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Clinical Cases in Internal Medicine: Learning Through Practice
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AWARD-WINNING CASES

1ST PLACE: GIL VERSCHULDEN

Significant response to dabrafenib in a patient with Erdheim–Chester disease with BRAF^{V600E} mutation: a case report and literature review

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BACKGROUND Erdheim–Chester disease (ECD) is a rare type of non–Langerhans cell histiocytosis, with vemurafenib as the only agent approved by the Food and Drug Administration in November 2017 for the treatment of patients with ECD with the BRAF^{V600E} mutation. ECD is classified by the World Health Organization as a lymphoproliferative disorder; it affects about 500 cases worldwide. Skeletal involvement is the most frequent ECD-related manifestation (74.1%). The disease is often diagnosed at a late stage after initial clinical manifestation and is associated with poor prognosis. The somatic BRAF^{V600E} mutation is found in about half of ECD cases. BRAF inhibition has shown efficacy in targeting BRAF^{V600E} in several ECD cases.

CASE A 57-year-old male patient with a 10-year history of retroperitoneal fibrosis of unknown origin was admitted with fever, dysphagia, dysphonia, and weight loss. ECD was diagnosed on the basis of radiologic and histologic findings. The patient was treated with a combination therapy with a BRAF inhibitor (dabrafenib) and a mitogen-activated protein kinase/ERK inhibitor (trametinib) as the first-line treatment. As the patient presented with severe arthralgias, dabrafenib was continued as monotherapy after a dose reduction. We noted a clinical and radiological improvement after 3 years of treatment.

CONCLUSION We reported a case of a dramatic recovery of a patient with symptomatic ECD after treatment with dabrafenib. The BRAF pathway inhibition seems to be an effective treatment of ECD, even in patients with a long disease history and severe morbidity.

Key words

BRAF mutation, dabrafenib, Erdheim–Chester disease, non–Langerhans cell histiocytosis, trametinib

2ND PLACE: SEUNG SHIN PARK

Catatonia as a presenting symptom of ectopic adrenocorticotrophic hormone syndrome caused by thymic carcinoid tumor

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INTRODUCTION Ectopic adrenocorticotrophic hormone (ACTH) syndrome due to atypical thymic carcinoid tumor is a rare cause of Cushing syndrome. It can be manifested as atypical cushingoid features due to acute hypercortisolemia. We report a case of ectopic Cushing syndrome due to thymic carcinoid tumor with an unusual presentation of acute psychotic symptoms such as catatonia.

CASE DESCRIPTION A 57-year-old previously healthy Korean man presented with lower extremity edema and fatigue, which started a few months earlier. Cushingoid appearance was not observed. However, he had been recently diagnosed with diabetes mellitus and hypertension. At an outpatient clinic, a chest X-ray revealed well-defined opacity at the right cardiac border and chest computed tomography showed a mass of approximately 6.5 cm in size along

the anterior mediastinum. The patient was admitted for further diagnostic workup. On admission, he developed hallucinations and delusions. Five days after admission, he fell into stupor and had rigid posture. He did not respond to external stimuli, and the Glasgow Coma Scale score was E1V1M1. There was no evidence of acute cerebral hemorrhage or infarction. Laboratory tests showed morning cortisol levels of 107.0 µg/dl (reference range, 5–25 µg/ml), ACTH levels of 241.5 pg/ml (0–60 pg/ml), aldosterone levels of 10.2 ng/dl (3.7–24 ng/dl), renin levels of 0.27 ng/ml/h (0.3–2.9 ng/ml/h), and severe hypokalemia (2.5 mmol/l). To control acute psychiatric symptoms and hypercortisolemia, continuous intravenous etomidate infusion was started in the intensive care unit. Serum cortisol levels decreased to 25.0 µg/dl one day after etomidate infusion at 2.5 mg/h. Psychiatric symptoms improved. Total thymectomy with video-assisted thoracoscopic surgery was performed on hospital day 8. After the surgery, plasma ACTH and serum cortisol levels decreased to 2.8 pg/ml and 14.2 µg/dl, respectively. Pathological findings were consistent with atypical thymic carcinoid with ACTH production. The patient subsequently developed fungal pneumonia and *Pneumocystis jiroveci* pneumonia. Pneumonia resolved with proper management, and the patient is followed on an outpatient basis with a supplemental dose of hydrocortisone.

DISCUSSION Thymic carcinoid tumors can cause ectopic ACTH syndrome, but they are responsible for only 10% of all cases. Other causes are small-cell lung cancer, gastrointestinal neuroendocrine tumors, and ovarian tumors. Cushing syndrome can manifest typically as facial plethora, increased abdominal and face fat, wide purplish striae, and bruising. However, it is unusual for patients with Cushing syndrome to present with acute psychotic symptoms such as catatonia. Catatonia is a psychomotor immobility, which is manifested by stupor. The optimal treatment of ectopic ACTH syndrome is surgical resection of the ACTH-producing tumor. However, acute psychosis is an emergency that requires urgent medical treatment. Among adrenal steroidogenesis blockers, such as ketoconazole, mitotane, and metyrapone, etomidate is the most rapid-acting drug in reducing serum cortisol levels. In this case, dramatic improvement of acute psychosis was achieved through etomidate continuous infusion.

LESSONS TO BE LEARNED FROM THE CASE Our case shows that ectopic Cushing syndrome can manifest with atypical psychotic symptoms such as catatonia, and intravenous etomidate could be a useful approach to an immediate reduction of cortisol levels.

Key words

catatonia, ectopic Cushing syndrome, etomidate, thymic tumor

3RD PLACE: LORIS AZOYAN

A 76-year-old man with fever, arthralgia, aphthae, anemia, and life-threatening gastrointestinal bleeding

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Several cases reported the association between gastrointestinal Behçet disease-like symptoms and myelodysplastic syndromes (MDSs) or chronic myelomonocytic leukemia (CMML) with trisomy 8. This association is rare, making it difficult to diagnose, and even more difficult to manage and treat.

A 76-year-old man of Lebanese descent presented with a history of fatigue, painful mouth ulcers, weight loss, and anemia. On examination, he revealed fever, large oral ulcers, multiple small inflammatory subcutaneous nodules of the limbs, arthralgias of the knees, elbows and ankles, as well as hepatosplenomegaly. Blood tests showed no-regenerative normocytic anemia, monocytosis, and inflammatory syndrome. CMML with trisomy 8 was diagnosed on the basis of bone marrow aspirate. The patient's condition deteriorated within a month, with recurrent life-threatening

gastrointestinal bleedings requiring 2 ileal resections. Histopathological examination revealed ulcerations of different ages, most probably secondary to vasculitis. The main hypothesis was that of gastrointestinal Behçet disease-like symptoms with pan-intestinal ulcers secondary to CMML with trisomy 8. Azacitidine treatment was initiated and led to regression of the ulcers, no recurrence of intestinal bleeding, resolution of monocytosis, and remission on the bone marrow aspirate. Nine months after the end of 4 azacitidine cycles, the patient presented with intestinal bleeding, and the bone marrow aspirate showed signs of dysmyelopoiesis, an increase in the blast count, and the recurrence of trisomy 8, leading to new cycles of azacitidine. Disease control was achieved until it progressed to acute myeloid leukemia 14 months later, associated at the same time with recurrence of severe oral ulcers and intestinal bleeding.

In patients with MDS or CMML, trisomy 8 seems to have a causal role in the occurrence of gastrointestinal Behçet disease-like symptoms. Thus, in a patient presenting with such symptoms, the differential diagnosis should include an MDS or CMML with trisomy 8, and a specific hematologic treatment, such as azacitidine, needs to be considered promptly to control gastrointestinal disease, even in the absence of hematologic indications.

Key words

azacitidine, Behçet disease, chronic myelomonocytic leukemia, trisomy 8

BAKHITAH ALEID

Plasmablastic lymphoma in an immunocompetent patient: a case report and literature review

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INTRODUCTION Plasmablastic lymphoma (PBL) is considered a rare aggressive variant of diffuse large B-cell lymphoma that commonly develops in immunocompromised patients, particularly in HIV-positive individuals. This report describes the development of this malignancy in an immunocompetent male patient. It outlines the atypical clinical presentation, difficulty in establishing diagnosis, and pathological features of the disease. Overall prognosis and response to chemotherapy differ depending on the presence of immunosuppression. We summarize the epidemiologic, clinical, and immunohistochemical characteristics of PBL based on a comprehensive review of the reported cases in immunocompetent individuals.

CASE DESCRIPTION We describe a case of a 55-year-old man without any known chronic illness, who received rivaroxaban treatment for unprovoked deep venous thrombosis (DVT) of the right lower extremity. Three months after DVT diagnosis, he presented with progressive swelling and pain of the right leg that was previously diagnosed as DVT. On physical examination, he presented with a large mass in the middle thigh, measuring 19×18×35 cm in size. The mass was warm, extremely tender, erythematous with dilated superficial veins, not attached to the skin. Pitting edema in the legs was observed; however, peripheral pulses were palpable. Imaging tests revealed a large heterogeneous mass in the soft tissue, occupying all compartments of the thigh and showing an intra-articular extension to the knee joint with bone destruction. Immunohistological studies confirmed the diagnosis of PBL.

CONCLUSION PBL should be considered in a differential diagnosis of rapidly progressing soft-tissue masses in immunocompetent individuals to facilitate earlier diagnosis and initiation of treatment, possibly resulting in better outcomes. The use of cell block technique for fine-needle aspiration biopsy is a reliable diagnostic method.

Key words

deep vein thrombosis, immunocompetent, plasmablastic lymphoma

AGATA HANNA BRYK

Congenital disorders of glycosylation type 1 as a cause of transient antithrombin deficiency in a woman with pulmonary embolism and gastrointestinal disorders

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Inherited antithrombin deficiency is associated with the highest risk for venous thromboembolism among all thrombophilias. The molecular basis of antithrombin deficiency is defect in the encoding gene *SERPINC1*. In some cases, this defect is absent. We present a case of a 31-year-old woman with a history of Crohn disease and high-risk unprovoked pulmonary embolism at the age of 22, treated with embolectomy. She also suffered from transient antithrombin

deficiency. Electrophoretic and high-performance liquid chromatography pattern of antithrombin showed increased portion of hypoglycosylated forms identical to those of patients with congenital disorders of glycosylation type 1. Analysis of genes involved in the plasma protein N-glycosylation pathway revealed 2 variations in mannose 6-phosphate isomerase. Treatment with D-mannose alleviated gastrointestinal symptoms and increased antithrombin activity. To summarize, antithrombin deficiency caused by its hypoglycosylation may be present in patients with venous thromboembolism but without mental disorders and with no symptoms during childhood typical for congenital disorders of glycosylation type 1.

Key words

antithrombin deficiency, congenital disorders of glycosylation type 1, pulmonary embolism, thrombophilia

JIN-JU CHOI

Differential diagnosis of gallbladder cancer and xanthogranulomatous cholecystitis using multiple percutaneous biopsies

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INTRODUCTION Xanthogranulomatous cholecystitis (XGC) is an uncommon form of cholecystitis presenting with nonspecific symptoms that mimic gallbladder cancer. Although the clinical and radiologic features of XGC have been described in detail, the exact diagnosis of XGC is virtually impossible. Fine-needle biopsy plays an important role in differentiating between gallbladder cancer and XGC.

CASE DESCRIPTION A 72-year-old man was admitted to the hospital with complaints of spiking fever and dyspepsia lasting 1 month. Blood sample was sent for bacterial culture, and the patient was given parenteral piperacillin/tazobactam. Abdominal computed tomography (CT) was performed to identify the focus of intra-abdominal infection. The result of bacterial culture was negative, and CT showed irregularly shaped gallbladder with asymmetric wall thickening suggestive of cancer. Infiltration of the surrounding hepatic parenchyma and a low attenuated mass-like lesion in liver segment 7 suggested cancer invasion and metastasis. On physical examination, the abdomen was soft without tenderness. Blood tests showed a white cell count of $10.31 \times 10^5/L$, C-reactive protein (CRP), 14.10 mg/dl; total bilirubin, 1.0 mg/dl. Liver function tests were normal. Serum levels of CEA were 1.7 ng/ml and CA 19-9, 33.4 U/ml. The patient received antibiotics for 2 weeks, which resolved fever and reduced CRP levels to 1.12 mg/dl. However, follow-up abdominal CT showed constant lesion with the same morphology and size as the previous scan. Percutaneous liver biopsy of the lesion on segment 7 was performed, and the specimen showed nonneoplastic hepatic parenchyma with minimal portal inflammation. The percutaneous biopsy of the thickened gallbladder wall again showed nonneoplastic hepatic parenchyma. The results of acid-fast staining and polymerase chain reaction for *Mycobacterium tuberculosis* were negative. The patient underwent the third percutaneous biopsy of the hepatic hilar mass, which revealed xanthogranulomatous inflammation. He was discharged on oral antibiotics. He received multiple endoscopic biliary procedures for benign biliary stricture. Follow-up CT scan showed improvement of gallbladder mass, inflammation, and biliary stricture.

DISCUSSION XGC is an uncommon disease that mimics gallbladder cancer. Radiologic findings were highly suggestive of malignancy but 2 needle biopsies failed to confirm the diagnosis. XGC can coexist with, and may predispose to, gallbladder cancer. Complete resection

of gallbladder is the treatment of choice, but it can be difficult due to severely destructive inflammation around the vessel and the bile ducts, as in our case. Careful monitoring for subsequent obstructive cholangitis or malignancy is needed.

LESSONS TO BE LEARNED FROM THE CASE Gallbladder masses are commonly encountered in clinical practice and have numerous causes. Physicians and radiologists should be familiar with the differential diagnosis to avoid misdiagnosis. Clinical suspicion is very important and early multidisciplinary cooperation is needed in this complex case.

Key words

benign gallbladder mass, gallbladder cancer, xanthogranulomatous cholecystitis

JUSTYNA CHUDIO

Bean syndrome: a rare cause of iron deficiency anemia

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Bean syndrome, or blue rubber bleb nevus syndrome, can be an unusual cause of iron deficiency anemia resulting from gastrointestinal bleeding. The etiopathogenesis of this rare vascular malformation syndrome is unclear. It occurs mostly sporadically as autosomal dominant inheritance has also been reported. It is characterized by multiple recurrent hemangiomas in the skin and visceral organs. There have been only about 200 case reports of Bean syndrome since the first description in 1860.

We report a case of a 32-year-old woman with Bean syndrome, who was referred to our department with severe iron deficiency anemia and vascular malformations in the skin and oral cavity. The patient was treated with iron supplementation for many years, and the cause of anemia remained undiagnosed. Gastroscopy, colonoscopy, and ^{99m}technetium-labelled red blood cell scintigraphy revealed hemangiomas in the esophagus, ascending colon, and rectum. Magnetic resonance imaging of the head did not show hemangiomas in the central nervous system, but malformations in the subcutaneous tissue of the left temporal region and the right long muscle were revealed. After 2 endoscopic interventions, the level of hemoglobin was satisfactory, and the patient required only oral iron supplementation and close follow-up.

Patients with Bean syndrome usually live a long life, but its quality might be limited due to episodes of gastrointestinal bleeding, oral drug therapy, and blood transfusion. Because Bean syndrome is extremely rare, there is no standard of care. In most cases symptomatic treatment is sufficient. Attempts are also made at pharmacological treatment. Unfortunately, its sustained long-lasting effects are doubtful. In the most severe cases, a segmental resection of the intestine might be necessary. Since multiple recurrent hemangiomas are highly likely, examination of other systems and organs, as well as close follow-up, is crucial in every case of Bean syndrome. Occasionally more sophisticated diagnostic procedures might be required to localize the lesions.

We decided to report this case to raise awareness of Bean syndrome and to emphasize that the cause of anemia should be thoroughly investigated in every case of iron deficiency anemia. Even though gastrointestinal bleeding is not likely to be the reason of anemia, it should be carefully considered.

Key words

Bean syndrome, blue rubber bleb nevus syndrome, iron deficiency anemia, vascular malformations

PAULINA GORZELAK-PABIŚ

From angioma to sarcoma: a very rare and aggressive splenic neoplasm with poor prognosis

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Primary splenic angiosarcoma is an extremely rare neoplasm of mesodermal origin with a high degree of malignancy and rapidly developing metastases. The metastatic index ranges from 69% to 100%, and the main metastatic sites are the liver (89%), lungs (78%), lymph nodes (56%), and bones (22%).

This report presents a case of a 78-year-old patient hospitalized for nonspecific pain in the left upper quadrant of the abdomen. Laboratory tests on admission showed mild anemia (hemoglobin, 10.9 g/dl), thrombocytopenia (98×10^9 /l), erythrocyte sedimentation rate of 160 mm, as well as D-dimer levels of 5055 ng/ml and alkaline phosphatase levels of 277 U/l. During hospitalization, abdominal ultrasound was performed twice due to severe pain in the left subcostal region. It revealed an enlarged spleen with angioma-like lesions, with the largest one of $81 \times 54 \times 68$ mm in size. The diagnostic workup was extended by a computed tomography scan of the abdomen and thorax and bone marrow biopsy. The patient was consulted by a surgeon and after 3 weeks of hospitalization in our department, he was transferred to the Department of Vascular Surgery, where he was diagnosed with splenic angiosarcoma based on a histopathological examination following splenectomy.

Three weeks after the diagnosis, the patient was admitted to the Department of Chemotherapy. A computed tomography scan showed multiple small liver metastases and infiltration in the gastric and peritoneal region. Due to an advanced stage of the disease and serious clinical condition, as well as low potential responsiveness to chemotherapy and a very high risk of complications, the patient was not considered eligible for chemotherapy. He was transferred to the Department of Palliative Care for further symptomatic treatment.

The reported case proves that clinical and radiologic symptoms of splenic angiosarcoma are very atypical. This makes the diagnosis particularly difficult and challenging. The annual incidence rate of this malignancy is between 0.14 and 0.25 of cases per one million, with the peak incidence in patients aged 50 to 60 years. The differential diagnosis of splenomegaly accompanied by nonspecific pain in the left abdominal region and decreased morphological parameters should also include angiosarcoma.

Key words

angiosarcoma, primary, spleen, tumor

ALONSO HEUDEBERT

Azathioprine-induced DRESS mimicking ANCA vasculitis

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INTRODUCTION DRESS syndrome (drug reaction with eosinophilia and systemic symptoms) is a rarely described complication of azathioprine therapy. The diagnosis was particularly challenging in the reported case as it mimicked the patient's previous antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis.

CASE A 60-year-old male patient with a history of renal-limited perinuclear ANCA (P-ANCA)-positive vasculitis, treated with azathioprine, presented to a referring hospital with malaise, fever, and rash. One year prior to presentation, the patient was diagnosed with renal vasculitis based on laboratory studies showing acute kidney

injury, red cell casts in urine, and P-ANCA positivity. A renal biopsy was deferred as the patient was on treatment with clopidogrel to maintain patency of a recently placed drug-eluting coronary stent for acute coronary syndrome. For renal vasculitis, he was treated with 10 cycles of intravenous cyclophosphamide. Three days prior to presentation at the referring hospital, he began azathioprine maintenance therapy. His initial evaluation revealed significant leukocytosis and relative hypotension. He was started on broad-spectrum antibiotics. Over the next 2 days, his rash progressed. He was treated with intravenous methylprednisolone and transferred to Barnes-Jewish Hospital. On arrival, initial laboratory test results were notable for acute kidney injury with elevated creatinine levels to 3.1 mg/dl (baseline ~2.0) and troponin levels to 0.3 ng/ml, neutrophil predominant leukocytosis of 12.2 K/cumm (down from peak of 24 at the referring hospital), and mild right-sided infiltrates on chest X-ray. Physical examination was notable for erythematous macules and papules coalescing across the trunk. Urine obtained from the patient and examined under microscopy revealed white blood cell casts but no dysmorphic red blood cells. A chest computed tomography showed ground glass opacities in the right middle lobe. The patient was consulted by a dermatologist and a skin biopsy was obtained, which revealed dermal edema and inflammatory infiltrates composed of lymphocytes, neutrophils, and eosinophils. The overall picture was consistent with DRESS syndrome involving the kidney, lungs, and heart. The patient's clinical course showed improvement in the subsequent days. He was transitioned from azathioprine to mycophenolate and discharged on a slow prednisone taper. Subsequent outpatient follow-up approximately 20 days after initial presentation showed excellent control of autoimmune disease, rash resolution, and renal function approaching baseline.

DISCUSSION This case illustrates a rare complication of azathioprine use and provides an interesting diagnostic challenge as the presentation could have been mistaken for a relapse of previous renal-limited ANCA-associated vasculitis. Finally, failure to identify the primary process and discontinue the offending agent could have resulted in delay of care.

Key words

azathioprine, clinical reasoning, DRESS

TEREZA HLAVATA

Unexpected cause of acute kidney injury

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Leptospirosis is a worldwide zoonosis presenting by a wide spectrum of clinical manifestations that range from subclinical infection to multiorgan failure and death. The most common triad of symptoms includes fever, jaundice, and acute renal failure. A severe form of leptospirosis is known as Weil disease and reported mortality rates range from 10% to 15%. It is common in tropical and subtropical environments but rare in Central Europe. The disease is transmitted by contact with water or soil contaminated by the urine of *Leptospira*-infected animals. Humans are infected with the contaminated environment through the skin defect or by gastrointestinal route. The diagnosis is established on the basis of clinical presentation and detection of *Leptospira* by serological methods and polymerase chain reaction–DNA analysis.

Here, we present a case of 41-year-old man with a recently diagnosed, untreated hepatitis C, presenting with fever, diarrhea, and vomiting. Initial assessment revealed inflammatory syndrome and acute kidney injury. Within the subsequent 24 hours, signs of rhabdomyolysis, hemolysis, and worsening of kidney function had developed. The patient received 3 sessions of hemodialysis with

prompt renal recovery. The first-line empirical antibiotic treatment with ciprofloxacin and metronidazole had to be adjusted due to persistent fever and high inflammatory parameters. On day 4, cefotaxime was added. After a few days on treatment, we observed a major clinical improvement with a drop of inflammatory parameters and full recovery of renal function with normal diuresis. At discharge, after successful antibiotic treatment, platelet count and hemoglobin levels normalized, and a presumptive diagnosis of leptospirosis was established on the basis of positive microscopic agglutination test results with elevated titer and a paradox reaction typical for an early phase of leptospirosis. The diagnosis of Weil disease was confirmed on the basis of another 2 blood samples within 2 weeks after discharge, which showed increased titer of *Leptospira interrogans* serovar *icterohaemorrhagiae* antibodies to 1:6400 and 1:12800, respectively.

Leptospirosis is an underreported infectious disease and should be considered as a cause of acute kidney injury, especially if a triad of symptoms of fever, jaundice, and acute kidney injury are present. The physician should be alert to the probable diagnosis of leptospirosis especially in countries where the disease is considered as low risk and can be easily overlooked. Importantly, even some leptospirosis cases can present as anicteric, and still they can have a serious course. Prompt recognition and administration of the right antimicrobial therapy is the key to reducing the risk of mortality.

Key words

acute kidney injury, hemodialysis, leptospirosis, Weil disease

LEONEL LANGELOTTI

Autoimmune encephalitis hiding behind psychosis

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INTRODUCTION Limbic encephalitis is a rare disease characterized by an acute or subacute clinical condition presenting with affective, amnestic, neurovegetative, and sleep disorders, all of which are associated with inflammatory signs in the cerebrospinal fluid, electroencephalographic disturbances, and limbic gyrus injuries. Although initially the disease was related to neoplasms, such as paraneoplastic autoimmune encephalitis, cases of an autoimmune nonparaneoplastic origin have been recently reported.

CASE DESCRIPTION A 25-year-old man with a history of mild smoking, currently in a stable relationship and with no family history of limbic encephalitis, presented to the emergency room due to high temperature, generalized tonic-clonic seizures, amnestic gaps, disorientation to time, place, and person, and psychomotor agitation. A brain computed tomography (CT) scan on presentation showed no abnormal findings, lumbar puncture with clear and colorless cerebrospinal fluid (CSF), and normal protein and glucose concentrations with no cells. Meningoencephalitis was suspected, and the patient was placed in an isolated room and administered empirical treatment with ceftriaxone and acyclovir, following a 48-hour treatment with negative cultures. During the hospital stay, the patient showed no convulsive episodes and remained afebrile, although the appearance of suicidal ideation led to a psychiatric consultation, which resulted in the diagnosis of the first signs of schizophrenia. Treatment with haloperidol was provided, and the patient was discharged 72 hours after admission.

The patient was readmitted 48 hours later due to a new episode of generalized tonic-clonic seizures associated with visual and auditory hallucinations. Laboratory tests and brain CT revealed no lesions, but CSF obtained by lumbar puncture, although still clear and colorless, showed a much higher protein concentration, normal glucose concentration, and negative staining. Encephalitis was suspected and treatment with acyclovir and diphenylhydantoin was started, but right faciobrachial dystonic crisis persisted. In addition,

physical examination showed disorientation to time and place as well as psychomotor agitation. The patient was negative for hepatitis B virus, hepatitis C virus, and HIV infection. Rheumatic and CSF viral panel was also negative, while electroencephalography showed evidence of diffuse slowing at 6 Hertz, with paroxysmal discharges of generalized sharp waves, spike-wave type, at 3 Hertz. In addition, magnetic resonance imaging showed abnormal hyperintense signals in T2-weighted and FLAIR sequences in the parainsular area and the hippocampus head, with an asymmetric pattern. Brain, thoracic, abdominal, and pelvic CT revealed no signs of neoplasms. Due to suspicion of immune-mediated encephalitis, treatment with methylprednisolone was started. The patient's condition improved, with positive results for anti-voltage-gated potassium channel antibodies in CSF.

DISCUSSION A diagnostic workup in a patient suspected of autoimmune encephalitis must include electroencephalography, imaging tests, lumbar puncture, serologic tests for infectious diseases and presence of antibodies against NMDA, AMPA, GABA-r, HU, CV2, VGKC (LGI1, CASPR2), MA2, as well as a thoracic CT scan. The benign evolution of numerous types of limbic encephalitis leads to the suspicion of reversible pathogenic mechanisms. Nevertheless, a neoplasm must be excluded in every case.

CONCLUSION Autoimmune encephalitis is a rare disease that is often misdiagnosed and treated as a psychiatric disease, thus delaying the proper diagnosis and favoring progression of the condition and development of its sequelae.

Key words

autoimmune encephalitis, limbic encephalitis

NÓRA LEDÓ

Unusual manifestation of antineutrophil cytoplasmic antibody-associated vasculitis in a young man

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INTRODUCTION Systemic vasculitides associated with antineutrophil cytoplasmic autoantibodies (ANCA) have an extremely wide variety of symptoms; therefore, fast and proper diagnosis is difficult to establish even for experienced physicians. Gastrointestinal manifestations in ANCA-associated granulomatosis with polyangiitis (GPA) may be present; however, severe, life-threatening complications (such as perforations) are rare.

CASE DESCRIPTION An 18-year-old previously healthy white man was admitted to our nephrology department with elevated serum creatinine levels. His symptoms started 2 weeks earlier with severe abdominal pain, vomiting, hematemesis, and diarrhea, when inflammatory bowel disease was diagnosed. On admission, the patient still had moderate umbilical abdominal pain with the clinical symptoms of rapidly progressive glomerulonephritis. Urgent kidney biopsy and laboratory tests were performed, including autoimmune panels. Anti-proteinase 3 ANCA positivity (461 U) was confirmed, while pauci-immune, crescentic glomerulonephritis with the signs of vasculitis was detected in kidney biopsy. Based on these results, GPA with rapidly progressive glomerulonephritis was diagnosed and high-dose intravenous methylprednisolone, intravenous cyclophosphamide, and plasma exchange therapy were started, while the previously administered antihypertensive and proton-pump inhibitor therapies were continued. Because of progressive renal function decline, the patient was put on hemodialysis. He developed severe anemia (with a hemoglobin level of 67 g/l); therefore, he underwent examinations to exclude extrarenal bleeding. Upper gastrointestinal endoscopy was performed, which showed normal esophagus, stomach, and duodenum without any signs of bleeding or inflammation.

Colonoscopy was also scheduled in 2 weeks. Chest X-ray and high-resolution computed tomography scan excluded bleeding from the respiratory tract. The patient was discharged on the 24th day after admission with stable hemoglobin levels. Three days later, he was readmitted to our hospital in a rapidly progressive hemorrhagic shock with hematochezia and hematemesis. Gastroscopy revealed arterial bleeding of the duodenum. The bleeding was controlled with 3 clips placed on the artery. Two days later, the bleeding restarted next to the clips, and the patient needed surgery to suture the artery. Three days after the first surgery, a reoperation was needed because of duodenal perforation. After an additional 22 days of care, the patient recovered from the second surgery, and could be discharged with continuing immunosuppressive therapy.

CONCLUSIONS Although it is known that gastrointestinal manifestations are possible in GPA, only a few cases have been reported with gastrointestinal symptoms as the first signs of the disease. In our patient, gastrointestinal symptoms (abdominal pain, vomiting, diarrhea) were the first and, for several days, the only signs of GPA. The initial diagnosis of inflammatory bowel disease delayed the administration of proper immunosuppressive therapy, which might have contributed to the rare and life-threatening complication of arterial duodenal bleeding with perforation. Therefore, we suggest that the differential diagnosis of such abdominal symptoms should include laboratory measurement of ANCA levels.

Key words

ANCA, gastrointestinal manifestation, glomerulonephritis, vasculitis

BRETT MANSFIELD

Spinal cystic echinococcosis

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INTRODUCTION Cystic echinococcosis (CE), or hydatid disease, is a neglected tropical disease affecting predominantly rural communities throughout many parts of the world. The disease remains dormant for many years and only comes to medical attention when it leads to compression of surrounding structures or cyst rupture. CE most commonly occurs in the liver and lungs. On rare occasions, it may manifest in the spine. Management of spinal CE is difficult. Surgery is the mainstay of treatment, and concomitant long-term antiparasitic agents are required. The disease is associated with high rates of morbidity and recurrence.

CASE REPORT A 38-year-old male patient presented with lower back pain, which had been present for 14 months. The pain was initially intermittent but had worsened and began radiating down the back of his legs. He had no past medical or surgical history. He had grown up in Port Elizabeth in the Eastern Cape province but had moved to Johannesburg 7 years earlier.

Spinal magnetic resonance imaging showed vertebral body destruction with multiple cysts attenuating the spinal cord. The patient underwent a vertebral corpectomy, and a vertebral cage was inserted. Histopathological examination confirmed the presence of hydatid cysts. Echinococcus serology was positive, at a titer of 128. The patient was started on albendazole, 400 mg orally twice daily. He was discharged home 2 weeks after surgery and was followed on an outpatient basis.

DISCUSSION CE, which affects more than a million people worldwide, is caused by the larval stages of the cestode *Echinococcus granulosus*. The parasite's life cycle involves sheep as the intermedi-

ate host and the dog as the definitive host. Humans are incidental intermediate hosts. Hydatid cysts most commonly occur in the liver (70%) and lungs (20%). Rarer manifestations of the disease include the kidneys, brain, heart, ovaries, peritoneum, and bone. Fifty percent of CE affecting bone occurs in the vertebral column. Radiologically, spinal CE may appear as a “bunch of grapes”. This is not pathognomonic of spinal CE, but vertebral body destruction with paravertebral extension as well as contiguous extension to a rib is very suggestive of the disease. Surgery is the first line of treatment. Expert consensus recommends at least 6 months of albendazole. The disease has high rates of morbidity. Recurrence rates may be as high as 48% over a 2-year period.

CONCLUSION Spinal CE is a rare manifestation of hydatid disease but should be considered in those known to have lived in highly endemic regions who have suggestive radiological features on computed tomography or magnetic resonance imaging scan. Many patients require multiple surgical procedures and poor outcomes result in many of them being left with some degree of neurological fallout. Concomitant albendazole is recommended for at least 6 months. Spinal CE has high rates of recurrence, and long-term follow-up with serial imaging is required.

Key words

cystic echinococcosis, hydatid, spine

MAŁGORZATA MARCZEWSKA

A 62-year-old man with pruritic skin rash, lymphadenopathy, fever, and renal and hepatic involvement: challenges in the diagnosis of DRESS syndrome

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A 62-year-old man with bipolar disorder and hypertension, previously treated with systemic and topical glucocorticoids at a dermatology department for allergy-related diffuse pruritic skin lesions, fever, and lymphadenopathy, was admitted to an internal medicine department with suspicion of cutaneous T-cell lymphoma. Three days after discharge from the dermatology department, he developed skin lesions, along with thickening of the facial features. In addition, biochemical evidence of renal and hepatic injury was observed. Laboratory, imaging, and histopathologic examinations excluded cutaneous lymphoma (Sézary syndrome, mycosis fungoides), supporting the initial diagnosis of adverse drug reaction: carbamazepine-induced drug reaction with eosinophilia and systemic symptoms (DRESS syndrome). On discontinuation of all previous medications, skin, renal, and hepatic abnormalities resolved. This case shows the difficulty in diagnosing allergic drug-reactions involving multiple organ systems, as well as the difficulty in identifying the culprit medication. Further research and standardization of this syndrome are warranted.

Key words

carbamazepine, drug eruptions, drug-induced hypersensitivity syndrome, drug reaction with eosinophilia and systemic symptoms

JIN HEE NOH

Malignant melanoma on the pancreas and liver mimicking a neuroendocrine tumor

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INTRODUCTION Malignant melanoma is a malignant tumor originating from the melanocyte derived from the neural crest. It is mainly found in the skin and is not common in non-skin organs such as the eyes, gastrointestinal tract, genitourinary tract, and pleura. A case of malignant melanoma involving the gastrointestinal tract without skin lesions or history of melanoma has not been reported previously in Korea. We report a rare case of malignant melanoma only involving the pancreas and liver without the primary site, which was difficult to differentiate from a pancreatic neuroendocrine tumor with liver metastasis.

CASE DESCRIPTION A 65-year-old man reported with a chief complaint of anorexia and body weight loss for 1 month. Abdominal ultrasound performed in a local clinic showed suspicious findings of pancreatic cancer with liver metastasis. The patient was transferred to our hospital. On admission, general physical examination and vital signs were normal. His history was unremarkable except for the use of antihypertensive medication. He was a social drinker and a nonsmoker. Computed tomography (CT) and contrast-enhanced endoscopic ultrasonography (EUS) revealed hepatic and pancreatic hypervascular tumors. Based on imaging findings, he had been provisionally diagnosed with pancreatic neuroendocrine tumor with liver metastasis. He did not have any lesions of the skin, eye, or gastrointestinal tract on physical examination, upper endoscopy, and whole-body positron emission tomography. Eventually, EUS-guided pancreatic core biopsy and percutaneous ultrasonography-guided liver biopsy confirmed malignant melanoma of the pancreas and liver. The patient was transferred to the oncology department for systemic chemotherapy and was administered pembrolizumab, an immune checkpoint inhibitor. He received the second cycle of medication and is currently under regular follow-up at the outpatient clinic.

DISCUSSION The primary lesions cannot be found in about 10% of malignant melanomas. There are several hypotheses that explain the cause of malignant melanoma of unknown primary site, with the spontaneous regression of the primary site as the most widely accepted theory. When malignant melanoma has metastasized to other organs, the primary lesions can spontaneously regress. In this case, the dermatologist and ophthalmologist examined the patient and the upper gastrointestinal endoscopy was performed, but the primary site was not confirmed. Abdominal CT showed multiple hypervascular masses in the pancreas and liver. EUS showed a clear hypoechoic mass in the pancreatic body. On contrast-enhanced EUS, the mass was enhanced homogeneously in the arterial phase and washed out in the venous phase. The hypervascular mass such as neuroendocrine tumor may demonstrate similar imaging. Therefore, we strongly suspected that this was a neuroendocrine tumor with liver metastasis. However, the pancreas and liver biopsy specimens did not confirm this.

LESSONS TO BE LEARNED FROM THE CASE We presented a rare case of malignant melanoma, which only involved the pancreas and liver without primary lesions, mimicking a neuroendocrine tumor on imaging. It is important to consider malignant melanoma when imaging findings reveal pancreatic tumors such as neuroendocrine tumor. Although melanoma is a rare metastatic pancreatic tumor, it should be included in a differential diagnosis.

Key words

malignant melanoma, neuroendocrine tumors, pancreatic metastasis

Sheehan syndrome presenting as postpartum hyponatremia

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INTRODUCTION Sheehan syndrome, a postpartum pituitary necrosis, may cause different pituitary hormonal deficits, depending on which pituitary area is malfunctioning. The syndrome has low incidence in developed countries. On the other hand, hyponatremia is a common laboratory finding in multiple conditions, including hormonal deficiency.

CASE DESCRIPTION A healthy 24-year-old woman in the 39th week of pregnancy (G2P2 according to the gravida/para/abortion system), with a normal pregnancy follow-up, presented to the obstetric emergency department due to uterine contractions. Due to no progress of labor, labor induction was performed. Because of a nonreassuring fetal heart rate, the patient underwent an emergency caesarean section. On the second postoperative day, she complained of severe headache, fatigue, and failure to lactate. Brain computed tomography (CT) scan demonstrated an abnormal unspecific suprasellar mass. On day 5, the patient developed fever of 39°C for the first time during hospitalization. Abdominal CT scan excluded an abdominal source of infection. Blood and urine cultures were negative. A routine biochemical investigation revealed new-onset hyponatremia (sodium level, 124 mEq/l; on admission, 146 mEq/l; reference range, 135–145 mEq/l). Urine analysis showed osmolality of 572 mOsmol/kg and a sodium level of 149 mEq/l consistent with the syndrome of inappropriate antidiuretic hormone secretion. She was treated with fluid restriction, furosemide, salt tablets, and eventually, hypertonic saline. Sodium levels failed to increase. Physical examination on admission showed weakness, lethargy, and altered mental status. Blood pressure was at the low end of the reference range (90/50 mm Hg). Thyroid function tests were consistent with central hypothyroidism. A complete hormonal workup demonstrated anterior hypopituitarism. Sheehan syndrome and lymphocytic hypophysitis were considered, and brain magnetic resonance imaging (MRI) was performed to differentiate between the 2 conditions. The patient was initially treated with intravenous hydrocortisone followed by thyroid hormone replacement, which resulted in a rapid clinical and biochemical response, including reversal of hyponatremia. Brain MRI on day 17 was consistent with Sheehan syndrome. Several days after discharge, the patient developed polyuria and nocturia. Dehydration test was consistent with partial central diabetes insipidus, with a good response to desmopressin. On follow-up visits in the endocrine clinic, estrogen-based hormone replacement therapy and growth hormone treatment were added.

DISCUSSION Although rarely, Sheehan syndrome may develop without any obvious postpartum bleeding and in a developed country with modern medical care, as in the presented case. The condition should be differentiated from lymphocytic hypophysitis. The differential diagnosis is primarily based on the presence of a major postpartum hemorrhage, distinctive brain MRI findings, higher rates of posterior pituitary failure, and a concurrent autoimmune disease in lymphocytic hypophysitis. Sheehan syndrome may lead to abnormalities in serum sodium levels. Hyponatremia may develop in the setting of posterior pituitary necrosis and central diabetes insipidus. It is a more common manifestation and occurs when adrenal insufficiency or hypothyroidism develops, as they are both etiologies of euvolemic hyponatremia. Hyponatremia, and specifically euvolemic hyponatremia, may have numerous etiologies and all must be considered in a diagnostic workup. As common findings may be caused by rare conditions, clinicians should always consider an alternative diagnosis upon treatment failure.

Key words

adrenal insufficiency, central hypothyroidism, hyponatremia, hypopituitarism, Sheehan syndrome

PETER UHRÍK

Complicated diagnosis of insulinoma localization

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INTRODUCTION Insulinoma is a rare pancreatic neuroendocrine tumor (PNET), with an incidence of 1 to 4 cases per million persons per year. Clinically, insulinoma is characterized by the Whipple triad (presence of hypoglycemia with clinical symptoms and its resolution after glucose administration). Biochemical diagnosis is based on blood glucose, insulin, C-peptide, and proinsulin hydroxybutyrate levels, and absence of sulphonylurea metabolites in plasma or urine. The recommended diagnostic algorithm includes ultrasound, computed tomography (CT), magnetic resonance imaging (MRI), positron-emission tomography (PET), and endoscopic ultrasound (EUS). EUS can be extended by elastography (EUS-EG) and contrast agent administration (CE-EUS).

CASE REPORT A 15-year-old previously healthy woman was initially admitted to an emergency department with less serious signs of hypoglycemia. Laboratory studies showed a glucose level of 1.2 mmol/l and an insulin level of 9.6 µU/ml. Complete blood count, metabolic profile, C-reactive protein activity, and urine drug test results were normal. Glucose treatment led to symptom relief. A fasting test confirmed biochemical signs of insulinoma. In June 2016, the first CT, multiphase CT, and EUS were performed with negative or less convincing results. The diagnostic workup was gradually extended by PET-CT and MRI using fluorodopa and ⁶⁸Ga-DOTATOC. Only atypical changes of the pancreatic head were recorded. In March 2017, we implemented an extended EUS. EUS-EG showed a consistent reduction in elasticity in the particular region of interest in the pancreatic head; the strain ratio index was 10.38. After administration of a contrast agent (sulfur hexafluoride), we observed hypoenhancement of the lesion on CE-EUS and hypervascularization on conventional Doppler-mode EUS. We concluded that the insulinoma was localized in the pancreatic head. To confirm these findings, somatostatin receptor scintigraphy (SRS) and ⁶⁸Ga-DOTATATE PET-CT were performed, but no abnormalities in the pancreatic tissue were found. However, the site and type of the tumor were confirmed after a successful operation.

DISCUSSION Insulinomas are relatively small at the time of diagnosis. Despite advances in imaging techniques and the development of new localization procedures, tumors of less than 2 cm in size remain difficult to localize by conventional methods. The diagnostic algorithm recommends moving from less invasive imaging techniques to more invasive examinations. Transabdominal ultrasound has extremely low sensitivity in the diagnosis of PNETs (9%–22%). There are no selective data on the accuracy of CT and MRI in localization of insulinomas; nevertheless, their diagnostic sensitivity ranges from 54% to 94%. According to published data, the sensitivity of SRS is less than 50%, and of F-DOPA PET-CT, lower than 63%. They are not generally recommended for diagnosing insulinoma. A better option is to fuse CT or MRI images with PET ⁶⁸Ga-DOTATOC or DOTATATE. The published data show the high specificity and sensitivity of EUS in the diagnosis of PNETs (95%–97% and 67%–76%, respectively). CE-EUS can be used for differentiation between adenocarcinoma and other pancreatic tumors, with a high specificity (94%) and

sensitivity (89%). It can be very difficult to precisely localize an insulinoma by conventional imaging methods. EUS-EG and CE-EUS seem to be accurate and useful tools for this purpose.

Key words

contrast enhancement, elastography, endoscopic ultrasound, insulinoma

KATARZYNA WAWRZYCKA-ADAMCZYK

Recognized for best poster

How to look ataxia-telangiectasia syndrome in the eye?

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Ataxia-telangiectasia is a rare disorder caused by the *ATM* gene mutation, presenting with ataxia, immunodeficiency, telangiectasia, and increased incidence of cancer. We present a case of a 28-year-old woman with suspected immunodeficiency. Her medical history included the diagnosis of cerebral palsy, chronic bone infections leading to severe forearm deformation, and premature deaths in first-degree family members. Further tests revealed low immunoglobulin concentrations and CD4⁺ T-cell depletion. Complex immunodeficiency syndrome and secondary pyoderma gangrenosum were diagnosed, and a combination treatment with steroids and monthly intravenous immunoglobulin was administered. Skin ulcerations resolved. After 3 years, the patient presented with neck lymphadenopathy, diagnosed as peripheral T-cell lymphoma. Childhood medical history revealed that she developed gait impairment at the age of 3, which progressed in her teens. Neurological examination confirmed cerebellar ataxia and mild telangiectasias in the eyes. Genetic tests revealed an extremely rare coexistence of ataxia-telangiectasia syndrome and the *UNC119* mutation (idiopathic CD4 lymphocytopenia).

Key words

ataxia-telangiectasia syndrome, immunodeficiency

JI WON LEE

Primary mesenteric choriocarcinoma in a 26-year-old man

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INTRODUCTION Choriocarcinoma is a germ cell tumor containing cells of trophoblastic origin. It rarely occurs in the extragonadal sites such as the mediastinum, lung, or retroperitoneum, especially in men. Gynecomastia is a benign proliferation of the glandular tissue of male breast. It is usually the result of an increased estrogen to androgen ratio. Certain medical conditions can predispose to gynecomastia, such as use of medications, malnutrition, hyperthyroidism, hypogonadism, and tumors.

CASE DESCRIPTION INCLUDING CLINICAL SITUATION, COURSE OF EVENTS, AND CLINICAL RESOLUTION A 26-year-old man was admitted to our hospital, because of an abdominal mass. Six months before admission, the patient had undergone a left mastectomy for gynecomastia. On physical examination, he had a palpable, solid, and irregular mass in the periumbilical area without abdominal tenderness. An abdominal computed tomography (CT) scan demonstrated a mass of 78 mm in size, with hypervascularity in the small bowel mesentery. A lymph node of 10 mm in size was also found in the aortocaval area. The patient underwent a laparoscopic excision of

the mass and lymph node, and a histologic examination revealed syncytiotrophoblasts and cytotrophoblasts in both specimens. Immunohistochemical stains were positive for β -human chorionic gonadotropin (β -HCG), glypican, and GATA3. The mesentery mass was finally diagnosed as primary mesenteric choriocarcinoma. Metastatic evaluation by positron-emission tomography CT (PET-CT) showed a hypermetabolic lymph node in the left supraclavicular, aortocaval, retrocaval, left paraaortic, and iliac bifurcation space. After the surgery, serum β -HCG and estradiol levels were 38 668 mIU/ml and 102 pg/ml, respectively. The patient was treated with 5 cycles of palliative chemotherapy with cisplatin, methotrexate, dactinomycin, and etoposide. After the chemotherapy, β -HCG and estradiol levels normalized (0.4 mIU/ml and 8.2 pg/ml, respectively). PET-CT showed complete resolution of the hypermetabolic lymph nodes, and gynecomastia in the right breast also improved. There is no evidence of cancer progression or relapse at 1-year follow-up.

DISCUSSION Primary choriocarcinoma in the gastrointestinal tract is extremely rare. Only 16 cases of primary colonic choriocarcinoma were reported in the literature. To the best of our knowledge, there have been no previous reports of primary choriocarcinoma of the mesentery.

Several hypotheses have been suggested as the pathogenesis of primary extragonadal choriocarcinoma, including dedifferentiation of a preexisting adenocarcinoma; development from an underlying teratoma; metastasis from a latent primary unknown lesion in the genitalia; and development from primordial germinal cells migrated abnormally during the embryogenesis. In this case, choriocarcinoma might have developed from a malignant lesion in the ectopic chorion or totipotent cells.

Prognosis of extragonadal choriocarcinomas is extremely poor. Most treatment options are based on regimens designed to treat choriocarcinoma of gestational trophoblastic neoplasia. The survival rate in patients who received chemotherapy was significantly higher than that for patients without chemotherapy.

LESSONS TO BE LEARNED FROM THE CASE It should be recognized that choriocarcinoma can affect the mesentery and it should be considered as a rare cause of gynecomastia.

Key words

choriocarcinoma, gynecomastia, mesentery

ERIKA ZECCA

First report of meningitis due to *Staphylococcus condimenti* in a human

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Staphylococcus condimenti (*S. condimenti*) is a coagulase-negative member of the bacterial genus

Staphylococcus, generally regarded as not pathogenic. Indeed, *S. condimenti* derives its name from the fact that it has been isolated from starter cultures of fermented sausage, as well as from fish and soy sauces. To date, to the best of our knowledge, only 2 cases of human infection caused by this bacterium have been reported. Here, we present a case of meningitis due to *S. condimenti* in a 65-year-old woman who was brought to hospital after having been found unconscious at home. On presentation, she had fever and type 1 respiratory failure. The Glasgow Coma Scale score was 3, with muscular flaccidity and absence of meningeal signs. There were no other abnormalities except for scattered rhonchi in both lungs. Cerebrospinal fluid examination showed a mildly increased white blood cell count, with normal glucose and protein concentrations. Paired cultures in blood and liquor samples revealed *S. condimenti*. Targeted antibiotic treatment with ceftriaxone led to complete recovery.

The route by which our patient acquired the infection is unclear. She had psychiatric problems, lived alone, and there were no witnesses to what had happened in the last 2 days before her admission; therefore, we had little details on her recent medical history. Our hypothesis is that she might have had an alimentary toxoinfection followed by blood invasion and dissemination to the central nervous system. Alternatively, the infection may have originated from periodontitis, which was diagnosed in the preceding weeks and was supported by an orthopantomogram confirming multiple foci of osteolysis.

Meningitides due to coagulase-negative *Staphylococci* are rare, except for those related to ventriculoperitoneal shunt placement. They have a good prognosis in comparison with other bacterial meningeal infections.

S. condimenti is often considered as contaminant, and potential virulence factors have been described. Proteins with similarity to leucocidin have been identified by gene sequencing, and they appear to be involved in immune evasion and to possess proinflammatory and cytolytic activity. In our case, the findings at presentation and the isolation of the same bacterium in blood and liquor samples were in stark contrast to the hypothesis of contamination, prompting us to start antibiotic treatment. The use of matrix-assisted laser desorption ionization time-of-flight mass (MALDI-TOF) spectrometry led to an accurate and fast isolation of the pathogen.

S. condimenti infections have been described as susceptible to all antibiotics tested. In our case, the bacterium was resistant to fosfomycin. This peculiarity may herald the development of further antibiotic resistance to this coagulase-negative *Staphylococcus*. Our unique case expands our knowledge on *S. condimenti* as a pathogenic bacterium.

Key words

antibiotic, coagulase-negative *Staphylococci*, contaminant, meningitis, *S. condimenti*

ALANOOD AHMED ASIRI

Acquired methemoglobinemia without a culprit

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INTRODUCTION Methemoglobin is a form of hemoglobin in which the iron of the heme group is oxidized from a ferrous to a ferric state. Its level is normally maintained at no more than 1% of total hemoglobin by endogenous enzymes, and an increase of its level is defined as methemoglobinemia. The ferric hemes are unable to carry oxygen; moreover, increased affinity of the accompanying ferrous hemes in the remaining normal hemoglobin causes a left shift in the oxygen dissociation curve and hypoxia. Methemoglobinemia has many potential causes and its outcomes could be fatal if left untreated. We report a case of a 44-year-old woman with methemoglobinemia without clear evidence of an acquired cause.

CASE REPORT A 44-year-old woman with a history of recurrent venous thromboembolism and diagnosed with protein S deficiency presented to her local hospital with deep vein thrombosis (DVT) of her right leg while on dabigatran. She was managed and then discharged on enoxaparin, but within a couple of weeks she developed left-leg DVT, dyspnea, chest pain, peripheral cyanosis, lightheadedness, and platypnea-orthodeoxia syndrome. SpO₂ was 88% in room air, with no abnormalities on cardiopulmonary examination; the arterial blood gas test showed a pH of 7.57, pCO₂ of 40 mm Hg, pO₂ of 167 mm Hg, and HCO₃ of 28 mEq/L. Further studies showed a methemoglobin level of 1.1%, a negative lower-limb Doppler and V/Q scan for filling defects, negative bubble echocardiogram and contrast-enhanced computed tomography of the chest and abdomen for atrioventricular shunts, and normal pulmonary function test, high-resolution computed tomography, and right heart catheter results. Based on these findings and the discrepancy between her pulse oximetry reading and arterial oxygen tension, we considered dysmethemoglobinemia. A methemoglobin level of 11.9% measured in an external laboratory supported methemoglobinemia; a repeated measurement after 8 weeks showed the level of 0.5%. With time, the patient showed significant improvement; however, a detailed history did not reveal a clear cause of acquired methemoglobinemia. Therefore, we postulated that she likely had acquired methemoglobinemia and may have had some form of cytochrome b5 reductase enzyme deficiency.

DISCUSSION Most cases of methemoglobinemia are due to acquired causes and can occur in healthy individuals of any age in the presence of severe oxidative stress. Conversely, inherited methemoglobinemia commonly manifests in infancy.

The *CYB5R3* gene encodes the cytochrome b5 reductase enzyme responsible for reducing methemoglobin to hemoglobin. In type I deficiency, which is a mild form, the enzyme is produced at a normal rate but is unstable, leading to a lower threshold for acquiring methemoglobinemia if exposed to oxidative stress, but not low enough to cause clinical symptoms otherwise.

It is important to have a high index of suspicion of methemoglobinemia in patients presenting with hypoxia where there is discordance between the oxygen level measured by pulse oximetry and arterial oxygen tension measured by arterial sampling. It is crucial to be aware of constraints in the local laboratory and to have readily available methemoglobin tests for the rapid diagnosis and treatment of methemoglobinemia, which was a limitation in our facility.

We advised our patient to avoid common medications and situations that could potentially cause methemoglobinemia.

Key words

cytochrome b5 reductase, hypoxia, methemoglobinemia

IMRE BODO

A 61-year-old man with fever, weight loss, and new-onset headache

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A 61-year-old man presented with fever up to 38.4°C, night sweats, and a new-onset unilateral headache for the past month. He experienced a weight loss of 15 kg. His medical and travel histories were unremarkable. Physical examination revealed moderate obesity without any other abnormal findings.

Baseline laboratory tests showed isolated normocytic anemia (hemoglobin, 106 [g/L]) with mild thrombocytosis (platelet count, 408 [G/L]), normal levels of lactate dehydrogenase ([U/L]) and thyroid-stimulating hormone ([mU/L]), and markedly elevated levels of inflammatory markers (erythrocyte sedimentation rate [ESR], 98 [mm/h]; C-reactive protein [CRP], 199 [mg/L], and ferritin, 1933 [μg/L]). Electrolyte levels as well as renal and liver function were normal. Serum electrophoresis showed a slightly elevated immunoglobulin G level with a polyclonal elevation in the γ-globulin fraction. Chest x-ray and head, chest, abdominal, and pelvic computed tomography (CT) scans were normal. Blood cultures were negative, and a transthoracic echocardiogram was normal. On repeat interview, the patient reported jaw pain. At this point, giant cell arteritis (GCA) was suspected. Histopathological examination of temporal artery biopsy showed no abnormalities.

A differential diagnosis of a febrile illness lasting several weeks and associated with weight loss as well as elevated ESR and CRP levels includes infectious, malignant, and immune disorders. Negative blood and urine cultures, normal procalcitonin levels, and a normal echocardiogram exclude systemic infection, such as infective endocarditis. Due to new-onset headache, we considered advanced malignancy with brain metastasis or an abdominal abscess, but they were both excluded by negative CT scans. On the other hand, fever, severe new-onset headache, and jaw claudication, especially in association with markedly elevated ESR and CRP levels, strongly suggested GCA and/or polymyalgia rheumatica, which was evaluated in this case by a temporal artery biopsy. Temporal artery biopsies are reported to have a sensitivity of 70% to 87%, with very high specificity if GCA is histologically confirmed. Thus, a negative biopsy is not enough to exclude the diagnosis if the pretest probability is sufficiently high. A positron emission tomography scan showed a high metabolic activity of the aorta and its thoracic branches. These images strongly suggested large artery vasculitis with Takayasu aortitis and GCA as primary considerations.

Vasculitis of the aorta and its branches raises the possibility of Takayasu aortitis or GCA. These 2 disorders used to be considered as separate entities, with Takayasu aortitis typically affecting young Asians (<40 years), whereas GCA, elderly Caucasian patients (>50 years). However, several recent reviews concluded that the 2 entities may be very closely related, probably as 2 ends of a single spectrum. Finally, it should be noted that sudden onset blindness, the most dreaded complication of GCA, has practically been absent in patients with negative temporal artery biopsy results.

In our patient, GCA with aortic, carotid, and subclavian artery involvement was diagnosed. He was started on a moderate-dose steroid regimen (oral methylprednisolone, 32 mg). He showed a prompt clinical improvement. As GCA tends to recur, the steroid should be tapered according to the patient's response.

Key words

fever of unknown origin, giant cell arteritis, headache, vascular

Fecal microbiota transplantation for severe alcoholic hepatitis: experience in our center

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Severe alcoholic hepatitis is one of the most serious forms of alcoholic liver disease; it is associated with high mortality, particularly in patients who are not suitable for corticosteroid (CS) treatment or in CS nonresponders as rated by the Lille model. Unfortunately, the number of these patients is increasing and there is no alternative treatment. More recently, attention has been paid to a possibly effective method of fecal microbiota transplantation (FMT). In our report, we discuss a case of a young woman with severe alcoholic hepatitis, a CS nonresponder with a high risk of short-term mortality. For the first time at our institution, we used FMT. Based on our case report, FMT appears to be a beneficial method that improves short-term survival in this patient population.

Key words

alcoholic liver disease, fecal microbiota transplantation, severe alcoholic hepatitis

FRANCISCO CONSIGLIO

A patient with AIDS: breaking the uncausal theory by means of clinical reasoning

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INTRODUCTION HIV infection leads to a progressive decrease in lymphocyte count and an increase in viral load. Without treatment, it evolves to severe immunosuppression and AIDS, predisposing an individual to multiple opportunistic infections and neoplastic diseases.

CASE REPORT A 30-year-old male patient with HIV infection was admitted with a 15-day history of functional class II dyspnea associated with fever and weight loss. On physical examination, normal blood pressure, sinus tachycardia, and tachypnea were observed. Oral candidiasis and palatal ecchymotic lesions were detected. Erythematous macular lesions were seen on the thoracic wall. Vesicular breath sounds were diminished in both lungs. Neurological examination was normal. The abdomen was soft and nontender. Laboratory tests showed normal hemogram, renal and liver function, hyponatremia, erythrocyte sedimentation rate (104 mm/h), C-reactive protein (214 mg/l), and lactic dehydrogenase (1369 IU/l). The CD4 lymphocyte count was 36 cells/mm³. Chest X-ray showed bilateral diffuse interstitial infiltrates. On funduscopic examination, whitish lesions with clear edges in the posterior pole were observed. Chest computed tomography (CT) revealed a diffuse bilateral ground glass pattern. Empirical treatment with ampicillin-sulbactam, clarithromycin, and trimethoprim-sulfamethoxazole was started. Subsequent bronchoalveolar lavage (BAL) showed *Pneumocystis jiroveci* and *Cryptococcus neoformans*; the latter was also isolated in the spinal fluid and blood cultures. Disseminated cryptococcosis with *Pneumocystis pneumonia* was diagnosed. Amphotericin deoxycholate was added to treatment. The patient developed an acute abdomen. CT showed involvement of the distal ileum, cecum, and ascending colon. Hemicolectomy and colostomy were performed, because several perforations were detected. Pathological examination revealed *Histoplasma capsulatum*, cytomegalovirus, and *Mycobacterium*

tuberculosis. In addition, a biopsy of a thoracic skin lesion showed Kaposi sarcoma.

COMMENT A single cause for the whole clinical presentation has been for decades the paradigm of clinical reasoning. This concept can be presented as the uncausal theory. Such a way of thinking has been severely questioned by reality, but it remained in the clinician's brain almost unchanged until the appearance of HIV in the early 1980s. HIV infection represents the typical change of a medical paradigm, leaving aside the uncausal theory. Without treatment, it predisposes to multiple infections and neoplasia with high morbidity and mortality. In our patient, the diagnosis of disseminated cryptococcosis with meningeal involvement was made because of the finding of fungus in BAL. The absence of neurologic symptoms in this setting is quite unusual. Nevertheless, cerebrospinal fluid examination was indicated. Besides, in the absence of digestive symptoms, the patient developed an acute abdomen related to bowel perforation, and multiple coexisting opportunistic infections were diagnosed on a pathological basis. The Kaposi sarcoma was an additional skin finding.

CONCLUSION It is important for physicians to be aware of the possible multiple opportunistic infections and neoplasms in immunocompromised patients, because presentations are usually nonspecific. Early indication of invasive diagnostic procedures is essential to prescribe adequate treatment and to avoid complications.

Key words

AIDS, AIDS-associated neoplasia, HIV, opportunistic infections

DIANA FERNANDES

More than pneumoniae

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The majority of patients presenting to the emergency department with acute chest pain have nonsevere etiologies. Nevertheless, life-threatening causes must be considered in the differential diagnosis. One of them is esophageal perforation caused by a foreign body, which is a rare condition.

A 77-year-old Caucasian man was admitted to the intermediate care unit, with a 4-day history of dyspnea, pleuritic chest pain, and dyspepsia with a diagnosis of pneumoniae with hypoxemic respiratory insufficiency. Despite first-line empirical antibiotic therapy and supplemental oxygen, the patient remained very symptomatic with abundant purulent sputum and persistent fever. A thoracoabdominal computed tomography scan showed signs suggestive of esophageal perforation and mediastinitis. In clinical history, the patient reported that the symptoms started after choking on a small chicken bone, which left him with the feeling of a bloated stomach. Upper digestive endoscopy was performed, which showed esophageal perforation that was closed with clips. The patient was treated with broad-spectrum antibiotics, and treatment success was evaluated with an esophageal transit and computed tomography scan with oral contrast, both of which revealed favorable imaging evolution, with no leakage. He was discharged 40 days after admission with complete symptom resolution.

The diagnosis of perforation by foreign bodies is based on the patient's dietary history and symptoms. Foreign body sensation and localized pain could be the main complaints in the early period, but systemic symptoms prevail as soon as inflammation develops. Treatment depends on the etiology, site, and size of perforation, time elapsed between perforation and diagnosis, underlying esophageal disease, local complications, systemic impact, and the overall health status of the patient.

Key words

esophageal perforation, foreign bodies, mediastinitis

ALEJANDRO FROLA

Merkel cell carcinoma in a renal transplant recipient

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Merkel cell carcinoma (MCC) is an uncommon and highly aggressive neuroendocrine tumor of the skin. The primary risk factors for MCC development include older age, exposure to UV radiation, and immunosuppression. Solid organ transplant recipients constitute an important subset of affected patients.

We report a case of a 69-year-old male patient with Fitzpatrick skin type III with a history of kidney transplant in 2011 due to hepatorenal polycystic disease. He was managed with azathioprine, cyclosporine, and meprednisone. Six years after transplantation, the patient presented with a 5-cm, red-purplish, painless, dome-shaped tumor in his left buttock. He also was noted to have pink-red papulonodules forming a painless 7-cm plaque in his left hip. A histologic analysis of both lesions showed layers of small-medium round blue cells with numerous mitotic figures infiltrating the dermis and extending into the subcutaneous tissue. The histologic pattern was solid with evidence of widespread necrosis. The cells tested positive for cytokeratin AE1-AE3, CK 20, synaptophysin, and chromogranin and negative for lymphocyte marker and TTF1.

A whole-body computed tomography scan showed supraclavicular, retroperitoneal, inguinal, and bilateral iliac chain lymphadenopathy, consistent with nodal metastasis. The case was presented at a multidisciplinary tumor board, and the patient was considered a poor surgical candidate, given the rapid growth and limited chance for surgical control. No chemotherapy was initiated due to a poor performance status of the patient. He died of his underlying conditions 20 days after diagnosis.

The diagnosis of MCC must be considered when nontender, rapidly growing nodular lesions develop in the skin of an immunocompromised host. Therapeutic options depend on the presence or absence of metastatic disease to the local lymph basin. Surgical excision with negative margins remains the preferred treatment for local disease, and radiation therapy is recommended for both regional and local tumors with high-risk features. Chemotherapy has a high response rate but a limited duration of response in metastatic disease. Regular total body skin examination, sun protection, and sunscreen use are advisable in the prevention of MCC. Whenever possible, reduction of immunosuppression to the lowest level maintaining good graft function should be considered in all solid organ transplant recipients.

Key words

kidney transplant, merkel cell carcinoma, therapy

CLARISA M. GASHU

Liver abscess and endophthalmitis due to hypermucoviscous *K. pneumoniae*: an emergent infection

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INTRODUCTION A distinctive syndrome of community-acquired invasive *K. pneumoniae* infection, was described in China 2 decades ago. It involves pyogenic liver abscess often complicated by metastatic endogenous endophthalmitis and/or meningitis in young

and healthy individuals. Although these infections were reported initially in Southeast Asia, an increasing number of cases were reported around the world, indicating that this unique syndrome is becoming a globally emerging disease. This strain of *Klebsiella pneumoniae* (*K. pneumoniae*), which is highly virulent, phenotypically shows hypermucoviscosity and its diagnosis requires a string test (colonies forming a string >5 mm in length when touched with a loop). This type of hepatic abscess is not related to gastrointestinal or biliary disease.

OBJECTIVE To present a case of endogenous endophthalmitis secondary to hepatic abscess due to hypermucoviscous *K. pneumoniae*.

CASE A previously healthy 71-year-old man was admitted due to 15 days of fever associated with ocular and abdominal pain. Physical examination showed pain at right hypochondrium and a painful/red right eye with blurred vision; no hypopyon was observed. Ocular echography revealed vitritis and pneumatization of the anterior camera with retinal detachment. Intraocular digital pressure was high. Endophthalmitis was diagnosed. Laboratory tests showed leukocytosis (white blood cell, 11 300/ μ L) and slightly abnormal liver function. Abdominal computed tomography revealed a hepatic hypodense loculated lesion of 90 mm, compatible with abscess. Blood cultures were negative; cerebrospinal fluid was normal. A diagnostic hepatic puncture revealed hypermucoviscous (positive string test) *K. pneumoniae*. Intravenous and intravitreal ceftazidime was started adjusted to antibiogram; a percutaneous catheter in hepatic abscess was placed under ultrasound control. The patient became afebrile and the hepatic lesion improved, but right eye enucleation was needed.

CONCLUSION In the presence of hepatic abscess by *K. pneumoniae* in a previously healthy patient, a string test should be ordered to confirm the hypermucoviscous variant. Increased awareness about the high prevalence of metastatic foci could allow an earlier detection and optimal treatment.

Key words

endophthalmitis, hypermucoviscous, *Klebsiella pneumoniae*, liver abscess

MARGARITA GROMOVA

Ischemic bowel disease complicated by perforation in a patient with tophaceous gout

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INTRODUCTION It is well known that patients with metabolic syndrome, type 2 diabetes, arterial hypertension, hyperuricemia, dyslipidemia, and obesity have a high risk of cardiovascular diseases associated with atherosclerotic vascular lesions. The most frequent types of cardiovascular diseases are ischemic heart disease, cerebrovascular diseases, and atherosclerosis of the arteries of the lower extremities. However, there is also a rare form of atherosclerotic disease, namely, ischemic bowel disease. Due to the rarity of this pathology, it is difficult to diagnose.

CASE PRESENTATION A 64-year-old Caucasian man presented with tophaceous gout, hypertension, type 2 diabetes, obesity, and chronic kidney disease with acute pain in the upper abdomen. The patient underwent a number of examinations: several blood and urine analyses, chest and abdominal x-ray, as well as abdominal computed tomography and ultrasound. Laparotomy was also performed. The operation resulted in an opening of the intestinal abscess and resection of the jejunum with the formation of entero-entero anastomosis. A biopsy was taken, which revealed severe inflammation of fibers and vessels, foci of necrosis, and atherosclerotic arteries with a narrowed and deformed lumen due to thrombus. The patient

developed acute mesenteric ischemia with perforation of the small intestine and a complication in the form of peritonitis.

DISCUSSION This case is untypical in that ischemic bowel disease was the first manifestation of atherosclerosis. The patient did not receive adequate therapy aimed at preventing the development of cardiovascular events, as he had no manifestations of coronary heart disease, cerebrovascular disease, or atherosclerosis of the lower extremity arteries, which is most often detected in patients with metabolic risk factors for cardiovascular disease.

CONCLUSION This case demonstrates the relationship between gout, diabetes, and atherosclerotic bowel disease, followed by the development of acute ischemia of the intestinal vessels, which is a rare complication of atherosclerotic disease. Once diagnosed, the pathology should be treated by a multifactorial therapy, including statins and antiplatelet drugs.

Key words

gout, ischemic bowel disease, perforation

SEDAT IRMAK

Patient without a history: a challenging case of infective endocarditis

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Infective endocarditis is a difficult diagnosis that is rarely made at an emergency department. We present a challenging case of infective endocarditis complicated with septic embolism. A 22-year-old Syrian man with altered mental status, confusion, and drowsiness was transferred by ambulance to the emergency department. He had severe hypotension and tachycardia. Because of the language barrier, we did not learn his medical history. During follow-up, we realized that he had irregular blood pressure, which led us to search for disorders of the central nervous system. Laboratory results revealed no abnormalities except inflammation. After exclusion of other conditions, the final diagnosis turned out to be infective endocarditis.

Key words

confusion, infective endocarditis, thalamic stroke

VIKTORIJA JERINA

A case report of generalized tetanus in an adult patient

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Tetanus is a potentially fatal infection caused by the *Clostridium tetani* bacterium. It is a rare disease, largely due to a highly protective effect of immunization in developed countries. That is why, the low incidence of tetanus has resulted in a decreased alertness to this disease. The risk group includes patients older than 65 years of age, patients with diabetes, injecting drug users, people who had the last booster more than 10 years ago, and unvaccinated people. Tetanus is a central nervous system disorder characterized by muscle spasms that are caused by the toxin-producing anaerobe *Clostridium tetani*. There is no existing laboratory test to confirm tetanus and diagnosis depends on bacteriologic confirmation. The diagnosis is based on clinical presentation: risus sardonicus, trismus, and painful muscular contractions. With disease progression, autonomic dysfunction, respiratory failure, and cardiac arrest can occur. It is crucial to recognize the clinical symptoms of tetanus at the early

stages. Timely treatment gives an opportunity to avoid complications and determines the prognosis.

We report a case of a 68-year-old woman with a 1-week history of general malaise, pain throat, difficulties with mouth opening, and headache. She was treated for nasopharyngeal infection by her family doctor. Later, she developed dysphagia, opisthotonos, and generalized spasms. Based on clinical signs and a history of injury, tetanus was suspected. Unfortunately, tetanus unrecognized in the prodromal period progressed into a critical condition with severe and intensely painful muscle spasms and autonomic dysfunction. The patient was intubated, transferred to an intensive care unit with the diagnosis of tetanus, and treated with tetanus toxoid, tetanus immune globulin, antimicrobial agents, sedatives, analgesics, and muscle relaxants. She fully recovered 3 months after admission.

Key words

dysphagia, opisthotonos, risk group, tetanus immune globulin, tetanus toxoid

MARIYA KHUDYAKOVA

Recognized for best poster

A 71-year-old man presenting with symptomatic anemia and rash

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INTRODUCTION We find this complex case of a patient with a presenting symptom of anemia interesting because of numerous diagnostic and therapeutic challenges.

CASE PRESENTATION A 71-year-old Caucasian man with a history of marginal zone lymphoma and hemolytic anemia (lost to follow-up for the 2 previous years and thought to be in remission) was admitted to our department for investigation of anemia with a hemoglobin level of 7.1 g/dl. On his left upper arm, a raised red lesion of 3.5 × 5 cm was noticed. Blood tests on admission showed no findings that would indicate anemia. Gastroscopy and colonoscopy were performed earlier due to melena; however, the results were normal. Fluorescent activated cell sorting from peripheral blood was not diagnostic. Whole-body computed tomography (CT) revealed no signs of malignancy. The patient underwent a bone marrow biopsy and a skin biopsy (the latter was not immediately performed because the lesion was first diagnosed as a hematoma).

During hospitalization, the patient developed hoarseness. Biopsy samples from the swollen nasopharynx and an enlarged right tonsil were obtained. Two days after the biopsy, the patient developed septic shock. The second whole-body CT scan revealed bilateral lung consolidations and multiple new brain infarctions. Transesophageal echocardiogram demonstrated a large patent foramen ovale. The patient died despite optimal antibiotic therapy and hemodynamic and ventilatory support. The pathological results were as follows: skin, findings compatible with marginal zone lymphoma; tonsil and nasopharynx, low-grade B-cell lymphoma; bone marrow, no definite evidence of bone marrow involvement by a lymphoproliferative neoplasm.

DISCUSSION We faced numerous challenges during the management of this patient. Firstly, his medical history and lack of specific symptoms at presentation resulted in a very broad differential diagnosis on admission. Secondly, the occurrence of melena was misleading and led to unnecessary invasive diagnostic procedures (gastroscopy and colonoscopy), while the patient did not have signs of iron deficiency. Thirdly, the cutaneous lesion was not immediately examined, thus delaying tissue diagnosis. Fourthly, the patient

was treated with multiple blood transfusions because of unstable hemoglobin levels; they were preceded by intravenous steroids due to a Coombs-positive reaction. At a certain point, it became very difficult to establish a venous access. Still many questions remain unanswered in this case: Was hospital-acquired sepsis a result of relative immune deficiency because of relapsed lymphoma? Did multiple brain infarcts on the CT scan result from embolism of cardiac origin after anticoagulation had been discontinued at presentation? Would it be reasonable to restart anticoagulation therapy after obtaining the normal results of gastroscopy and colonoscopy although the reason for anemia was still unknown? And last but not least, what in fact was the cause of the presenting symptom of anemia?

LESSONS TO BE LEARNED FROM THE CASE 1) Objective signs on patient's examination are not always more reliable than subjective ones; 2) patients with a history of oncologic disease who present with cutaneous lesions should undergo prompt tissue biopsies to facilitate a faster diagnosis.

Key words

anemia, biopsy, lymphoma

PAWEŁ KUCZIA

Macrophage activation syndrome with features of myositis and acute pancreatitis as primary manifestation of systemic lupus erythematosus

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Macrophage activation syndrome (MAS) is a rare, life-threatening condition of overwhelming hyperinflammation. It shares many common features with hemophagocytic lymphohistiocytosis (HLH) and is characterized by an uncontrolled activation of T lymphocytes and macrophages, with substantially elevated proinflammatory cytokine levels. Typical features of HLH include high fever, cytopenias, hyperferritinemia, hepatomegaly, splenomegaly, hypertriglyceridemia, hypofibrinogenemia, and hemophagocytosis. In general, HLH can be categorized as either primary (familial) or secondary. The latter can be caused by infections, neoplasms, or autoinflammatory rheumatic diseases, including systemic lupus erythematosus (SLE). The particular form of HLH in the course of rheumatic disorders is often termed macrophage activation syndrome.

Here, we report a case of MAS as a primary manifestation of SLE, with rare and potentially misleading features of autoinflammatory muscle and pancreatic damage, as well as its treatment considerations.

A 35-year-old patient presented with fever of unknown origin, arthralgias, abdominal discomfort, and unintentional weight loss. Physical examination revealed aphthous stomatitis and enlarged submandibular lymph nodes. Initial blood tests showed mild pancytopenia and normal C-reactive protein levels. Blood and urine cultures along with serology tests for HIV, Epstein-Barr virus, hepatitis B and C virus were negative. The patient tested slightly positive for anti-cytomegalovirus (CMV) IgM and CMV IgG antibodies, but negative for CMV as determined by polymerase chain reaction. There were no abnormalities on a cytological examination of the bone marrow. On the following days, a rapid deterioration of the patient's general condition with altered mental status, nonspecific abdominal pain, intensive myalgias, and mild respiratory insufficiency was observed. Further laboratory tests showed progressive pancytopenia and increasing levels of muscular and pancreatic damage markers. In addition, high concentrations of ferritin and triglycerides, along with low concentrations of fibrinogen and albumin, were found. A computed tomography scan of the abdomen revealed moderate peritoneal fluid accumulation and mild edema

of iliopsoas muscles bilaterally, consistent with muscular inflammatory changes. Cervical lymph node biopsy and repeated bone marrow biopsy showed hemophagocytic macrophages with no evidence of malignancy. Subsequent immunological tests were strongly positive for antinuclear antibodies ($>1:20480$) and revealed low plasma concentrations of complement C3 and C4 compounds, leading to a final diagnosis of SLE with secondary MAS. High-dose intravenous methylprednisolone pulses and cyclosporine were initiated, resulting in a rapid resolution of symptoms and decrease in muscle and pancreatic damage markers within a few days. The patient has been followed-up for 4 years with no flare of disease.

Currently, there are no guidelines on how to manage patients with SLE-associated MAS depending on the severity of disease. There is only scarce evidence of pancreatitis in the setting of MAS in SLE. Furthermore, elevation of alanine and aspartate aminotransferases is often observed in MAS, but they are commonly considered a consequence of liver damage. In this case, muscular damage was confirmed by high levels of myoglobin and kinase creatinine.

In conclusion, it should be remembered that the development of MAS can initially manifest as fever of unknown origin. Moreover, SLE-associated MAS can be accompanied by autoinflammatory muscle or pancreatic damage. In this type of manifestation, remission can be achieved with steroids and cyclosporine, supporting the use of cyclophosphamide or etoposide in more severe, resistant, or recurrent cases.

Key words

acute pancreatitis, hemophagocytic lymphohistiocytosis, macrophage activation syndrome, myositis, systemic lupus erythematosus

SABRINA LEMME

When fever is the only sign

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INTRODUCTION Fever of unknown origin is a rare clinical condition, associated with a wide spectrum of diseases ranging from mild to life-threatening. Its diagnostic management is challenging and varies significantly across regions and populations.

CASE DESCRIPTION We present a case of a 58-year-old woman who was admitted to our hospital after experiencing 40 days of fever. Due to fever and dry cough, she first consulted an outpatient physician. She was diagnosed with infectious respiratory process and received antibiotics for 1 week. Dry cough subsided but fever continued and night sweats occurred. Her fever spiked 3 times a day and did not have a specific pattern. She lived in Argentina all her life. She had no history of recent travels or high-risk sexual contacts. She worked as a secretary in a health center. Her past medical history included Graves-Basedow disease, melanoma, ovarian cancer, and active smoking. Her physical exam was unremarkable. Her temperature was 38.6°C. Baseline blood tests revealed a hematocrit of 25% and a platelet count of $717\,000 \times \text{mm}^3$. Ferritin levels were 1378 ng/ml. She had mild elevation of liver enzymes (alkaline phosphatase 339 U/l; γ -glutamyl transferase, 91 U/l; aspartate transaminase, 49 U/l). Erythrocyte sedimentation rate (ESR) exceeded 100 mm/h, and C-reactive protein levels (CRP) were 17.87 mg/dl. Her blood and urinary sediments were negative. Serologic tests for HIV, syphilis, toxoplasmosis, tuberculosis, and viral hepatitis were negative. Chest and abdominal computed tomography scans were normal. A transesophageal echocardiogram showed no vegetations. Bone marrow and liver biopsies were negative for an infectious disease, granulomatous processes, or neoplasia. An autoimmune diagnostic panel was also negative. A temporal artery echo-Doppler was done. It showed a deep caliber decrease as the only positive sign. Because of the negative

results of all tests and persistent fever, ^{18}F -fluorodeoxyglucose positron emission tomography–computed tomography (FDG-PET/CT) was performed. It showed diffuse parietal thickening of the thoracic and ascending aortic arch, supra-aortic vessels, descending thoracic and abdominal aorta, proximal superior artery, and primitive iliac artery, suggestive of giant cell arteritis. She was started on prednisone and her fever subsided. During outpatient visits, she reported a notable improvement. Laboratory tests after 1 month of steroid treatment showed an ESR of 31 mm/1st h, C-reactive protein of 1.18 mg/dl, and ferritin of 130.4 ng/ml; hemogram and liver function were completely normal. FDG-PET-CT after 6 months showed a marked reduction in the thickness of the parietal vascular wall.

DISCUSSION Our case illustrates the importance of PET-CT in patients with fever of unknown origin as the only sign of giant cell arteritis. We would like to emphasize the value of a systematic approach to working with this type of a diagnostic challenge.

Key words

arteritis, fever of unknown origin, positron emission tomography–computed tomography, temporary arterial biopsy, vasculitis

INNA LIPNITZKI

Treatment of severe autoimmune hepatitis with repeated plasmapheresis

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INTRODUCTION Autoimmune hepatitis (AIH) is a form of chronic hepatitis of unknown etiology. Generally, the disease is characterized by circulating autoantibodies and a high serum globulin concentration.

The general approach to treatment is based on glucocorticoids, either as a monotherapy or in combination with azathioprine. The optimal treatment of resistant disease is not well established.

CASE DESCRIPTION A 69-year-old woman presented to an internal ward with symptoms of active hepatitis. She was diagnosed with autoimmune hepatitis (AIH) 12 years prior to the last admission. Based on positive anti-smooth muscle antibody titers, exacerbation of AIH was diagnosed. Other infectious, toxic, vascular, and cholestatic etiologies of her disease were excluded. In the last year, her liver disease was complicated by liver cirrhosis. Her additional medical conditions included idiopathic thrombocytopenic purpura and polyclonal hypergammaglobulinemia. On presentation, she had rectal and vaginal bleeding, severe polyclonal hypergammaglobulinemia, and profound thrombocytopenia. The vaginal bleeding was nonresponsive to gynecological treatment.

Treatment with glucocorticoids was started but without success. Due to prominent hypergammaglobulinemia and its possible influence on disease pathogenesis, after a consultation with a hematologist we decided to complement the treatment with 3 cycles of plasmapheresis. The therapy resulted in good clinical and laboratory response, with a reduction of serum gamma globulin levels, elevation of platelet count, and bleeding cessation. The patient was discharged several days later.

DISCUSSION There are scarce literature data on the role of plasmapheresis in patients with AIH. Because of combined morbidity in our patient, a reduction of globulin levels had clinical reasons. The use of plasmapheresis as an optional treatment in AIH with significant hypergammaglobulinemia should be investigated.

Key words

autoimmune hepatitis, immunoglobulin, plasmapheresis

DÁNIEL NÉMETH

Anabolic steroid–induced acute liver injury combined with nephropathy, pancreatitis, and cardiomyopathy

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INTRODUCTION Drug-induced liver injury (DILI) is one of the most important adverse drug reactions. With the increasing number of prescriptions or over-the-counter medications, herbal products, and dietary supplements, the risk of DILI has become a growing public health problem. Some patients have nonspecific symptoms, while others may present with typical clinical forms. If hepatocellular injury is combined with increased bilirubin levels, the case fatality rate is high.

CASE DESCRIPTION A 41-year-old male patient was admitted to our hospital because of deep jaundice, pruritus, and sexual dysfunction. He had taken intramuscular anabolic steroid injections (trenbolone and stanozolol) in the last 3 months. Four weeks prior to admission, he had undergone a surgical intervention because of an intramuscular abscess and was treated with ciprofloxacin.

On admission, the serum bilirubin level was 1011 $\mu\text{mol/L}$. Plasmapheresis treatment was started because of the increasing intrahepatic cholestasis. A total of 37150 ml of plasma was exchanged in 11 cycles. Hemodialysis was performed twice because of bile acid–induced cholemic nephropathy. On day 8, intravenous fluid supplementation and transitional fasting were applied due to acute pancreatitis. Imipenem and cilastatin therapy was initiated because of leukocytosis and elevated C-reactive protein levels. The decreased albumin and cholinesterase levels and the prolonged international normalized ratio were suggestive of decreased synthetic capacity of the liver; however, no sign of hepatic encephalopathy could be detected. Blood transfusion was also administered for anemia. Echocardiography showed concentric hypertrophy of the left ventricle with grade II diastolic dysfunction. A complex treatment resulted in improvement of acute kidney and liver injury, and pancreatitis resolved. However, the serum bilirubin level normalized only 4 months later.

DISCUSSION Although the liver is frequently affected by serious adverse events as a central organ responsible for the metabolism of toxic agents, in many cases other organ systems can be also damaged, as seen in this report. Besides hepatotoxicity, acute kidney injury, pancreatitis, and sexual dysfunction also developed.

DILI may take a cholestatic and a hepatocellular form. Anabolic steroids mainly induce cholestasis; however, in our patient hepatocellular necrosis was already present, which worsened his chance of survival. According to the Hy's law, hepatocellular necrosis in combination with cholestasis indicates poor prognosis. In our case, the need for liver transplantation was also considered, yet the patient's condition improved after plasmapheresis and conservative supportive treatment.

The diagnosis of DILI may be challenging. Since it can present in any form of liver disease and there are no specific diagnostic tests, other possible causes of liver damage or cholestasis have to be carefully considered and excluded. On the other hand, most patients take several drugs, which makes it difficult to identify the culprit drug. Based on this case, we emphasize the risk of the uncontrolled use of anabolic–androgenic steroids.

Key words

acute pancreatitis, anabolic-androgenic steroid, bile acid–induced cholemic nephropathy, drug-induced liver injury

Endovascular thrombolysis and stenting in a patient with May–Thurner syndrome presenting with phlegmasia cerulea dolens

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May–Thurner syndrome is a rare condition that causes extensive thromboembolic disease. Various treatment methods have been reported, of which intravascular stenting has been found to be the most effective.

A 52-year-old male smoker with metabolic syndrome presented with an edematous, purple, tender, and blistering left leg with skin lesions extending up to the groin. He also presented with diabetic ketoacidosis and hypertensive urgency. Venous and Doppler ultrasound of his leg revealed an extensive thrombus with complete occlusion of the left popliteal, superficial femoral, common femoral, and external iliac veins. The clinical diagnosis was phlegmasia cerulea dolens secondary to extensive iliofemoral thrombosis. Thrombosis was treated with a catheter-directed infusion of tissue plasminogen activator over a 24-hour period. A follow-up venogram showed resolution of the thrombus but May–Thurner syndrome was diagnosed, defined as an anatomical anomaly whereby the left iliac vein is compressed by the right common iliac artery. The patient's common iliac vein was subsequently balloon-dilated and stented. Long-term anticoagulation with warfarin and aspirin was scheduled.

This case highlights the presentation of this rare disease and explores treatment options and complications.

Key words

catheter-directed thrombolysis, endovascular stenting, May–Thurner syndrome, phlegmasia cerulea dolens

LENNA ÖRD

An 80-year-old patient with exertional chest pain: an unusual outcome

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INTRODUCTION Apical hypertrophic cardiomyopathy (AHCM) is an uncommon morphologic variant of hypertrophic cardiomyopathy (HCM), first described in Japan 40 years ago. It is suggested that the prevalence outside Japanese population is 1% to 3% among patients with HCM. AHCM can be of various etiology but is most commonly associated with sarcomere protein gene mutations, which are inherited as an autosomal dominant trait. AHCM is characterized by giant negative T-waves in the left precordial leads on electrocardiogram and an “ace-of-spades” configuration of the left ventricular cavity during diastole on echocardiogram or ventricular angiogram. Left ventricular hypertrophy predominantly affects the apex, which does not cause left ventricular outflow tract obstruction, but may be complicated by midventricular obstruction with cavity obliteration and/or development of an apical aneurysm.

CASE REPORT AND DISCUSSION We present a case of an 80-year-old female patient with a history of exertional chest pain, fatigue, and dyspnea lasting a few weeks. Her electrocardiogram was consistent with left ventricular hypertrophy; deep inverted T-waves were present in anterolateral leads. Cardiac markers showed moderately elevated

troponin T levels and creatine kinase MB isoenzyme mass. Since she also had a swollen left leg, she was initially investigated for pulmonary embolism. A computed tomography with pulmonary angiography was performed. No pulmonary embolism was found, but deep-vein Doppler-ultrasound revealed thrombosis of the left femoral vein. She was admitted to the cardiology department with a working diagnosis of unstable angina. Echocardiography gave the suspicion of AHCM and the diagnosis was confirmed by cardiac magnetic resonance imaging, which showed left ventricular apical hypertrophy with a maximum wall thickness of 16 mm; the apical and medial wall ratio was 2:1 and left ventricular ejection fraction was 60%. Myocardial fibrosis was present in the apical segment of the anterior wall and in the medial segment of the inferolateral wall. Because the patient also experienced exertional chest pain distinctive of coronary artery disease, a coronary angiography was performed. Atherosclerotic coronary artery disease was eliminated with the procedure: our patient's angina was probably the result of small-vessel disease with intramural coronary artery narrowing as is characteristic of HCM. As a complication of AHCM the patient experienced asymptomatic nonsustained ventricular tachycardia episodes for which she was prescribed a β -blocker since her estimated risk for sudden cardiac death was low. In cases of high risk for sudden cardiac death, implantation of a cardioverter–defibrillator device should be considered. Despite the risk of developing diastolic heart failure or life-threatening ventricular arrhythmias, AHCM is generally associated with a better prognosis than other forms of HCM.

LESSONS TO BE LEARNED This case illustrates that one should keep an open mind: it is important to remember that frequently seen nonspecific symptoms may not always be caused by a common disease.

Key words

apical hypertrophic cardiomyopathy, hypertrophic cardiomyopathy, unstable angina

ALICE PIROVANO

A 28-year-old woman with ascites and anasarca

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INTRODUCTION Serous effusions often complicate the course of non–Hodgkin and Hodgkin lymphomas. In the rare occasions in which lymphomas present with ascites, the ascitic fluid is usually chylous, because of the obstruction of the lymphatic trunks caused by enlarged lymph nodes.

CASE PRESENTATION We report a rare case of a 28-year-old woman admitted to the internal medicine division of an academic hospital in northern Italy, with complaints of progressive abdominal distention and fatigue, a substantial weight gain, and anorexia over the previous 6 months. On physical examination, she was afebrile, with anasarca, marked swelling and erythema of the lower legs, no enlarged or palpable lymph nodes, protuberant abdomen, with central tympany and flank dullness on percussion. Full blood count and coagulation index were normal, serologic tests for HIV, hepatitis B virus, and hepatitis C virus were negative. A paracentesis was performed with aspiration of a yellow-colored turbid fluid, with serum to ascites albumin gradient of 1.1 g/dl, indicative of portal hypertension, negative culture and negative amplification test for *Mycobacterium tuberculosis*. Then, a pig-tail catheter was left in the abdomen of the patient for symptomatic relief. A computed tomography scan of the abdomen and chest and positron emission tomography confirmed massive ascites, liver enlargement without focal lesions, enlarged spleen, and multiple enlarged retroperitoneal lymph nodes. Bone marrow aspiration and biopsy were indicative

of a nonspecific lymphoproliferative disease, but not diagnostic. A decision was made to proceed to diagnostic splenectomy.

DISCUSSION Self-reported conditions remained stable, but the patient developed disseminated intravascular coagulation, as a complication of lymphoma. Although disseminated intravascular coagulation has been reported to worsen the clinical outcome and to be associated with an elevated risk of surgery, it has been demonstrated that this is not so prohibitive and that the only way to determine its resolution is to treat the underlying disorder. Intravenous corticosteroids and fresh frozen plasma infusions were started, and surgical splenectomy was performed. Through a histologic examination of the spleen, diffuse large B-cell lymphoma was diagnosed and R-CHOP chemotherapy was started. A gradual improvement in the patient's clinical condition was observed, and the pig-tail catheter was removed. She was discharged and is currently followed in a hematology unit and in our ward.

Key words

ascites, anasarca, lymphoma, splenectomy, disseminated intravascular coagulation

MAREK PRZYBYSZOWSKI

Clinical case of a patient with severe allergic asthma treated with omalizumab.

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INTRODUCTION The role of immunoglobulin E in an allergic reaction has resulted in the development of a new medication: omalizumab. It has become more available in Poland since 2013 thanks to the Polish National Health Fund program. The program financed by the National Health Fund covers patients older than 12 years of age, with severe uncontrolled bronchial asthma, who are allergic to perennial allergens.

CASE DESCRIPTION In March 2013, our 47-year-old woman with severe uncontrolled allergic asthma met the criteria for inclusion in the program. Since diagnosis, her asthma had been difficult to control, and for about 10 years, she had been on regular oral corticosteroid treatment (mean dose, 10 mg equivalent to methylprednisolone) and with maximum doses of inhaled medications (4500 µg equivalent to fluticasone: the mean dose of inhaled corticosteroids in µg at the time of referral for treatment).

For more than 1 year prior to admission, the patient reported numerous disease exacerbations requiring either hospitalization or increased doses of systemic steroids. She had been treated with omalizumab in our department for 3 years. During treatment, we observed a significant reduction in the number of exacerbations and the dose of systemic steroids, as well as improvement in asthma control and quality of life. Unfortunately, complete discontinuation of systemic steroids was not possible. Moreover, she developed worsening pain in the interphalangeal joints when the dose of oral steroids was tapered due to omalizumab therapy. The patient was hospitalized twice in the rheumatology department. A radiological examination revealed symmetrical destructive changes in the joints of the hands and feet, including the metacarpophalangeal joints of the right and left hands, with exudation in the joints revealed on ultrasound. Laboratory tests in 2015 showed that she was positive for rheumatoid factor (RF) (80.2 IU/ml), while in 2016, she was negative for RF and anti-citrullinated protein antibodies (the tests were performed during long-term steroid therapy). The levels of antinuclear and anti-neutrophil cytoplasmic antibodies, as well as C-reactive protein and complete blood count, were not elevated (either in 2015 or 2016).

Based on the interview, symptoms (recent-onset joint symptoms lasting more than 6 weeks, symptoms mainly from the metacarpophalangeal joints, the greatest severity of symptoms in the morning), and laboratory test results, seronegative rheumatoid arthritis was diagnosed. The patient was referred for immunosuppressive treatment with methotrexate, and omalizumab was discontinued.

phalangeal joints, the greatest severity of symptoms in the morning), and laboratory test results, seronegative rheumatoid arthritis was diagnosed. The patient was referred for immunosuppressive treatment with methotrexate, and omalizumab was discontinued.

DISCUSSION AND LESSONS TO BE LEARNED FROM THE CASE As omalizumab is a relatively new medication, each change in the health status of the patient during biological treatment requires a thorough diagnostic management. It is difficult to unequivocally assess whether the reported symptoms result from chronic oral corticosteroid therapy itself, biological asthma treatment, or additional development of autoimmune joint disease (previously masked by higher doses of oral corticosteroids, which are reduced in anti-IgE therapy). The association of omalizumab treatment with rheumatoid arthritis in our patient seems to be unlikely, yet we cannot fully exclude it.

Key words

asthma, biological treatment, omalizumab, rheumatoid arthritis

JAKUB RUSZKOWSKI

Atypical hemolytic uremic syndrome: the first Polish adult patient treated with eculizumab

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A 33-year-old woman suffered from pyrexia, chills, and muscle pain, followed by fatigue, nausea, and vomiting. Two weeks later, laboratory tests showed features of hemolytic anemia with negative Coombs test results, thrombocytopenia, and acute kidney injury with proteinuria. Based on these results, nonimmune hemolytic anemia was recognized, and thrombotic microangiopathy (TMA) was suspected. A nonspecific treatment with plasma exchange and glucocorticoids was introduced.

The most common causes of TMA are thrombotic thrombocytopenic purpura and hemolytic uremic syndrome (HUS). The latter can be caused by Shiga toxin-producing *Escherichia coli* (STEC-HUS) or other triggers (atypical HUS). We excluded TTP and STEC-HUS. In renal biopsy, acute TMA was diagnosed.

After ineffective nonspecific treatment of atypical HUS with plasma exchange and glucocorticoids, eculizumab was implemented. The clinical and laboratory tests confirmed therapy effectiveness, and no adverse effects were observed. Our patient was the first Polish adult with atypical HUS treated with eculizumab.

Key words

atypical hemolytic uremic syndrome, eculizumab

ALICJA RYTA-ŻYGOWSKA

Infective endocarditis and bicuspid aortic valve in a patient after prostatectomy

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The bicuspid aortic valve (BAV) is the most common congenital heart disease affecting approximately 1% to 2% of the population. Its main complications are valve dysfunction and aortopathy; there is also an increased risk of infective endocarditis. A 69-year-old man with hypertension, an abdominal aortic aneurysm, and suspicion of bicuspid aortic valve presented to the general practitioner due to weakness and persistent fever lasting for about a month after a

radical prostatectomy with pelvic lymphadenectomy for glandular cancer.

On physical examination, apart from moderate cachexia, no abnormalities were found. Laboratory tests revealed signs of urinary tract infection and elevated C-reactive protein (CRP) levels. A microbiological examination of urine showed the presence of *Escherichia coli* and *Enterococcus sp.* Empiric treatment with cefuroxime was started, resulting in a transient improvement. Due to persistent hoarseness, the patient was consulted by a laryngologist, but no abnormalities were found. Due to recurrence of fever, he was hospitalized. Additional tests revealed elevated CRP levels without leukocytosis. Abdominal computed tomography revealed pelvic structure with an unclear character and hypodense lesion in the spleen. On transthoracic echocardiography, features of infective endocarditis were not clearly observed. Due to suspicion of inflammatory changes on computed tomography, antibiotic treatment with metronidazole, amoxicillin with clavulanic acid, and additional fluconazole was started. The treatment was continued for 10 days, resulting in resolution of fever and reduction of CRP levels. On control abdominal ultrasound, no focal lesions in the spleen or any other changes that might correspond with inflammatory changes were observed. In the following month, the patient again observed feverish states, and laboratory tests revealed a further increase in CRP levels. Metronidazole and fluconazole were administered, but due to deterioration of the general condition the patient was referred to the hospital. During hospitalization, endocarditis caused by *Enterococcus faecalis* was diagnosed. Finally, transesophageal echocardiography confirmed the diagnosis of bicuspid aortic valve with inflammatory changes.

Key words

bicuspid aortic valve, infective endocarditis

NATALIA SUVOROVA

A clinical case of primary antiphospholipid syndrome

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Antiphospholipid syndrome often presents as a multisystem disorder characterized by recurrent arterial and venous thrombosis and pregnancy loss, early atherosclerosis, renal failure, and arterial hypertension.

We report a rare case of a 49-year-old male patient, whose previous medical history was notable for several cardiovascular events. He had long-standing arterial hypertension, atherosclerotic aortic valve defect, and a history of symptomatic epilepsy and 2 episodes of stroke. Moreover, he had symptoms of angina and signs of previous myocardial infarction. At that stage, the patient presented to the department with deep vein thrombosis of the upper limb, renal insufficiency, and thrombocytopenia. Despite anticoagulant therapy, he developed phlegmasia of the right upper limb. As ischemia of the right upper limb progressed, amputation was performed according to vital indications. Laboratory tests revealed elevated levels of anticardiolipin antibodies (IgG class), anti-b2-glycoprotein antibodies (IgG class), lactate dehydrogenase, D-dimer, and lupus anticoagulant; anti-double-stranded DNA and antinuclear antibodies were negative. The patient was newly diagnosed with primary antiphospholipid syndrome. Fresh frozen plasma transfusions (10 ml/kg/d) were started, and anticoagulant therapy was continued. It prevented the recurrence of thrombotic episodes and aggravation of renal function. Over the next 3 weeks, the combination therapy led to clinical, hematological, and renal improvement.

This case shows that it took clinicians over 15 years to establish the primary clinical diagnosis. If antiphospholipid syndrome was

diagnosed earlier, it might have been possible to avoid such serious cardiovascular events as stroke or heart attack, renal insufficiency, deep and superficial vein thrombosis of the upper limb with subsequent amputation, and final disability of the patient. The case shows that it is important to perform a comprehensive diagnostic workup in patients with multiorgan manifestations, especially in young people.

Key words

acute renal failure, anticardiolipin antibody, plasma exchange, primary antiphospholipid syndrome

JOHN THADATHILANKAL-JESS

Recognized for best poster

Cranial nerve fallout as presentation of hypertrophic pachymeningitis associated with systemic lupus erythematosus

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Hypertrophic pachymeningitis (HP) is a rare clinical disorder that is characterized by either a focal or diffuse thickening of the dura mater secondary to underlying chronic inflammation. It may be idiopathic or secondary to underlying pathological processes. The latter include infectious processes (commonly tuberculosis, neurosyphilis, fungal infections), underlying neoplasms (commonly lymphomas), autoimmune conditions (rheumatoid arthritis, polyarteritis nodosa, antineutrophil cytoplasmic antibody-associated vasculitis), or systemic illness (sarcoidosis, tuberous sclerosis, IgG4-related systemic disease). The link between systemic lupus erythematosus (SLE) and HP is extremely rare, with only 6 cases having been reported in the literature according to our knowledge. We describe a case of recurrent presentations for complications related to HP underlying SLE, the current presentation being due to multiple cranial nerve fallout. This case highlights the need to maintain a high index of suspicion for HP in patients with SLE who present with any neurological fallout. Steroids remain the backbone of treatment for HP in SLE, with success rates reported in isolated cases. It is imperative that the diagnosis is made early and treatment is initiated promptly to prevent further neurological fallout.

Key words

pachymeningitis, systemic lupus erythematosus

JEAN-BAPTISTE VULSTEKE

Thymoma-associated systemic lupus erythematosus triggered by hepatitis E

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Thymomas are associated with autoimmune disease in up to 55% of cases, with myasthenia gravis being the most frequent. Other diseases, such as systemic lupus erythematosus (SLE), can also occur and should always be considered.

We report a case of a 45-year-old male patient who presented with central chest pain during exercise, reduced exercise tolerance,

and swelling of the hands and feet. A tentative diagnosis of thymoma was made based on chest radiography. An upfront resection was performed and the results confirmed thymoma. A few months later, the patient consulted his general physician because of persistent fatigue and facial edema. Physical examination was notable for an elevated jugular vein. Laboratory results showed elevated liver enzyme levels in a predominantly hepatocellular pattern. Additional viral serology was positive for hepatitis E virus infection. Chest and abdominal computed tomography showed a bilateral pleural and pericardial effusion. Pericardiocentesis and thoracentesis were performed. No malignant cells were found. The patient was positive for antinuclear antibody titers (1:320) and antineutrophil cytoplasmic antibody titers (1:5120, pANCA pattern), but negative for anti-myeloperoxidase antibodies. A tentative diagnosis of thymoma-associated SLE was made, probably triggered by an acute hepatitis E infection.

Various autoimmune diseases besides myasthenia gravis can be associated with a thymoma. Thymoma-associated SLE is rare, with a co-occurrence rate of 1.5% to 2.3%. It has a similar presentation to regular SLE. In this case, it was probably triggered by a viral infection. Thymoma is associated with an increased risk of infection. This case illustrates that autoimmunity and an increased risk of infection should always be considered in patients with thymoma.

Key words

autoimmune disease, hepatitis E, paraneoplastic, systemic lupus erythematosus, thymoma

KADRI VÄLJATAGA

Allopurinol-induced toxic epidermal necrolysis

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INTRODUCTION Allopurinol is a xanthine oxidase inhibitor used in treating gout. The most serious adverse effect of allopurinol is toxic epidermal necrolysis (TEN): a rare, potentially life-threatening condition. The clinical manifestations are erythematous patches, bullae, erosions, and ulcers. The majority of cases present with mucosal involvement. The exact name of the disease depends on the percent of body surface area with epidermal detachment: <10%, Stevens-Johnson syndrome (SJS); 10%–30%, SJS/TEN overlap; and >30%, TEN. TEN is thought to be an immune reaction initiated by cytotoxic lymphocytes. Skin biopsy usually shows total keratinocyte necrosis.

CASE REPORT A 65-year-old woman was admitted with infection of unknown origin and kidney failure. General redness of her skin was also noticed. After a few days, the condition of the skin rapidly worsened. The patient was referred to an intensive care unit. Almost 95% of the body surface area was erythematous with exfoliation of the epidermis leaving behind erosions. Vesicles and bullae were formed on the legs. The patient had conjunctivitis, and the oral mucosa had painful erosions. The diagnosis of TEN was made. Skin biopsy showed total necrosis of the epidermis. The patient was previously diagnosed with idiopathic pulmonary hypertension and right-sided heart failure, type 2 diabetes, autoimmune thyroiditis, and hypertension. The latest drug added to her medication list was allopurinol for asymptomatic hyperuricemia. Allopurinol as a presumable cause was discontinued. Based on the anamnesis and laboratory values, we calculated a severity-of-illness score for TEN—the probability of death was 58%. The treatment was multidirectional and involved replacement of fluid loss, vasopressor support, empiric antibiotic therapy, intubation, sedation and mechanical ventilation, continuous renal replacement therapy, plasmapheresis, and intravenous immunoglobulin. Despite aggressive treatment, the patient's clinical condition deteriorated and she eventually died.

DISCUSSION Drugs are the leading trigger of TEN. In our case, the culprit drug was allopurinol, which is most commonly associated with SJS/TEN. Indication for allopurinol in the treatment of asymptomatic hyperuricemia is unclear and more studies are needed on this topic. Supportive care is the most accepted treatment intervention for TEN. Adjunctive therapies most commonly mentioned are systemic corticosteroids, intravenous immunoglobulin, cyclosporine A, and plasmapheresis. Decisions regarding those treatments should be made on a case-by-case basis. TEN can cause multiple-organ failure by influencing the skin, lungs, kidneys, and gastrointestinal tract. Infection and sepsis are the leading causes of death. In our case, the patient had sepsis on top of her chronic respiratory and cardiac failure; development of TEN added remarkably to the risk of the worst outcome.

CONCLUSION It is important to know the basic characteristics of TEN to ensure early diagnosis. Supportive care is the cornerstone of treatment, while specific therapies need more investigation. Our case report illustrates the fact that we should think twice before prescribing allopurinol for asymptomatic hyperuricemia.

Key words

adverse drug reactions, allopurinol, asymptomatic hyperuricemia, toxic epidermal necrolysis

ANNA WITKOWSKA

Pylephlebitis in the course of appendicitis caused by *Streptococcus pseudoporcinus*: a case study

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INTRODUCTION Pylephlebitis is thrombosis of the portal vein, associated with an intra-abdominal infection. It is characterized by nonspecific symptoms. It can contribute to hepatomegaly, splenomegaly, and ascites. Laboratory tests show inflammation and damage to hepatic cells.

CASE PRESENTATION A 35-year-old patient was admitted to the hospital due to fever up to 39°C. The symptoms had started 3 weeks prior to admission. Initially, he complained of epigastric pain, but on admission, he denied abdominal pain. Physical examination revealed a distended abdomen, audible and painless peristalsis, and enlarged liver. Laboratory tests revealed a considerable increase in the levels of inflammatory markers without leukocytosis and increased D-dimer levels. *Streptococcus pseudoporcinus* was cultured from both hands with the use of the VITEK 2 Compact system. An abdominal ultrasound revealed signs of portal vein thrombosis (PVT), which was confirmed by computed tomography. The appendix appeared to be thickened and infiltrated by the surrounding tissues.

A differential diagnosis did not confirm changes that might have contributed to the reported complaints. After a surgical consultation, an invasive treatment of the inflamed appendix was not ordered. The patient was administered a targeted antibiotic therapy with meropenem and long-term antithrombotic therapy. The treatment resulted in an improvement of the patient's general condition and normalization of inflammatory parameters.

The patient was hospitalized again 2 months later. He underwent a control abdominal computed tomography. It did not show thrombotic changes. In the place of a previously observed infiltrate, a single slight fibrotic band was noted. The patient was consulted by a hematologist, who excluded coagulation abnormalities. The antithrombotic therapy was continued until a hematologic diagnosis was established. The clinical presentation showed that *Streptococcus pseudoporcinus*-induced septic appendicitis was a cause of pylephlebitis.

DISCUSSION Pylephlebitis constitutes about 10% of cases of PVT; 60% of cases are concomitant with sepsis and 19% are caused by appendicitis. *Streptococcus pseudoporcinus* is a rare species. It was first described in 2006 as a cause of infections of the female reproductive tract. So far, only 2 cases of male infection have been reported. The first one was a purulent infection of the thumb, and the other, bacteremia with endocarditis of unknown etiology, not related to any infection of the female reproductive tract.

In our case, the patient presented with pylephlebitis in the course of *Streptococcus pseudoporcinus*-induced septic appendicitis, not diagnosed before. Considering that PVT can be caused by neoplastic diseases and congenital thrombophilias, the final diagnosis was made after exclusion of the above causes of thrombosis.

LESSONS TO BE LEARNED FROM THE CASE An optimal management of pylephlebitis caused by acute appendicitis requires a prompt diagnosis and implementation of a broad-spectrum antibiotic therapy and antithrombotic treatment.

Key words

appendicitis, portal vein thrombosis, pylephlebitis, sepsis, *Streptococcus pseudoporcinus*