POLISH ARCHIVES OF
Internal Medicine
POLSKIE ARCHIWUM MEDYCYNY WEWNĘTRZNEJ

ESTABLISHED 1923 VOL.129 (2019) Special Issue 4 WWW.MP.PL/PAIM

SPECIAL ISSUE

Abstract proceedings of the best case report contest 2019

Clinical Cases in Internal Medicine: Learning Through Practice (McMaster International Review Course in Internal Medicine, 9–11 May 2019, Kraków, Poland)

IMPACT FACTOR 2018
2.882

medycyna praktyczna

POLISH ARCHIVES OF
Internal Medicine
POLSKIE ARCHIWUM MEDYCYNY WEWNĘTRZNEJ

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

PUBLISHER
Medycyna Praktyczna

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

Print ISSN: 0032-3772

Online ISSN: 1897-9483

Impact Factor 2018 = 2.882

The journal receives funding from the Ministry of Science and Higher Education for promoting scientific research (agreement no. 595/P-DUN/2019).

ABSTRACT PROCEEDINGS OF THE BEST CASE REPORT CONTEST 2019

Clinical Cases in Internal Medicine: Learning Through Practice
(McMaster International Review Course in Internal Medicine,
9–11 May 2019, Kraków, Poland)

AWARD-WINNING CASES

1ST PLACE: SHARON COWLEY

Macrophage activation syndrome in a patient with systemic lupus erythematosus

Sharon Cowley*, Sekharipuram Ramakrishnan
Rheumatology Department, Our Lady's Hospital, Navan, Ireland
*Corresponding author: sharoncowley111@gmail.com

INTRODUCTION We report a case of macrophage activation syndrome (MAS) associated with systemic lupus erythematosus (SLE). This is an underrecognized hyperinflammatory syndrome of excessive immune activation. It is characterized by an inappropriate survival of histiocytes and cytotoxic T cells, leading to a cytokine storm, hemophagocytosis, and multiorgan damage. It is a life-threatening condition if not promptly identified and treated.

CASE DESCRIPTION A 37-year-old woman presented to the Emergency Department with a 6-week history of new-onset generalized nonpuritic erythematous rash covering her face, hands, arms, trunk, and lower limbs. She reported intermittent fevers over the same 6-week period, with a weight loss of 5 kg. On physical examination, she had a widespread erythematous macular rash with skin sloughing on the fingertips. There were also areas of pigmentation on the trunk. She had no synovitis. Diffuse hair loss was noted but no alopecia. Cardiac, respiratory, gastrointestinal, and neurologic examinations were unremarkable. She was on no regular medications.

Extensive laboratory investigations were performed. Hemoglobin levels dropped to 8.5 g/dl (reference range, 11.5–16.5 g/dl) on the third day of admission, with a mean corpuscular volume of 74 fl (reference range, 78–97 fl) and mean corpuscular hemoglobin of 23 pg/cell (reference range, 26–34 pg/cell). Platelet count dropped to 69×10^9 (reference range, $150–145 \times 10^9$/l) in the first week. Liver function tests on admission showed evidence of transaminitis. Ferritin levels on day 3 were elevated at 6703 ng/ml (reference range, 5–204 ng/ml). Total cholesterol and triglyceride levels were elevated at 8 mmol/l (<5 mmol/l) and 15.08 mmol/l (reference range <1.7 mmol/l), respectively.

The patient was positive for antinuclear antibodies (ANA; 9.9, reference range <0.7), anti-Ro antibodies (240 U/ml, reference range <0.7 U/ml), and anti-La antibodies (84 U/ml, reference range <0.7 U/ml). Anti-RNP antibody titers were elevated (63 U/ml, reference range <0.7 U/ml). Complement studies showed reduced C3 levels of 0.78 g/l (reference range, 0.83–1.8 g/l) and normal C4 levels of 0.22 g/l (reference range, 0.14–0.54 g/l).

Thoracic, abdominal, and pelvic computed tomography was negative for any malignancy. Bone marrow biopsy showed disorganized hematopoiesis and evidence of hemophagocytosis. Dermatologic examination revealed skin rash consistent with acute lupus. Skin biopsy was completed; the results were not typical for but in keeping with acute lupus.

The patient met the 2012 Systemic Lupus International Collaborating Clinics Criteria for SLE (positive ANA, positive lupus anticoagulant, thrombocytopenia, leukopenia, and low C3 levels). Macrophage activation syndrome secondary to acute SLE was diagnosed on the basis of full blood workup and fulfillment of HScore criteria or criteria for secondary hemophagocytic lymphohistiocytosis / MAS.

The patient was treated with methylprednisolone (500 mg 3 times/d) for the first 3 days plus intravenous immunoglobulins (1 g/kg) for 2 consecutive days. Ferritin, lactate dehydrogenase, and full blood count levels were monitored daily. On day 4, the patient was switched to oral dexamethasone (50 mg). After 14 days, she was commenced on mycophenolate mofetil (500 mg/d; titrated up to 1 g twice daily), and steroids were slowly tapered.

DISCUSSION Secondary hemophagocytic lymphohistiocytosis is a clinical syndrome that mimics numerous other systemic diseases. Pyrexia of unknown origin is the cardinal feature of the syndrome. As this is a nonspecific sign, extensive investigations are required to establish the diagnosis. Hyperferritinemia is a key laboratory feature and was a key diagnostic clue in this case. Other notable blood abnormalities include cytopenias, hypertriglyceridemia, and hypofibrinogenemia.

Key words
hemophagocytic lymphohistiocytosis, hyperferritinemia, pyrexia of unknown origin, systemic lupus erythematosus

2ND PLACE: LE GOUEFF ANOUK

Concomitant relapsing polychondritis and immunoglobulin G4–related disease: a diagnostic challenge

Anouk Le Goueff[1]*, Frédéric Vandergheynst[2], David Jayne[3]
1 St Pieters Hospital, Brussels, Belgium
2 Erasmus Hospital, Brussels, Belgium
3 Addenbrookes Hospital, Cambridge, United Kingdom
*Corresponding author: anouk.le.goueff@ulb.be

Relapsing polychondritis (RP) and immunoglobulin G4–related disease (IgG4-RD) are rare multiorgan immune-mediated diseases. Both conditions can present with nonspecific symptoms such as fatigue, weight loss, arthralgia, elevated levels of inflammatory markers, or chronic anemia, and diagnostic delay is common. Early recognition and diagnosis are important to minimize morbidity and risk of disability, as both diseases show a potentially good treatment response to steroids with or without immunosuppression or immunomodulation.

We present a case of a 78-year-old man with a 3-year history of fatigue, arthralgia, weight loss, anemia, and increased levels of inflammatory markers. The patient presented first with extensor tenosynovitis and arthritis of the left wrist, followed by episodes of perichondritis of the ears, nasal tenderness, and hoarse voice. He was subsequently repeatedly admitted due to shortness of breath and productive cough, followed by sensorineural deafness and neuronal vestibulitis, then acute kidney injury with tubulointerstitial nephritis and high titers of IgG4 plasma cell infiltration, and, finally, submandibular sialadenitis, while being treated with subtherapeutic doses of prednisolone for 1 year. After all those complications, prednisolone was increased to 15 mg/d, and rituximab, 1 g, was added twice, at day 1 and day 15, leading to resolution of all constitutional symptoms as well as other abnormalities described above, including renal impairment.

The 3 years of morbidity and progressive disability suggest that the patient presented with typical signs and symptoms of relapsing perichondritis and IgG4-RD in an alternating fashion. He fulfilled the latest diagnostic criteria for both these entities. Poor response to a steroid trial can be explained by subtherapeutic doses as well as the presence of all risk factors for refractory relapsing polychondritis: male sex, old age at diagnosis, cutaneous involvement, general symptoms, and myelodysplasia. Despite a lengthy period of diagnostic uncertainty that delayed effective therapy, he responded well to rituximab, which reinforces the rationale for using this biological drug in relapsing polychondritis and IgG4-RD.

In a complex multisystem presentation of an inflammatory disease, a broad diagnostic approach is required to include IgG4-RD. With an improved understanding of pathogenic mechanisms and identification of newer phenotypes, traditional approaches to definition and classification of multisystem disease are being questioned. This represents a challenge for the medical community to collate large-scale cohorts and continue to refine classification and diagnostic pathways.

Key words
IgG4-related disease, relapsing polychondritis, vasculitis, rituximab

3RD PLACE: JAARIKA JÄRVISTE

A large benign left atrial mass

Jaarika Järviste*

North Estonia Medical Centre, University of Tartu, Tartu, Estonia

*Corresponding author: jaarika.jarviste@gmail.com

INTRODUCTION A differential diagnosis of large benign intracardiac masses can be difficult, considering numerous overlapping characteristics between the 2 main suspects: a thrombus and myxoma. In this report, we describe a benign mass in the left atrium, which was conclusively diagnosed by transesophageal echocardiography (TEE) on the operating table before a planned heart surgery.

CASE DESCRIPTION A 73-year-old woman presented with clinically decompensated heart failure and a left atrial mass as an accidental finding on computed tomography (CT).

She was diagnosed with permanent atrial fibrillation and a mild--moderate mitral stenosis over 10 years earlier, and since then, she received anticoagulant treatment with warfarin. Later, she admitted that she had not been fully adherent to treatment.

Transthoracic echocardiography (TTE) and magnetic resonance tomography (MRT) were performed. The results differed to some extent but were characteristic either for a thrombus or myxoma. It was decided that a biopsy was needed for a conclusive diagnosis. Because the patient also had a moderate-severe mitral stenosis, an elective surgical removal of the mass and repair of rheumatic mitral valve disease were planned. Two months later, TEE was performed on the operating table immediately before the surgery. The left atrial mass had shrunk 3 times in size following an adequate warfarin treatment. The mitral valve pathology was no longer considered severe enough, and the surgery was cancelled. Three months later, the mass was no longer observed on TTE, and the mitral stenosis was graded as moderate.

DISCUSSION There have been quite a few case reports describing difficulties in differentiating myxomas from thrombi. Although there are specific characteristics that guide us to one or the other diagnosis, the final decision may still be hard to make. Getting the diagnosis right is very important, considering different treatment strategies: anticoagulation for a thrombus versus surgical removal for a myxoma. In our patient, the mass was located in the left atrial septal wall, had lobulated structure, smooth edges, was oval in shape, and rather large. All of these features can be characteristic for any of the 2 masses.

LESSONS TO BE LEARNED In our case, there were slightly different findings on TTE as well as CT and MRT, and different radiologists were involved. This report suggests that an interdisciplinary team could be valuable in more complex cases requiring a differential diagnosis of intracardiac masses. It also suggests that an attempt at anticoagulant treatment and repeated radiologic evaluation of the left atrial mass and mitral valve pathology should be considered before making the final diagnosis, in order to avoid an unnecessary cardiac surgery.

Key words

atrial fibrillation, left atrial mass, mitral stenosis, myxoma, thrombus

ORAL PRESENTATIONS

ANAT ARBEL

An 82-year-old woman presenting with discoloration and pain of the fingers and toes

Anat Arbel*, Adi Kibari, Shai Cohen

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

*Corresponding author: freund.anat@gmail.com

INTRODUCTION This case is remarkable due to its rarity and the diagnostic challenge it posed.

CASE PRESENTATION An 82-year-old Caucasian woman was admitted to our department for investigation of discoloration and pain of the fingers and toes that had progressed over 1 week. Physical examination on admission revealed cyanosis of fingers II and III (right hand) and of toes (both feet) as well as a few maculopapular lesions on the feet. Laboratory tests showed mild normocytic anemia, mild leukocytosis, as well as elevated C-reactive protein and erythrocyte sedimentation rate levels. Chest and abdominal computed tomography angiography revealed an irregular enlargement of the cecum and small splenic infarcts.

During hospitalization, the patient developed ischemia of fingers II–IV (left hand) and of the first toes (both feet). She was also diagnosed with a dropped wrist (right) and with the first episode of atrial fibrillation.

At this point, the differential diagnosis included a neoplastic, inflammatory, infectious, and thromboembolic disease. Therefore, an extensive diagnostic workup was carried out. Positive electromyography results (showing polyneuropathy with radial nerve involvement), abnormal colonoscopy findings (showing ischemic colitis), and splenic infarcts, together with a rapid clinical progression, strongly suggested an inflammatory etiology, most probably vasculitis. Accordingly, pulse steroid therapy was initiated. On the basis of angiography (which showed obstruction of the distal right radial artery) and skin lesion biopsy (which revealed fibrinoid necrosis of small-sized blood vessels), polyarteritis nodosa (PAN) was eventually diagnosed. Following initial therapy, treatment was switched to oral prednisone and cyclophosphamide, resulting in a marked clinical and laboratory improvement.

DISCUSSION The diagnosis of PAN is challenging because the disease is rare, has a gradual course, and has no specific symptoms (especially at presentation) or diagnostic laboratory tests. In our case, we also encountered some other unique challenges. First, the differential diagnosis was broad, and no single disease could account for all the findings, which made it crucial to distinguish between relevant and incidental findings. Second, one of the early findings was an irregularly enlarged cecum. Considering the suspicion of a neoplastic process, colonoscopy was deemed critical for establishing the diagnosis. Yet, given the patient's fragile condition and preferences, there was a risk that the test would not be performed. Eventually, colonoscopy was carried out, revealing an inflammatory rather than neoplastic disease, which holds a completely different clinical course, treatment, and outcome. Finally, some of the disease manifestations seemed unusual for PAN, including atypical age (the disease usually occurs in the 40s and 50s); atypical sex (men are twice as likely to be afflicted); atypical course (the disease usually progresses over weeks to months); and atypical angiographic findings (aneurysms are the most specific finding).

LESSONS TO BE LEARNED The lessons to be learned from the case are as follows: 1) not all diagnostic clues can be found on presentation, as some develop over time and must be considered in the workup; 2) it is essential to differentiate potential diagnostic clues from the misleading ones by adopting a systematic approach; and 3) the diagnosis of PAN relies on clinical, angiographic, and histologic findings.

Key words
ischemia, polyarteritis nodosa, vasculitis

ELENA BOIN *(PRESENTED BY VERONICA VASSIA)*

Velvet eyes, long dark hands, and a life-threatening misdiagnosis: an unusual coexistence of Marfan syndrome and Addison disease

Boin Elena*, Casciaro G. Francesco, Vassia Veronica

S.C.D.U. Medicina Interna I, A.O.U. Maggiore della Carità – Novara, Università del Piemonte Orientale, Novara, Italy

*Corresponding author: elena.boin@tiscali.it

INTRODUCTION Marfan syndrome is a genetic disorder that results in connective tissue abnormalities, mainly manifesting as skeletal, cardiovascular, and ocular alterations. On the other hand, Addison disease is a disorder characterized by an insufficient cortisol production. The diagnosis of Addison disease is difficult, as this condition usually develops slowly and shows a certain variety of symptoms that might be misinterpreted. Several precipitating factors, notably concurrent gastrointestinal diseases, may lead to an adrenal crisis. Under such conditions, it is particularly important to establish a prompt diagnosis, as the disease is associated with high morbidity and mortality.

We report a case of Addison disease in a young patient with Marfan syndrome, who presented to the emergency room due to vomiting, diarrhea, and fever. He was initially diagnosed with septic shock due to evidence of hypotension nonresponsive to fluid repletion, but he eventually turned out to have an adrenal crisis. This case highlights the difficult diagnosis of an acute presentation of Addison disease and its risks, especially in patients with comorbidities.

CASE DESCRIPTION A 25-year-old man with Marfan syndrome was admitted to our Internal Medicine Department, complaining of fever, vomiting, and diarrhea. He also reported hyporexia and progressive fatigue, which had made him bedridden after having undergone right foot arthrodesis 3 months earlier. On admission, his vital signs were as follows: blood pressure, 70/30 mm Hg; heart rate, 112 bpm; oxygen saturation, 99% on room air; and temperature, 37.8°C. The patient was drowsy, confused, and extremely asthenic and cachectic (body mass index, 12.4 kg/m^2). Blood tests revealed anemia, neutrophilic leukocytosis, hypoglycemia, and renal impairment with mild hyponatremia. Chest radiography and abdominal computed tomography did not show significant abnormalities. Based on a presumptive diagnosis of septic shock, intravenous fluids, norepinephrine, and empiric antibiotic therapy were administered after collecting blood and fecal cultures, which eventually yielded negative results. The patient's condition improved, but hypoglycemia, hypotension, and weakness persisted. Hyperpigmentation of the skin was noted, leading to the suspicion of an adrenal crisis in chronic Addison disease. Hence, the serum levels of adrenocorticotropic hormone and cortisol were measured, confirming severe adrenal insufficiency. Intravenous hydrocortisone was started, later switched to oral therapy with cortone acetate, which resulted in a rapid improvement of clinical condition. To confirm an autoimmune etiology, blood samples were analyzed for the presence of anti–21-hydroxylase antibodies, yielding positive results.

DISCUSSION The diagnosis of Addison disease can be challenging, especially because of nonspecific manifestations such as fatigue, weight loss, and gastrointestinal symptoms. They are easy to be misdiagnosed and thereby treated as gastroenteritis or septic shock, potentially leading to death. This particularly applies to patients with Marfan syndrome, who are extremely fragile because they are already prone to other potentially life-threatening complications.

Key words
Addison disease, adrenal insufficiency, Marfan syndrome, septic shock

UDDALAK CHAKRABORTY *(PRESENTED BY TANUKA MANDAL)*

An interesting case of fever, rash, and polyarthritis

Uddalak Chakraborty*, Tarun Paria, Jyotirmoy Pal

Department of General Medicine, R. G. Kar Medical College and Hospital, Kolkata, India

*Corresponding author: uddaalakchakraborty@gmail.com

Poncet disease has been reported to be associated mainly with extrapulmonary tuberculosis but very rarely with pulmonary tuberculosis. Its association with immunoglobulin A (IgA) vasculitis is even rarer. We report a case of a 15-year-old male patient who presented to our department in June 2018 with low-grade fever, cough with mucoid expectoration, and bilateral symmetrical nonerosive polyarthritis along with palpable purpura of the lower limbs and buttocks. After extensive workup, he was diagnosed with pulmonary tuberculosis on the basis of a sputum sample analyzed using the cartridge-based nucleic acid amplification test. Polyarthritis was attributed to Poncet disease, while skin biopsy revealed IgA vasculitis. The rash resolved and polyarthritis markedly improved with antitubercular therapy. Our case illustrates a rare association of Poncet disease with pulmonary tuberculosis and IgA vasculitis (Henoch–Schönlein purpura).

Key words
IgA vasculitis, Poncet disease, pulmonary tuberculosis

EUNHO CHOI

Recognized for best poster

Pituitary invasive aspergillosis with lung aspergilloma presenting as headache and hyponatremia

Eunho Choi, Sun Bean Kim, Min Ja Kim

Department of Internal Medicine, Korea University Anam Hospital, Seoul, South Korea

*Corresponding author: buzzchoikr@gmail.com

Diseases affecting the pituitary gland can manifest with hormonal disorders, headaches, or visual impairment. Fungal infections involving the pituitary gland are rarely reported. We present a case of invasive aspergillosis involving the pituitary gland and multiple sinus cavities with hypopituitarism.

A 75-year-old man with a previous history of hypertension and diabetes mellitus presented with a nonspecific headache lasting several months and hemoptysis lasting 1 week. On admission, he was alert and afebrile. Neurologic examination was normal. Laboratory tests showed hypo-osmolar hyponatremia due to syndrome of inappropriate antidiuretic hormone. Chest computed tomography performed due to hemoptysis showed a 23-mm nodule with pleural invasion in the right upper lung, which was highly suspicious of lung cancer. Positron emission tomography revealed a hypermetabolic mass in the right upper lung and a nodular lesion in the pituitary gland. A video-assisted thoracoscopic wedge resection of the lung mass was performed, and a pathologic examination confirmed it to be aspergilloma. No further treatment was administered. However, headache and visual impairment worsened. For further evaluation, a pituitary hormone test was done, and hypopituitarism was diagnosed. Pituitary magnetic resonance imaging showed enhancement in the bilateral sphenoid sinus, right orbital apex, cavernous sinus, and pituitary gland. Leptomeningeal enhancement was not definite, and cerebrospinal fluid examination was normal. With underlying diabetes and rapidly progressing symptoms, an invasive fungal disease including aspergillosis or mucormycosis was suspected. An endoscopic biopsy of the sphenoid sinus was performed, and empirical intravenous amphotericin B was given. The pathological diagnosis was invasive aspergillosis. Amphotericin B was switched to oral voriconazole, which the patient has been currently taking for 3 months.

Invasive aspergillosis involving the pituitary gland should be considered in the differential diagnosis of a pituitary mass in an immunosuppressed patient, because the mortality rate is high if untreated. Simultaneous intensive antifungal therapy with aggressive histopathologic diagnosis is needed for a successful treatment.

Key words
aspergillus, pituitary gland, pulmonary aspergilloma, sinusitis, voriconazole

INGA CHOMICKA

Is sport healthy enough? Rhabdomyolysis with acute kidney injury requiring dialysis after a marathon run

Inga Chomicka*, Małgorzata Kępska-Dzilińska

Department of Nephrology, Dialysis and Internal Diseases, University Clinical Center, Medical University of Warsaw, Warsaw, Poland

*Corresponding author: inga.chomicka@gmail.com

INTRODUCTION Rhabdomyolysis is a clinical syndrome caused by muscle cell damage, which leads to a release of toxic intracellular content into the circulatory system. The breakdown of skeletal muscle tissue can be induced by numerous factors, including extreme physical effort. This syndrome can be life threatening. The main complications include acute kidney injury, electrolyte disorders, and disseminated intravascular coagulopathy. The major goal in the management of rhabdomyolysis is to prevent kidney injury, which can be achieved by fluid resuscitation. In some cases, renal replacement therapy is required to manage complications.

CASE DESCRIPTION A 36-year-old man was admitted to our department with a complaint of acute abdominal pain and anuria. Two days before hospitalization, he fainted during a marathon. On admission, abdominal tenderness with a positive Blumberg sign was observed. Based on laboratory test results (sCK, 254 060 U/l; myoglobin, 268 600 ng/ml), he was diagnosed with rhabdomyolysis, acute kidney injury (creatinine, 6 mg/dl; urea, 115 mg/dl; potassium, 6.3 mmol/l), liver damage, and massive inflammation (white blood cell count, 18.38 t/l, C-reactive protein, 204.1 mg/l; procalcitonin, 157 mg/ml). Abdominal radiography and computed tomography revealed intestinal occlusion suggesting an ischemic etiology. The patient underwent a surgery with a segmental resection of the small intestine. During the procedure, no blood clot was found in the arteries. Conservative treatment was insufficient; therefore, renal replacement therapy was introduced. Once the patient's condition stabilized, further radiologic, serologic, and coagulation tests were performed to exclude potential complications, such as coagulopathy. The results showed no abnormalities. The applied treatment resulted in normalization of kidney function and a reduction in the levels of inflammatory parameters. The patient was discharged in good general condition.

DISCUSSION Our case illustrates severe rhabdomyolysis with life-threatening complications accompanied by intestinal occlusion and suspicion of ischemic colitis. It alerts clinicians to rare complications of rhabdomyolysis, such as disseminated intravascular coagulopathy.

Key words
acute kidney injury, bowel obstruction, hemodialysis, rhabdomyolysis

HYUNJI CHUNG

Ventricular tachycardia in a patient with acute overdose of mycophenolate mofetil

Hyunji Chung*, Kyung Ann Lee, Sang-Heon Lee

Department of Internal Medicine, Konkuk University Medical Center, Konkuk University School of Medicine, Seoul, South Korea

*Corresponding author: 20150117@kuh.ac.kr

INTRODUCTION Mycophenolate mofetil (MMF) is an immunosuppressive drug used to prevent rejection in organ transplant recipients or to manage autoimmune diseases, such as systemic lupus erythematosus. Common adverse effects of MMF include gastrointestinal and hematologic manifestations. Cardiac toxicity, such as sustained ventricular tachycardia (VT), has not been reported before.

CASE DESCRIPTION A 20-year-old woman with lupus nephritis was found unresponsive presented to the emergency department after ingestion of 34 g of MMF in a suicide attempt. At 2 hours from ingestion, electrocardiography showed sustained monomorphic VT with hypotension. The arrhythmia spontaneously converted to sinus VT within 1 minute, and blood pressure normalized after a resuscitation bolus of normal saline. At 12 hours, ventricular bigeminy was reported without hemodynamic instability. Echocardiography revealed no evidence of structural heart disease. Symptomatic treatment with antiemetics and potassium replacement therapy were administered. We ordered oral cholestyramine to reduce enterohepatic recirculation (8 g 3 times/d for 11 days). The total white blood cell count and serum mycophenolic acid (MPA) levels were regularly monitored for 5 days from ingestion. The white blood cell count was within the reference range for 5 days. The MPA level was 7.25 μg/ml at 12 hours and decreased to 1.5 μg/ml at 72 hours from ingestion. The patient was discharged after 5 days without any sequelae.

DISCUSSION Most of the side effects of MMF described in the literature were mild. Apart from a case of hypotension that occurred 8 hours after taking 9 g of MMF, there have been no reports of acute cardiotoxicity, such as VT. It may be assumed that hypokalemia due to vomiting caused a prolonged QT interval and sustained monomorphic VT. This is different from the majority of recognized drug-induced ventricular arrhythmias related to a prolonged QT interval causing polymorphic VT.

Also, it cannot be excluded that an acute overdose of MMF may have directly induced the VT, considering that ventricular bigeminy was documented at 12 hours when serum potassium concentrations were maintained within the reference range with adequate potassium chloride supplementation. Plasma MPA reaches the maximum concentration within 1 hour from ingestion, with a secondary increase due to enterohepatic recirculation at about 6 to 12 hours. It can be assumed that in our case, the MPA level reached the first peak within 2 hours from ingestion, leading to sustained VT with hypotension, and the secondary peak at 12 hours, causing ventricular bigeminy. Considering that the MPA level at 12 hours was 7.25 μg/ml and the half-life of MPA is 16 hours, the baseline MPA level must have been much higher than the optimal therapeutic range (1~3.5 μg/ml).

CONCLUSION Our case suggests that MMF overdose can induce VT. Long-term electrocardiographic monitoring and measurement of MPA levels are critical in patients with a rare cardiac complication of MMF overdose. Cholestyramine is helpful in reducing enterohepatic recirculation of MPA.

Key words
intoxication, mycophenolate mofetil, systemic lupus erythematosus, ventricular tachycardia

FRANCISCO JAVIER CONSIGLIO

Celiac disease and ascites: an unusual association and etiology

Francisco Consiglio*, Roberto Parodi, Alcides Greca

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

*Corresponding author: franciscojconsiglio@gmail.com

INTRODUCTION Celiac disease is a common condition; however, the finding of ascites represents a diagnostic challenge. Clinical reasoning following diagnostic clues can lead to establishing the etiology.

CASE REPORT A 28-year-old male patient was admitted because of a 3-month history of celiac disease, diarrhea lasting 1 month, and pedal edema. Seven days before admission, he also experienced abdominal distension and pain. On physical examination, normal blood pressure and heart sounds without murmurs were detected. Vesicular breath sounds and neurologic examination results were normal. The abdomen was painful and distended, with signs of ascites. Lower limb edema with a positive Godet sign was present. Laboratory tests showed thrombocytosis with normal erythrocyte and leukocyte counts. No significant 24-hour proteinuria was detected, and chest radiography and echocardiography were normal. Abdominal fluid revealed a white blood cell count of 1800/mm^3 (70% granulocytes). Serum-to-ascites albumin gradient was 0.6 g/dl (<1.1 g/dl). Spontaneous bacterial peritonitis was diagnosed, and antibiotic treatment was indicated. A gluten-free diet was started. Diuretic treatment was needed because edema did not resolve. On the basis of hypoalbuminemia, cholestasis, prolonged prothrombin time, and spontaneous bacterial peritonitis, hepatopathy was suspected. An HIV test, as well as hepatitis C and B viruses, yielded negative results. Ferritin and thyrotropin levels were normal. Antinuclear, antimitochondrial, anti-smooth muscle, and antineutrophil cytoplasmic antibody titers were negative. Ascite cultures did not reveal any bacterial, mycobacterial, or fungal agent. A computed tomography scan showed no tumor or adenopathy, but hypertrophy of the caudate lobe of the liver was detected. Budd–Chiari syndrome was suspected, and a Doppler ultrasound confirmed the diagnosis. A thrombophilic condition was investigated. Laboratory studies included bone marrow examination, anticardiolipin antibodies, peripheral blood cytometry, *JAK2* and *G20210A* gene mutations, homocysteine, antithrombin III activity, as well as protein C and S levels. All parameters were within the reference ranges. A heterozygous factor V Leiden mutation was demonstrated, and oral anticoagulant treatment was started. As ascites and edema did not improve, a portocaval shunt was performed by an interventional radiologist, with resolution of the clinical picture.

COMMENTS Three clues in this case were fundamental for the diagnosis: ascites in a patient with celiac disease, the presence of hepatopathy, and thrombophilia. The factor V Leiden mutation is a common cause of Budd–Chiari syndrome. A recent literature review described only 28 cases of celiac disease and Budd–Chiari syndrome reported so far. Most of these reports concerned patients of North African origin, and less than 50% of the cases presented underlying thrombophilia. In our patient with factor V Leiden mutation, thrombosis of the suprahepatic veins was probably triggered by celiac disease.

LESSON TO BE LEARNED FROM THE CASE In our case, clinical reasoning following diagnostic clues allowed for a useful workup. We could identify an unusual association and etiology, choose an adequate treatment, and finally reach a favorable result.

Key words
Budd–Chiari syndrome, celiac disease, factor V Leiden mutation

KESHIA DE FREITAS

Mushrooms in the forest: what can they be? A case of undiagnosed diffuse skin lesions in a seropositive patient

Keshia De Freitas*, Melissa McDonald, Zahira Khalid

Georgetown Public Hospital Corporation, Georgetown, Guyana

*Corresponding author: keshiadefreitas@gmail.com

A 34-year-old Afro-Guyanese male patient, who had been diagnosed with HIV in 2007 but was lost to follow-up in the clinic, presented with generalized weakness, shortness of breath on exertion, and diffuse generalized skin lesions. Approximately 1 month earlier, he presented to his HIV clinic, where he underwent a diagnostic workup for tuberculosis and treatment for pruritic papular eruptions. On physical examination, he showed generalized hyperpigmented papules extending from the head to the extremities. He was admitted to our hospital for investigation of his skin lesions and generalized weakness, and he underwent treatment for community-acquired pneumonia, anemia with hyponatremia, and cutaneous lesions. However, he did not respond to treatment and his condition deteriorated. Skin scraping and biopsy were performed, but the patient died before the results were available. Biopsy showed patches of lymphoid infiltration. According to the biopsy report, the findings suggested mycosis fungoides, which is a rare diagnosis in patients with HIV.

Key words
HIV, mycosis fungoides, papules, seropositive, skin lesions

KHALID ELKHOLY

Dermatology meets medicine: from a skin tissue to a multi--visceral issue

Khalid Elkholy[1]*, Qamarul Razli[2], Bairbre Wynne[2], Dierdre O'Riordan[1]

1 Department of Respiratory Medicine, St. James's University Hospital, Dublin 8, Ireland

2 Department of Dermatology, St. James's University Hospital, Dublin 8, Ireland

*Corresponding author: dr.khalid.elkholy@gmail.com

INTRODUCTION In 1977, 3 physicians, Arthur Birt, Georgina Hogg, and James Dubé, described a large Canadian family with 3 generations of individuals who presented with tiny, smooth, flesh-colored papules on the face and neck, called fibrofolliculomas. From then on, these skin lesions have been seen in association with what has since been called Birt–Hogg–Dubé syndrome (BHDS), a rare hereditary condition with an autosomal dominant inheritance pattern.

Birt–Hogg–Dubé syndrome is primarily characterized by these multiple benign skin tumors and other hamartomas, thin-walled pulmonary cysts (>80% of cases), and an increased risk of various types of tumors, including a high prevalence of renal tumors. The syndrome's exact incidence is unknown; however, it has been reported in more than 400 families worldwide. It has been shown to be caused by germline mutations in the folliculin (*FLCN*) gene.

Patients with BHDS have an elevated risk of life-threatening complications. Therefore, recognition of this syndrome is critical to timely introduce appropriate surveillance, therapy, and prevention of potential complications, as well as to provide genetic counseling to closely related family members.

CASE We describe a 39-year-old woman who attended her surveillance appointments at a dermatology clinic. She had a previous history of a superficial malignant melanoma (pT3a according to the American Joint Committee on Cancer Melanoma Staging System) of her left shoulder (Breslow thickness, 2.7 mm) in 2013, for which she underwent a wide local excision with negative sentinel lymph node biopsies at the time. During these visits, she had concerns over numerous subtle, but subjectively disfiguring, bumps on her forehead and cheeks, developing over a few months.

On examination, these bumps were 1 to 3 mm in size. On closer inspection, they appeared as white to flesh–colored, smooth, dome--shaped papules, easily dismissible by clinicians. Skin biopsies were undertaken due to diagnostic uncertainty, and a histopathologic diagnosis of fibrofolliculoma was established. Due to a relevant association with BHDS, molecular genetic testing for the *FLCN* gene mutation was ordered to confirm the diagnosis. This identified a missense mutation in exon 13, which reflects a pathogenic variant.

A thoracic computed tomography was performed, which revealed multiple bilateral thin-walled lung cysts. There was no evidence of a pneumothorax, but the patient was referred for a consultation with respiratory specialists and remained under their close supervision.

Given the high risk of renal tumors, abdominal magnetic resonance imaging (MRI) was also performed. Fortunately, it revealed no kidney abnormalities and no evidence of neoplastic lesions. An annual surveillance with abdominal MRI was subsequently planned.

DISCUSSION Birt–Hogg–Dubé syndrome is a rare inherited multiorgan condition with oncogenic predisposition due to germline mutations in the tumor suppressor *FLCN* gene.

Undiagnosed patients or those with a delayed diagnosis of BHDS have an elevated risk of life-threatening complications, including spontaneous pneumothorax, skin tumors, malignant renal tumors, and various other neoplastic manifestations.

In terms of the management, a multispecialty and multidisciplinary approach is essential, involving dermatologists, pulmonologists, and nephrologists among other medical specialists, as deemed relevant to the patient's presenting complaints. Recognition of this clinical entity is critical in order to timely introduce appropriate management and provide early genetic counseling for the potentially affected family members.

Key words
Birt–Hogg–Dubé syndrome, fibrofolliculoma, folliculin, pneumothorax, tumors

COLM KERR

A retiree, a chronic cough, and a diagnosis of cystic fibrosis

Colm Kerr*, David Morrissy, Mary Horgan, Barry Plant

Cork University Hospital, Cork, Ireland

*Corresponding author: colmkerr@gmail.com

Cystic fibrosis is the most common life-limiting autosomal recessive genetic disorder among white populations. The majority of cystic fibrosis cases are diagnosed in childhood; however, an increasing number of adults are being diagnosed with the condition. We report a case of a 65-year-old Irish woman presenting with a cough productive of green sputum on a background of a 5-month history of recurrent respiratory tract infections requiring multiple hospital admissions. *Staphylococcus aureus*, *Scedosporium apiospermum*, and *Stenotrophomonas maltophilia* were grown from bronchoalveolar lavage, raising suspicion for cystic fibrosis. Genetic testing subsequently revealed the presence of ΔF508 and R117H cystic fibrosis mutations. In this case, the microbiological results were key to arriving at a diagnosis of cystic fibrosis.

Key words
CFTR potentiators, cystic fibrosis, *Scedosporium apiospermum, Staphylococcus aureus*, *Stenotrophomonas maltophilia*

MARCO MORETTI

Late-onset Lemierre syndrome: a case report

Marco Moretti*, Sabine D. Allard

Department of Internal Medicine and Infectious Disease, Vrije Universiteit Brussel, UZ Brussel, Belgium

*Corresponding author: marco.moretti9@gmail.com

Lemierre syndrome is a septic thromboembolic complication of an oropharyngeal or neck infection, primarily caused by *Fusobacterium* species. Although it usually affects young healthy patients, some case reports described this syndrome in elderly individuals. We report a case of a 66-year-old patient with comorbidities, diagnosed with Lemierre syndrome caused by *Fusobacterium nucleatum*. Beside bilateral internal jugular vein thrombosis, distant thromboembolism to the lungs, central nervous system, and liver were observed, which initially obscured the diagnosis and delayed the treatment of Lemierre syndrome. Lemierre syndrome should also be considered in elderly patients with an oropharyngeal infection and systemic involvement.

Key words
Fusobacterium nucleatum, infectious disease, internal jugular vein thrombosis, Lemierre syndrome, septic embolism

DOMINIKA MULAWKA

Atrial septal defect: a rare cause of pulmonary artery hypertension

Dominika Mulawka[1]*, Paulina Mulawka[2]

1 Department of Cardiology, Provincial Specialist Hospital, Lublin, Poland
2 Department of Internal Medicine, Medical University of Lublin, Lublin, Poland

*Corresponding author: dominikamulawka@gmail.com

An atrial septal defect (ASD), one of the most common congenital heart diseases in adults, rarely presents late as pulmonary artery hypertension (PAH). The hemodynamic definition of PAH is a mean pulmonary artery pressure of 25 mm Hg or higher measured by right heart catheterization. Pulmonary artery hypertension is characterized by a progressive increase of pulmonary pressure, which can lead to right-sided heart failure and death. Patients with ASD complicated by PAH require a different medical approach due to the considerable effect of PAH on management, morbidity, and mortality.

Nowadays, prenatal screening and fetal echocardiography enable an early diagnosis and timely intervention. Early detection and closure of a significant ASD might prevent some of the associated complications including arrhythmias, paradoxical embolism, brain abscess, stroke, PAH, pulmonary artery aneurysm, right ventricular failure, and, finally, Eisenmenger syndrome. However, there are also patients who present with ASD as an incidental finding or with its subsequent complications for the first time in adult life. In some cases of ASD, closure might be contraindicated because of a delayed diagnosis. In these patients, alternative management should be considered. A detailed clinical interview and physical examination, together with competent interpretation of a routine chest radiograph and electrocardiogram, can facilitate a proper diagnosis of ASD complicated by PAH, thus improving clinical outcomes.

We present a case of a 29-year-old woman from southeastern Poland who was referred to our Department of Cardiology 8 years earlier with malnutrition, cyanosis, and right heart failure symptoms just after her second childbirth. Right heart catheterization, which is the gold standard procedure, confirmed the initial suspicion of undiagnosed ASD complicated by PAH, which led to cardiac cachexia. Unfortunately, the hemodynamic examination was complicated by paradoxical embolism and stroke, which resulted in left-sided hemiparesis and left facial nerve paralysis. Our patient was immediately transferred to the Department of Neurology, where she was successfully treated with thrombolytic drugs. After 1 year of targeted treatment for PAH, the patient changed her functional class from the fourth to first according to the World Health Organization classification and returned to her normal life. She suffered an episode of depression, which was treated with a selective serotonin reuptake inhibitor (fluoxetine). She also developed chronic obstructive pulmonary disorder because of smoking. Currently, she requires chronic targeted treatment for PAH and close follow-up in our center.

We believe that this case report will help raise the awareness of PAH and facilitate timely diagnosis, as the delay might reduce the length and quality of life of these patients.

Key words
atrial septal defect, Eisenmenger syndrome, paradoxical embolism, pulmonary artery hypertension

PABLO PUIG

A chagoma and Chagas disease in a patient with systemic lupus erythematosus

Pablo Puig[1]*, Pablo Guerrero[1], Alfredo Palavecino[2]

1 Department of Internal Medicine, Hospital San Bernardo, Salta, Argentina
2 Department of Neurology, Hospital San Bernardo, Salta, Argentina

*Corresponding author: puigpabloi@gmail.com

INTRODUCTION Chagas disease (also referred to as American trypanosomiasis) is a parasitic infection endemic in Latin America. It is caused by the flagellate protozoan parasite *Trypanosoma cruzi*. The disease is associated with several typical syndromes. The reactivation of the infection during the chronic phase may occur in immunosuppressive states, with severe clinical manifestations including central nervous system compromise and myocarditis. Chagas disease reactivation with a concomitant diagnosis of autoimmune disease without previous immunosuppressive drug therapy has not been reported before.

CASE REPORT A 63-year-old male Wichí patient from Pichanal (Salta, Argentina), with a history of thyroid insufficiency and hypertension, was referred to our unit with ascending muscle weakness in the lower limbs and fatigue, with suspicion of Zika virus infection in the setting of prolonged fever syndrome. On admission, the patient was in good mental state and presented with muscle weakness predominantly affecting the proximal lower limbs, loss of bilateral patellar reflexes, as well as hypesthesia and hypopalestesia in the lower extremities. Brain computed tomography (CT) was unremarkable. A spinal tap revealed no abnormalities. Electromyography showed symmetrical sensory and motor polyneuropathies, myelin with secondary axonal compromise in the lower limbs, undetected denervation, and absent F waves. Zika virus–related Guillain-Barré syndrome was suspected. Immunoglobulin therapy was administered for 5 days, resulting in improved muscular strength and subsequent impaired sensorium. The brain CT was repeated, revealing a hypodense lesion in the pons. Magnetic resonance imaging [MRI] showed hypointensity in T1-weighted images and hyperintensity in FLAIR and T2-weighted images, with ring-enhancing lesions revealed in the right pons after administration of an intravenous contrast agent. Isolated hyperintensity in FLAIR and T2-weighted images of the periventricular white matter was also identified. At the same time, negative cerebrospinal fluid (CSF) cultures were received (polymerase chain reaction [PCR] and serology of CSF samples negative for Zika infection and tuberculosis). Relatives were interviewed and reported to have had a 3-month history of large-joint polyarthralgia and diffuse hair loss. The association of encephalopathy and polyneuropathy due to an autoimmune disease or infection was suspected (Sjögren syndrome vs systemic lupus erythematosus). Laboratory findings were as follows: rheumatoid factor of 1898 UI/ml; negative antinuclear factor, ANA-DNA, anti-La antibodies, IgG anticardiolipin, anti-neutrophil cytoplasmic antibody titers and cryoglobulins; reduced levels of complements C3 and C4; positive anti-Ro, anti-Sm, and IgM anticardiolipin antibody titers;

and peroxidase antibodies of 1/50. On serologic testing, the patient was positive for Chagas immunohistochemical assay/enzyme-linked immunosorbent assay and toxoplasma IgG antibodies but negative for toxoplasma IgM, HIV, syphilis, bovine herpes virus, canine herpes virus, alcelaphine herpesvirus, mononucleosis, and hydatid disease. He was also negative for human T-cell leukemia virus 1 and 2. Fungal assay results were negative. A tentative diagnosis of systemic lupus erythematosus led to pulse therapy and maintenance steroid treatment. On follow-up magnetic resonance imaging, an increased size and number of lesions in the corpus callosum were noted. Brain biopsy revealed multifocal lymphohistiocytic inflammation with organisms consistent with amastigotes. The PCR of an encephalic tissue sample was positive for *Trypanosoma cruzi*. A therapy with nifurtimox for 30 days and benznidazole for 20 days was started. As no neurologic improvement was obtained, brain MRI was repeated, revealing new abscess lesions. The patient presented with multiple infections, which led to his death.

DISCUSSION Although Chagas disease occurs mainly in patients with HIV coinfection and transplant recipients, other conditions that lead to immunosuppression (such as autoimmune diseases) also deserve special attention, particularly in disease-endemic areas. It is important to develop diagnostic protocols as well as therapy and follow-up of patients with this untypical presentation with the view to improving their management and prognosis.

Key words
Chagas disease, chagoma, meningoencephalitis, reactivation, systemic lupus erythematosus

SOO MYOUNG SHIN

Recognized for best poster

Food-dependent exercise-induced anaphylaxis confirmed with an alcohol challenge test: a case report

Soo Myoung Shin[1*], Sung Won Chung[1], Hye-Ryun Kang[1,2]

1 Department of Internal Medicine, Seoul National University College of Medicine, Seoul, South Korea
2 Institute of Allergy and Clinical Immunology, Seoul National University Medical Research Center, Seoul, South Korea

*Corresponding author: smshin992@gmail.com

Food-dependent exercise-induced anaphylaxis (FDEIA) is a systemic allergic food reaction triggered by physical exercise. Beside physical exercise, medications such as nonsteroidal anti-inflammatory drugs or aspirin, alcohol, and physical or atmospheric conditions can trigger allergic reactions to food ingestion. Although skin prick tests to food allergens and the measurement of serum food-specific immunoglobulin E (IgE) levels are helpful in evaluating sensitization status, a food and exercise challenge test is the gold standard in the diagnosis of FDEIA.

A 56-year-old man presented to the allergy clinic with urticaria, palpitation, and dyspnea, which developed when he was taking a walk after eating noodles and pork fries for breakfast. He had a history of chronic urticaria. Moreover, 6 years earlier, he experienced an episode similar to the current one, combined with profound hypotension, which resolved with epinephrine injection at a local hospital. Laboratory tests revealed a total IgE antibody titer of 123 kU/l, while serum specific IgE levels measured by the ImmunoCAP system showed positivity to wheat, gluten, and ω5-gliadin. Food challenge tests with bread were negative both with and without exercise. On additional and meticulous history taking, we found that these episodes were related to his drinking alcohol liquor on the previous day. Based on this information, another food and exercise challenge test was scheduled, in which bread ingestion was followed by alcohol ingestion and exercise. After the challenge with bread, the patient drank 3 glasses of alcohol. Urticaria developed on his neck and back on a short walk to the treadmill room. After 10 minutes of walking on a treadmill, his systolic blood pressure suddenly decreased to 90/56 mm Hg. We stopped the test and administered an intramuscular epinephrine injection as well as intravenous fluid. The patient's condition returned to normal, and he was finally diagnosed with FDEIA. He was instructed to avoid exercise after eating wheat, especially with simultaneous alcohol drinking. We prescribed self-injectable epinephrine as a rescue medication. Over the 3 years of regular follow-up, he reported no episode of anaphylaxis.

Our case of wheat-dependent exercise-induced anaphylaxis triggered by alcohol ingestion suggests that the food and exercise challenge test with specific consideration of the triggering factor such as alcohol may help enhance the diagnostic sensitivity of the challenge test and confirm a food-related allergic disease.

Key words
alcohol, anaphylaxis, exercise, food hypersensitivity, wheat hypersensitivity

MUHARREMI SHKELQIM

A case of seronegative pulmonary–renal syndrome: a diagnostic and therapeutic challenge

Shkelqim Muharremi[1*], Gjulsen Selim[2], Zvezdana Petronijevik[2], Vangjelka Kovaceska[1], Liljana Tozija[2], Muhamed Saidi[2], Adrijana Spasovska[2], Nikola Gjeorgjievski[2], Ana Stojanoska[2], Bojana Poposka[2], Angela Kabova[2], Mimoza Milenkova[2]

1 Institute of Nephrology, Struga, Macedonia
2 University Clinic of Nephrology, Skopje, Macedonia

*Corresponding author: shkelqimmuharremi@hotmail.com

Pulmonary–renal syndrome (PRS) is most commonly used to describe a combination of glomerulonephritis and pulmonary hemorrhage as a manifestation of a multisystem autoimmune disease. It is usually associated with antineutrophil cytoplasmic antibody (ANCA)–associated vasculitis and anti–glomerular basement membrane (GBM) disease.

We report a case of a 51-year-old man who was initially suspected of Goodpasture syndrome and pneumonia, but later lung imaging studies revealed diffuse alveolar hemorrhage. Renal biopsy showed crescent glomerulonephritis. We were unable to classify the patient into any known subgroups because all commercially available serologic tests were negative. As a result, PRS was diagnosed on the basis of clinical manifestations and pathological findings. 24-hour proteinuria was measured several times and was used as a unique parameter to monitor the disease activity. As the patient's condition worsened, we administered corticosteroid pulse therapy and cyclophosphamide, followed by 5 courses of plasmapheresis starting on day 4. During the first hospitalization, the patient developed chronic kidney disease stage 4, with maintained diuresis and no need of hemodialysis. However, 4 weeks later, he was hospitalized again and despite 9 more courses of plasmapheresis, an increase in the corticosteroid dose, and switching cyclophosphamide to mycophenolate mofetil, his kidney function and general condition did not improve. Therefore, we decided to put the patient on hemodialysis.

Diffuse alveolar hemorrhage in a patient with pauci-immune glomerulonephritis and negative ANCA and anti-GBM antibody titers is a rare condition. Our case has important clinical implications because unclassified PRS without the presence of ANCA and anti-GBM antibodies is extremely rare and is associated with high morbidity and mortality rates.

Key words
ANCA vasculitis, plasmapheresis, pulmonary–renal syndrome, seronegative, 24-hour proteinuria

PAULA ANTONELLA SILVA

Pericardial effusion as the first manifestation of giant cell arteritis

Paula Antonella Silva*, María Emilia Manzanares Ciancio, Enrique Mario Baldessari

Internal Medicine Department, University Hospital Favaloro Foundation, Buenos Aires, Argentina

*Corresponding author: psilva@ffavaloro.org

Giant cell arteritis (GCA) is a chronic inflammatory disease of the medium and large blood vessels, which mainly affects cranial branches of the arteries originating from the aortic arch. Patients usually present with headache, fever, weight loss, symptoms of polymyalgia rheumatica, dilation of the temporal artery, claudication of the jaw, and blindness. Pericardial effusion is an extremely rare manifestation of GCA, and it should always be included in the differential diagnosis of idiopathic pericarditis, especially in patients over 50 years of age.

We present a case of a 59-year-old woman with a history of Fabry disease without systemic involvement, consulted for anorexia, early satiety, nausea, fever, and dry cough of 1 month of evolution. Thoracic computed tomography showed moderate pericardial effusion and minimal bilateral pleural effusion. She reported predominantly nocturnal fever, which she had consulted in our institution, and received several antibiotic schemes without improvement of symptoms. Physical examination revealed skin pallor and dilation of both temporal arteries, which were painless on palpation. She denied mandibular claudication or visual alterations. Laboratory tests showed hypochromic normocytic anemia (hematocrit, 26%; hemoglobin, 8.2 g/dl), erythrocyte sedimentation rate greater than 120 mm, and C-reactive protein levels of 18.8 mg/l. Doppler echocardiography showed moderate pericardial effusion without compromise of the cavities. Blood cultures, serology, and the profile of autoimmune diseases were negative. Doppler echocardiography of the temporal arteries showed both vessels to be permeable, without signs of periluminal halo. Given the nonspecific findings, whole-body positron emission tomography was performed, which showed a marked increase in the diffusion of the metabolic activity of the thoracoabdominal aorta wall, from the supra-aortic branches, subclavian, axillary, and primitive carotid arteries to its bifurcation. At the left temporal level, there was superficial vascular thickening with metabolic uptake. Metabolic findings were compatible with arteritis.

Treatment with deltisone (1 mg/kg/d) was started. Left temporal artery biopsy reported hyperplasia and myointimal fibrosis with luminal occlusion of 25% to 50%, without parietal inflammatory infiltrate. The case was interpreted as GCA.

Given the age older than 50 years, erythrocyte sedimentation rate greater than 50 mm/h, and the report of the temporal artery biopsy, the diagnosis of GCA was made. The patient received 1-month treatment with steroids, and clinical and laboratory parameters improved (hematocrit, 42%; erythrocyte sedimentation rate <25 mm; C-reactive protein, 0.5 mg/l; mild pericardial effusion on repeat Doppler echocardiogram).

Giant cell arteritis rarely causes heart problems, such as thoracic aortic aneurysms or aortic dissection. The prevalence of these findings is quite low at the time of diagnosis but is higher during follow-up. However, over the last 40 years, fewer than 30 cases have been reported, and the exact prevalence is unknown. There is evidence that these symptoms improve after corticosteroid therapy. Giant cell arteritis is usually asymptomatic and carries a favorable prognosis. According to the literature, in cases that present with pericardial effusion, diagnosis is often delayed due to atypical presentation, without headache and with clinically normal temporal arteries. The simultaneous presence of pericardial and pleural effusion has been extremely rarely reported. Although the association is uncommon, GCA should always be considered as a probable diagnosis in an elderly patient with pericardial effusion and an unexplained elevation of inflammatory biomarkers.

Key words
arteritis, erythrocyte sedimentation rate, pericardial effusion

OTHER PRESENTATIONS

ANNA BRASZAK

A difficult road to diagnosis: a link between chronic diarrhea and fever of unknown origin

Anna Braszak*, Marta K. Walczak-Galezewska, Wiesław Bryl

Department of Internal Diseases, Metabolic Disorders and Hypertension, Poznan University of Medical Sciences, Poznań, Poland

*Corresponding author: anna.braszak@gmail.com

INTRODUCTION Fever of unknown origin (FUO) remains a medical diagnostic challenge. The most common causes of FUO are infectious diseases, especially tuberculosis, but also connective tissue diseases and neoplasms (particularly non–Hodgkin lymphomas). Despite continued progress in medicine, the etiology of a significant number of FUO cases is considered idiopathic. We present this case to show the importance of a patient's history when available diagnostic methods have been exhausted.

CASE DESCRIPTION A 76-year-old woman was admitted with a recurrent fever lasting over a year accompanied by strong chills, weakness, weight loss of about 10 kg, dehydration, and malnutrition. During the year, the patient had been hospitalized several times for a diagnostic workup of fever; however, the etiology was not established despite a series of advanced diagnostic tests, including magnetic resonance enterography. The patient had undergone partial resection of the small intestine due to perforation and abscess (13 years prior to admission, no histopathologic examinations were performed at the time). Three months prior to admission, the patient underwent surgery for vesicoenteric fistula. She also reported periodic abdominal pain and diarrhea in recent years.

On clinical examination, the patient was fully conscious and stable; cachexia and dehydration were noted. Baseline laboratory tests showed elevated levels of alkaline phosphatase, lactate dehydrogenase, C-reactive protein, anemia of chronic disease (with significantly elevated ferritin levels), and hypoalbuminemia. Microbiologic tests revealed no abnormalities. Chest computed tomography (CT) confirmed pneumonia. An abdominal CT scan revealed a lymph node (20 × 12 mm in size) to the left of the aorta, an increased density of the periaortic fat tissue, as well as free fluid on the border of the abdominal cavity and pelvis. The findings were described as postoperative, inflammatory, or cardiac failure–related changes. Empiric broad-spectrum antibiotic therapy was administered, and the patient was scheduled for ^{18}F-fludeoxyglucose positron emission tomography.

The patient died on the 27th day of hospitalization due to progressive respiratory failure. A repeat histopathologic evaluation of the samples obtained during the fistula surgery revealed a malignant lymphoma of the small intestine. The autopsy showed the presence of lymphoma in the mesentery of the small intestine, uterus, cecum, lung, and mediastinal lymph nodes, classified as peripheral T-cell lymphoma not otherwise specified.

DISCUSSION We strongly believe that the lymphoma was induced by long-term, undiagnosed celiac disease. Despite our growing knowledge of this disease and its increased visibility, it remains significantly underdiagnosed. It has been shown that 2% to 3% of patients with celiac disease will develop an intestinal lymphoma, of which 65% have a T-cell origin. The diagnosis of lymphomas remains difficult, and there are questions about the diagnostic path to be followed when the commonly available laboratory tests and imaging methods do not help determine the diagnosis.

LESSONS TO BE LEARNED FROM THE CASE This case shows that the complications of celiac disease, which is still insufficiently diagnosed, are serious and difficult to diagnose. It also emphasizes the importance of careful monitoring and treatment of symptoms reported by the patient.

Key words
celiac disease, fever of unknown origin, lymphoma

JIMENA CARRIZO

Weight loss and fever concealing central nervous system autoimmunity

Jimena Carrizo*, Clarisa Taffarel, Ramiro Larrea

Internal Medicine Department, Hospital Central de San Isidro, Buenos Aires, Argentina

*Corresponding author: jimenacarrizo@live.com.ar

INTRODUCTION Behçet disease was first described in 1937 by Hulusi Behçet. The disease is characterized by recurrent canker and genital sores, ocular, gastrointestinal, and neurologic involvement, vascular disease, and/or arthritis. Its prevalence varies from 1 in 15 000 to 1 in 500 000 people in North America and North Europe, respectively. The disease mainly affects young adults (aged 20–40 years) and is rare in children. It seems to be more severe in young male patients from Central Asia or the Far East, and the effect on the nervous system is uncommon (less than 5% of cases).

CLINICAL CASE A 52-year-old male patient with a history of smoking as well as marijuana and cocaine abuse presented for a consultation due to persistent febrile syndrome and weight loss. Physical examination showed canker sores in the oral mucosa and hypopigmented lesions on the scrotum. Imaging and laboratory tests revealed the following results: leukocytes, 13 000/µl; hemoglobin, 10.9 g/dl; hematocrit, 33.6%; platelets, 295 000/µl; lactate dehydrogenase, 700 U/l; sodium, 137.1 mmol/l; and erythrocyte sedimentation rate, 43 seg. The patient tested negative for HIV, hepatitis B virus, nonreactive hepatitis C virus, syphilis, or the presence of any bacteria. Brain, thoracic, abdominal, and pelvic computed tomography revealed no abnormalities. Antibiotic therapy with ceftriaxone, ampicillin, acyclovir, tuberculostatic drugs, and dexamethasone was started but was discontinued after excluding infectious causes. Brain magnetic resonance imaging (MRI) showed a lesion (13 × 21 × 17 mm) in the right posterior parasagittal section of the pons, hyperintense in T2- and FLAIR-weighted images and hypointense in T1-weighted images, with mild enhancement after gadolinium administration and multiple focal hyperintense T2- and FLAIR-weighted images with the corona radiata and semioval centers, one of which was located on the medial end of the left globus pallidus.

After a new episode of fever and arthralgia, knee puncture showed inflammatory liquid without microbiological isolation in culture. Pathergy test results were positive.

Based on the presence of clinical, imaging, and laboratory findings compatible with Behçet disease, treatment with high doses of corticosteroid and cyclophosphamide was started. An improvement in neurologic symptoms and resolution of the lesions in the brain stem and basal ganglia were evidenced by MRI performed 15 and 30 days after treatment. The patient remains under motor rehabilitation, and outpatient follow-up shows good clinical evolution.

DISCUSSION The neurologic involvement in Behçet disease is uncommon and usually preceded by other forms of the disease, associated with higher morbidity and mortality. The most frequently affected area is the central nervous system—from 20% to 60% of cases show involvement of the brain parenchyma, with inflammatory lesions in the brain stem and basal ganglia associated with pyramidal symptoms or behavioral disorders, as well as movement disorders and epilepsy due to cortical involvement.

MRI represents a gold standard and allows a differential diagnosis from other neurologic disorders. Cerebrospinal fluid study usually shows hypercellularity with high protein and normal glucose levels.

CONCLUSION Behçet disease is a complex condition given the fact that patients do not present with pathognomonic symptoms, which tends to delay diagnosis and treatment. Neurologic involvement is uncommon but may be associated with worsening prognosis. Therefore, an early diagnosis is the key to improving morbidity and mortality rates.

Key words
autoimmunity, Behçet disease, fever

GIUSEPPE FRANCESCO CASCIARO

A case of severe Wernicke encephalopathy with atypical magnetic resonance imaging findings

Giuseppe Francesco Casciaro*, Veronica Vassia, Elena Boin

S.C.D.U. Medicina Interna 1, A.O.U. Maggiore della Carità di Novara, Università del Piemonte Orientale, Novara, Italy

*Corresponding author: fra.casciaro90@gmail.com

INTRODUCTION Wernicke encephalopathy (WE) is a neurologic disorder induced by thiamine deficiency, usually associated with chronic alcoholism and malnutrition. The classic triad of symptoms includes oculomotor dysfunction, gait ataxia, and encephalopathy. Magnetic resonance imaging (MRI) is the most sensitive imaging technique in detecting neurologic lesions, which can be located in typical (medial thalamus, aqueduct and third ventricle, tectal plate, mamillary bodies, and dorsal medulla) and atypical (cerebellum, cranial nerve nuclei, dentate nuclei, caudate nuclei, red nuclei, splenium, and cerebral cortex) sites. The atypical sites are more frequently involved in nonalcoholic patients.

CASE REPORT A 56-year-old Caucasian man was admitted to the Internal Medicine Department of Academic Hospital (Novara, Italy) because of severe hyporexia along with worsening of general condition, which made him unable to do basic and instrumental activities of daily living. His medical history included chronic alcohol abuse (although he had abstained from drinking for 2 years), depression with several suicide attempts, and gastroesophageal surgery after caustic ingestion.

On admission, his vital signs were as follows: blood pressure, 85/55 mm Hg; heart rate, 65 bpm; oxygen saturation, 97% on room air; temperature, 36.5°C, and Glasgow Coma Scale score (GCS), 10. During the first few days of hospitalization, we observed a rapid deterioration of consciousness (GCS score, 7) in association with the development of hypothermia (temperature, 34°C), pinpoint pupils, severe hyposthenia of the upper and lower limbs, and loss of deep tendon reflexes. In addition, the patient was hypotensive and bradycardic (heart rate, 45 bpm). Electrocardiography showed QT prolongation (corrected QT, 490 ms) and J waves. As sepsis was suspected, we administered intravenous fluids and empiric antibiotic therapy after collecting blood and urine cultures (both negative), but no improvement was observed. Due to the persistence of hypotension and hypothermia, norepinephrine and warm fluid infusion were started, leading to an increase in pressure values.

As pupillary abnormalities suggested pons damage, a neurologic consultation was requested. Electroencephalography showed mild epileptiform abnormalities. Brain MRI highlighted typical bilateral T2-weighted signal alterations in mammillary bodies, posterior mesencephalic regions, cerebral peduncles, periaqueductal area, third ventricular walls, and thalami. Notably, it also showed atypical lesions such as T2-weighted alterations in the posterior pons and bilateral point-like restricted diffusion areas in the cingulate and parietal postrolandic cortex on diffusion-weighted images. We eventually started a thiamine infusion (500 mg 3 times a day, then 250 mg once a day), which led to a slight improvement of mental status.

DISCUSSION Wernicke encephalopathy is a clinical diagnosis that can be missed when features of the classic triad are absent and the presence of other symptoms may suggest more common conditions. According to autopsy studies, Wernicke encephalopathy is not uncommon in the Western world. Therefore, the present case emphasizes that clinicians should maintain a high index of suspicion for this disease, especially if history provides clues for insufficient thiamine intake due to alcoholism, anorexia, and abnormal anatomy of the gastrointestinal tract.

Key words
atypical, brain MRI, hypothermia, thiamine, Wernicke encephalopathy

UDDALAK CHAKRABORTY

Recognized for best poster

An atypical manifestation of lateral medullary syndrome

Uddalak Chakraborty*, Biswajit Banik, Jyotirmoy Pal

Department of General Medicine, R. G. Kar Medical College and Hospital, Kolkata, India

*Corresponding author: uddaalakchakraborty@gmail.com

Lateral medullary syndrome (Wallenberg syndrome) is a common brainstem stroke associated with a classic triad of Horner syndrome (ptosis, miosis, and anhidrosis), ipsilateral ataxia, and loss of pain and temperature sensation in the ipsilateral face along with other symptoms, such as contralateral hemisensory loss of the trunk and extremities, vertigo, diplopia, nausea, vomiting, dysphagia, or hiccups. It most commonly occurs due to atherothrombotic vertebral artery occlusion. We report a case of a 49-year-old postmenopausal woman with diabetes and without hypertension, who presented with symptoms similar to Wallenberg syndrome, involving the left dorsal medulla along with right-sided hemiparesis and the upper motor neuron type of left facial palsy. Contralateral hemiparesis can be explained by a caudal extension of an infarct involving the pyramids before decussation at the medulla, known as Babinski–Nageotte syndrome. The upper motor neuron type of facial palsy can be attributed to the involvement of hypothetical aberrant supranuclear corticobulbar fibers of the facial nerve, which descend into the contralateral ventromedial medulla, decussate at the level of the upper medulla, and then ascend in the dorsolateral medulla to reach the facial nerve nucleus. Although these 2 entities have been each reported in association with Wallenberg syndrome, we are the first to show their simultaneous presence in a patient.

Key words
aberrant, Babinski–Nageotte syndrome, corticobulbar, Wallenberg syndrome

UDDALAK CHAKRABORTY

A case of fever, jaundice, and altered sensorium in a patient with alcoholism

Uddalak Chakraborty*, Tanuka Mandal, Jyotirmoy Pal

Department of General Medicine, R. G. Kar Medical College and Hospital, Kolkata, India

*Corresponding author: uddaalakchakraborty@gmail.com

Scrub typhus is a rickettsial disease with protean manifestations, which can affect all age groups. This zoonotic disease is caused by *Orientia tsutsugamushi*, transmitted by an arthropod vector (ticks) of the Trombiculidae family, and humans are accidental hosts. In India, studies from the 1960s and 1970s revealed the prevalence of this disease in some states and union territories; however, the disease disappeared gradually over the next few years. In recent years, there has been a resurgence of scrub typhus, along with considerable mortality and morbidity. The clinical spectrum of this disease varies from a self-limiting course to life-threatening complications often leading to death. Apart from the acute febrile illness with rash, myalgia, nausea, or vomiting, scrub typhus can present with florid manifestations involving the central nervous system, cardiovascular system, or gastrointestinal system.

We report a case of a 30-year-old man without hypertension or diabetes and with a history of chronic alcohol use for the past 10 years. He presented with high-grade fever, acute onset of jaundice, ascites with hepatosplenomegaly, and cervical lymphadenopathy. A day later, his sensorium deteriorated, and he developed meningeal signs. An extensive workup revealed a high serum–ascites albumin gradient accompanied by lymphocytic pleocytosis and marginally elevated protein levels in cerebrospinal fluid. Other investigations suggested

acute liver failure with meningoencephalitis. Serology testing was ordered. The Weil–Felix test revealed OXK antigen titers of 1:320, while an enzyme-linked immunosorbent assay for scrub typhus revealed immunoglobulin M antibody titers of 1:320, which confirmed the diagnosis. The patient was started on doxycycline and recovered gradually within the next few days.

Neurologic manifestations of scrub typhus have been increasingly reported in the past few years. In the present case, the diagnostic challenge was to establish the cause of fever, jaundice, and altered sensorium in an alcoholic patient with suspicion of acute liver failure.

Key words
alcoholic, IgM, meningoencephalitis, scrub typhus

MICOL GIULIA CITTONE

Systemic capillary leak syndrome leading to aseptic meningitis in a patient with undifferentiated connective tissue disease

Micol Giulia Cittone*, Giuseppe Francesco Casciaro

S.C.D.U. Medicina Interna 1, Azienda Ospedaliero Universitaria Maggiore della Carità di Novara, Università del Piemonte Orientale, Novara, Italy

*Corresponding author: micolcittone@gmail.com

Idiopathic systemic capillary leak syndrome (ISCLS) is characterized by recurrent acute episodes of increased vascular permeability associated with edema, tissue hypoperfusion, hemoconcentration, and low blood albumin levels.

We report the case of a 24-year-old Pakistani woman hospitalized due to the acute onset of undifferentiated connective tissue disease (UCTD), associated with recurrent episodes of hypotension, dyspnea, tachycardia, and facial edema. Blood tests revealed hematocrit of 46%, albumin levels of 26 g/l, and total serum protein levels of 88 g/l with polyclonal gammopathy. Due to the onset of headache and nuchal rigidity, a lumbar puncture was performed, and the diagnosis of aseptic meningitis was established. Given the suspicion of ISCLS, an intravenous infusion of immune globulins was started, resulting in an immediate positive response.

We speculate that the autoimmune process (or processes) responsible for UCTD may have triggered ISCLS. Aseptic meningitis might be part of the spectrum of ISCLS.

Key words
aseptic meningitis, edema, leak capillary syndrome, undifferentiated connective tissue disease

RECIE DAVERN

A case of "simple Conn's"

Recie Davern*, Mensud Hatunic

Department of Endocrinology, Mater Misericordiae University Hospital, University College Dublin, Dublin, Ireland

*Corresponding author: reciedavern@rcsi.ie

Conn syndrome is a common secondary cause of hypertension. We present a unique case of Conn syndrome that we think also illustrates the work needed to improve the awareness of the condition among general medical physicians.

We report the case of a 52-year-old man with a 9-year history of hypertension, hypokalemia, right macular edema, and a splenectomy for spherocytosis at the age of 11. His treatment included telmisartan (80 mg/d), lercanidipine (20 mg/d), and potassium chloride extended--release tablets (2 tablets 3 times/d). Potassium levels were low since November 2015, with the lowest level of 2.5 mmol/l, and remained low despite increasing doses of potassium replacement.

Baseline laboratory investigations revealed renin levels of 8.6 mIU/l (reference range, 9.0–103.5 mIU/l); aldosterone, 567 pmol/l (138–670 mIU/l); sodium, 144 mmol/l (133–146 mmol/l); potassium, 3.3 mmol/l (3.3–5.0 mmol/l); urea, 5.2 mmol/l (2.8–8.6 mmol/l); creatinine, 95 μmol/l (65–107 μmol/l); ARR, 50.3 pmol/l:mIU/l; TFT, normal; and cortisol, 275 nmol/l (150–455 nmol/l). Plasma methanephrines/normetanephrine levels were normal.

Antihypertensive drugs were changed to verapamil to facilitate investigation with a saline suppression test. At time zero, the test showed cortisol levels of 199 nmol/l; aldosterone, 553 pmol/l; and renin, 11.0 mIU/l. After 4 hours, the cortisol level was 68 nmol/l; nonsuppressed aldosterone, 249 pmol/l; and renin, 9.3 mIU/l.

Adrenal computed tomography showed a 2.1-cm nodule in the left adrenal gland with a Hounsfield unit of 5, consistent with an adenoma, and a 2.7-cm enhancing mass arising from the left kidney as well as 2.5-cm enhancing mass arising from the right kidney, consistent with renal cell carcinoma. The patient underwent a laparoscopic left partial nephrectomy and left adrenalectomy with normalization of potassium levels and a significant improvement in BP. Four months later, a laparoscopic right partial nephrectomy was performed. Histology confirmed bilateral renal cell carcinoma and left adrenal adenoma.

This is the first reported case of bilateral renal cell carcinoma and adrenal adenoma, making the case clinically unique. This patient was also seen for a number of years with hypertension and hypokalemia by a number of physicians without any testing for Conn syndrome. This shows that the knowledge of the condition among general physicians needs to be improved.

Key words
adrenal, carcinoma, Conn syndrome, hypertension, hypokalemia

AGATA DUTKOWSKA

Toxic interstitial pneumonia after "fake hash" smoking

Agata Dutkowska*, Adam Antczak, Maciej Ciebiada

Department of General and Oncological Pulmonology, Medical University of Lodz, Łódź, Poland

*Corresponding author: agata.dutkowska@umed.lodz.pl

Designer drugs (DDs) are dangerous substances that cause addictions among young people. Due to the rapid invention of new analogues of psychoactive substances, the side effects after DD use are becoming more and more unpredictable. The present case report shows the irreversible health consequences after chronic inhalation of synthetic cannabinoids, manifesting as toxic interstitial pneumonia in a young man addicted to DDs.

Key words
ARDS, designer drugs, interstitial pneumonia, psychoactive substances, synthetic cannabinoids

KHALID ELKHOLY

Pseudo-koebnerization of the bone: a rare and intriguing case of extrapulmonary sarcoidosis

K. Elkholy, M. Ul Rasool, P. Kerr, J. Feeney, R. Mullan, S. Donnelly

Tallaght University Hospital, Dublin, Ireland

*Corresponding author: dr.khalid.elkholy@gmail.com

INTRODUCTION While sarcoidosis is recognized as an orphan disease, Ireland has one of the highest prevalence globally. Sarcoidosis of the bone occurs in only 5% of patients with sarcoidosis. Although bone involvement may be the earliest manifestation of sarcoidosis, it is usually accompanied by infiltrative skin lesions. Scar sarcoidosis may be more common than recognized because some lesions may be misdiagnosed as hypertrophic scars or keloids.

The well-recognized phenomenon of koebnerization (the appearance of skin disease at sites of skin injury) may be a factor in some cases of scar sarcoidosis. We shed light on a uniquely

novel and intriguingly rare entity of extrapulmonary sarcoidosis, namely, pseudo-koebnerization of the bone, a condition where sarcoid lesions initiate or recur at inflammatory sites of injury or trauma.

CASE We present a case of a 33-year-old Irish soldier with a known diagnosis of biopsy-proven pulmonary sarcoidosis made 2 years earlier. During his routine respiratory clinic review, he reported complaints of an unresolving right foot pain following minor trauma 2 years earlier.

On assessment, he had right first metatarsophalangeal joint tenderness, swelling, and erythema. In addition, he reported recurrent chronic sinonasal symptoms on a background of prior functional endoscopic sinus surgery. Given the length and refractory nature of his symptoms, he was admitted to undergo further investigations.

Biochemical tests revealed normal levels of inflammatory markers, including serum angiotensin-converting enzyme, calcium, and C-reactive protein. As part of the multidisciplinary approach, a rheumatologic and dermatologic expert opinion was obtained, and the diagnosis of cutaneous and osteo-sarcoidosis was hypothesized. A plain film radiography followed by magnetic resonance imaging (MRI) of the right foot subsequently demonstrated features compatible with skeletal sarcoidosis. In addition, delayed whole--body bone imaging and single-photon emission computed tomography–computed tomography revealed significant bone abnormalities in keeping with multifocal osseous sarcoidosis. Lastly and in the context of the sinonasal complaints, computed tomography followed by MRI of the sinuses revealed appearances that represent sarcoidosis of the paranasal sinuses and the floor of the anterior cranial fossa.

An intravenous pulse steroid therapy was administered over 5 days, followed by 30 mg of oral prednisolone along with gastro- and osteoprotective cover. Follow-up reviews were arranged with respiratory, ear, nose and throat, and maxillofacial surgical services.

DISCUSSION Our case highlights how diffuse bone lesions represent an "osseous pseudo-koebnerization" phenomenon, in which new lesions of the pre-existing condition manifest at sites where the skin (or the skin and bone as in our case) is traumatized. This typically follows mechanical insults such as stretching, friction, or compression, which may very likely reflect the nature of our patient's triggering event, as he was a military person.

The development of sarcoidosis in a scar or bone trauma site may be the initial sign of sarcoidosis in some patients. Clinicians should be aware of the possibility of sarcoidosis in patients with a changing or persistently inflammatory scar or bone injury. It is unknown whether environmental factors that come in contact with the skin and/or bones serve as a nidus for granuloma formation or stimulate an aberrant immune response through other mechanisms.

Key words
koebnerization, osseous sarcoidosis, sarcoidosis, scar sarcoidosis

MARGARITA GROMOVA

AA amyloidosis induced by long-term recurrent gout

Margarita Gromova*

Department of Faculty Therapy, Faculty of Medicine, Pirogov Russian National Research Medical University, Moscow, Russia

*Corresponding author: margarita-gromov@mail.ru

INTRODUCTION Although amyloidosis may complicate many chronic infections or inflammatory diseases, including rheumatoid arthritis, ankylosing spondylitis, and psoriatic arthritis, its association with gout is extremely rare. We report a case of a patient with amyloidosis secondary to gout, in whom amyloid A (AA) protein was found in the kidneys, intestines, and adrenal glands.

CASE PRESENTATION In 2018, a 62-year-old Caucasian man was hospitalized because of severe impairment of renal function. For over 26 years, he had been suffering from recurrent gout attacks. He was on allopurinol treatment (300 mg/d) for the past 10 years and did not follow the diet. During a gout attack, he used nonsteroidal anti-inflammatory drugs. Previous diseases included persistent atrial fibrillation, arterial hypertension, and urolithiasis. Due to increased proteinuria, elevated creatinine and C-reactive protein (CRP) levels, and a tendency to hypotension, the patient was referred for further examination in the therapeutic department. Laboratory tests revealed the following findings: hemoglobin, 108 g/l; erythrocyte sedimentation rate, 66 mm/h; creatinine, 269 μmol/l; total protein, 53.7 g/l; albumin, 19 g/l; uric acid, 402 μmol/l; CRP, 99.1 mg/l; proteinuria in the morning urine sample, 3.0 g/l; pyuria, 750 cells/μl; and hematuria, 37 cells/μl. An enzyme immunoassay of serum and urinary proteins revealed no pathological gradients of monoclonal secretion. Blood pressure was controlled at 90/50 mm Hg. Considering the nephrotic syndrome, chronic gout, and hypotension, the patient was suspected to have amyloidosis. Histologic examination of a subcutaneous fatty tissue biopsy and esophagogastroduodenoscopy with an intestinal mucosal biopsy under polarized light showed extensive deposition of Congo red material, indicative of severe amyloidosis. On immunoperoxidase staining, the deposition proved to be type AA amyloidosis.

The patient was eventually diagnosed with tophaceous gout, chronic gouty arthritis, relapsing synovitis of the knee joints, chronic kidney disease stage IV, kidney cysts, urolithiasis, secondary arterial hypertension, amyloidosis of the gastrointestinal tract and adrenal glands, mild severity anemia, persistent atrial fibrillation, and chronic heart failure.

Since 2018, he has been administered subcutaneous canakinumab (150 mg once a month), prednisone (5 mg/d), colchicine (1 mg/d), allopurinol (100 mg/d), and sotalol (40 mg twice a day). His condition is currently stable.

DISCUSSION The present case is an example of a rare development of AA amyloidosis as a consequence of chronic gout with the formation of tophi due to insufficient treatment. In our opinion, the main pathogenic mechanism was associated with chronic inflammation. Long-term recurrent gout and the lack of proper treatment in our patient led to an increase in the synthesis of acute phase markers, which are part of amyloid.

CONCLUSION Clinicians should be aware that patients may present with atypical comorbidities. Our case raises the awareness of such a rare association of diseases and emphasizes the importance of considering the diagnosis of secondary amyloidosis in patients with gout.

Key words
AA amyloidosis, Congo red, gout, hyperuricemia

ANGELA KANNUKENE

Bilateral lung abscesses in association with the opportunistic gram-negative pathogen *Morganella morganii*

Angela Kannukene[1,2]*

1 University of Tartu, Tartu, Estonia
2 South Estonian Hospital, Võru, Estonia

*Corresponding author: angela.kannukene@gmail.com

INTRODUCTION Pulmonary abscess is usually a complication of pneumonia in patients with risk factors for aspiration, often caused by anaerobic flora from gingival cavities. Early pulmonary abscess is clinically identical to pneumonia, but the course of the infection as well as the etiological microbes are different.

Although *Morganella morganii*, a facultative anaerobic gram--negative bacterium, is widely available in nature, infections with this pathogen are uncommon and often associated with the patient's immunocompromised status. We present a case of a male homeless patient with alcoholism, who developed large bilateral pulmonary abscesses while being hospitalized for pneumonia.

CASE DESCRIPTION A 60-year-old man presented to the emergency department with a 3-day history of sudden-onset sharp pain in the right chest wall and dry painful cough. During the previous 2 years, he had been hospitalized for pneumonia twice. He had a known history of chronic alcoholism and homelessness. He had been smoking 2 packs of cigarettes a day for 45 years. During hospitalization, he continued smoking in a designated area.

In the emergency department, his analyses showed high levels of inflammatory markers and alcohol in blood. Radiography showed bilateral pneumonia. Empiric treatment was started with intravenous amoxicillin/clavulanic acid and oral clarithromycin.

During hospitalization, the patient initially showed improvement, but after 8 days of treatment both inflammatory markers and fever increased again. On day 9, chest computed tomography revealed several bilateral pulmonary abscesses, the largest measuring 8×6 cm on the right side and 3×3 cm on the left side.

On admission, DNA panel for common respiratory pathogens (*Bordetella pertussis*, *Chlamydophila pneumoniae*, *Legionella pneumophila*, *Mycoplasma pneumoniae*, *Streptococcus pneumoniae*, and *Bordetella parapertussis*) was taken from the nasal passages. It tested positive for the DNA of *Haemophilus influenzae*. An HIV blood test was negative. Sputum and blood cultures taken on admission were negative. Sputum analyses performed on days 9 and 10 of hospitalization showed the growth of *Morganella morganii*, susceptible to piperacillin/tazobactam. After the initiation of piperacillin/tazobactam treatment, C-reactive protein levels and body temperature normalized. After 10 days, the chest radiography showed improvement on the left side but not on the right side.

Bronchoscopy (performed on the 21st day of hospitalization) showed copious purulent secretion in bilateral bronchial tree with secretions obstructing the bronchi of the right central lobe and left lower lobe anteromedial segment.

The patient was discharged to a nursing care unit to continue oral antimicrobial therapy for 3 weeks with trimethoprim/sulfamethoxazole (as per antimicrobial susceptibility results), with another radiograph and blood analyses scheduled afterwards.

CONCLUSION *Morganella morganii* remains a possible opportunistic pathogen. It is important to remember that people with lowered immunity are at higher risk of developing pneumonia complications, and the possibility of rarer pathogens should be considered.

Key words
bilateral lung abscess, *Morganella morganii*

DEARBHLA M. KELLY

Chronic kidney disease due to high-output ileostomy

Dearbhla M. Kelly*, Gasim Ahmed, Liam Casserly

Department of Nephrology, University Hospital Limerick, Dooradoyle, Limerick, Ireland

*Corresponding author: dearbhla.kelly@ndcn.ox.ac.uk

Volume depletion, electrolyte abnormalities, acid-base disorders, and acute kidney injury (AKI) are well-described complications of high-output ileostomies. Chronic kidney disease (CKD) is an underrecognized consequence of cumulative AKI episodes in this context. We report a case of a 65-year-old woman with CKD as a result of pervasive and recurrent extracellular fluid volume depletion due to an end ileostomy. We review the literature and describe the possible pathogenesis, natural history, and management of this important phenomenon.

Key words
acute kidney injury, chronic kidney disease, ileostomy, stoma

COLM KERR

The patient clinicians catch the worm: a case of periduodenal cysticercosis in Ireland

C. Kerr[1], E. O'Donovan[2], M.A. Ahmad[3], E. Aher[3]

1 Department of Medicine, Trinity College Dublin, Dublin, Ireland
2 Department of Pathology, Mater Misericordiae University Hospital, Dublin, Ireland
3 Department of Geriatrics and General Medicine, St. Luke's General Hospital, Kilkenny, Ireland

*Corresponding author: colmkerr@gmail.com

Cysticercosis is caused by the accidental ingestion of eggs of the pork tapeworm (*Taenia solium*). Cysticercosis is endemic in parts of Latin America, Sub-Saharan Africa, the Indian subcontinent, and Southeast Asia. *Taenia solium* is believed to be eradicated from Western Europe. We report a case of a patient who presented to an Irish hospital with abdominal pain and anorexia following travel to the Indian subcontinent. Radiology findings revealed a periduodenal hypoechoic cystic mass initially concerning for tuberculosis and lymphoma. A fine-needle aspirate demonstrated numerous rounded structures of approx. 35 μm in diameter with a radially stratified shell consistent with ova of *Taenia* spp. This supported a diagnosis of cysticercosis. Treatment with an antihelminthic led to the complete resolution of the patient's symptoms and improvement of imaging findings. This is the first reported case in the literature of a diagnosis of extraneural cysticercosis in Ireland.

Key words
abdominal mass, cysticercosis, periduodenal, *Taenia solium*

JI YOON KIM

A 48-year-old man with rare complications of atypical Lemierre syndrome

Ji Yoon Kim*, Min Jae Kim, Sung Han Kim

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

*Corresponding author: jiykims1234@naver.com

INTRODUCTION Lemierre syndrome is a rare disease characterized by septic thrombophlebitis of the internal jugular vein with septicemia following a recent oropharyngeal infection. If the disease remains undiagnosed and untreated, it can result in severe complications and even death. We report a patient who developed rare complications of Lemierre syndrome.

CASE DESCRIPTION A 48-year-old man was admitted with a 2-week history of fever and right frontotemporal headache. He had poorly controlled diabetes. He usually had been suffering from periodontal infection. He had no meningeal irritation sign, and cerebrospinal fluid (CSF) examination revealed no abnormality. His brain magnetic resonance imaging (MRI) and transesophageal echocardiography showed no definitive abnormality. Blood cultures grew *Streptococcus constellatus* in 2/3 bottles. Ceftriaxone was continued, but his headache worsened. Suddenly, diplopia and right-sided ptosis developed at hospital day 5. His mental status deteriorated to confusion. The repeated CSF examination revealed pleocytosis (15530/ml; neutrophil, 91%). Orbit MRI showed thrombophlebitis of the bilateral cavernous sinuses and right internal jugular vein as well as diffuse meningeal enhancement. The septic thrombophlebitis of the cavernous sinuses and meningitis were diagnosed, and atypical presentation of Lemierre syndrome was considered. Meropenem, teicoplanin, and low-molecular-weight heparin were administered. Two days later, the patient suddenly showed ataxia and nystagmus. Brain MRI demonstrated multifocal acute infarctions, including the right lateral medulla. Repeated echocardiography showed no vegetation. After 4 weeks of antibiotic treatment, brain imaging

showed recanalization of the right internal jugular vein and cavernous sinus. The patient was discharged on warfarin.

DISCUSSION Because Lemierre syndrome can present with nonspecific symptoms, a high index of suspicion is very important. When the cavernous sinus is additionally involved, serious central nervous system complication can occur. Repeated CSF examination and brain imaging are crucial when the headache aggravates despite initial treatment. The delayed diagnosis can result in severe bacterial meningitis and brain infarction.

Key words
cavernous sinus thrombosis, cerebral infarction, Lemierre syndrome, meningitis

MARÍA AGUSTINA MARCO

Recognized for best poster

Renal involvement in a patient with non–Hodgkin lymphoma: a well-known pathology and an atypical presentation

Marco María Agustina*, Cámera Luis Alberto, Pollán Javier Alberto

Hospital Italiano de Buenos Aires, Buenos Aires, Argentina

*Corresponding author: maria.marco@hospitalitaliano.org.ar

Acute kidney failure is a common condition in patients with lymphoma, and it can result from a broad spectrum of causes. Although the prevalence of interstitial kidney involvement by lymphomas is high, the presence of acute kidney failure is rare.

A 75-year-old man with a history of chronic lymphocytic lymphoma and mild chronic renal failure presented to our hospital with asthenia, hyporexia, frequent vomiting, and weight loss. Laboratory tests showed leukocytosis, thrombocytosis, and acute renal failure with a creatinine level of 2.3 mg/dl. Uric acid and lactate dehydrogenase levels were significantly elevated. Physical examination showed a palpable mass in the right hemiabdomen. Ultrasound and computed tomography (CT) showed severe deformation of the right kidney and a mild left pelviectasis.

During the first days of hospitalization, the patient received parenteral fluids and allopurinol with a slight improvement in renal function. However, 48 hours later, a further and more significant increase of creatinine levels was observed. This progressive nonoliguric acute renal failure presented urinary parameters that were not compatible with a prerenal etiology, and the urine sediment was not pathologic. Uric acid levels had rapidly normalized after admission, and laboratory tests did not show any other parameter suggesting tumor lysis syndrome. A Doppler ultrasound was performed, and it revealed no changes in the urinary tract or in the renal vessels.

Due to increasing serum creatinine levels, non–Hodgkin lymphoma of diffuse large B cells was diagnosed by percutaneous biopsy of the right kidney. As creatinine levels reached a maximum value of 4.5 mg/dl, prephase chemotherapy with vincristine and cyclophosphamide was initiated based on the suspicion that kidney interstitial involvement was causing the progressive renal failure.

On the day of starting the prephase chemotherapy, a positron emission tomography (PET) scan for staging was performed. It revealed a high fluorodeoxyglucose uptake in the right mass involving the kidney and in the left renal sinus. An ultrasound showed a left pelvicalyceal dilation of 25 mm and a double-J catheter was placed in the left ureter. After the intervention, creatinine levels decreased abruptly in the next days. The patient was started on complete R-CHOP chemotherapy and was discharged with normalized renal function.

Renal involvement secondary to lymphomas is frequent. However, it rarely becomes clinically significant. Secondary kidney lymphomas may present in different patterns, and interstitial infiltration may have a silent evolution or present with nonspecific symptoms, making its diagnosis more difficult. Asymmetric kidney infiltration and sinus invasion as shown in this clinical case is an atypical presentation, and consequent obstructive renal failure is even less common.

Although kidney biopsy is the gold standard, imaging studies such as CT or PET can help diagnose renal involvement by lymphoma. As a more sensitive and detailed imaging test, PET-CT may not only be useful for assessing the extension and pattern of the disease, but it may also elucidate the pathophysiologic mechanisms underlying kidney failure. In our case, it turned out to be the key to this challenging diagnosis.

Key words
acute renal failure, lymphoma, PET-CT, urinary tract obstruction

HANNA M. MUISCHNEK

A 41-year-old man with basal ganglia calcifications and low serum calcium levels

Hanna M. Muischnek*

Department of Endocrinology, Faculty of Medicine, Tartu University Hospital, University of Tartu, Tartu, Estonia

*Corresponding author: hannamaria.muischnek@kliinikum.ee

Pseudohypoparathyroidism (PHP) is a rare group of disorders characterized by hypocalcemia, hyperphosphatemia, and high parathyroid hormone (PTH) levels, reflecting end-organ resistance to PTH. A 41-year-old man presented to the emergency department following a tonic seizure. His serum calcium levels were 1.3 mmol/l (reference range, 2.20–2.69 mmol/l). He was referred to the endocrinology department for further evaluation. Ionized calcium levels were 0.56 mmol/l (1.16–1.32 mmol/l); phosphate, 2.07 mmol/l (0.8–1.45 mmol/l); and PTH, 32.9 pmol/l (1.6–6.9 pmol/l). Head computed tomography demonstrated multiple subcortical and diffuse basal ganglia calcifications. Based on these findings, the patient was diagnosed with pseudohypoparathyroidism, and treatment with calcitriol and calcium carbonate was started. The patient was referred to a medical geneticist for further evaluation. The causes of hypocalcemia should be considered in the diagnostic workup of patients with seizures. The diagnosis of PHP type 1B is often difficult but the overall prognosis is good.

Key words
Fahr disease, hypocalcemia, parathyroid, pseudohypoparathyroidism

TUULI PUNGAS

Listeria monocytogenes sepsis as an adverse event of glucocorticoid therapy for amiodarone pneumonitis

Tuuli Pungas[1,2]*

1 Centre of Internal Medicine, East-Tallinn Central Hospital, Tallinn, Estonia
2 Faculty of Medicine, University of Tartu, Tartu, Estonia

*Corresponding author: tuuli.pungas@gmail.com

INTRODUCTION Nonspecific interstitial pneumonia (NSIP) is a diffuse parenchymal lung disease with a suggested initial treatment with systemic glucocorticoids. Glucocorticoid therapy is associated with a variety of adverse events, including sepsis. Balancing between benefits and potential side effects is a serious challenge to physicians.

CASE DESCRIPTION An 81-year-old Caucasian man with a history of dyspnea for 1 week presented to the emergency department. Thoracic computed tomography (CT) showed diffuse interstitial and alveolar consolidation with ground-glass opacities. Radiologically, it was interpreted as interstitial lung disease, mainly indicating NSIP. As the patient had a 2-year history of amiodarone

use, an initial diagnosis of amiodarone pneumonitis (a subgroup of NSIP) was made. After discontinuing treatment with amiodarone and initiating treatment with systemic glucocorticoids, the patient's symptoms and abnormal radiologic findings started to resolve. After 1.5 months of prednisone treatment (originally 50 mg/d, then reduced), the patient developed high fever and low blood pressure with a syncope. Sepsis with acute kidney failure and respiratory failure was diagnosed. The patient developed multiple infections including *Listeria monocytogenes* septicemia, multi-drug–resistant *Enterococcus faecium* urinary tract infection, and fungal pneumoniae of *Clavispora lusitaniae*. Prednisone therapy was slowly tapered. With prolonged intravenous antimicrobial treatment including different β-lactam antibiotics, linezolid, and fluconazole, the patient started to show signs of improvement. As respiratory failure persisted, repeat thoracic CT was scheduled. It showed progression of interstitial disease compared with initial hospitalization. Since prednisone had been tapered earlier and amiodarone pneumonitis may recur if treatment is discontinued too early, the most likely diagnosis remained amiodarone pneumonitis. Idiopathic NSIP still remained a potential differential diagnosis but because treatment for both diseases is the same, glucocorticoid therapy was restarted. Considering the patient's age, comorbidities, and functional status, a lung biopsy to confirm the diagnosis was considered unsafe and therefore was not performed. Even though glucocorticoid treatment had some effect in terms of dyspnea, the patient had suffered a vast loss of respiratory function and therefore required home oxygen therapy. He was discharged from the hospital on low-dose glucocorticoid therapy to receive rehabilitation.

CONCLUSION This case illustrates that the development of severe *Listeria monocytogenes* sepsis due to the use of systemic glucocorticoids for amiodarone pneumonitis may exacerbate interstitial disease while tapering glucocorticoids. Although glucocorticoids are the first-line treatment for NSIP, potential benefits and risks should always be weighed before and during therapy to ensure the best outcome for the patient.

Key words
amiodarone pneumonitis, glucocorticoids, *Listeria monocytogenes*, nonspecific interstitial pneumonia

MANN RANDARU

Complicated diverticulitis: a difficult road to diagnosis

Mann Randaru*

Tartu University Hospital, University of Tartu, Tartu, Estonia

*Corresponding author: mann.randaru@gmail.com

INTRODUCTION Diverticulosis is a common problem in Western countries. The prevalence is age dependent, with the elderly being more commonly affected. Due to population aging, the condition is becoming more frequent.

CASE DESCRIPTION A 64-year-old man was admitted to the hospital due to a suspicion of an unknown infection. He complained of high fever that had lasted for 1.5 months, enlarged abdomen, and postprandial abdominal pain. He also reported a history of alcohol abuse. During the examination, there were no signs of peritonitis. Blood analysis showed a probable bacterial infection, with neutrophilic leukocytosis and high C-reactive protein levels. In addition to the probable infection, he also had anemia of chronic disease. A computed tomography (CT) scan showed ascites, multiple enlarged lymph nodes in the mesenteric fatty tissue, and infiltration of the greater omentum, indicating inflammation. To analyze the abdominal fluid, an ultrasound-guided paracentesis was planned. It revealed lack of considerable ascites, but there was an encapsulated mass (about 16 cm × 8 cm × 10 cm in size) in the lower abdomen. Drainage was performed, yielding about 600 ml of white-yellow exudate. Since the main source of infection was still unclear, a new CT was planned, this time with the administration of intravenous and oral contrast media. It revealed diverticula in the sigmoid colon, with possible diverticulitis and diverticular abscess. The patient was treated with antibiotics (cefuroxime and metronidazole) and percutaneous abscess drainage, with good outcome. He was discharged after 3 weeks in good condition.

DISCUSSION The main problem in this case was to reach the diagnosis. The first CT scan finding was misinterpreted as ascites, and since the patient had a history of alcohol abuse, it was mainly associated with liver damage. After the sonography, it was clear that the patient had no significant ascites but an abscess in front of the bowels, which was confirmed by paracentesis. Since the patient had received antibiotics for a week before the paracentesis, the cultures were negative. A definitive diagnosis was reached with a new CT scan with both intravenous and oral contrast media.

LESSONS TO BE LEARNED FROM THE CASE When the condition of the patient and the findings on the CT scan do not seem to match, it might be a good idea to study the scans one more time. In the case of a large abscess, antibiotic therapy is usually not enough, and the abscess requires drainage.

Keywords
abdominal pain, diverticulitis, fever

ELEONORA RIZZI

An unusual case of abdominal pain

Eleonora Rizzi*, Giulia C. Micol, Francesca M. Giulia

S.C.D.U. Medicina Interna 1, Azienda Ospedaliero Universitaria Maggiore della Carità di Novara, Università del Piemonte Orientale, Novara, Italy

*Corresponding author: eleonora.rizzi18@gmail.com

A 30-year-old Caucasian man presented to the emergency department of an academic hospital with abdominal pain accompanied by vomiting and diarrhea lasting 1 week. In the preceding 3 weeks, the patient experienced fever, profuse sweating, pruritus, and unintentional weight loss of approximately 10 kg. His past medical history was notable for hypertriglyceridemia, isolated thrombocytopenia of undetermined origin, and remote Epstein–Barr virus infection. On physical examination, blood pressure was 125/85 mm Hg; heart rate, 96 bpm; and oxygen saturation, 99% on room air. Abdominal palpation revealed enlargement of the liver and spleen. Complete blood count showed slight anemia (hemoglobin, 11.2 g/dl), mild thrombocytopenia (110 000/μl), and elevated lactate dehydrogenase levels. Coagulation tests were normal and HIV serology was negative. An abdominal computed tomography (CT) scan with a contrast agent was performed. It confirmed hepatosplenomegaly and showed slight perihepatic ascites without deep lymphadenopathies. Most importantly, CT sequences demonstrated the absence of contrast agent diffusion in the hepatic veins. A color doppler examination confirmed the diagnosis of hepatic vein thrombosis (Budd–Chiari syndrome); thus, anticoagulation with enoxaparin was started. Thrombophilia screening including lupus anticoagulant, factor II, factor VIII, proteins S and C, and factor V Leiden was negative. The results of antinuclear antibody testing, an extractable antinuclear antibody panel, as well as the search for *JAK2* mutations were also negative. Further examinations showed Coombs-negative hemolytic anemia, while flow cytometry revealed a deficient expression of CD55 and CD59, consistent with paroxysmal nocturnal hemoglobinuria. Treatment with eculizumab, a humanized monoclonal antibody targeting the complement component C5 and preventing its cleavage by convertases, was started. At the last follow-up visit (8 weeks later), he reported that anemia and fatigue had improved, with no recurrence of abdominal pain or B symptoms.

Paroxysmal nocturnal hemoglobinuria (PNH) is a rare acquired, life-threatening hematologic disease that has fascinated clinicians for more than a century because of its diverse manifestations and intricate pathophysiology. The pathogenesis involves hematopoietic

stem cells, and, in most cases, somatic mutations in the *PIGA* gene are present. This gene encodes a protein essential for the synthesis of the glycosylphosphatidylinositol (GPI) anchors that allow the decay-accelerating factor to inhibit the complement cascade. In patients with PNH, a disrupted GPI anchor leads to dysregulated complement activation. In addition to other well-known hematologic manifestations (eg, Coombs-negative hemolytic anemia), PNH encompasses a broad spectrum of signs and symptoms, such as abdominal pain, thrombosis, fatigue, red urine, and erectile dysfunction, which makes the diagnosis challenging. The present case highlights the importance of considering PNH in the differential diagnosis when an unprovoked thrombosis at an unusual site develops in a young man without a family history of thrombophilia.

Key words
Budd–Chiari syndrome, eculizumab, hepatosplenomegaly, paroxysmal nocturnal hemoglobinuria, thrombosis

MARTA SKOCZYŃSKA

Recognized for best poster

Nearly fatal thrombotic microangiopathy complicating systemic lupus erythematosus: challenges in diagnosis and treatment

Marta Skoczyńska[1,2]*, Małgorzata Chowaniec[2,3], Monika Ponikowska[2]

1 Department and Clinic of Rheumatology and Internal Medicine, Wroclaw Medical University, Wrocław, Poland
2 Department of Rheumatology and Internal Medicine, Wroclaw University Hospital, Wrocław, Poland
3 Department of Hygiene, Wroclaw Medical University, Wrocław, Poland

*Corresponding author: marta.skoczynska@gmail.com

INTRODUCTION We report a case of thrombotic microangiopathy complicating systemic lupus erythematosus (SLE) that occurred in a young female patient. It highlights the differential diagnosis of a life-threatening thrombotic microangiopathy and illustrates a successful use of biologic treatment in a patient with poor prognosis.

A 35-year-old woman with SLE was admitted to the hospital with acute kidney injury in the course of azathioprine-induced pancreatitis. In the therapy of SLE, the patient received steroids, antimalarial drugs, and azathioprine, which was stopped shortly before the patient's admission. In the past, the patient was triple positive for antiphospholipid antibodies (APLAs) but did not meet the clinical criteria for antiphospholipid syndrome (APS).

During hospitalization, the patient became septic and developed a severe intestinal hemorrhage requiring a life-saving laparotomy, complicated by multiorgan failure. Laboratory tests revealed prolonged activated partial thromboplastin time, hypofibrinogenemia, thrombocytopenia, and decreased levels of complement components C4 and C4. Despite a prolonged treatment with heparin, blood preparations, steroids, and plasmaphereses, her condition worsened and she developed an ischemic stroke. As a last resort, eculizumab administration was started.

After therapy with eculizumab, the patient's condition began to improve and she was discharged in a state of partial recovery, with dialysis-dependent renal failure.

DISCUSSION The exact diagnosis in this complicated case of SLE was challenging because it might have been a result of overlapping conditions. The initial clinical picture (thrombotic microangiopathy associated with thrombocytopenia and bleeding, potentially induced by infection) was strongly suggestive of disseminated intravascular coagulation, but together with the pronounced renal failure, it might have also indicated atypical hemolytic uremic syndrome. The major thrombotic event, namely, ischemic stroke, that occurred later on, in the light of the past triple positivity for APLA, might have indicated APS. Although rare, catastrophic APS. should be suspected in all patients with SLE and multiorgan failure affecting more than 3 organs, particularly in the presence of APLAs. Primary thrombosis prevention in patients with triple APLA positivity may include aspirin and hydroxychloroquine administration, or heparin in higher-risk cases. Laboratory findings of our patient suggested the involvement of complement pathway activation. Eculizumab is a novel antibody targeted against complement C5, used both in the treatment of hemolytic uremic syndrome and, experimentally but with promising results, in the treatment of catastrophic APS.

LESSONS TO BE LEARNED FROM THE CASE In the case of combined antibiotic, steroid, heparin, and plasmapheresis therapy failure, in patients with thrombotic microangiopathy of possible multiple and overlapping causes, biological treatment as a life-saving therapeutic option should be considered.

Key words
antiphospholipid syndrome, eculizumab, systemic lupus erythematosus, thrombotic microangiopathy