proper clinical examination and biochemical evaluation are useful tools in early diagnosis.

Key words
central hypothyroidism, hyponatremia, obtundation, panhypopituitarism, pituitary

EKATERINA TOLKACHEVA

Malaria and acute kidney injury

Ekaterina Tolkacheva*

University of Tartu, Faculty of Medicine, Institute of Clinical Medicine, Tartu, Estonia

*Corresponding author: ekaterina.tolkachev@gmail.com

CASE DESCRIPTION This report presents the case of malaria in a nonendemic region, with acute kidney injury resulting from severe course of the disease. It also gives an overview of certain treatment measures depending on clinical events and pathologic manifestations during the course of the disease. Based on the statistical data of the Estonian Health Board, malaria is a rare disease in Estonia, brought from endemic regions, which makes the presented case particularly interesting for a young specialist. A proper travel history can provide information important for differential diagnosis of the disease. This case shows the importance of considering the characteristics of endemic areas and the clinical findings of common infectious diseases. It also emphasizes the importance of timely diagnostics and the need for cooperation between several treatment centers.

Key words
acute kidney injury, malaria

SARAH VAN OSTAEYEN (*PRESENTED BY VEERLE WIJTVLIET*)

Emphysematous osteomyelitis caused by *Escherichia coli*: a rare cause of septic shock

Sarah Van Ostaeyen*, Veerle Wijtvliet, Walter Verbrugghe

Department of Intensive Care Medicine, Antwerp University Hospital, Antwerp, Belgium

*Corresponding author: sarah.vanostaeyen@student.uantwerpen.be

CASE DESCRIPTION We report a case of a 60-year-old man who presented at the emergency department with septic shock, which during clinical workup turned out to be emphysematous osteomyelitis (EO). EO is a rare disease characterized by bacterial osseous inflammation. EO can be caused by various bacteria, with anaerobic strains and *Enterobacteriaceae* being the most prevalent. Appropriate management through adequate antimicrobial treatment, and in selected cases surgical treatment, is indicated to maximize the chances of survival.

Key words
emphysematous osteomyelitis, septic shock

IREN ZARGARI

A 24-year-old woman presenting with periorbital edema

Iren Zargari[1]*, Nizar Hijazi[2], Michal A. Peleg[2]

1 Department of Internal Medicine B, Lady Davis Carmel Medical Center, Haifa, Israel
2 Rheumatology Unit, Lady Davis Carmel Medical Center, Haifa, Israel

*Corresponding author: irenzargari@gmail.com

INTRODUCTION We report an unusual manifestation of hydroa vacciniforme-like lymphoproliferative disorder (HV-LPD) with predominant periorbital edema. This is the first report in Israel. HV-LPD is a rare cutaneous form of Epstein–Barr virus (EBV)-associated T/NK-cell lymphoproliferative disease, usually affecting Asian and South American children.

CASE DESCRIPTION A 24-year-old Druze woman with Behcet disease, manifested by oral and genital ulcers, erythema nodosum, and arthralgia, presented with left periorbital swelling, purulent rhinitis, and fever. Periorbital cellulitis was diagnosed, secondary to sinusitis. She received antibiotics and dexamethasone for reactive edema.

As there was no improvement, computed tomography (CT), technetium mapping, and magnetic resonance venography were performed, demonstrating left periorbital inflammation without venous thrombosis or osteomyelitis. The swelling was attributed to her inflammatory baseline disease. Prednisone, colchicine, and azathioprine were prescribed. One month later, she presented with bilateral periorbital swelling, along with subcutaneous nodules over the mandible and upper extremities, erythemal chest rash, and oral aphthous ulcers. Anakinra was initiated for a suspected autoinflammatory disease. A genetic evaluation revealed a heterozygous mutation in the *MEFV*, *NLRP12*, and *LACC1* genes. A biopsy of the subcutaneous nodule showed leukocytoclastic vasculitis. Total body CT demonstrated multiple perivascular lung nodules, hepatosplenomegaly, and portal hypertension. Cyclophosphamide was initiated without significant improvement. Re-evaluation of the skin biopsy revealed atypical lymphocytes in clusters, vasculitic changes, and lymphocyte vascular invasion. Immunophenotyping demonstrated CD3+, CD20–, CD4+/–, CD56+/–, CD30+/–, EBV in situ positive, Ki67 –80% in atypical cells, compatible with a diagnosis of HV-LPD. Serum EBV polymerase chain reaction demonstrated 174 000 copies. Bone marrow involvement was not evident.

Following diagnosis, the patient was first treated with valacyclovir and cyclosporine, but her condition continued to worsen, suggestive of an aggressive form of cutaneous T-cell lymphoma. She was then treated with chemotherapy. Despite numerous lines of chemotherapy, her disease continued to progress, and allogeneic stem cell transplantation was planned. Sadly, she developed hemophagocytic lymphohistiocytosis syndrome and transfusion-associated lung injury in the setting of acute infection and succumbed to her illness 9 months following diagnosis.

CONCLUSIONS Apart from being associated with a worse prognosis in HV-LPD patients, periorbital edema can be misdiagnosed, making it an important differential diagnosis in young patients. Cases are rare, and long-term data are lacking. Awareness of such cases promotes further research and treatment guidelines development.

Key words

Epstein–Barr virus, hydroa vacciniforme, T-cell lymphoma

OTHER PRESENTATIONS

SEUNG-MIN BAEK

A case of hereditary hemorrhagic telangiectasia presenting as chronic hypoxemia

Seung-min Baek[1]*, Jong Sun Park[2]

1 Department of Internal Medicine, Seoul National University Bundang Hospital, Seongnam, Republic of Korea
2 Division of Pulmonary and Critical Care Medicine, Department of Internal Medicine, Seoul National University College of Medicine, Seoul National University Bundang Hospital, Seongnam, Republic of Korea

*Corresponding author: 54531@snubh.org

INTRODUCTION Hereditary hemorrhagic telangiectasia (HHT, known as Osler–Weber–Rendu syndrome) is a rare autosomal dominant vascular malformation disorder with an estimated prevalence of 1 in 5000, usually presenting as epistaxis, gastrointestinal bleeding, or mucocutaneous telangiectasia. Especially, arteriovenous malformations (AVMs) are common and frequently affect the pulmonary, hepatic, and / or cerebral circulation. HHT is usually underdiagnosed or the diagnosis is severly delayed. Patients with HHT can develop serious complications, such as severe life-threatening hemorrhage, stroke, or brain abscess.
CASE DESCRIPTION We report a case of a 43-year-old woman referred to an outpatient clinic due to mild dyspnea and decreased oxygen saturation. Her oxygen saturation was 84% on room air. She had mild chronic shortness of breath and frequent epistaxis but those were not disturbing her regular life. Physical examination revealed telangiectasia on the tongue and skin. Chest computed tomography (CT) scan showed multiple, numerous pulmonary AVMs in both upper and lower lung lobes, and contrast echocardiography showed grade III/IV extracardiac shunt. According to the Curaçao clinical criteria, the patient was diagnosed with HHT. A total of 11 coil embolization procedures for pulmonary AVMs were performed, her shortness of breath improved, and oxygen saturation increased.
CONCLUSIONS This case shows that in patients with HHT with diffuse, numerous pulmonary AVMs, multiple coil embolization is feasible and helpful in improving the symptoms. If a patient has hypoxemia, and pulmonary AVMs are visible on chest CT, clinicians should carefully look for the symptoms and signs of HHT, such as epistaxis, gastrointestinal bleeding, mucocutaneous telangiectasia, and other visceral organ AVMs. As HHT can be inherited as an autosomal dominant disease, genetic testing and family genetic counseling are important.

Key words
chronic hypoxemia, coil embolization, hereditary hemorrhagic telangiectasia, Osler–Weber–Rendu syndrome, pulmonary arteriovenous malformations

HERMAN BAGULA

A young adult with leptospirosis-associated acute inflammatory demyelinating polyneuropathy: a case report

Herman Bagula*, Ismail A. Banderker, Muhammed S. Moosa

Department of Medicine, New Somerset Hospital, University of Cape Town, Cape Town, South Africa

*Corresponding author: herman.bagula@gmail.com

INTRODUCTION Leptospirosis is a zoonotic disease that commonly affects the liver and the kidney. It can rarely affect the neurological system with aseptic meningitis being the most common neurological presentation. We present a case of a patient with leptospirosis complicated by acute inflammatory demyelinating polyneuropathy.
CASE DESCRIPTION A 41-year-old man with no significant past medical or surgical history and no known allergies presented to a regional hospital in Cape Town, South Africa, with a 5-day history of nonbloody and nonmucoid diarrhea with associated lower limb weakness. He was hypotensive (86/54 mm Hg) and jaundiced on general examination, with no lymphadenopathy. He had decreased tone in both lower limbs with symmetrical weakness (Medical Research Council grade 2/5), extending from L1 to S1 myotomes. His ankle and patellar reflexes were bilaterally absent, and he experienced a loss of sensation (light touch and pain) extending up to L1 dermatome in both lower limbs. His upper limb power and reflexes were normal, anal sphincter tone was normal, and he had no cerebellar signs. His cardiovascular, respiratory, and abdominal examinations were unremarkable. His blood results showed bicytopenia, acute kidney injury, conjugated hyperbilirubinemia, and mild transaminitis. His international normalized ratio, vitamin B_{12}, and thyroid functions were normal, and serologic testing for viral hepatitis A, B, and C was negative. His sputum geneXpert test (Cepheid, California, United States) was negative for *Mycobacterium tuberculosis*, and his blood cultures were negative. His cerebrospinal fluid showed an elevated protein level of 1.31 g/l (reference range [RR], 0.25–0.35 g/l), glucose of 3.9 mmol/l (RR, 2.75–4.45 mmol/l), polymorphs, 4/µl, lymphocytes, 7/µl, erythrocytes, 25/µl (RR, 0/µl for all 3). The gram stain, bacterial culture, tuberculosis culture, and tuberculosis geneXpert test on his cerebrospinal fluid were negative. Serum immunoglobulin M enzyme-linked immunosorbent assay for *Leptospira* was positive (titer of 3.8). Computed tomography scans of his head, chest, and abdomen were unremarkable, and magnetic resonance imaging of the spine showed no signs of arachnoiditis or other pathologies. Nerve conduction study findings were compatible with sensorimotor polyradiculopathy possibly due to acute inflammatory demyelinating polyneuropathy. He was treated with intravenous ceftriaxone, intravenous fluids, and his complete blood count, renal function, and liver profile were monitored on a regular basis. Furthermore, he received daily physiotherapy, thromboprophylaxis, and was closely monitored for any indication for hemodialysis. He was able to walk by day 2 in the hospital and by the time of discharge, approximately 2 weeks after the admission, his renal function and liver profile had improved significantly.
CONCLUSIONS Neurological manifestations of leptospirosis are rare but should be considered in any patient diagnosed with leptospirosis and presenting with focal neurologic signs. Furthermore, because of its rarity, clinicians should carefully exclude alternative causes for neurologic symptoms and signs in patients with leptospirosis.

Key words
acute inflammatory demyelinating polyneuropathy, leptospirosis, polyneuropathy

ADAM CELER

Successful endoscopic treatment of insulinoma in an elderly patient

Adam Celer*, Michal Krcma

Department of Internal Medicine, Faculty of Medicine in Pilsen, Pilsen University Hospital, Charles University, Pilsen, Czech Republic

*Corresponding author: celera@fnplzen.cz

INTRODUCTION Insulinoma, a tumor belonging to the neuroendocrine tumor (NET) family, is a very rare cause of hypoglycemia. In most cases, the tumors are benign, located in the pancreas, and they can be sporadic or occur in the course of multiple endocrine neoplasia type 1 syndrome. The major symptom of insulinoma is persistent, often severe hypoglycemia. The primary treatment is surgery, although there are also pharmacological procedures, with a number of side effects.
CASE DESCRIPTION An 80-year-old woman with a history of type 2 diabetes mellitus was admitted with recurrent symptomatic hypoglycemia with confusion, syncopes, headache, sweating, and

hand tremor. The patient had no history of hypoglycemia-causing medication (previous treatment with gliptin), liver disease, alcohol intake, adrenal insufficiency, or cancer.

She had high plasma levels of insulin, C-peptide, and low blood sugar level during the 72-hour fasting test. Computed tomography imaging revealed a small tumor 8 mm × 9 mm × 10 mm between the body and tail of the pancreas, without satellite nodules or lymphatic infiltration. The patient was found unfit for radical surgery due to comorbidities. Ethanol ablation was performed during endoscopic ultrasonography with a prompt resolution of hypoglycemia without any need for enteral nutrition or any other treatment. The effect still lasts after 3 months. Biopsy showed tumor cells compatible with a diagnosis of high-grade NET.

CONCLUSIONS This case demonstrates a rare but important cause of severe recurrent hypoglycemia. It shows that a minimally invasive method can be an effective and safe alternative to surgery for the treatment of insulinomas in elderly patients at high surgical risk. Admittedly, data on long-term effectiveness of this approach are scarce.

Key words
endoscopic ablation, hypoglycemia, insulinoma

SHAKIRA DAWOOD

Papillary muscle rupture

Shakira Dawood*

University of Cape Town Medical School, Cape Town, South Africa

*Corresponding author: sdawood1212@gmail.com

INTRODUCTION Papillary muscle rupture is a rare complication of acute myocardial infarction with an excessive mortality rate. Optimal outcomes require a rapid diagnosis and prompt surgical referral. Papillary muscle rupture is a rare but devastating sequela of myocardial necrosis.

CASE DESCRIPTION We report successful treatment of a 51-year-old woman, after a challenging diagnosis of mitral valve regurgitation, resulting from a complete rupture of the posterior papillary muscle. The patient with cardiogenic shock was an emergency admission. Transthoracic echocardiography revealed a prolapsed mitral valve scallop, a tethered posterior mitral valve leaflet with severe posterior directed mitral regurgitation. Immediate coronary angiogram showed changes in the posterior lateral branch of the right coronary artery.

After careful investigation, our patient was diagnosed with acute-on-chronic mitral regurgitation, complicated by posterior papillary muscle and cord rupture secondary to missed inferior myocardial infarction.

She was referred for cardiothoracic surgery, and the mitral valve replacement was performed. It was noted at the surgery that she had anterior mitral valve leaflet prolapse, and ruptured posterior medial papillary muscle. She had an uneventful surgical course and recovered well in the intensive care unit at Groote Schuur Hospital.

Key words
acute complications, acute myocardial infarction, mitral regurgitation, papillary muscle rupture, pulmonary edema

MAXIMILIAAN DE PRETER

Approach to and management of simultaneous invasive nocardiosis, aspergillosis, and *Verruconis gallopava* central nervous system infection

Maximiliaan De Preter[1,2*], Heleen Van Mieghem[1], Eric Florence[1,3]

1 University Hospital of Antwerp, Antwerp, Belgium
2 Faculty of Medicine and Health Sciences, University of Antwerp, Antwerp, Belgium
3 Institute of Tropical Medicine, Antwerp, Belgium

*Corresponding author: maximiliaan.depreter@uza.be

CASE DESCRIPTION A 43-year-old heart transplant recipient who had undergone transplantation 6 months before was admitted to the hospital after a routine endomyocardial biopsy with fever, cough, and dyspnea. Multisensitive *Candida albicans* was cultured in blood, *Aspergillus fumigatus* complex (AFC) was cultured in bronchoalveolar lavage (BAL), and computed tomography (CT) of the thorax showed bilateral lung consolidations with cavitations suggestive of invasive pulmonary aspergillosis. Despite therapy with voriconazole and caspofungin, the patient deteriorated and redeveloped fever and increasing respiratory distress. *Nocardia nova* was cultured in the second BAL sample, and trimethoprim-sulfamethoxazole (TMP-SMX) was added. However, recurrent fever was present. Additional cerebral imaging was requested to investigate possible invasive cerebral nocardiosis. CT and magnetic resonance imaging of the brain revealed 3 focal hypointense lesions with a thick hyperintense rim and surrounding parenchymal edema, suggestive of cerebral abscesses. The patient had no neurological complaints at that time. Stereotactic brain biopsy was performed. Meropenem, amphotericin B, and caspofungin were added empirically to cover cerebral nocardiosis and aspergillosis. After a few weeks, an unusual fungus called *Verruconis gallopava* was cultured in the biopsy specimen. Only a few case reports exist that describe *V. gallopava* as a cause of disseminated disease with cerebral abscesses in immunosuppressed patients. AFC and *N. nova* were not found on the brain biopsy.

Therefore, selecting an appropriate antimicrobial regimen proved to be difficult. Additionally, the AFC fungiogram was panresistant to all available azoles. Achieving adequate central nervous system (CNS) penetration is essential for the treatment of cerebral abscesses. While there are sufficient data to show that certain antibiotics have excellent tissue penetration, only limited data exist about the CNS penetration of antifungal agents. The isolated AFC was not sensitive to voriconazole, which the recommended first-line therapy for invasive aspergillosis proposed by the Infectious Diseases Society of America. Ultimately, TMP-SMX, meropenem, posaconazole, liposomal amphotericin B, and caspofungin were chosen as antimicrobial therapy. The patient responded remarkably well and subsequent brain imaging showed clear partial regression of the abscesses. However, multiple side effects of his therapy occurred, including acute kidney insufficiency (requiring temporary hemodialysis), pancytopenia, and microangiopathic hemolytic anemia. Amphotericin B, TMP-SMX, and tacrolimus were regarded as suspected culprits and promptly discontinued. TMP-SMX was replaced by ceftriaxone and minocycline, and tacrolimus was substituted with sirolimus. The patient recovered and was put on a long-term ambulatory therapy.

LESSONS TO BE LEARNED Learning points were ample in this complex case. Symptoms of opportunistic and invasive disease are often nonspecific or absent due to underlying immunosuppression. Cerebral abscesses can be present even without neurologic symptoms. The isolation of certain pathogens, such as *N. nova* and AFC should prompt further investigation to identify disseminated disease, particularly the CNS involvement. Selecting an appropriate antimicrobial regimen is often difficult and depends on the isolated pathogens, their in vitro susceptibility pattern, involved infection sites, and required tissue penetration. Finally, special consideration should be given to possible drug interactions and side effects. When complications arise during therapy, suspected culprit drugs should be discontinued and replaced.

Key words
aspergillosis, cerebral abscess, heart transplant, nocardiosis, *Verruconis gallopava*

JANINA DI PIETRO

Acute leukemia of ambiguous lineage: a case report

Janina Di Pietro[1,2]*, Veronica Canosa[1,2], Patricia Gisbert[1,2]

1 Medical Clinic Service, Lagomaggiore Hospital, Mendoza, Argentina
2 Faculty of Medical Sciences, National University of Cuyo, Mendoza, Argentina

*Corresponding author: janina.dipietro@gmail.com

INTRODUCTION Mixed or ambiguous lineage acute leukemia (MPAL), is a rare disease that represents 2% to 5% of all leukemias. It is characterized by the associated expression of 2 or more markers of different blast cell lines. This pathology has a poor prognosis, so there is little information regarding its management.

CASE DESCRIPTION A 41-year-old white man, with a history of G3 obesity, hepatic steatosis, and type 2 diabetes mellitus, presented with 4-day long progressive dyspnea, cystic fibrosis grade II-III, associated with headache, asthenia, nasal congestion, odynophagia, and fever. Laboratory tests showed pancytopenia, with critical neutropenia, so it was interpreted as febrile neutropenia. The patient was hemodynamically stable with generalized hypoventilation. The ocular examination showed multiple intraretinal hemorrhages. Hemogram performed by a hematologist revealed hematocrit of 19.5%, hemoglobin level 6.6 g/dl, platelet count 12 000/mm^3, white blood count 1910/mm^3, blasts 48/mm^3. Abdominal ultrasound showed mild to moderate liver steatosis, a partially calcified cyst in segment 7, spleen size of 148 mm. Chest computed tomography (CT) showed no evidence of infiltrates. On admission, piperacillin-tazobactam was administered empirically after blood and urine culture. Afterward, a bone marrow biopsy (BMB) was performed that confirmed leukemia of ambiguous lineage. Cytometry detected a population of myeloid and T / myeloid blasts. Monocytic series expressed CD56, and granulocytic series had altered CD1b/CD13/CD16 differentiation patterns. Genetic tests revealed karyotype 46 XY, no mutations in the *CEBPA*, *MLL*, *BCR/ABL*, *FLT3 ITD*, or *FLT3 D835* genes, and mutated *NPM 1* gene.

Considering the diagnosis and blood test results, hyper-CVAD chemotherapy was started. Fever reoccurred, and another chest CT scan showed bilateral ground-glass infiltrates with a tendency to consolidation. In addition, the patient presented nasal congestion and headache in the frontal lobe, for which CT of paranasal sinuses was performed, revealing sinus polyposis involving maxillary sinuses, ethmoidal cells, frontal sinuses, and sphenoidal sinus bilaterally, involvement of the ostium, mucosal hypertrophy of polypoid appearance with osteolysis of the nasal turbinates. Anterior rhinoscopy demonstrated erythematous, thickened, and heterogeneous mucosa. Samples were taken for the culture of *Fusarium solani*, for which the patient received treatment, and during hospitalization he presented multiple infectious recurrent lesions that were treated according to microbiologic findings.

After no response to hyper-CVAD, the IDA-FLAG regimen was introduced. Sinusitis due to *F. solani* reoccurred and was treated accordingly.

After a cycle of IDA-FLAG, BMB was repeated and demonstrated blasts with myeloid differentiation 8.5% of medium size and moderate internal complexity. Due to the lack of response and poor clinical evolution, supportive and palliative care was implemented and maintained until death 10 months after diagnosis.

CONCLUSIONS MPAL is a rare type of presentation, of which there is limited literature, with uncertain course and a poor prognosis aggravated by infections with opportunistic fungal strains, such as *Fusarium*. Therefore, diagnostic alertness, correct classification, and appropriate therapy are essential to provide better care to patients with this disease.

Key words
acute leukemia of mixed or ambiguous lineage, *Fusarium*

TOM ELMEDVI

A 61-year-old woman presenting with fever of unknown origin and elevated lactate dehydrogenase values

Tom Elmedvi[1,2]*, Anat Arbel[1,2], Ibrahim Zoubi[1,2]

1 Department of Internal Medicine B, Lady Davis Carmel Center, Haifa, Israel
2 Rappaport Faculty of Medicine and Research Institute, Technion Institute of Technology, Haifa, Israel

*Corresponding author: tmelmedvi@gmail.com

CASE DESCRIPTION A 61-year-old woman was admitted to our department for investigation of fever of unknown origin (FUO) accompanied by elevated lactate dehydrogenase (LDH) and liver enzyme levels over a period of 3 weeks.

Upon admission the patient's physical examination revealed no findings. Her laboratory tests showed normal complete blood count and elevation of cholestatic liver enzymes as well as LDH (up to 3000 U/l; reference range, 140–280 U/l) and C-reactive protein levels, and erythrocyte sedimentation rate. Her chest X-ray was unremarkable and abdominal ultrasonography revealed liver steatosis. Chest and abdomen computed tomography (CT) scan revealed nodules and ground-glass opacities in the lungs and splenomegaly.

At this point, the differential diagnosis was still broad. Further workup was guided by clinical clues. To address the liver enzyme elevation, we performed liver biopsy. To investigate the constellation of elevated LDH level and splenomegaly, despite the lack of lymphadenopathy, we performed bone marrow biopsy.

During her hospitalization, the patient gradually developed dyspnea and desaturation. Pulmonary function tests revealed low diffusion capacity. Chest CT angiography was negative for pulmonary embolism. Bronchoscopy with transbronchial biopsy was carried out.

Later, bone marrow, liver, and transbronchial biopsy results demonstrated intravascular infiltrates of atypical lymphoid cells with high mitotic rate, suggesting the diagnosis of intravascular large–B-cell lymphoma (IVLBCL). An urgent positron emission tomography-CT scan revealed disseminated disease involving bone marrow, lungs, liver, spleen, and kidneys.

R-CHOP (rituximab, cyclophosphamide, vincristine, prednisone) regimen was initiated immediately, resulting in marked clinical and laboratory improvement.

DISCUSSION IVLBCL is a rare lymphoma entity characterized by predominant growth of large cells within the lumen of different-sized blood vessels. The spectrum of its clinical presentation is heterogeneous, ranging from monosymptomatic to polysymptomatic forms, with FUO being by far the most common symptom.

In this particular case, we faced some unique challenges on our way to diagnosis. First, the differential diagnosis was broad and no single disease could account for all findings. Second, despite elevated LDH level, the lack of lymphadenopathy rendered the diagnosis of lymphoproliferative disease less probable. Finally, common organ involvement (eg, skin and central nervous system) and laboratory abnormalities (eg, anemia, leukopenia, and thrombocytopenia) typical of IVLBCL were absent in our patient.

LESSONS TO BE LEARNED

- The clinical presentation of IVLBCL is heterogenous and often mimics other more common conditions.
- In the case of a complex presentation with multiorgan involvement, it is crucial to adopt a systematic approach.
- Upon histopathologic diagnosis, IVLBCL must be considered as an aggressive disseminated disease and therapy should be initiated immediately.

Key words
fever, intravascular, lactate dehydrogenase, lymphoma

RAMONA GOVENDER

RECOGNIZED FOR BEST POSTER

Refractory adult-onset Still disease with arthritis mutilans

Ramona Govender*, Bridget Hodkinson

Division of Rheumatology, Groote Schuur Hospital, Cape Town, South Africa

*Corresponding author: ramona.govender3@gmail.com

CASE DESCRIPTION Adult-onset Still disease (AOSD) is a rare polygenic autoinflammatory disorder associated with 2 major phenotypes a predominantly systemic subtype and a chronic articular subtype. Arthritis can become severe and progress to erosive disease with ankylosis, leading to significant functional impairment and disability. We present a patient with a refractory chronic course and predominantly articular involvement evolving into arthritis mutilans, despite stepwise escalation of the therapy with conventional synthetic disease-modifying antirheumatic drugs and biologic agents.

Key words

arthritis mutilans, autoinflammatory disease, chronic articular subtype, refractory adult-onset Still disease

BINNAZ İĞRET

Fatal complication during vaso-occlusive pain crisis in sickle cell disease: thrombotic microangiopathy

Binnaz Igret*, Sukriye M. K. Bozgul, Devrim Bozkurt

Faculty of Medicine, Department of Internal Medicine, Ege University, Izmir, Turkey

*Corresponding author: binazigret@gmail.com

INTRODUCTION Thrombotic microangiopathy (TMA) with multiorgan involvement has been described in patients with sickle cell disease (SCD) as a rare complication of unknown etiology. Here, we report a patient with vaso-occlusive crisis (VOC) of SCD complicated by TMA and treated with eculizumab, a monoclonal antibody of C5, which blocks terminal complement activation.

CASE DESCRIPTION The patient was a 31-year-old man previously diagnosed with sickle cell β-thalassemia who was on hydroxyurea. He was admitted to an intensive care unit (ICU) due to severe chest and abdominal pain. He received red cell exchange (RCE) transfusion as a result of vaso-occlusive pain crisis. After RCE, his pain decreased but he developed respiratory distress. At the 36th hour, liver and kidney were impaired on functional tests, lactate dehydrogenase (LDH) and indirect bilirubin increased, thrombocytopenia developed, the amount of urine output decreased, and schistocytes were observed in the peripheral smear. Immune function tests and the direct Coombs test were negative. The percentage of CD55 and CD59 erythrocytes in total erythrocyte pool was below 0.5% each. The preliminary diagnosis was thrombotic microangiopathy (TMA). Therapeutic plasma exchange (TPE) and hemodialysis (HD) and were initiated on the 2nd day. On the 3rd day of the ICU follow-up, the patient was intubated due to increased respiratory distress. TPE and HD were continued due to acute kidney injury with anuria. On the 3rd day, 900 mg of eculizumab was administered after ADAMTS-13 activity was found to be normal with the diagnosis of complement system mediated hemolytic uremic syndrome. Liver functional tests, LDH, pulmonary infiltrates, and lactate level regressed 3 days after implementation of eculizumab treatment. On the 10th day, a second dose of eculizumab was administered. The patient received a total of 3 sessions of TPE and 6 sessions of HD. On the 14th day, the patient was conscious and cooperative, and he was extubated. During the follow-up, urine output started. However, septic shock developed and the patient died on the 16th day of the follow-up.

DISCUSSION Complement system alternative pathway activation in SCD may result from the effect of hem on endothelial surfaces or from the effect of changes in erythrocyte membrane phospholipids. In addition, overreactive, ultralarge von Willebrand factor (VWF) multimers released from damaged endothelial cells or caused by inability of ADAMTS-13 to cleave hemoglobin-bound VWF may contribute to the pathogenesis of TMA in SCD. In this report, we want to emphasize the importance of early diagnosis of TMA in SCD, as early appropriate treatment can prevent chronic multiorgan damage and death.

CONCLUSIONS Clinicians, especially internal medicine specialists should be aware of TMA when hemolytic anemia, thrombocytopenia, altered consciousness, liver and kidney dysfunction develop during VOC of sickle-cell anemia. TPE should be started immediately. Multiple organ failure and death can be prevented with early eculizumab treatment.

Key words

atypical hemolytic uremic syndrome, sickle cell disease, thrombotic microangiopathy

JI-YOON IM

Amyloid light-chain amyloidosis initially presenting as isolated tricuspid regurgitation

Ji-Yoon Im[1]*, Seung-Ah Lee[2], Hee-Sang Hwang[3]

1 Department of Internal Medicine, Asan Medical Center, University of Ulsan College of Medicine, Seoul, Republic of Korea

2 Department of Cardiology, Asan Medical Center, University of Ulsan College of Medicine, Seoul, Republic of Korea

3 Department of Pathology, Asan Medical Center, University of Ulsan College of Medicine, Seoul, Republic of Korea

*Corresponding author: imji0818@gmail.com

INTRODUCTION Early diagnosis is key to managing amyloid light-chain (AL) amyloidosis, but vague symptoms and signs make early recognition challenging. Here we report a difficult case of AL amyloidosis initially assessed as isolated tricuspid regurgitation (TR).

CASE DESCRIPTION A 49-year-old man was referred to our hospital complaining of bilateral leg edema and dyspnea for 5 months. Transthoracic echocardiography (TTE) revealed severe eccentric TR with annulus dilation. The systolic function of both ventricles was normal. Poorly responsive to diuretics, the patient proceeded to video-guided endoscopic tricuspid annuloplasty for isolated TR. We expected the symptoms to improve, but he still had abdominal bloating and leg edema. Postoperative TTE revealed right ventricular (RV) dysfunction, although TR was corrected. To identify the cause of RV dysfunction, we reviewed preoperative images. Interestingly, noncontrast T1 mapping values of cardiac magnetic resonance imaging were diffusely increased in the whole myocardium, suggesting early fibrosis. Based on this, images of late gadolinium enhancement were thoroughly reevaluated. Diffuse late gadolinium enhancement was identified in the right heart, including the tricuspid valve and the left atrium, suggesting infiltrative cardiomyopathy. Serum and urine protein electrophoresis with immunofixation showed monoclonal α free-light chain (FLC). Amyloid deposits were confirmed on myocardial and random stomach biopsies; thus, AL amyloidosis at cardiac stage 3 (troponin T, 0.093 ng/ml; brain natriuretic peptide [BNP], 666 pg/ml; N-terminal pro–B-type natriuretic peptide, 5275 pg/ml) was diagnosed. After 4 cycles of bortezomib, thalidomide, and dexamethasone, the differential FLC level decreased, but the BNP level was still high and symptoms related to the right heart failure persisted. Heart transplantation was carried out for advanced cardiac amyloidosis. The patient resumed outpatient chemotherapy. He was asymptomatic and had no signs of the organ rejection. One year after the heart transplantation, he underwent sequential autologous stem cell transplantation.

CONCLUSIONS AL amyloidosis can involve the valve first and present as isolated TR. Considering the rapid progression and poor prognosis of the disease, clinicians should keep amyloidosis in mind

as an etiology of heart failure even without prominent ventricular wall thickening.

Key words
amyloidosis, disease awareness, tricuspid regurgitation

SUNGGU KIM

RECOGNIZED FOR BEST POSTER

A case of cervical spinal gout with meningitis-like symptoms

Sunggu Kim[1], Jeong Rae Yoo[1,2]*

1 Department of Internal Medicine, Jeju National University Hospital, Jeju, Republic of Korea
2 Department of Internal Medicine, Jeju National University College of Medicine, Jeju, Republic of Korea

*Corresponding author: mdyoojr@gmail.com

INTRODUCTION Gout is characterized by the deposition of monosodium urate (MSU) crystals and by acute and chronic inflammation in response to the deposited crystals. Deposition of MSU in the spine is a rare manifestation of gout, but the actual prevalence of spinal involvement is unknown. We present a case of vertebral osteomyelitis and spinal meningitis misdiagnosed as cervical spinal gout.

CASE DESCRIPTION An 85-year-old man was admitted due to a 5-day history of fever and neck pain. On admission, physical examination revealed neck pain and stiffness. His blood pressure was 140/102 mm Hg, pulse rate of 95 bpm, respiration rate of 18 breaths/min, and body temperature 38 °C. Laboratory workup revealed white blood cell count (WBC) of 11 200/mm^3 (reference range [RR], 4000–10 000) and C-reactive protein level of 12.26 mg/dl (RR, 0–0.3). Magnetic resonance imaging findings suggested cervical osteomyelitis and meningitis. Cervical spine computed tomography (CT) revealed curvilinear calcifications of the transverse ligament of the atlas. After assessing blood cultures, empirical antibiotics were administered intravenously. On the 2nd day of admission, cerebrospinal fluid (CSF) analysis did not reveal meningitis (WBC 8/µl; glucose and protein levels 79 and 168 mg/dl, respectively), and CSF culture and molecular tests were negative for bacteria and mycobacteria. The patient was diagnosed with crowned dens syndrome, received 20 mg of prednisolone, and his symptoms dramatically improved. However, CSF examination under a polarizing microscope revealed MSU crystals, and dual-energy CT confirmed the calcifications to be MSU crystals.

DISCUSSION Crowned dens syndrome is a rare finding in calcium pyrophosphate deposition disease, and may be clinically similar to meningitis, so it should be suspected when the evidence for infection is unclear. Our patient had gout history, and MSU crystals were confirmed by CSF analysis and dual-energy CT, suggesting that the symptoms were caused by gout flare in the cervical spine.

CONCLUSIONS Physicians should consider that intrathecal MSU crystal formation can cause meningitis-like symptoms in patients with gout history.

Key words
meningitis, monosodium urate crystal, spinal gout

FABIAN KOSKO

Co-occurrence of granulomatosis with polyangiitis and lung carcinoid tumor

Fabian Kosko*, Piotr Skonieczny, Alicja Dębska-Ślizień

Department of Nephrology, Transplantology and Internal Medicine, Medical University of Gdansk, Gdańsk, Poland

*Corresponding author: fkosko@gumed.edu.pl

INTRODUCTION Granulomatosis with polyangiitis (GPA) is a rare systemic disease, which is classified as a type of primary systemic vasculitis and affects multiple organs. Pulmonary carcinoid tumors are also uncommon entities that account for less than 1% of all lung cancers. They derive from neuroendocrine cells and are divided into 2 subcategories: typical and atypical carcinoids.

Here we discuss a case of co-occurrence of these 2 unusual entities and describe difficulties with diagnosis and treatment.

CASE DESCRIPTION A 35-year-old man, with no previous medical history except for smoking, initially presented to a hospital with acute kidney injury (AKI). Immunologic tests showed high titers of antineutrophil cytoplasmic antibodies (ANCA). Chest computed tomography (CT) scan showed infiltration of the inferior lobe of the left lung. Due to clinical picture of rapidly progressive glomerulonephritis, lung lesions on imaging studies, and ANCA positivity, GPA was diagnosed and the patient received high doses of methylprednisolone, 1 dose of cyclophosphamide (CP), and due to still rising serum creatine level, renal replacement therapy was initiated. Due to unclear character of the lung lesions bronchoscopy was performed and revealed the presence of obturating endobronchial mass in the left lower lobe. Unexpectedly, a biopsy showed features of typical carcinoid tumor.

Then the patient was admitted to our department for further diagnosis and treatment. During the hospitalization, he received another pulse of CP, and a permanent catheter for hemodialysis was inserted. High-resolution chest CT confirmed the pathologic mass corresponding to a carcinoid tumor and showed regression of inflammatory lesion from the lower lobe of the left lung in comparison with previous CT. Finally, a multidisciplinary medical team evaluated the patient, and he was qualified for sleeved lobectomy of the lower lobe of the left lung with lymphadenectomy. The histopathologic specimen of the lung tissue confirmed features of typical carcinoid and fortunately there were no metastases in the lymph nodes.

The treatment is continued as 1-day-long hospitalizations at our department and the patient remains dialysis-dependent.

DISCUSSION Diagnosis of GPA is difficult and a coexisting carcinoid tumor makes it even more challenging. According to the available literature, there has been only 1 case described where these 2 diseases occurred together. However, there have been 2 cases reported where carcinoid coexisted with giant cell arteritis. Moreover, vasculitis was described as a possible complication of COVID-19. Treatment of a patient suffering from those 2 entities requires cooperation of a multidisciplinary medical team. Additionally, immunosuppressive drugs make the surgery more prone to complications, such as infections or impaired wound healing.

CONCLUSIONS Co-occurrence of lung carcinoid tumor and GPA, especially in a young patient, makes diagnosis difficult. The treatment consists of immunosuppressive drugs and surgery, which might be complicated by infections or impaired wound healing. So far, there have been only 3 cases of coexisting vasculitis and carcinoid tumors described in the available literature. Thus, it is important to report more such cases to increase awareness of the physicians.

Key words
granulomatosis with polyangiitis, lung carcinoid tumor

IVETA KROUPOVÁ

Hepatopulmonary syndrome as a rare complication of liver cirrhosis

Iveta Kroupová*, Kateřina Bláhová

Znojmo Hospital, Znojmo, Czech Republic

*Corresponding author: iveta.kroupova@nemzn.cz

INTRODUCTION This case report focuses on hepatopulmonary syndrome, a serious and rare complication of advanced liver disease. This syndrome manifests itself by functional intrapulmonary

shunt caused by intrapulmonary vascular dilatations resulting in ventilation-perfusion mismatch and hypoxemia.

CASE REPORT A 66-year-old woman with liver cirrhosis (Child-Pugh B) of combined (autoimmune and nonalcoholic fatty liver disease) etiology, on a waiting list for liver transplant, presented to our hospital with dyspnea. Clinical examination revealed hypoxemia, central cyanosis, and lower limb edema. Chest high-resolution computed tomography and spirometry did not explain these clinical findings. Lung perfusion scan was performed with no signs of pulmonary embolism; however, it demonstrated an abnormal accumulation of the radiopharmaceutical in the organs supplied with the systemic circulation. This finding made us suspect a right-to-left shunt. The diagnosis was confirmed by contrast transthoracic echocardiography. Severe hypoxemia required oxygen therapy to improve the symptoms. Hospital stay was complicated by SARS-CoV-2 infection, which, though with mild overall symptoms, led to progression of the liver failure. After 1 month the patient was discharged to home care with an oxygen concentrator. Due to the abovementioned complications the patient was withdrawn from the liver transplant waiting list and later died of terminal liver failure.

DISCUSSION Dyspnea is a common but nonspecific finding in many pathologic conditions accompanying liver failure. They can be distinguished by clinical examination and imaging modalities. Severe hypoxemia in the absence of coexisting cardiopulmonary disease in this case is strongly suggestive of hepatopulmonary syndrome. Conservative treatment involves symptomatic oxygen therapy, and only liver transplant is to provide long-term survival benefits.

CONCLUSIONS Correct recognition and early diagnosis of this syndrome are crucial. Patients with hepatopulmonary syndrome receive additional points to their model for end-stage liver disease score and are prioritized at the liver transplant waiting list. In some cases (as in our patient), the clinical condition caused by the syndrome can easily exhaust the patient's physical reserves, and make them ineligible for such a serious intervention as the liver transplant undoubtedly is. In such cases, we must make the difficult choice and ask ourselves if the transplantation is likely to be successful or if we rather save the limited resources for more suitable candidates.

Key words
hepatopulmonary syndrome, hypoxemia, liver failure, right-to-left shunt

ARITRA KUMAR RAY

A rare case of altered sensorium in a young man

Aritra Kumar Ray[1]*, Soumitra Ghosh[2], Sk Tahsin Islam[2]

1 Department of General Medicine, Kharagpur Subdivisional Hospital, West Bengal, India
2 Institute of Post Graduate Medical Education & Research, Kolkata, India

*Corresponding author: aritra121@gmail.com

INTRODUCTION Meningitis is a disease that causes grave complications if its diagnosis and treatment are delayed. It is usually caused by bacteria, viruses, and fungi. Parasites are rare etiologic agents of meningitis and *Acanthamoeba*-caused meningitis is even rarer. *Acanthamoeba* is a free living ameba common in freshwater bodies. It may cause cutaneous lesions, granulomatous encephalitis, and keratitis. Here we report a rare case of Acanthamoeba-related meningitis.

CASE DESCRIPTION A 33-year-old nondiabetic, normotensive young adult, migrant laborer by occupation, presented with complaints of low- to moderate-grade fever with headache and nausea for 14 days. Fever was intermittent in nature and associated with myalgia. The patient also had a history of altered sensation for the last 7 days prior to hospitalization. On admission, he was febrile and appeared confused, but had stable hemodynamic parameters. On detailed neurologic assessment, he was found to have stiff neck with positive Kernig and Brudzinski signs. Both pupils were mid-dilated and sluggishly reacting to light. Keeping in mind the clinical scenario, neuroimaging of the brain and cerebrospinal fluid (CSF) sampling were planned. The CSF examination revealed low glucose and raised protein levels with a large number of trophozoites of *Acanthamoeba* species. On further enquiry, the patient admitted to having taken frequent baths in the water bodies of Kerala for the last couple of months. He was put on rifampicin, trimethoprim sulfamethoxazole, fluconazole, and miltefosine to which he promptly responded. His sensation improved within the next few days with gradual subsidence of fever and headache.

DISCUSSION *Acanthamoeba* is usually found in freshwater bodies, such as lakes, rivers, and swimming pools. It is transmitted either by inoculation through skin lesions or by inhalation. Immunocompromised individuals or patients on chemotherapy are more prone to develop *Acanthamoeba* infection. Though the patient presented in this case report was immunocompetent, he was probably infected while bathing in the ponds. *Acanthamoeba*-related meningitis can be diagnosed using the Wright-Giemsa stain or the CSF wet mount. It can also be cultured in non-nutrient agar with overlay of *Enterobacter aerogenes* or *Escherichia coli*. Though there are no standard treatment regimens, studies have shown good outcome with a combination of trimethoprim sulfamethoxazole, rifampicin, and ketoconazole.

CONCLUSIONS It is of utmost importance to identify *Acanthamoeba* infection of the central nervous system (CNS), as early diagnosis and treatment prevent serious complications and reduce morbidity and mortality. Misdiagnosis with other infectious diseases is very common and physicians should be aware of *Acanthamoeba* as a potential agent causing CNS infections.

Key words
Acanthamoeba species, meningitis, immunocompetent, water bodies

MELVIN LEE YOONG ZHER

RECOGNIZED FOR BEST POSTER

Nonislet-cell tumor hypoglycemia: a rare cause of persistent hypoglycemia

Melvin Lee Yoong Zher*

Department of Diabetes and Endocrinology, Division of Internal Medicine, Pinderfields General Hospital, Wakefield, United Kingdom

*Corresponding author: melvinleeyz@gmail.com

INTRODUCTION Nonislet-cell tumor hypoglycemia (NICTH) is a rare cause of hypoglycemia. There is no single pathogenic mechanism that can explain all cases of NICTH, which makes this condition difficult to treat.

CASE DESCRIPTION A 59-year-old man with known metastatic prostate adenocarcinoma presented with symptoms of hypoglycemia with blood glucose level of 1.8 mmol/l (reference range, 4–7 mmol/l). He received emergency treatment for his hypoglycemia, however the hypoglycemic episodes recurred persistently. Other glucose-stabilizing treatments were initiated, such as dexamethasone, octreotide injections, and diazoxide. They only had a transient effect on maintaining euglycemia. Serum C-peptide, insulin, and urine sulfonylurea levels tested during one of the hypoglycemic episodes revealed that the hypoglycemia was nonhyperinsulinemic and exogenous in origin. The patient had an elevated insulin-like growth factor (IGF)-2/IGF-1 ratio, suggesting that the hypoglycemia could be due to NICTH. His hypoglycemia persisted and the patient unfortunately succumbed 10 days thereafter.

CONCLUSIONS NICTH is a rare and serious complication of malignancy. The efficacy of medical therapies for this condition has not been well established. With this case we wish to highlight the complexity in diagnosis and management of this condition.

Key words
hypoglycemia, insulin-like growth factor 2, nonislet-cell tumor hypoglycemia

TETYANA MALSKA

Leontiasis ossea

Tetyana Malska*, Liliya Martynyuk

Third Department of Internal Medicine, I. Horbachevsky Ternopil National Medical University, Ternopil, Ukraine

*Corresponding author: malysyka_tleo@tdmu.edu.ua

INTRODUCTION Leontiasis ossea, also known as leontiasis or lion face, is a very rare syndrome characterized by face deformity due to overgrowth of the facial and cranial bones. Incidence of this syndrome in chronic kidney disease is even less frequent, probably due to dialysis and medical advances in the treatment of secondary hyperparathyroidism. We present the case of a 56-year-old woman on hemodialysis who developed leontiasis ossea.

CASE DESCRIPTION A 56-year-old woman on ambulatory hemodialysis for 4 years experienced during the last 6 months mouth discomfort, hypertrophy of the oral mucosa, and difficulties with speaking and swallowing of food. She was treated by a dental surgeon; resection of the oral mucosa was performed several times. The treatment results were unsuccessful.

X-ray revealed hyperostosis of the maxilla and mandible bones. Biochemical analyses showed serum calcium level consistently within the normal range (NR) (between 2.28 and 2.54 mmol/l) but exceeding the recommended target level of 2.1 mmol/l, serum phosphate was elevated between 2.25 and 2.75 mmol/l (NR, 0.79–1.49 mmol/l). The patient also had elevated alkaline phosphatase of 998–1250 U/l, (NR, 34–160 U/l), and intact parathormone (iPTH) between 1185–5300 pg/ml (NR, 10–69 pg/ml). Ultrasound revealed enlarged adenomatic parathyroid glands. The patient was treated with calcium-based phosphate binders. She was noncompliant with phosphate-lowering medications and active metabolites of vitamin D treatment. At that time, calcimimetics were not available in our country.

Parathyroidectomy was performed without any complications and side effects during intra- and postoperative period. The patient was prescribed a low-phosphate diet. Alphacalcidol was administered orally at 0.5–0.75 mg, 3 times per week after the hemodialysis session. After 6 months the patient's condition improved, there was no prominent discomfort in the mouth, and no mucosal hypertrophy. Calcium, phosphorus, and PTH levels were within target ranges.

DISCUSSION Leontiasis ossea with craniofacial bone lesions is often a difficult diagnostic dilemma in patients with end-stage renal disease (ESRD). Such diseases as fibrous dysplasia, giant cell tumor, Paget disease, or ossifying tumors are more common reasons of this syndrome, and may have similar radiologic picture. Anamnestic, clinical, and laboratory data may improve the accuracy of diagnosis. Our patient had a history of ESRD and poorly controlled mineral metabolism with elevated serum phosphorus, alkaline phosphatase, and iPTH levels. According to our data, it is possible that the patient has developed tertiary hyperparathyroidism, which requires surgical treatment. The presented case report highlights the importance of early diagnosis and treatment of secondary hyperparathyroidism. The data confirm the idea of early control of mineral metabolism in hemodialysis patients with phosphate binders, active vitamin D metabolites, and calcimimetics.

CONCLUSIONS Nowadays leontiasis ossea is a rare medical syndrome in ESRD patients. However, it may develop in noncompliant patients with poor control of mineral metabolism and secondary hyperparathyroidism.

Key words
leontiasis ossea, secondary hyperparathyroidism, end-stage renal disease

MAHLATSE MANKGELE

An initially missed diagnosis of a venous thromboembolic event in adult-onset Still disease: a case report and literature review

Mahlatse Mankgele[1]*, Delisa Hlawe[2], Mpoti Seboka[3]

1 Department of Clinical Haematology, Groote Schuur Hospital, Cape Town, South Africa
2 Division of Internal Medicine, Faculty of Health Sciences, School of Clinical Medicine, University of Witwatersrand, Johannesburg, South Africa
3 Department of Rheumatology, Charlotte Maxeke Johannesburg Academic Hospital, Johannesburg, South Africa

*Corresponding author: mahlatsemankgele55@gmail.com

INTRODUCTION Still disease is a rare multisystemic autoinflammatory disorder. The diagnosis of adult-onset Still disease (AoSD) can be challenging due to its rarity and features overlapping with many other systemic disorders. One of the least documented hematologic complications of AoSD are thromboembolic events.

CASE DESCRIPTION We present a case of a 43-year-old woman with a known diagnosis of AoSD, whose disease-modifying antirheumatic drugs (DMARDs) had been tapered and stopped due to low disease activity. She presented with respiratory symptoms and features of an AoSD flare. As she did not completely improve on antibiotic therapy and reinitiated DMARDs, we sought an alternative/concurrent diagnosis. The workup yielded pulmonary embolism without other risk factors for thrombosis.

CONCLUSIONS In the reviewed literature, there is a close association between hyperferritinemia and AoSD complicated with venous thromboembolisms (VTE). A rigorous search for alternative diagnoses should be performed and other potential uncommon complications of AoSD should be kept in mind when assessing patients with AoSD, especially those who are not improving on therapy. Given the rarity of AoSD, meticulous data collection may be useful in gaining understanding of the pathophysiology and features of the disease presentation, including complications such as VTE.

Key words
adult-onset, Still disease, thrombosis

LUCÍA B. MOLINERO

RECOGNIZED FOR BEST POSTER

Hepatic injury induced by "cow's hoof"

Lucía B. Molinero*, Andrés Serra, Roberto Parodi

Internal Medicine Division, Hospital Provincial del Centenario, National University of Rosario, Rosario, Santa Fe, Argentina

*Corresponding author: lucia.molinero94@gmail.com

INTRODUCTION The use of herbs is wrongly considered a natural alternative therapy with no adverse effects. There are no known regulations, epidemiologic data, or gold standard tests for diagnosing the hepatic injury they can cause, which is known as herb-induced liver injury (HILI). Establishing the causal relationship is a challenge that requires a high level of clinical suspicion, and its diagnosis is usually based on exclusion. *Bauhinia forficata*, also known in Argentina as "cow's hoof", is an herb that can be found in Latin America and bought in dietary stores. It is used as an infusion due to its supposed beneficial properties. We present a case of an acute-on-chronic liver injury induced by this herb.

CASE DESCRIPTION A 56-year-old man with hepatic cirrhosis, Child-Pugh score C secondary to alcohol consumption with later cessation of the habit, was admitted with 5 days of abdominal pain and distention, jaundice, and choluria. He admitted to consuming 3 l of the *Bauhinia forficate* infusion 15 days ago. He had ascites and no signs of encephalopathy. Laboratory workup showed total bilirubin of 19.3 mg/dl, direct bilirubin 16.6 mg/dl, aspartate ami-

notransferase 110 UI/l, alanine aminotransferase 49 UI/l, alkaline phosphatase 198 UI/l, γ-glutamyl transferase 143 UI/l, prothrombin time 16.6 s (rate 40%), factor V 45%, eosinophilia 683/mm^3, negative viral serology (HIV, hepatitis B surface antigen, hepatitis C, E, and A viruses, immunoglobulin M, cytomegalovirus, Epstein-Barr virus, parvovirus), and negative immunological panel. Magnetic resonance cholangiopancreatography was performed with evidence of chronic hepatopathy. The patient developed coagulopathy, progressive increase in bilirubin level, factor V consumption, and hepatorenal syndrome. Liver biopsy was performed and showed micronodular cirrhosis, eosinophils that damaged the bile ducts and bile plugs. Due to these findings and the Model for End-Stage Liver Disease score of 37 the patient was included on the transplant list. He presented the following complications: septic shock secondary to a nosocomial pneumonia, renal failure with hemodialysis requirement, grade IV hepatic encephalopathy, and a need for mechanical ventilation. Finally, the patient died from multiple organ failure.

CONCLUSIONS It is important to consider the consumption of *Bauhinia* as a cause of HILI, specially in individuals with chronic liver disease. The cases must be included in the national system of pharmacovigilance, to enable comprehension of possible cause and effect relationships between the consumption of herbs and clinical presentations.

Key words
acute-on-chronic, herb induced liver injury, liver failure

MARTA OREL

A case of thyroid storm

Marta Orel*, Larysa Martynyuk

Department of Emergency Medical Care, Faculty of Medicine, I. Horbachevsky Ternopil National Medical University, Ternopil, Ukraine

*Corresponding author: orel_ma@tdmu.edu.ua

INTRODUCTION Thyroid storm, also known as thyrotoxic crisis, is an acute, life-threatening complication of hyperthyroidism. The mortality associated with thyroid storm is 8% to 25%. As fever is the most common presentation of numerous conditions, differential diagnosis for other diseases with similar symptoms and signs should be performed as soon as possible and treatment should start even before obtaining laboratory results. We present a rare case of Graves disease decompensation at the time of acute illness.

CASE DESCRIPTION A 32-year-old woman suddenly developed psychomotor excitation, jaundice, and fever on the 7th day of SARS-CoV-2 infection. She was hospitalized at an intensive care unit. According to her family members, for the last 5 days she was suffering from nausea, vomiting, diarrhea, psychomotor excitation, impatience, muscle tremor, and insomnia. During the last 6 months she had lost 6 kg in weight, and had a history of autoimmune thyroid disease. On examination, the patient was agitated, easily irritable, the skin was warm, moist, with a shade of jaundice. Eyes were red, watery, and bulging. Her body temperature was 39.5 °C. The neck was visually thickened. The thyroid gland was enlarged to the 2nd degree due to thickening of both lobes and the isthmus. It was painless and movable on palpation, of elastic consistency, a systolic murmur was heard over the gland when auscultating. Breath rate was 20/min, blood pressure 80/45 mm Hg, heart rate 135 bpm, heart tones were loud, systolic murmur and extrasystoles were present. Lower liver edge was 2 cm below the ribs. Electrocardiography showed atrial fibrillation with heart rate 135 bpm and frequent ventricular premature beats.

DISCUSSION Different diagnostic algorithms can be applied to diagnose thyroid storm. According to Burch–Wartofsky Point Scale (BWPS), our patient had elevated temperature (20 points), mild central nervous system (CNS) dysfunction (10 points), tachycardia (20 points), atrial fibrillation (10 points), jaundice (20 points), and a precipitating factor in the form of SARS-CoV-2 infection (10 points), a total of 90 points that is highly suggestive of thyroid storm. According to the Japanese Thyroid Association (JTA) scoring system, she had CNS manifestation, fever, tachycardia, and hepatic manifestation, which accounted for definite thyroid storm. A score of 45 or more based on the BWPS scoring system is more sensitive, but less specific than the JTA scoring system to detect thyroid storm cases. A BWPS score of 25 to 45 may suggest an impending storm. Treatment involves thyrostatic therapy (eg, propylthiouracil), iodine to reduce thyroid hormone release, adrenergic antagonists (β-blockers), and dexamethasone to provide adrenal support and block peripheral conversion of thyroxine to triiodothyronine. Supportive measures such as intravenous fluids, oxygen, cooling blankets, and acetaminophen might be are needed.

CONCLUSIONS Thyroid storm is a rare, life-threatening endocrine emergency that may be difficult to recognize and fatal if left untreated. Complications include arrhythmias, heart failure, seizures, delirium, coma, jaundice, abdominal cramps, atrial fibrillation, and thromboembolism. The cause of death may be heart failure, arrhythmias, or multiple organ failure. However, with treatment, most patients show improvement within 24 hours.

Key words
atrial fibrillation, fever, goiter, jaundice, thyroid storm

MARTA OREL

Unusual manifestation of polyglandular autoimmune syndrome type 2

Marta Orel*, Larysa Martynyuk

Department of Emergency Medical Care, Faculty of Medicine, I. Horbachevsky Ternopil National Medical University, Ternopil, Ukraine

*Corresponding author: orel_ma@tdmu.edu.ua

INTRODUCTION Polyglandular autoimmune syndrome type 2 (PAS-2) is an autoimmune disease with polygenic inheritance, which leads to lymphocytic infiltration causing organ-specific damage. While PAS-2 is the most common type of PAS, we here present a case with atypical course of the disease: coexistence of Addison disease and autoimmune thyroid disease with hypothyroidism and type 1 diabetes mellitus (T1DM) in a middle-aged woman.

CASE REPORT A 35-year-old woman developed brain fog and was confused when answering questions. Ten years ago she was diagnosed with autoimmune thyroiditis with hypothyroidism, and was taking L-thyroxine at 75 mg/day. Two years later T1DM was diagnosed, and insulin glargine at 36 U before dinner, and insulin glulisine at 8-14-10 U before each meal were implemented. The patient had no history of allergies. Her condition aggravated after influenza, when she noted loss of appetite, general weakness, nausea, and vomiting. The patient's height was 165 cm, weight 56 kg. Her skin was dry, moderately pigmented for the type of sun tan with areas of hyperpigmentation in axillar and inguinal regions and skin folds. The face was motionless, with swollen eyelids. Pulse was 60 bpm, rhythmic, blood pressure 75/40 mm Hg. Heart tones were weakened. The thyroid gland was of a normal size, movable, and painless when palpating. Biochemical blood tests showed total bilirubin of 0.4 mg/dl, alanine transaminase 18.7 U/l, aspartate transferase 19 U/l, glucose 84.68 mg/dl, total protein 7.54 g/dl, creatinine 1.01 mg/dl, urea 18.7 mg/dl, total cholesterol 223.94 mg/dl, potassium 6.3 mEq/l, sodium 124 mEq/l. Glycemic profile was 70.27–91.89–77.48 mg/dl; glycated hemoglobin, 7.8%; thyroid-stimulating hormone, 11.2 mIU/ml (reference range [RR], 0.5–5 mIU/ml); and free thyroxine, 0.42 ng/dl (RR, 0.9–2.4 ng/dl). Thyroid gland ultrasound revealed a typical location and hypoechoic patchy structure with fibrous masses.

DISCUSSION PAS-2 is diagnosed based on at least 2 out of 3 manifestations in the same patient, including primary adrenal insuf-

ficiency (Addison disease), autoimmune thyroid disease causing Graves disease or hypothyroidism and T1DM. Other endocrine and nonendocrine manifestations of PAS-2 are primary hypogonadism, myasthenia gravis and celiac disease, alopecia, vitiligo, pernicious anemia, idiopathic heart block, stiff person syndrome, immunoglobulin A deficiency, serositis, dermatitis herpetiformis, idiopathic thrombocytopenia, and hypophysitis. Patients with PAS-2 and their family members should be monitored on long-term basis for symptoms related to adrenal, thyroid, and endocrine pancreatic dysfunction. Organ-specific antibodies, such as 21-hydroxylase antibody for Addison disease, an antibody against glutamic acid decarboxylase antibody for T1DM, thyrotropin receptor antibody, and thyroid peroxidase antibodies for autoimmune thyroid disease can be investigated. However, the presence of autoantibodies to the thyroid, adrenal glands, and islets of Langerhans does not predict glandular failure.

CONCLUSIONS The diagnosis of PAS-2 is often delayed. Usually, the patients present with isolated endocrine dysfunction and later develop other endocrine and nonendocrine diseases. Ignoring the signs of PAS-2 entails a high risk of severe hypothyroidism, adrenal crisis, and diabetic ketoacidosis. The patients with PAS have a 2.5-fold higher risk of adrenal crisis. The effective treatment is replacement of appropriate hormones.

Key words
Addison disease, type 1 diabetes mellitus, hypothyroidism, polyglandular autoimmune syndrome type 2

MARTA OREL

Polyglandular autoimmune syndrome type 1 in an adult man

Marta Orel*, Larysa Martynyuk

Department of Emergency Medical Care, Faculty of Medicine, I. Horbachevsky Ternopil National Medical University, Ternopil, Ukraine

*Corresponding author: orel_ma@tdmu.edu.ua

INTRODUCTION We present a case of a rare polyglandular autoimmune syndrome type 1 (PAS-1), also known as autoimmune polyendocrinopathy candidiasis ectodermal dystrophy, which manifested in an adult man with skin hyperpigmentation and focal alopecia.

CASE DESCRIPTION A 37-year-old patient received treatment in an endocrinology department. Upon admission, he complained of headache, dizziness, tinnitus, visual impairment, periodic palpitations and pain in the heart region, constipation, low blood pressure, morning sickness, recurrent seizures, and general weakness. The patient has considered himself ill for about 8 years, since he noticed his skin hyperpigmentation. He underwent treatment with hydrocortisone 20 mg, L-thyroxine 150 mg, and calcium 2000 mg daily. His height is 176 cm and he weighs 70 kg, his body mass index is 22.6 kg/m^2. Visible symptoms included skin and mucous membrane hyperpigmentation with areas of vitiligo and focal alopecia. His body temperature was 36.7 °C. Thyroid gland was of normal size. Pulse was 72 bpm, rhythmic, blood pressure 80/60 mm Hg. The lower liver edge was up to 2 cm below the ribs, slightly sensitive when palpating. The patient suffered from frequent constipation. Laboratory workup revealed complete blood count within normal range. Biochemical blood tests showed total bilirubin of 0.15 mg/dl, alanine transaminase 25.7 U/l, aspartate transferase 29.1 U/l, total protein 6.77 g/dl, creatinine 1.01 mg/dl, urea 21 mg/dl, total cholesterol 212.36 mg/dl, calcium 4 mg/dl, potassium 4.7 mEg/l, iron 63.13 mg/dl, amylase 116 U/l, γ-glutamyl transferase 34 U/l, uric acid 5.55 mg/dl, alkaline phosphatase 115 U/l, chloride 100 mEq/l, cortisol 1.72 μg/dl (reference range [RR], 8–20 μg/dl), thyroid-stimulating hormone 61 mIU/ml (RR, 0.5–5 mIU/ml), free thyroxine 0.88 ng/dl (RR, 0.9–2.4 ng/dl), antithyroid peroxidase 375 IU/ml (RR, <35 IU/ml). Electrocardiography showed regular sinus rhythm with 69 bpm.

DISCUSSION Our patient had PAS-1, which is an autosomal recessive syndrome due to mutation of the *AIRE* gene, characterized by a sequential or simultaneous deficit in the function of several endocrine glands. It results in hypoparathyroidism, adrenal insufficiency, hypogonadism, vitiligo, mucocutaneous candidiasis, and other conditions. Although the etiology is most often autoimmune, the triggers include drugs, viral infections, and dietary and environmental factors. Because of a huge variety of symptoms, it can be initially difficult to recognize. A diagnosis of PAS-1 is suggested clinically and confirmed by detecting deficient hormone levels. Patients with autoimmune manifestations should be carefully examined. PAS-1 is defined based on the presence of at least 2 of the following: chronic mucocutaneous candidiasis, hypoparathyroidism, or adrenal insufficiency.

PAS-1 is associated with autoantibodies against type 1 interferons and the presence of these antibodies suggests the diagnosis, which can be verified by mutational analysis of the *AIRE* gene. Detecting autoantibodies to each affected glandular tissue can help to differentiate polyglandular deficiency syndromes from the other causes. Treatment usually includes hormone replacement and antifungal therapy (oral fluconazole or ketoconazole).

CONCLUSIONS Polyglandular autoimmune deficiency syndromes should be early diagnosed and treated. Polymorphic symptoms that they cause lead to significant impairment of health and quality of life, while some of the symptoms, such as unrecognized hypoparathyroidism or adrenal insufficiency, can be life-threatening.

Key words
adrenal insufficiency, hormone replacement, hypothyroidism, polyglandular autoimmune syndrome type 1

ANKAN PATHAK

Unusual case of fever with arthralgia in a patient with diabetes: going beyond the usual suspects

Ankan Pathak[1]*, Aparna Tewari[2], Nirmalya Roy[3]

1 Department of Cardiology, Apollo Multispecialty Hospital, Kolkata, India
2 Department of Anaesthesia and Critical Care Medicine, Medical College, Kolkata, India
3 Department of Medicine, KPC Medical College and Hospital, Jadavpur, India

*Corresponding author: ankan.manu@gmail.com

INTRODUCTION Tropical pyomyositis is an infectious disease that affects skeletal muscles and may appear as a diffuse inflammation or a rapidly progressing myonecrotic process. *Staphylococcus aureus* is the most common causative microorganism, accounting for 90% of cases. The diagnosis is sometimes late, because patients usually do not seek help with their initial symptoms, and because it is a rare disease, doctors are still not familiar with it.

CASE DESCRIPTION A 56-year-old type 2 diabetic man with grossly uncontrolled hyperglycemia developed tropical pyomyositis with muscle abscess that required prolonged antibiotic therapy and surgical drainage.

Based on the presentation, a provisional diagnosis of primary pyomyositis due to uncontrolled hyperglycemia was set. Incision and drainage of the abscesses were planned after obtaining proper glycemic control (basal bolus). Around 50 ml of pus were aspirated from the trapezius muscle and sent for cultures. The cultures revealed the presence of *Staphylococcus aureus* sensitive to piperacillin + tazobactam and clindamycin. The patient was put on antibiotic injections for 2 weeks. On repeated laboratory workup his complete blood count was 6300/mm, C-reactive protein concentration was 6 mg/dl, fasting glucose level was 135 mg/dl, and postprandial glucose level was 178 mg/dl.

CONCLUSIONS The initial signs and symptoms are nonspecific, which often makes primary pyomyositis underdiagnosed. Suspicions should be high in diabetic patients and / or otherwise compromised

individuals, mainly in the case of fever and myalgia without significant elevation in the muscle enzymes.

Key words
abscess, hyperglycemia, myonecrotic, pyomyositis, *Staphylococcus aureus*

MARTIN PEHR

Recurrent abdominal pain and fever as a presentation of autoinflammatory disease

Martin Pehr[1,2]*, Jan Škrha Jr[1,2]

1 Third Department of Internal Medicine, Endocrinology and Metabolic Diseases, General Teaching Hospital, Prague, Czech Republic
2 First Faculty of Medicine, Charles University, Prague, Czech Republic

*Corresponding author: martinpehr@gmail.com

INTRODUCTION Autoinflammatory diseases are rare and therefore should not be accounted for in initial differential diagnosis of suspected urinary tract infections, laryngitis, or acute abdomen. But when a patient presents repeatedly and frequently with such symptoms, rare diseases should be considered.

CASE DESCRIPTION A 28-year-old Israeli woman was referred by her general practitioner with a 2-week history of gradual worsening of abdominal pain. She also presented swelling of abdomen and legs upon eating or drinking. She had a history of frequent laryngitis and urinary bladder infections. She also had malaria several times from her 2-year stay in Mozambique.

Full physical examination revealed only slight abdominal discomfort on palpation. There was no asymmetry of the swollen legs and blood in the stool was not observed. The patient was very thin with body mass index of 16.8 kg/m^2. Complete blood count and biochemical workup revealed only slightly elevated C-reactive protein (13.6 mg/l; reference range, 0–5 mg/l) and slight anemia (hemoglobin 112 g/l; reference range, 120–160 g/l). Low human chorionic gonadotropin ruled out possible pregnancy. Abdominal X-ray and ultrasound ruled out acute abdomen and showed only slight thickening of the ascending colon. Significant thickening of the left kidney cortex to 2 cm was noticed. The patient was admitted to our internal department for further examination.

Extensive serologic and immunologic screening, computed tomography enterography, and colonoscopy with biopsy examinations were performed and found unremarkable. Serum amyloid A was within the reference range.

Further search of the patient's medical history during hospitalization and contact with a rheumatology department as well as her general practitioner in Israel revealed that as a teenager she was diagnosed with familiar Mediterranean fever, but she forgot the condition's name. The patient was offered colchicine treatment but refused it due to the fear of side effects and chose diet treatment instead.

She was treated symptomatically with gradual decrease of her symptoms and nutritional support was initiated.

DISCUSSION Familial Mediterranean fever is an autoinflammatory disease. It is a hereditary condition with autosomal recessive pattern, most prevalent among non-Ashkenazi Jews. Causative mutation in the marenostrin gene causes inadequate activation of innate immunity. It presents in episodes of a fever lasting for a few days and solving spontaneously. Sometimes it is accompanied by sterile inflammation of serous membranes and leg and abdominal pain. Due to its nature, it might be mistaken for cystitis, laryngitis, or acute abdomen. Empiric treatment with antibiotics might falsely confirm another diagnosis, while the symptoms would dimmish by themselves.

CONCLUSIONS The key to diagnosis was hidden in abundant medical history of the patient and countless symptoms presented at different occasions. Ample time and careful documentation are key to the diagnosis, and we must make time regardless of everyday rush or other factors. Some cases are easily manageable during hospitalization, as we simply do not have enough time, expertise, and examination equipment at the emergency or outpatient department.

Finally, no one can know all rare diseases and asking for advice from senior colleagues is always recommended.

Key words
autoinflammatory disease, familial Mediterranean fever, recurrent abdominal pain, recurrent infections

MARCELA ROMERO

An unwanted host in the bone marrow

Marcela Romero Caballero*, Agustina Varas Schaer, Dario Trela

Department of Internal Medicine, Hospital Escuela de Agudos Dr. Ramón Madariaga, Misiones, Argentina

*Corresponding author: romerocmarcela97@gmail.com

INTRODUCTION Visceral leishmaniasis (VL) is an infection spread by the protozoa *Leishmania donovani* and *Leishmania infantum*. The clinical manifestations usually involve prolonged febrile syndrome, fatigue, weight loss, hepatosplenomegaly, and lymph node enlargement. Laboratory findings may demonstrate the presence of pancytopenia, abnormal liver function, and hypergammaglobulinemia. Over 90% of HIV-infected patients affected by leishmaniasis have less than 200 CD4 lymphocytes/mm^3, so the parasite has favorable conditions for uncontrolled multiplication. This means that in these patients severe and atypical forms of the disease can occur.

CASE DESCRIPTION A 23-year-old man was consulted for wasting syndrome and 2-month long fever. Abdominal examination revealed hepatomegaly and splenomegaly. Laboratory workup on admission demonstrated pancytopenia, hypertransaminasemia, and electrophoretic proteinogram showed polyclonal increase in γ-globulins. Serology confirmed reactive HIV, with viral load of 2 630 000 copies/ml, CD4 count 5 cells/mm^3, nonreactive qualitative veneral disease research laboratory / unheated serum reagin, nonreactive hepatitis B surface antigen, nonreactive hepatitis C anti-immunoglobulin (Ig) G, nonreactive indirect hemagglutination assay for Chagas disease, toxoplasmosis IgG antibodies above 250 (reactive), nonreactive toxoplasmosis IgM antibodies, nonreactive Epstein–Barr virus IgM, Epstein–Barr virus IgG 2.2 (reactive), nonreactive cytomegalovirus IgM, cytomegalovirus IgG 8.07 (reactive), and Rk39 immunochromatography test was requested but it was not available at our center. Blood cultures were negative for common pathogens. Thoracic and abdominal computed tomography demonstrated hepatomegaly with slightly enlarged spleen. Bone marrow aspirate was performed with medulogram confirming preserved cellularity, but the presence of intracellular parasites matching leishmania was observed. Polymerase chain reaction was positive for leishmaniasis. We assumed the condition to be visceral leishmaniasis, and began treatment with amphotericin B for 14 days.

DISCUSSION Visceral leishmaniasis is an infectious disease with a low prevalence in our environment, especially in patients with HIV infection. When a patient reports with a prolonged febrile syndrome with organomegaly, leishmaniasis should be considered in the differential diagnosis. Given that early detection of the disease was achieved, it was possible to eradicate it in the patient, with subsequent improvement of his symptoms and laboratory parameters.

CONCLUSIONS This case is presented due to the low prevalence of visceral leishmaniasis in our environment. Although a wide variety of opportunistic pathogens should be suspected in patients with AIDS, the presence of parasites should not be neglected.

Key words
hepatosplenomegaly, HIV infection, prolonged febrile syndrome, visceral leishmaniasis

SUMAYYA SAMI

Cryptococcal IRIS in a patient with HIV infection leading to adrenal insufficiency

Sumayya Sami[1]*, Faisal Ali[2], Syed F. Mahmood[1]

1 Department of Medicine, Aga Khan University Hospital, Karachi, Pakistan
2 Department of Gastroenterology, Shifa International Hospital, Islamabad, Pakistan

*Corresponding author: dr.sumayyasami@gmail.com

INTRODUCTION Immune reconstitution inflammatory syndrome (IRIS), is a paradoxical worsening of clinical symptoms caused by increased inflammatory processes toward an underlying infection, following the initiation of antiretroviral therapy (ART) in HIV-infected individuals. There are 2 forms of IRIS: "unmasking" IRIS, defined as a flare-up of a previously undiagnosed, preexisting infection, after the initiation of ART, and "paradoxical" IRIS, referring to deterioration of a previously diagnosed and treated infection after the implementation of ART. Cryptococcal meningitis (CM) in an HIV-infected patient is a deadly and common disease. Noteworthy of CM is its association with the IRIS leading to cryptococcal IRIS (C-IRIS). C-IRIS usually occurs paradoxically, within 4 to 9 weeks after the start of ART. Here, we describe a case of a 59-year-old man, HIV-positive and with CM, who developed paradoxical IRIS that was manifested as adrenal insufficiency after the beginning of ART.

CASE DESCRIPTION The patient was diagnosed with CM and HIV 8 weeks prior to this presentation and was started on antifungal therapy at that time. Four weeks later, ART was initiated as his cerebrospinal fluid became negative for *Cryptococcus neoformans*. Four weeks after the initiation of ART, he presented to our hospital with an acute history of slurred speech and abnormal behavior followed by altered level of consciousness. Examination revealed Glasgow Coma Scale (GCS) of 3/15 with blood pressure 84/60 mm Hg, and heart rate of 96 bpm. Random plasma glucose was very low. The patient was treated with intravenous 25% dextrose, which markedly improved his GCS to 15/15. Fluid resuscitation improved his hemodynamics intermittently and his hypoglycemia was recurrent. Laboratory workup revealed serum morning (8 AM) cortisol of 165.5 nmol/l (reference range, 140–690 nmol/l). Furthermore, the short synacthen test confirmed adrenal insufficiency. Intravenous hydrocortisone (50 mg, every 8 h) was started, and later switched to equivalent oral prednisolone dosage, which remarkably improved hypoglycemia and hypotension. The patient is currently able to perform his daily activities.

DISCUSSION This case highlights the occurrence of paradoxical C-IRIS in the form of adrenal insufficiency in an adult, HIV-infected patient. The patient developed features of hypoglycemia and hypotension that persisted despite recurrent fluid resuscitation and intravenous 25% dextrose water administration, and resolved only when corticosteroids were initiated. Recurrent CM, paradoxical C-IRIS with neurologic manifestation or meningitis due to other infective etiologies were other differential diagnoses in this patient; however, the refractory nature of his hypotension and hypoglycemia made us suspect adrenal insufficiency secondary to paradoxical C-IRIS. The diagnosis of adrenal insufficiency caused by adrenal cryptococcosis was supported by the results of morning cortisol and the short synacthen test. Hence, in an HIV-infected patient with CM, IRIS should be suspected with its various manifestations. The presentation of adrenal insufficiency is rare and difficult to diagnose, but it is potentially treatable with high index of suspicion leading to early diagnosis and adequate treatment.

LESSONS TO BE LEARNED

- Refractory hypotension and hypoglycemia can be manifestations of adrenal insufficiency in paradoxical C-IRIS.
- The recommended time to start ART after implementation of CM treatment is 4 to 6 weeks.
- ART should not be discontinued because of IRIS.

Key words
adrenal insufficiency, cryptococcal meningitis, HIV, IRIS

JURAJ SMAHA

A rare cause of exudative pleural effusion in a patient with secondary biliary cirrhosis

Juraj Smaha*, Peter Jackuliak, Juraj Payer

Fifth Department of Internal Medicine, Faculty of Medicine, Comenius University and University Hospital Bratislava, Bratislava, Slovakia

*Corresponding author: smaha1@uniba.sk

INTRODUCTION Bilothorax is a rare cause of pleural effusion, defined by the presence of bile in the pleural space. There is a strong association between bilothorax and development of a pleural infection.

CASE DESCRIPTION We describe a case of a 48-year-old woman with cirrhosis who presented to the emergency department because of right upper quadrant pain, dyspnea, and cough. Ultrasound examination showed a large and complex pleural effusion, with undulating thick fibrinous septa within the fluid. Biochemical examination of the pleural fluid revealed bilothorax with progression to thoracic empyema. Prompt conservative therapy with intravenous antibiotics and chest tube drainage was started, but escalation to surgical management was later necessary.

CONCLUSIONS Pleural infections are associated with considerable morbidity and are life-threatening conditions requiring complex medical management. This case presents the application of thoracic ultrasound for a more detailed diagnosis than merely detecting the pleural fluid in the patient with exudative pleural effusion.

Key words
bilothorax, lung ultrasound, pleural effusion, thoracic ultrasound

LUKÁŠ ŠTOS

Extremely rare pancreatic extragastrointestinal stromal tumor: a case report

Lukáš Štos[1]*, Jaroslav Horáček[2], Tomáš Jonszta[3]

1 Department of Internal Medicine and Cardiology, University Hospital Ostrava, Ostrava, Czech Republic
2 Department of Clinical and Molecular Pathology and Medical Genetics, University Hospital Ostrava, Ostrava, Czech Republic
3 Department of Radiology, University Hospital Ostrava, Ostrava, Czech Republic

*Corresponding author: lukas.stos@email.cz

INTRODUCTION Mesenchymal tumors represent only 1%–2% of all gastrointestinal tumors. Gastrointestinal stromal tumor (GIST) is the most common type included in this rare group. It typically arises from the stomach, less often from the other sites of the gastrointestinal tract. In 5%–10% cases of GISTs, the tumors can be formed outside the gastrointestinal tract, they are so-called extragastrointestinal stromal tumors (EGISTs). EGISTs can arise from the mesenchymal tissue of the omentum, mesentery, or retroperitoneum, and sporadically from the other organs, such as the liver and hepatobiliary tract, spleen, pancreas, or urinary system. The last locations are only rarely described in case reports.

CASE DESCRIPTION We present a case of a 38-year-old man admitted for weight loss, fever, dyspepsia, and intermittent epigastric pain. Possibly malignant multiple focal hepatic lesions and pancreatic head tumor were found on abdominal sonography and subsequent abdominal computed tomography (CT) scan. However, multiple detailed histopathological and immunohistochemical examinations showed extremely rare primary EGIST of the pancreatic head with a low risk of progression. In the liver area, magnetic resonance imaging scans showed only abscess formations, which were subsequently successfully solved during CT-guided aspiration. This procedure was followed by several weeks of tailored intravenous antibiotic therapy.

No tumor cells were found in the abscess collections during both cytological and histological examination. Ultrasound check of the liver parenchyma showed no remaining abscess fluid. However, the etiology of liver abscesses has not yet been fully clarified, with the possibility of ascending bacterial infection of the biliary tree due to its partial obstruction by the pancreatic head tumor. After nutritional preparation, the patient underwent complete enucleation of the tumor without disrupting its capsule and without the need of hemipancreatoduodenectomy. During the 8-month long follow-up, the patient showed no evidence of the tumor recurrence. A whole-body positron emission tomography-CT examination using ^{18}F-fluorodeoxyglucose showed no pathology. The patient maintains a physically active lifestyle, he gained back 6 kilograms, and has had no health issues thus far.

CONCLUSIONS Only a few case reports describing primary EGIST of the pancreas can be found in the literature. This report brings another clinical case of this extremely rare tumor.

Key words
extragastrointestinal stromal tumor, gastrointestinal stromal tumor, pancreas

SVEN L. VAN LAER

Migration of a Hot AXIOS stent causing small bowel obstruction

Sven L. Van Laer[1,2*], Arne Ballet[1,2], Schalk W. Van der Merwe[3,4]

1 Department of Gastroenterology, St Dimpna Hospital Geel, Geel, Belgium
2 Faculty of Medicine and Health Sciences, University of Antwerp, Antwerp, Belgium
3 Department of Gastroenterology and Hepatology, University Hospitals Leuven, Leuven, Belgium
4 Faculty of Medicine, KU Leuven, Leuven, Belgium

*Corresponding author: Sven.VanLaer@uantwerpen.be

INTRODUCTION Lumen-apposing metal stents (LAMSs), such as Hot AXIOS stents, are devices approved for endoscopic drainage of a pancreatic pseudocyst, the gallbladder in patients with acute cholecystitis, and the bile duct after failed endoscopic retrograde cholangiopancreatography (ERCP). Due to their efficiency and safety, LAMSs are increasingly used for off-label indications including gastrojejunostomy, gastro-gastrostomy, and drainage of postsurgical collections. The complication rates for these off-label indications are not well-known. We present a case of a 68-year-old woman with small bowel obstruction caused by migration of LAMS following endoscopic gastro-gastrostomy with a Hot AXIOS stent after a Roux-en-Y gastric bypass.

CASE DESCRIPTION A 68-year-old woman presented to the emergency department with worsening abdominal pain and nausea. Her medical history included a Roux-en-Y gastric bypass, a laparoscopic hiatal hernia repair and blind loop resection of gastrojejunostomy, and nonalcoholic steatohepatitis liver cirrhosis treated with orthotopic liver transplantation, complicated by transverse colon perforation with the need for extended right hemicolectomy with terminal ileostomy. Five months before presentation she underwent endoscopic gastro-gastrostomy with a Hot AXIOS stent for a later ERCP. The patient described periumbilical cramping, nausea, and cessation of stool passage for 2 days. Physical examination revealed tachycardia with heart rate of 107 bpm, blood pressure of 136/85 mm Hg, dry mucous membranes, abdominal distention, tympanic percussion, and high-pitched bowel sounds on auscultation. Blood tests showed acute kidney injury and hyperkalemia. Abdominal X-ray showed dilated bowel loops with air-fluid levels, with a foreign object at the level of the right flank. Abdominal computed tomography scan confirmed migration of the Hot AXIOS stent to just before the ileostomy with the presence of fecal bowel sign. The patient underwent ileoscopy through the ileostomy, which confirmed the presence of the stent near the end of the ileostomy. The stent was rotated inside the bowel, with both ends facing the bowel wall, causing mechanical small bowel obstruction. It was endoscopically removed and rapid clinical recovery followed.

DISCUSSION The use of LAMSs has increased significantly in the recent years with a technical success rate of 98% and a clinical success rate of 95% for approved indications. These impressive findings explain the increase in off-label indications, such as gastro-gastrostomy, as in our case. Stent migration is a known adverse event occurring in approximately 3%–5% of patients. Its migration to the bowel, however, is a rare complication. Previously published case reports include a Hot AXIOS stent migration into the colon. Spontaneous evacuation occurred and no endoscopic intervention was required. Another case report describes the migration of a LAMS into the colon with elective endoscopic removal without intestinal obstruction.

CONCLUSIONS To our knowledge, this is the first reported case of a migrating LAMS causing an intestinal obstruction. Given the rapid increase in (off-label) indications for LAMS implantation, migration and subsequent possible intestinal obstruction should be considered in the differential diagnosis in patients with abdominal pain and a history of LAMS implantation.

Key words
abdominal pain, endoscopy, intestinal obstruction, self-expandable metallic stents

MARCELINA WILK

Radiologic suspicion of sarcoidosis in a young man: how to avoid diagnostic pitfalls?

Marcelina Wilk[1*], Anna Trojan-Królikowska[1,2], Krzysztof Składek[1,2]

1 Division of Pulmonology and Allergology, University Hospital, Kraków, Poland
2 Second Department of Internal Medicine, Jagiellonian University Medical College, Kraków, Poland

*Corresponding author: marwilk95@gmail.com

INTRODUCTION The diagnostic process of abnormal chest imaging findings is an everyday clinical scenario, yet many pitfalls remain. Respiratory complaints, such as cough or shortness of breath, reported by patients are nonspecific. It can take months or even years to make an accurate diagnosis. Malignant process is rarely suspected in young individuals, leading to delayed cancer diagnosis. Oncologic vigilance is a key, especially when the clinical picture is unclear.

CASE DESCRIPTION A 41-year-old white man, with no medical history or treatment was admitted to an emergency department due to severe epigastric pain, progressive exercise intolerance, and 1-month history of dry cough. He reported dull pain radiating to the left side of the chest and to the back, worsening in upright position, and occurring also during night rest. He confirmed occupational exposure to varnishes, paint, and dust in recent years. He was a past smoker (10 pack-years, he quitted 10 years ago). Standard chest X-ray was performed and revealed parenchymal opacities. Due to abnormalities on chest X-ray, elevated D-dimer concentration, and uncertainty of clinical image, the diagnostics was extended to computed tomography (CT) pulmonary angiography. There were no radiologic features of pulmonary embolism. However, hilar and mediastinal lymphadenopathy and small perilymphatic nodules predominantly in the perihilar and upper parts of the lungs were noted. The lesions formed 2 spicular infiltrations in the right lung. Moreover, multiple lytic, suspicious lesions, predominantly in the vertebral bodies of the thoracic spine were revealed. Due to radiologic suspicion of sarcoidosis, the patient was admitted to the pulmonology and allergology unit to continue the diagnostic process. Bronchoscopy was performed. Endobronchial biopsies from mucous membrane of the right secondary carina were taken for his-

topathologic analysis, which revealed normal findings. Metaplastic cells were found in the bronchial lavage, no microorganisms were isolated in the culture nor confirmed on molecular assays. Active tuberculosis was also excluded. Endoscopic ultrasound-guided needle aspiration of intrathoracic lymph nodes was performed. Cell-block material revealed cells of lung adenocarcinoma. Positron emission tomography-CT imaging detected metabolically active process in the right lung and dissemination to the skeletal system and lymph nodes. The patient's history was presented to a multidisciplinary team in cancer care to determine further treatment approach. He was qualified for combined treatment including chemotherapy and radiotherapy.

CONCLUSIONS A diagnostic pathway of radiologic abnormalities is often challenging, especially among younger patients, when we do not usually suspect a malignant condition. It is evident that oncologic alertness is essential also in this subgroup, especially when the clinical presentation is unclear and when "red flags" occur, as in the presented case. Detailed medical history assessment, with emphasis on environmental exposures and smoking, variety of imaging modalities and multidisciplinary, experienced decision-making team can improve care in such cases.

Key words
diagnostic pitfall, lung adenocarcinoma, sarcoid-like reaction to malignancy, sarcoidosis