An 81 year-old man presented to the emergency room with a history of fever, abdominal pain and bloody diarrhea. One week before his admission, he developed a skin rash on the lower extremities and buttocks and arthralgia in his knees and ankles. His medical history was unremarkable except for hypertension and a self-limited upper respiratory tract infection approximately one week before the onset of his complaints. Physical examination revealed diffuse abdominal rebound tenderness and palpable purpuric lesions predominantly on the lower extremities (Figure 1a). Laboratory investigations revealed a C-reactive protein of 77 mg/L, white blood cell count of 7,100/mm³, and platelet count of 66,000/mm³. His serum creatinine level was 1.9 mg/dL and the 24-hour urinary protein excretion was 4.2 g/day. Serological tests for anti-nuclear antibodies and anti-neutrophil cytoplasmic antibodies were negative and complement levels were within the normal range. Upper gastrointestinal tract endoscopy and colonoscopy revealed hyperaemic and ecchymotic lesions with irregular erosions and ulcerations at the level of the gastric mucosa, duodenum (Figure 1b), and sigmoid colon (Figure 1c). The skin biopsy showed a leukocytoclastic vasculitis with the deposition of IgA compatible with the diagnosis of Henoch-Schönlein purpura. During treatment with prednisone, skin and mucosal lesions disappeared but the patient developed sepsis and died, despite intensive antibiotic therapy.
Dramatic change in disease activity visualized by PET in a patient with sarcoidosis

Cihan Heybeli¹, İsmail Sarı², Recep Bekiş³, Servet Akar⁴

A 39-year-old female presented with malaise, weight loss, and generalized joint pain. Symptoms started 6 months prior to presentation and progressed over time. Laboratory tests revealed increased C-reactive protein (43.4 mg/L) and lactate dehydrogenase (330 U/L). Chest imaging showed mediastinal lymphadenopathy and nodular infiltration in the parenchyma of both lungs. Due to the subtle loss of weight and results of other imaging studies, positron emission tomography (PET) with fluorodeoxyglucose (FDG) was performed. Pathological FDG uptakes were observed in the cervical, hilar, and intraabdominal lymph nodes, lung parenchyma, liver, humerus, and ilium (Figure 1A). The patient was diagnosed as sarcoidosis according to the histopathological examination of a mediastinal lymph node. Corticosteroid and azathioprine were prescribed. Four months after treatment, a control PET scan was performed (Figure 1B). There was dramatic improvement in all anatomical sites compared with the first scan, correlating with symptom relief. In conclusion, PET scan may be a useful tool for monitoring disease activity in widespread sarcoidosis.

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A 32-year-old male patient, admitted to our outpatient service, complained of inflammatory back pain and knee pain that became progressively severe for 4 months. The patient had a history of paraplegia emerged after falling from a high place. In the radiographic evaluation, his x-ray results revealed deposition of bone around the iliac bone and femur neck. The sacroiliac joints were normal (Figure 1a). However, the triple phase bone scan demonstrated a heterotopic ossification (HO) (Figure 1b).

Heterotopic ossification is described as bone formation in extra-skeletal tissues and deposition of bone within the soft tissue around peripheral joints (1). It may occur especially after trauma to the brain and spinal cord injuries. Rarely, it is observed in non-traumatic central nervous system disorders and, in some cases, after joint replacement surgeries. In early stages, HO may resemble the presentation of back pain, cellulitis, osteomyelitis, or thrombophlebitis. Twenty percent of HO patients have mobility problems and inflammatory back pain complaints. The etiology and pathogenesis of HO are still unknown. Radiographic imaging, bone scanning, and especially triple phase bone scan are commonly accepted as the diagnostic methods in the diagnosis of HO. In the treatment process, non-steroidal anti-inflammatory drugs (NSAIDs) are thought to be efficient to prevent symptoms at the early stages, following the injuries observed. In progressive cases, surgical resection can be another effective treatment after radiation (2). However, NSAIDs can be used to prevent recurrence. This is why we used indomethacin 100 mg/day, and the symptoms of the patient relieved after 10 days of treatment.

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References
Role of PET/CT in the diagnosis of large vessel vasculitis in a patient with systemic inflammatory response syndrome

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A 57-year-old woman presented with systemic inflammatory response syndrome manifesting as high-grade fever over the past 2 months, unexplained fatigue, malaise, weight loss, night sweats, diffuse non-specific arthralgias and myalgias. The patient had a history of hypertension for 2 years. The physical examination, with a focus on pulse assessment, bruit auscultation, and inter-arm systolic blood pressure difference, was not suggestive of arterial disease. Laboratory results showed leukocytosis (12,100/μL), elevated erythrocyte sedimentation rate (72 mm/h), and C-reactive protein level (131 mg/L). No clear focus of infection was found. A transthoracic echocardiogram was negative for infective endocarditis. Contrast-enhanced computed tomography of the chest, abdomen, and pelvis did not reveal clinically important abnormalities. With a suspicion of an occult malignancy, the patient underwent whole-body fluorine-18 fluorodeoxyglucose positron emission tomography/computed tomography (18F-FDG PET/CT) that showed increased 18F-FDG uptake in the thoracic and abdominal aorta expanding to the subclavian, common carotid, and femoral arteries (Figure 1). In the setting of systemic inflammatory response syndrome, we judged these images to be compatible with large vessel vasculitis (LVV). The patient initiated treatment with methylprednisolone and methotrexate, resulting in significant improvement of symptoms and reduction of inflammatory markers.

LVV is characterized by predominant but not exclusive involvement of the aorta and its major branches, with Takayasu arteritis (TAK) and giant-cell arteritis (GCA) being the two major variants (1). The margin between GCA and TAK is blurred, and the histopathological findings are indistinguishable (2). This case illustrates the central role of 18F-FDG PET/CT findings in early diagnosis of LVV in a patient with unexplained systemic disease. PET/CT imaging reveals increased metabolic activity when morphological changes (like wall thickening, arterial stenosis, or dilatation) and overt vascular symptoms (such as pulselessness, bruits, blood pressure difference) are absent (3). Therefore, when diagnosed in an early stage, most patients would not meet the existing 1990 American College of Rheumatology classification criteria for either GCA or TAK, as these criteria are appropriate for advanced cases. Moreover, these criteria do not consider the possibility of using novel imaging modalities (4). Cases of early diagnosis of both GCA and pre-pulseless TAK with improved imaging techniques are increasingly reported (5). Therefore, we suggest that the time has come to update the classification criteria for LVV, which are currently not working at an early stage of the disease (3).
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Computed tomography angiography (CTA) findings of lupus-associated intestinal vasculitis

Türker Acar¹, Duran Efe¹, Melda Yıldız¹, Kazım Gemici², Serkan Güneyli³

Main text

A 31-year-old female was admitted to our institution with abdominal discomfort and right lower quadrant pain started 1 month ago. A slight defense was observed during physical examination, but no sign of rebound was detected. Her rectal examination and bowel sounds were within the normal limits. In her detailed history, she described systemic lupus erythematosus (SLE) anamnesis managed with a maintenance therapy of low-dose corticosteroids. Complete blood count, laboratory test, and stool sample analysis were within the normal limits, except for mildly decreased albumin levels (3.61 g/dL), increased C-reactive protein levels (18.4 mg/L), and elevated anti-double-stranded DNA antibody levels. Ultrasound examination revealed small bowel wall edema, splenomegaly, and diffuse ascites in the abdomen. Upon detection of small bowel wall edema and given the history SLE, the patient underwent computed tomography angiography (CTA) with a suspected diagnosis of mesenteric ischemia due to the vasculitic course of the disease. The visceral branches of the abdominal aorta and mesenteric and portal venous vessels were found to be intact; however, diffuse small bowel edema and mucosal enhancement giving a “target” sign were observed on CTA (Figure 1a, b). In addition to small bowel edema, considerable ascites was seen in the abdominal cavity (Figure 1a, b). Considering the history, laboratory test, and CTA findings, a diagnosis of lupus-associated intestinal vasculitis (LAIV) was made. The patient was referred to the internal medicine department and high-dose methylprednisolone (2 mg/kg body weight/day) was started, with the dose gradually tapered over a 2-week period. After medical treatment, her symptoms resolved progressively and control ultrasonographic evaluation showed absence of small bowel edema and ascites.

Globally, LAIV is seen up to 0.2%-9.7% of patients diagnosed with SLE (1). The manifestation of LAIV varies from mild to severe symptoms. Nonspecific abdominal pain, abdominal fullness, and diarrhea represent the mild symptoms. On the other hand, gastrointestinal bleeding and intestinal perforation due to necrosis are the severe symptoms that may result in acute surgical abdomen. Accurate and prompt diagnosis of...
LAIV is critical for reducing unnecessary surgical procedures. Because the laboratory and clinical symptoms are sometimes nonspecific, the diagnosis mainly depends on computed tomography (CT), which reveals not only the bowel wall but also the mesenteric vasculature successfully. Common CT findings that can be seen in LAIV include focal or diffuse bowel wall thickening, bowel wall contrast enhancement giving a “target” sign, stenosis or engorgement of mesenteric vessels, called “the comb sign,” and ascites (2). Immediate anti-inflammatory immunosuppressive therapy is the key to the management of LAIV (3).

Informed Consent: Written informed consent was obtained from patient who participated in this study.


Conflict of Interest: No conflict of interest was declared by the authors.

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References
Rheumatoid arthritis in a patient with bilateral congenital aplasia of the thumbs

Nassira Aradoini, Sofia Talbi, Fatima Zahra Abourazzak, Taoufik Harzy

Because congenital hypoplasia of the thumb represents 3.5% of all malformations of the upper limb, it is included in the category of malformations of the upper limb of group 5 (1). There may be a slight hypoplasia (type 1) to a total absence of the thumb (type 5) (2). This defect can be isolated or associated with other anomalies as part of a poly-malformation syndrome. Our patient, is 54 year-old, presented with seropositive rheumatoid arthritis that developed 2 years ago. Symptoms include pain, stiffness, swelling, and limited motion and function of many joints, especially, the small joints in the hands and feet. The laboratory workup revealed elevated erythrocyte sedimentation rate and C-reactive protein levels. The immunological test revealed positive rheumatoid factor and anti-citrullinated protein antibodies. The radiograph of the hands demonstrated juxta-articular osteopenia, loss of joint space, and erosions of the proximal and distal interphalangeal joints compatible with rheumatoid arthritis. The rest of the clinical examination found bilateral congenital aplasia isolated of type 5 from both thumbs. The patient’s karyotype showed no chromosomal instability. Written informed consent was obtained from the patient for the publication of the case (Figure 1, 2).

Informed Consent: Written informed consent was obtained from the patients who participated in this study.

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References
Tuberculous sacroiliitis: A cause of bone marrow edema in magnetic resonance imaging

Servet Akar¹, İsmail Safa Satoğlu², Berna Dirim Mete³, Öğür Tosun³

A 43-year-old female presented with progressive left buttock pain for 6 months. Her pain was worse at night and was not relieved by activity. She had morning stiffness for 5–10 min. She had no constitutional symptoms, history of peripheral arthritis, dactylitis, enthesitis, psoriasis, and inflammatory bowel disease. She denied having a past or family history of tuberculosis (Tb). She was administered with sulfasalazine and non-steroidal anti-inflammatory drugs (NSAIDs), but her condition did not improve. Laboratory examination revealed an ESR of 21 mm/h and a CRP of 1.43 mg/dL (normal range, 0.0–0.8 mg/dL). Pelvic antero-posterior radiograph showed minimal sclerosis with joint space changes of the left sacroiliac joint (SIJ). Parasagittal T1-weighted (Figure 1a) and short tau inversion recovery (STIR) (Figure 1b) magnetic resonance (MR) images revealed joint space enlargement with increased joint fluid in the left SIJ as well as heterogeneous bone marrow edema and hyperintense lesion spreading to periarticular soft tissues. Gadolinium-enhanced axial fat-suppressed T1-weighted MR image of SIJs (Figure 1c) showed a smooth thin-rimmed enhancement area extending anteriorly from the left SIJ to the iliococcygeus muscle and extending laterally to gluteal muscles that was compatible with cold abscess. Extensive destruction of both iliac and sacral bones of the left SIJ was also visualized in the axial computed tomography (CT) images (Figure 1d) obtained during CT guided biopsy procedure. Culture of the biopsy material yielded Mycobacterium tuberculosis, and the patient was administered with a four-drug anti-tuberculous therapy. Musculoskeletal involvement is uncommon and accounts for 1%–3% of all Tb (1) cases, and SIJ Tb was reported in approximately 10% (2) of

Figure 1. a-d. Para-coronal T1-weighted (a) and short tau inversion recovery (STIR) (b) magnetic resonance (MR) images of sacroiliac joint (SIJ) showed increased synovial fluid in the left SIJ and a hyperintense lesion spreading to periarticular soft tissues. Gadolinium-enhanced axial fat-suppressed T1-weighted MR image of SIJs (c) showed a rim enhancement area extending from the left SIJ to periarticular muscles compatible with cold abscess (d)
the musculoskeletal Tb cases. Early diagnosis of SI Tb is extremely difficult mainly because of the non-specific nature of the symptoms. Although the culture is the gold standard for the diagnosis, CT and MR may be complementary for the diagnosis. The severe joint destruction and a cold abscess with a smooth thin-rimmed enhancement may be suggestive of Tb (3).

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**References**

Scleredema of Buschke
Rajaie Namas¹, Ambreen Ashraf²

Main text
A 46-year-old African-American male with a history of type-1 diabetes (DM) was referred to the Rheumatology clinic for a further evaluation of possible systemic scleroderma. He reported a 3-year history of skin thickening and hyperpigmentation on his back and shoulders. Over the last 12 months, his symptoms progressed, involving the proximal arms, legs, and back with sparing of his hand and feet. His medical history was remarkable for insulin-dependent DM, which was complicated with diabetic retinopathy and peripheral neuropathy. Skin examination revealed a woody, hardened, indurated, hyperpigmented skin with a peau d’orange appearance involving the posterior aspect of the neck, shoulders, arms, legs, and mid-back (Figure 1a, b). The face, chest, forearms, hands, and feet were unaffected. There was no telangiectasia, sclerodactyly, fingertip ulceration, or calcinosis. The remainder of the physical examination was unremarkable. Laboratory evaluation revealed a normal complete blood count, comprehensive metabolic panel, Western sedimentation rate, C-reactive protein, creatine phosphokinase level, aldolase, and thyroid-stimulating hormone. Hemoglobin A1C level was 7.1 (normal <6). Immunofluorescence antinuclear antibodies, complement levels, and Scl-70 were all negative. Serum protein electrophoresis and urine protein electrophoresis were normal. Full thickness skin biopsy revealed a thickened dermis, with fenestrated collagen and mucin deposition in the deep dermis, which is consistent with the diagnosis of scleredema (Figure 2 a-c).

Scleredema is a sclerotic skin disease, which is a largely unknown pathogenesis, that generally occurs in association with DM, infection (particularly streptococcal infection of the upper respiratory tract), or monoclonal gammopathy. DM, particularly type II, is considered the most common form of the disease and primarily affects adults, particularly middle-aged obese individuals. In scleroderma associated with diabetes, an irreversible glycosylation of collagen as well as alterations in collagenase activity may lead to an excessive accumulation of collagen and mucin. The condition is characterized by firm, non-pitting edema that typically begins at the neck and spreads to the face, scalp, shoulders, and trunk. The hands and feet are characteristically not affected (1).

Several sclerotic disorders can share clinical features with scleredema, such as systemic sclerosis, scleromyxedema, and eosinophilic fascitis. Physical therapy has to be initiated as soon as possible to minimize functional limitations related to reduced joint mobility. Multiple treatment approaches have been studied,
including immunosuppressive agents such as cyclosporine, methotrexate, and systemic glucocorticoids (e.g., dexamethasone pulse therapy or high-dose intravenous glucocorticoids), and show promising outcomes, but further studies are required (2-4).

Ethics Committee Approval: N/A.

Figure 2. a, c. H&E staining of the skin biopsy represents (a), an unaffected epidermis, while the dermis is thicker than normal (the dermis can be up to four times thicker than normal). Edematous spaces between the collagen bundles are present at a microscopic magnification of 10x. Collagen fibers appear swollen and separated by wide spaces at a microscopic magnification of 40x (b), a focal lymphocytic infiltration of the reticular dermis with collagen bands separated by clear spaces are present at a microscopic magnification of 60x (c).

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References
A 47-year-old woman presented with an approximately 1.5-year history of swelling and pain in the hand, elbow, shoulder, knee, and temporomandibular joint; bruising (either spontaneously or following a trauma); and pruritus all over the body. She was diagnosed with rheumatoid arthritis (RA) and was prescribed immunosuppressive drugs. Although the patient regularly took these medications, her joint pain and limited movement increased daily over the course of 1.5 years. Physical examination revealed limited motion, swelling, and tenderness in her wrists, metacarpophalangeal joints, proximal interphalangeal joints, elbows, shoulders, and knees. The patient had subcutaneous nodular lesions and itching on her arms, back, and both hips and ecchymoses on her legs. Her laboratory test results were as follows: hemoglobin, 11.2 (12–15) g/dL; calcium, 10.5 (8.6–10.0) mg/dL; gamma-glutamyl transferase, 126 (5–36) U/L; alkaline phosphatase, 106 (35–104) U/L; erythrocyte sedimentation rate, 18 (0–20) mm/h; and C-reactive protein, <3.3 (0.0–3.5) mg/L, and rheumatoid factor, anti-cyclic citrullinated peptide antibodies, and anti-nuclear antibodies were negative. There were no erosive changes in her hand joints that were suggestive of RA. Knee magnetic resonance imaging (MRI) was performed because of pain, swelling, and flexion contracture in the knee joint; the findings are shown in Figure 1. Full-body scan and regional single-photon emission computed tomography (SPECT)/CT revealed heterogeneous and symmetrical involvement of bony substance in the soft tissue around both hip joints, in the gluteal region, and around both shoulder joints; increased osteoblastic activity that was compatible with degenerative/inflammatory changes in both knee joints, elbows, and ankle joints; involvement of bony substance in the soft tissue around the hip joint and medial border of both the lower and middle sections of the scapula; and concentration of symmetrical involvement of bony substance in the above-described areas that were compatible with heterotopic ossification (Figure 2). On the basis of clinical, radiographic, and SPECT/CT findings, the patient was determined to have heterotopic ossification.

Heterotopic ossification is characterized by new bone formation in the tissue in which ossification should not normally occur (1). In the acute phase, erythema, swelling, and warm are observed, and during the formation process, severe pain can be seen (2). In addition, limited movement and nerve entrapment can result in an impaired quality of life (3). Direct trauma to the muscle tissue and burns, fractures, surgery, and other causes of trauma may cause heterotopic ossification, which is usually classified into the following three groups: neurogenic, traumatic, and myositis ossificans progressiva (MOP) (4). MOP is a rare autosomal dominant disease, and radiological and laboratory findings can be helpful in diagnosis. Following a trauma, a decrease in the calcium level and a subsequent increase in the alkaline phosphatase level can be observed. Direct radiographs and bone scintigraphy are important; however, ultrasonography, CT, and MRI can also be helpful (5). Heterotopic ossification can be treated by protecting the affected joint’s range of motion, administering medical treatment such as non-steroidal anti-inflammatory drugs, performing radio-

Figure 1. a, b. Sagittal T2-weighted fat-suppressed (a) and T1-weighted (b) MRI showing a signal similar to that of the bone marrow in the region of the patellar apex corresponding to heterotopic ossification (thick arrow), suprapatellar bursal fluid (arrowhead), and a popliteal cyst (thin arrow).
therapy, and performing surgery of the mature bone tissue in joints with a limited range of motion.

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Figure 2. a-d. Anterior (a) and posterior (b) full-body scans show heterogeneous accumulation of 99 mTc-MDP in periarticular muscles of joints. In the axial SPECT/CT images, symmetrical accumulation of 99 mTc-MDP around the hip joints (c) and semitendinosus muscle at the proximal part of the femur (d) is observed.

References
Chikungunya and bilateral sacroiliitis—is there a link?

Muhammad K. Nisar, Christabel Packianathan

Chikungunya (CKG) is an arthritogenic mosquito-transmitted alphavirus that manifests itself as a febrile illness and often progresses to severe and incapacitating polyarthralgia (1). Several reports have demonstrated persistent polyarthritis akin to seronegative peripheral arthropathy (2). However, to our knowledge, imaging-confirmed axial disease has not been described in this context. We report the case of an Asian lady who developed axial spondyloarthropathy (SpA) after contracting CKG while visiting India during an outbreak. Informed consent was obtained from the patient.

A 51-year-old previously healthy woman developed CKG infection whilst traveling to India during a regional outbreak in an endemic area. It began with classic symptoms of acute fever and generalized myoarthralgia followed by residual pain primarily affecting the small joints and to a lesser extent, pain in the lumbar region and both hips. There was no family history of SpA. Initial investigations including inflammatory markers at the time were unremarkable. Dengue nonstructural protein 1 antigen and dengue IgM were negative. Thyroid function tests and bone profile were normal. Serum 25-OH vitamin D was within the reference range. X-rays of small joints at the time showed soft tissue swelling in the wrists and ankles. The X-ray of the lumbosacral spine showed minor degenerative change in L4/5 and L5/S1, whereas that of the sacroiliac joints was normal. The patient was commenced on regular non-steroidal anti-inflammatory drugs (NSAIDs). Following graded physical activity over six months, her symptoms improved, apart from mild episodic fatigue and lethargy.

A year later, she presented to our rheumatology department with a 2-month history of severe intermittent alternating pain in both hips and buttocks. She experienced low back pain in addition to widespread myalgia, malaise, and fatigue.

Investigations at the time of presentation showed a normal full blood count, serum biochemistry, C-reactive protein, liver functions, glucose, bone profile including vitamin D, thyroid function, negative antinuclear antibody, anti-cyclic citrullinated protein antibody, rheumatoid factor, and human leukocyte antigen B27. Her CKG IgG was strongly positive (>1:10,000 by an indirect fluorescent antibody test). Surprisingly, the bone scan showed increased uptake in the inferior aspect of the sacroiliac joints bilaterally with the right joint involved more than the left (Figure 1a). An magnetic resonance imaging (MRI) scan confirmed bilateral sacroiliitis with marked edema across the inferior aspects of the joints (Figure 1b). The patient was commenced on regular NSAIDs and physiotherapy with good symptomatic relief.

Figure 1. a, b. Bone scan shows increased uptake in the inferior aspect of the sacroiliac joints bilaterally (a), fat suppressed short inversion time inversion-recovery (STIR) magnetic resonance imaging (MRI) scan image confirms bilateral sacroiliitis with marked edema across the inferior aspects of the joints (b)
There are numerous reports suggesting CKG arthritis can mimic rheumatoid arthritis (2). However, to our knowledge, this is the first report of CKG-associated axial SpA, though causal relationship cannot be established. This could be a case of de novo sacroiliitis or axial SpA rather than as a consequence of viral infection. While most cases of CKG arthropathy recover within several weeks, up to 12% retain residual joint symptoms for months to years (3). At present, not much is known about the underlying immunopathophysiological processes by which CKG virus causes arthritis. A recent article has suggested that chronic musculoskeletal tissue pathology is associated with persistent CKG infection and is controlled by adaptive immune responses (2).

Currently, there is no approved anti-viral treatment for CKG. Management is mainly supportive, and NSAIDs tend to be the mainstay treatment for arthritis (4). Hence, as outbreaks of this debilitating disease become global, a high level of suspicion is required for early diagnosis and management, particularly in centers with little exposure to this disease.

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**References**


Breast calcinosis in a patient with Dermatomyositis

Rajaie Namas1,2, Nassar Beydoun3, Alireza Meysami2

A 71-year-old woman presented with a pruritic macular rash on her chest that progressed to involve the limbs, face, and scalp. In addition, the rash was associated with progressive proximal muscle weakness that was symmetrically distributed, dysphagia, and fatigue over the past 6 months. A skin examination revealed Gottron’s and shawl signs. She underwent a skin biopsy that revealed perivascular lymphocytic infiltration and interface dermatitis. This was consistent with the diagnostic findings of dermatomyositis (DM). Moreover, electromyography was consistent with a myopathic pattern, and the muscle biopsy revealed a well-defined perifascicular distribution of muscle fiber injury with sparse collections of inflammatory cells predominantly surrounding the perimysial vessels and muscle fibers. These characteristics were diagnostic of DM (1).

Routine and serological tests revealed that both erythrocyte sedimentation rate and C-reactive protein levels were elevated. The patient was reluctant to undergo prednisone or intravenous immunoglobulin therapy, and she was administered methotrexate instead. She exhibited a good response to treatment.

A mammogram revealed bilateral, unusual, coarse, heterogeneous, branched, and sheet-like dystrophic calcifications, which were regionally distributed at the top and toward the center at a posterior depth. The left side was affected to a greater extent than the right (Figure 1a, b).

Figure 1. a, b. Mediolateral oblique view (a) and craniocaudal view (b) of the mammography showing unusual, sheet-like dystrophic calcifications in the subcutaneous tissues, which are regionally distributed at the top and toward the center at a posterior depth. The left side was affected to a greater extent than the right.

Routine and serological tests revealed that both erythrocyte sedimentation rate and C-reactive protein levels were elevated. The patient was reluctant to undergo prednisone or intravenous immunoglobulin therapy, and she was administered methotrexate instead. She exhibited a good response to treatment. Age- and risk-appropriate screening was performed, which was negative for malignancy. Interestingly, a mammogram revealed bilateral, unusual, coarse, heterogeneous, branched, and sheet-like calcifications, which were regionally distributed at the top and spanned toward the center at a posterior depth. In addition, the left side was affected to a greater extent than the right, which is compatible with benign subcutaneous calcifications associated with DM (Figure 1a, b).

Diffuse dystrophic calcifications of the subcutaneous fat that become progressively coarser over time may be seen in collagen vascular diseases such as scleroderma, DM, and systemic lupus erythematosus. Patients with DM may develop unusual, sheet-like dystrophic calcifications in the subcutaneous tissue that could be...
either localized or extensive (2, 3), as observed in the mammogram of our patient.

Dermatomyositis is a heterogeneous disease of the connective tissue characterized by an inflammatory process involving the skin, skeletal muscles, and various connective tissues (4). Soft tissue calcifications termed “calcinosis” are common in approximately 10%-40% of patients with juvenile DM but are unusual in patients with adult-onset DM (5). Risk factors attributed to the development of calcinosis include young age and delayed diagnosis or therapy. Interestingly, the incidence of calcinosis was inversely proportional to creatinine phosphokinase levels (6). In summary, given the associated risk between DM and malignancy, physicians should be aware that this finding can occur within 4 months or up to 12 years from the onset of disease.

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References
Erosive cervical spine involvement in primary Sjögren’s syndrome

Marco Di Carlo¹, Marina Carotti², Francesco Sessa³, Daniele Roia¹, Marika Tardella¹, Fausto Salaffi¹

A 69-year-old woman, diagnosed with primary Sjögren’s syndrome that was characterized by the presence of sicca symptoms and anti-nuclear and anti-Ro antibodies and histologically confirmed with a minor salivary gland biopsy 11 years ago (Figure 1a), consulted our rheumatologic department for a recent onset of inflammatory neck pain. Physical examination revealed pronounced cervical stiffness, but no signs of synovitis in the peripheral joints. Magnetic resonance imaging of the cervical spine in sagittal T2-weighted scan showed the presence of synovitis of the atlantoaxial joint, with synovial pannus surrounding the odontoid process of the second cervical vertebra and compressing the cord (Figure 1b). Bone marrow edema and erosions were also detected in the odontoid process. Subsequent coronal (Figure 1c) and sagittal computed tomography scans (Figure 1d) confirmed the presence of well-defined multiple erosions at the base and apex of the odontoid process. Rheumatoid factor and anti-citrullinated protein antibodies were absent. X-rays of the hands, wrists, and knees revealed only mild signs of osteoarthritis.

Figure 1. a-d. Histological examination (a) of a minor salivary gland biopsy revealing a lymphocytic infiltrate suggestive of Sjögren’s syndrome, with a focus score of 4. Magnetic resonance imaging (b) (T2-weighted sagittal scan) shows synovial hypertrophy of the atlantoaxial joint with compression on the spinal cord and bone marrow edema of the odontoid process, whereas the coronal (c) and sagittal (d) computed tomography scans detail the presence of multiple erosions of the odontoid process.

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The atlantoaxial joint could be involved in different articular inflammatory diseases. Calcium pyrophosphate deposition disease could appear as calcifications in the ligaments surrounding the odontoid process (“crowned dens syndrome”) (1). However, the atlantoaxial joint is one of the chief target in rheumatoid arthritis (2). To our knowledge, this is the first depiction of inflammatory and erosive cervical spine involvement in primary Sjögren’s syndrome.

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References
Cutaneous calcinosis in a patient with limited scleroderma: CREST Syndrome

Nurşen Düzgün

Cutaneous calcinosis in a patient with limited scleroderma: CREST Syndrome

Calcinosis or dystrophic soft-tissue calcification occurs in damaged or devitalized tissues in the presence of normal calcium/phosphorus metabolism. It is a known manifestation in the subcutaneous tissues of patients with connective tissues diseases, especially scleroderma, systemic lupus erythematosus, or dermatomyositis, and may involve a relatively localized areas or present as widespread calcinosis (1). Little is known about its physiopathology. It occurs in tissues that are under chronic stress, such as local trauma or damage associated with underlying inflammatory processes (2).

Subcutaneous calcinosis occurs in all subsets of scleroderma but is more prominent in patients with limited scleroderma and in those with anticentromere antibody. CREST syndrome is a limited form of scleroderma, characterized by calcinosis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia (3).

A 46-year-old woman suffered from CREST syndrome for 15 years. She had sclerotic cutaneous findings on her face and fingers (sclerodactyly), prominent facial telengectasia, Raynaud's phenomenon, and esophageal reflux. There were also multiple hardened erythematous-whitish nodules, some having a chalky appearance, around her knees (Figure 1). X-ray showed extensive calcinosis in the soft tissue of the knees (Figure 2). Pulmonary arterial pressure without pulmonary fibrosis was found to be 40
mmHg using echocardiography. Antinuclear and anti-centromere antibodies were positive. Serum calcium and phosphorus levels were within the normal ranges. She had been treated with nifedipine, low dose aspirin, and colchicine for many years. Treatment was initiated with aluminum hydroxide. Medical therapy for cutaneous calcinosis is limited and has variable benefits. Multiple treatment approaches with diltiazem, disodium etidronate, probenecid, colchicine, minocycline, low-dose warfarin, and intralesional adrenal steroids have been explored, but no standard treatment has convincingly prevented or reduced calcinosis.

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**Conflict of Interest:** No conflict of interest was declared by the authors.

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A 10-month-old boy presented with a high-grade fever for 4 days. He also had bilateral, firm, tender anterior cervical node enlargement that was more prominent on the right side (Figure 1). No other lymph node groups were enlarged. Dryness, fissuring, and redness of the lips were noted at the time of admission. Skin desquamation with erythema was also noted in the perianal area (Figure 2), and hence, Kawasaki disease (KD) was considered a strong clinical possibility. There was no evidence of redness of the eyes, rash, erythema of the oral mucosa, or edema of the extremities. His blood count tests revealed leukocytosis (34.8×10⁹ cells/L) with neutrophilic predominance (56% polymorphs), anemia (hemoglobin, 79 g/L), and platelet counts of 106×10⁹ cells/L. Erythrocyte sedimentation rate (ESR) was 32 mm (1st hour) and C-reactive protein was 179.2 mg/L. Serum albumin was 2.3 g/dL. Echocardiography showed brightness of the left main coronary and left anterior descending arteries. There was, however, no dilatation. Blood cultures were sterile and anti-streptolysin O titer were normal. Intravenous immunoglobulin (IVIg) (Meglob; Synergy Diagnostics Pvt Ltd, Thane, Maharashtra, India) at 2g/kg infusion and aspirin at 50 mg/kg/day were initiated 5 days after onset. There was prompt defervescence of fever following IVIg therapy. Serial blood counts revealed progressive rise in platelet counts (251×10⁹ cells/L and 778×10⁹ cells/L on days 8 and 11 of illness, respectively). The baby subsequently developed periungual peeling over the fingers on day 10 of illness.

The American Heart Association (AHA) epidemiological definition for KD requires the presence of fever for at least 5 days and the presence of four of the following five clinical features: extremity changes, exanthematous rash, bulbar conjunctival injection, cervical adenopathy, and changes in lips and oral cavity (1). Apart from fever, the child had only three clinical features of the AHA Criteria for KD, namely anterior cervical adenopathy, lip changes, and periungual peeling at the subacute phase. As per the evaluation of suspected incomplete KD, the index child had additional features such as perianal peeling, coronary artery abnormalities in the echocardiogram, CRP of ≥3 mg/L, and ESR of ≥40 mm/h (1). Supplemental laboratory features for KD in the child were leukocytosis, anemia, elevation in platelet counts ≥450,000/mm³ after 7 days, and hypoalbuminemia (1). Moreover, the clinical course of the illness with the development of...
typical periungual peeling and progressive rise in platelet counts observed in the subacute phase of illness strongly suggests KD (1). Incomplete presentation of KD is well known in infants and carries a high risk of coronary artery abnormalities (2).

Perineal desquamation, a key clinical feature of KD, can be easily overlooked in the clinical evaluation of febrile children, unless meticulously looked for (3). Perineal or perianal peeling may be noted as early as the 6th day of fever and occurs much earlier than the periungual peeling. The mechanism for desquamation in KD is not completely understood. Release of toxins or superantigens and excess cytokine production by the immune cells in the skin are some of the proposed hypotheses for the skin peeling (4). Perineal skin peeling is not a part of the AHA clinical criteria for the diagnosis of KD (1). Although differential diagnoses for fever and cervical adenopathy are many, perianal desquamation seen in the index child was a strong clinical indicator of KD and enabled us to promptly initiate IV Ig (1). This report highlights the importance of perianal examination of a febrile child, in whom KD, an important medium vessel vasculitis that requires timely management, is suspected.

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References
Tropical pyomyositis caused by *Klebsiella pneumoniae* with rheumatoid arthritis

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The patient, a 33-year-old woman with rheumatoid arthritis, presented with a 3-month history of a painful nodule in the right thigh that progressively evolved to a large mass, accompanied by intermittent fever. Physical examination revealed a large fluctuating mass involving the entire anterior portion of the right thigh, which was tender and warm to palpation (Figure 1). Her medications included methotrexate 25 mg/week, prednisone 10 mg/day, [Infliximab 4mg/kg 6/6week] (total duration of treatment, 3 years). Laboratory tests revealed the following: hemoglobin 11.7 g/dL, white blood cell count 21.16×10⁹/µL with 85% neutrophils and 7% band cells, erythrocyte sedimentation rate 100 mm/h, and C-reactive protein level 15 mg/dL (normal<0.8 mg/dL). Blood culture was negative; Human Immunodeficiency Virus (HIV) and hepatitis serologies were non-reactive. Magnetic resonance imaging (MRI) of the right thigh revealed voluminous liquid collection involving the entire anterior compartment of the thigh with complete quadriceps musculature destruction (Figure 2, 3). A high volume of purulent material was drained after surgical incision of the abscess region, followed by necrotic muscle tissue debridement. Cultures from the abscess material and biopsy specimens of the muscles revealed *Klebsiella pneumoniae* with a multi-susceptible profile. Findings of fungal and mycobacterial tests were negative. Muscle biopsy showed abscedated nonspecific chronic myositis. Antibiotic treatment was initiated with imipenem for 14 days with a good clinical response. Tropical pyomyositis caused by gram-negative bacteria is uncommon and primarily seen in immunocompromised patients, being identified in only 11% of patients according to an American meta-analysis (1-3). The diagnosis may be difficult, particularly when we are faced with an atypical clinical case, such as this patient.

Figure 1. Large fluctuating mass involving anterior portion of the right thigh in all of its extension, intact skin presenting a shining erythematous aspect with collateral circulation

Figure 2. Axial T1-weighted magnetic resonance imaging (MRI) of right thigh revealed voluminous liquid collection involving all anterior compartment of the thigh with complete quadriceps musculature destruction
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References
Gouty tophi with purpura in a patient with ischemic heart disease and acute kidney injury

Manoj Karbhari Pawar

A 77-year-old male with a history of chronic poorly controlled hypertension (HTN) and ischemic heart disease (IHD) was referred to a dermatologist for the presence of red-brownish flat lesions over both forearms and the dorsum of hands since 2 weeks and localized yellowish elevated lesions that were hard in consistency over both 1st and 2nd toes and the left little finger since 8 months; these were associated with moderate-grade pain. The patient was on tablet nebivolol (5 mg) (Nebicip 5; Cipla, Mumbai, India) once daily and tablet clopidogrel (75 mg) plus aspirin (75 mg) (Clasprin; Biocon, Mumbai, India) for HTN and IHD. He had been admitted under the care of an internist for cellulitis of the left foot with acute kidney injury (AKI) 10 days previously. He was a chronic alcoholic and tobacco chewer. Skin examination revealed multiple purpuric lesions of varying sizes over both forearms and the dorsum of hands (Figure 1) along with multiple subcutaneous gouty tophi over both 1st toes and the left little finger (Figure 2, 3). On close inspection, there was extrusion of gouty material from the right great toe (Figure 4). Findings of the remaining cutaneous examination were unremarkable. Laboratory evaluation revealed leukocytosis with neutrophilia and elevated serum creatinine (2.3 mg/dL), urea (59 mg/dL), and uric acid (10 mg/dL) levels. The prothrombin time was also increased (16.5 s). Based on these findings, the patient was diagnosed as having gout with purpura co-existing with left foot cellulitis and AKI. X-ray of the right foot showed osteolysis with a coarse trabecular pattern, while that of the left forearm was normal (Figure 5). His chest X-ray revealed cardiomegaly with blunting of the left cardiopulmonary angle (Figure 6).

Gout is one of the most common debilitating types of arthritis, mostly present in males and postmenopausal females; it is characterized by increased serum uric acid levels (more than 6.8 mg/dL at 37°C, pH 7.4) and the deposition of monosodium urate crystals in and around the joints (1, 2). If this hyperuricemic stage persists for a long duration without treatment, gouty tophi develop and get deposited around joints, particularly the 1st metatarsophalangeal joint (podagra), and eventually damage the involved joint (1, 2). As most uric acid gets excreted through the kidney, any renal insufficiency that predisposes the patient to the development of gout and chronic hyperuricemia can also cause renal injury (1). Because our patient was suffering from chronic HTN, he was likely to have some degree of renal impairment, which was further worsened by preuremic (cellulitis-induced) AKI, leading to the development and aggravation of gout. Because the patient was taking anticoagulants, he developed iatrogenic purpuric lesions, which were possibly worsened by AKI and alcoholism (3). The gold standard for the diagnosis of gout is the identification...
of urate crystals in the tissue and/or synovial fluid of an inflamed joint (1). Patients presenting with gout often have co-morbidities such as hypertension, obesity, diabetes, chronic kidney disease, and IHD (1). The treatment of gout varies from the stage of the disease and level of uric acid. Acute flares are treated with nonsteroidal anti-inflammatory agents, colchicines, and steroids. Allopurinol, febuxostat, uricosuric drugs, and uricases reduce uric acid levels (1). Lifestyle modifications and concurrent management of co-morbidities help in treating gout and achieving remission.

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References
A 69-year-old woman presented with a 2-month history of bilateral morning stiffness and arthralgia of the wrists, shoulders, and ankles in November 2016. She had undergone total hysterectomy and bilateral salpingo-oophorectomy for endometrial cancer 7 years ago and had subsequently undergone chemotherapy. However, her response to chemotherapy had been poor, and she had developed metastatic lung cancer 4 years later. A physical examination showed clubbed fingers and toes (Figure 1a, b), swelling of both ankles, and bilateral tenderness of the forearms and shins. Blood test results showed mildly elevated C-reactive protein and plasma vascular endothelial growth factor (VEGF) levels. The results were negative for rheumatoid factor, anticitrullinated protein antibody, and antinuclear antibody tests. A plain radiograph of the tibias showed periosteal thickening, and power Doppler ultrasound signals over the periosteum of the radius, ulna, and tibia indicated periosteal inflammation (Figure 2 a-d). Bone scintigraphy revealed the linear uptake of technetium-99m-labeled methylene diphosphonate in the radii, ulnas, and particularly, the tibias (Figure 3). Hypertrophic osteoarthropathy (HOA) associated with metastatic lung cancer was diagnosed. After treatment with a nonsteroidal anti-inflammatory drug and intravenous zoledronic acid, the pain in her joints and bones improved with reduction in Doppler signals. However, the metastatic lung tumors slowly enlarged, and the pain in her joints and bones progressively worsened.

Hypertrophic osteoarthropathy is a syndrome that manifests as clubbing of the fingers, polyarthralgia of the large joints, and ostealgia of the tubular bones; HOA is characterized by periosteal proliferation (1-3). HOA is usually associated with lung cancer and cyanotic heart disease. Its pathogenesis has not been elucidated, but the roles of platelet-derived growth factor produced by non-fragmented megakaryocytes that bypass the pulmonary vascular bed and prostaglandins and VEGF produced by lung cancer have been suggested (4, 5). Radiography and bone scintigraphy are conventionally used as diagnostic tools to detect periosteal reactions. It is notable that Doppler ultrasound can detect periosteal inflammation, as was seen in the present case. Doppler ultrasound may be an alternative diagnostic tool, particularly in primary care settings, as it is an easy, cheap, and non-ionizing radiative procedure that can be beneficial for evaluating the efficacy of therapeutic procedures. In the management of HOA, treating the underlying disease is critical. When the underlying disorder cannot be treated, treatment with analgesics and bisphosphonates has been reported to relieve clinical symptoms (6).

Hypertrophic osteoarthropathy is a rare disease, but it can mimic an arthritic condition. If a patient presents with arthralgia and ostealgia with clubbed digits, physicians should suspect HOA and consider an underlying disorder such as an intrathoracic malignancy. Screening with plain radiographs of the chest and tubular
bones, as well as Doppler ultrasound to detect periosteal reactions and inflammation, may be helpful in the diagnosis of HOA.

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References
A 68-year-old male smoker presented with a painful mass on his right hand since the past 3 months. The mass, which mimicked an infectious, inflammatory, or metabolic disease, was swollen, erythematous, and located on the dorsum of his right hand. X-ray images of his hands revealed a massive and aggressive osteolytic lesion with cortical expansion, that destroys the fifth metacarpal (Figure 1). According to the patient’s radiological features, the diagnosis of a malignant tumor was strongly considered. Bony biopsy was performed, and a histopathological examination found a moderately differentiated carcinoma infiltrating the bone. Immunohistochemistry study results were negative for thyroid transcription factor-1, cytokeratin 20, prostatic-specific antigen, and thyroglobulin. The radiological findings and anatomopathological features highlighted the need to find an underlying malignant lesion in our patient. A thoraco-abdomino-pelvic computed tomography scan was performed, which revealed a right lobe lung mass (Figure 2). Biopsy of the lung tumor revealed adenocarcinoma. The patient was finally diagnosed as having lung carcinoma with acrometastasis. For that, he received palliative chemotherapy.
The occurrence of acrometastasis is very rare. Only 1% to 3% of cases of metastases occur in the hands (1). Moreover, acrometastasis as the first manifestation of carcinoma is very rare. Metastases in the hands preferentially reach the phalanges and then the metacarpals and carpals (2). Acrometastasis is most commonly observed secondary to lung cancer (40%-50%) (3). It is associated with a poor prognosis. Physiopathological mechanisms of acrometastasis are not well elucidated. The principal theory is the ability of tumor cells to migrate and invade the bone matrices of the hands under the influence of proinflammatory cytokines such as tumor necrosis factor, interleukin 6, and osteoclast-activating factors. Usually, acrometastasis presentations mimic an infectious, an inflammatory, or a metabolic disease. For this reason, its diagnosis is often delayed (4). The present case illustrates a rare and exceptional situation of acrometastasis of the metacarpals as the first manifestation of lung cancer.

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Dacrocystitis and orbital pseudotumor in a patient with Granulomatosis with polyangitis

Sajal Ajmani, Abhishek Zanwar, Pradeepta Patro, Able Lawrence

A 38-year-old male presented with decreased vision and diplopia in his right eye over the past month. On performing an examination, he had proptosis, periorbital puffiness, dilated episcleral vessels, sluggish pupillary reflex, external ophthalmoplegia, and diminished vision. He also had left dacryocystitis (Figure 1a). He had presented six years ago with bilateral hearing loss, recurrent sinusitis, and hemoptysis and was diagnosed with GPA (anti-PR3 titer >100 units/mL and a necrotizing granuloma in the nasal biopsy). He was initially treated with steroids and intravenous cyclophosphamide that were discontinued after the second dose as he developed pneumonia twice; he was subsequently treated with mycophenolate mofetil, followed by azathioprine and steroids. His current anti-PR3 titer was <3 units/ml. An MRI orbit showed an ill-defined T1/T2 isointense-to-hypointense lesion (pseudotumor) replacing the orbital fat that diffusely involved intra- and extraconal compartments (Figure 1b, arrows). The extraocular muscles were encased by the lesion and appeared bulky, with mild flattening of the posterior globe (Figure 1c, arrows). The lesion encased the optic nerve and mildly compressed the optic nerve in the orbital apex. The patient was administered three doses of methylprednisolone pulses and two doses of rituximab (500 mg) two weeks apart. He responded well to treatment; he showed resolution of dacryocystitis, periorbital puffiness (Figure 1d), and diplopia and improvement in his vision. GPA has a wide spectrum of orbital manifestations, which can occur in up to 52% of patients. Patients with GPA...
may develop conjunctivitis, corneal ulceration, episcleritis/scleritis, optic neuropathy, retinal vasculitis, and uveitis. In addition, a retro-orbital pseudotumor and nasolacrimal duct obstruction may occur (1, 2).

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References
A case of localized cervical bony ankylosis due to ulcerative colitis

Keiichi Iwanami

Localized neck pain is an uncommon presenting complaint of ulcerative colitis (1).

A 43-year-old man presented with a 10-year history of cervical pain. He had occasional diarrhea without bloody stool or abdominal pain for 20 years. As this abdominal symptom was mild, he did not seek medical attention. There was no family history of spondyloarthritis, psoriasis, or inflammatory bowel diseases. On examination, his neck was ankylosic and could not be moved in any direction. Peripheral joint examination revealed no enthesitis or synovitis. The modified Schober test result was negative. Skin and nail examinations indicated no evidence of psoriasis. Blood test results revealed elevated inflammatory marker levels (C-reactive protein level, 26.0 mg/L; erythrocyte sedimentation rate, 65 mm/h). Test results for the rheumatoid factor and anti-citrullinated protein antibody were negative. The test result for human leukocyte antigen (HLA)-B27 was negative, but the result for HLA-B60, which is associated with spondyloarthritis in the Asian population, was positive (2, 3). Computed tomography of the cervical spine was performed, revealing bony ankylosis of both facet joints (Figure 1). Magnetic resonance imaging revealed no evidence of sacroilitis or lumbar spondylitis. Underlying inflammatory bowel disease (IBD) was suspected; thus, colonoscopy was performed and showed mucosal edema and erythema of the rectum and sigmoid colon. Biopsy findings indicated lymphoid aggregates with crypt disarray. Accordingly, spondyloarthritis due to ulcerative colitis was diagnosed. After adalimumab treatment, his neck pain and diarrhea resolved, although his neck remained immobile.

Spinal involvement occurs in up to 20% patients with IBD, with the involvement of any facet joint (1, 4). Spinal symptoms may precede intestinal symptoms or develop later; spinal symptoms do not always correlate with intestinal symptoms. Spinal involvement is often silent. On the other hand, silent IBD can be found on biopsy in patients with spondyloarthritis, such as our case (5). Thus, colonoscopy with histological exploration should be performed in a case of spondyloarthritis with unknown origin.

In enteropathic spondyloarthritis, the prevalence of HLA-B27 is only between 53% and 75%; this is lower than that in ankylosing spondylitis (1). HLA-B60 is prevalent in patients with HLA-B27-negative ankylosing spondylitis and undifferentiated spondyloarthritis among the Asian population (2, 3). HLA-B60 may be correlated with enteropathic spondyloarthritis.

There are no specific blood tests for confirming a suspicion of IBD-related arthritis. This report highlights the importance of colonoscopy in addition to careful history and clinical examination of a patient in whom spondyloarthritis with unknown origin was found.

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Scurvy: A rare cause of arthritis in a child with neurologic disorder

Zeynep Küçükaydın1, İsmail Dursun1, Burcu Daldaban1, Alper Özcan2, Ekrem Ünal2

A 7-year-old boy presented with swelling in the knees, walking difficulty, petechial rashes on the lower extremity, and gum swelling and bleeding. His medical history was remarkable for mental retardation and autism. He was referred to our clinic with a differential diagnosis of bleeding disorder. His dietary history was positive for unbalanced nutrition (yogurt soup, chocolate, and wheat bread). On admission, his weight and height were normal, looked very ill, and had a body temperature of 38.3°C. His physical examination was remarkable for swollen and bleeding gum, follicular hyperkeratosis with perifollicular purpura at the lower extremities, and soft tissue swelling of both knees, which were painful during passive motion with bilateral 30° flexion contracture of the knees (Figure 1a and b). He had persistent and severe self-injurious behavior. Blood investigations showed anemia of chronic disease, elevated CRP level, and prolonged in vitro bleeding time. Bilateral knee diagraph showed a radio-dense band at the chondro-osseous junction Frankel’s line (Figure 2a). Magnetic resonance imaging demonstrated bright signal intensity on the metaphyses and juxtaosseous soft

Figure 1. a, b. Perifollicular hyperkeratosis on the lower extremity (a); gingival hypertrophy and gum bleeding (b)

Figure 2. a, b. Frontal radiograph of lower extremity shows increased density at the zone of provisional calcification (Frankel’s line) (a); coronal a T1-weighted image demonstrates bright signal intensity within the metaphyses of the distal femurs and proximal tibias (b)
Because we could not measure leukocyte vitamin C level, we measured serum vitamin C level, which was very low (<0.1 mg/dL). He was diagnosed with scurvy. His clinical and laboratory findings returned to normal with vitamin C supplementation.

Scurvy was first documented in the Ebers papyrus in 1550 BC (1) and became famous as a sailor disease after the death of at least two million sailors between the 16th and 18th centuries (2). Because scurvy is uncommon in pediatric patients, a high degree of suspicion is required to reach the diagnosis of scurvy, especially in children with severely restricted diets because of either developmental or psychiatric disturbances (1, 3-5). Clinicians should have an awareness of vitamin C deficiency as the differential diagnosis of musculoskeletal pain and purpura at-risk children with/without gingival bleeding and hypertrophy.

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References
Fever with multiple large vessel aneurysms: An unusual presentation of Takayasu arteritis in a child

Ashish Agarwal¹, Ankur K. Jindal¹, Sandesh Guleria¹, Tarun K. Jain², Bhagwant R. Mittal², Deepti Suri¹

A 12-year-old boy presented with a history of fever for one and a half months. On performing an examination, he had pulse rate of 90/min, respiratory rate of 20/min, and blood pressure of 110/70 mmHg; all peripheral pulses were equally palpable, pallor was noticed, and a bruit was heard in the bilateral carotid region. Examinations of the eyes revealed unremarkable findings. Laboratory investigations revealed anemia, a high erythrocyte sedimentation rate, and elevated C-reactive protein levels; work-up for infective causes was negative (Table 1). Doppler ultrasound revealed thickening of the internal lamina of the left common carotid artery, dilatation (1.2 cm), and a small saccular outpouching (8 mm) of the superior mesenteric artery (SMA) near its origin. Contrast-enhanced computed tomography angiography of the abdominal vessels revealed diffuse irregular and non-calcified mural thickening, vascular dilatation, and multiple small saccular aneurysms involving the abdominal aorta, proximal SMA, and proximal parts of the bilateral renal arteries (Figure 1, 2). Computed tomography and corresponding fluorodeoxyglucose positron emission tomography images revealed mural thickening of the proximal left subclavian artery, brachiocephalic artery, dilated abdominal aorta, bilateral proximal renal arteries, and proximal SMA (Figure 3-6). The boy was diagnosed as having Takayasu arteritis (TA) based on the European League Against Rheumatism / Paediatric Rheumatology International Trials Organisation / Paediatric Rheumatology European Society classification criteria for childhood TA (1). He was initiated on injections of methylprednisolone (30 mg/kg/day) that led to prompt defervescence. He was given five pulse doses of methylprednisolone and was subsequently initiated on oral prednisolone (2 mg/kg/day). He was also initiated on monthly pulse cy-

Table 1. Laboratory investigations

<table>
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<th>Result</th>
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<td>Hemoglobin level (gm/L)</td>
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<tr>
<td>White blood cell count (×109 cells/L)</td>
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<tr>
<td>Differential count (N/L/M/E)</td>
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<tr>
<td>Platelet counts (×109/L)</td>
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<tr>
<td>ESR (mm in the 1st hour)</td>
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<tr>
<td>CRP (mg/L) (N &lt; 6)</td>
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<td>Tuberculin skin test</td>
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<tr>
<td>Blood culture</td>
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<td>2-dimensional echocardiography</td>
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CRP: C-reactive protein; EBV-VCA: Epstein Barr Virus-viral capsid antigen; ESR: erythrocyte sedimentation rate; HBsAg: hepatitis B surface antigen; HCV: hepatitis C virus; HIV: human immunodeficiency virus; N/L/M/E: neutrophils/lymphocytes/monocytes/eosinophils; VDRL: venereal disease research laboratory
The natural course of Takayasu arteritis consists of two phases: an initial acute phase that represents the phase of inflammation, resulting in constitutional symptoms, and a second phase of vascular insufficiency, leading to symptoms such as claudication and diminished pulses. Studies on pediatric TA cases from India and USA have reported that hypertension and headache are the most common presenting manifestations (2-4). The index case had fever as the sole clinical presentation and had no signs of vascular insufficiency. Fever has been reported in approximately 4-45% of children with TA; however, fever as the only clinical presentation of TA in children is extremely uncommon (2-4).

Stenosis of large vessels is the most common angiographic abnormality associated with TA (5). Aneurysms have been reported in approximately 19-65% of children with TA (6). Aneurysms as the only angiographic abnormality of TA without stenosis or occlusion are extremely uncommon (7-8).

Figure 1. Contrast-enhanced computed tomography of the abdominal vessels shows dilatation of the abdominal aorta near the origin of the proximal superior mesenteric artery (black arrow) and small saccular aneurysms at the proximal parts of the bilateral renal arteries (white arrow).

Figure 2. Transverse cuts of contrast-enhanced computed tomography of the abdominal vessels show multiple aneurysms at the origin of the renal arteries (arrow).

Figure 3. Fluorodeoxyglucose (FDG) positron emission tomography images show FDG uptake (SUVmax: 1.5) at the thickened wall of the dilated left subclavian artery.

Figure 4. Fluorodeoxyglucose (FDG) positron emission tomography images show FDG uptake (SUVmax: 1.9) at the thickened wall of the dilated left subclavian artery.

Figure 5. Fluorodeoxyglucose (FDG) positron emission tomography trans-axial images at the abdominal level show FDG uptake at the thickened wall of the dilated abdominal aorta and superior mesenteric artery [SUVmax: 2.8 at the abdominal aorta (arrow) and 2.2 at the superior mesenteric artery].

Figure 6. Fluorodeoxyglucose (FDG) positron emission tomography coronal images at the abdominal level show FDG uptake at the thickened wall of the dilated left subclavian artery.

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Balanitis circinata
Abhishek Zanwar, Latika Gupta, Ramnath Misra

A 24-year-old boy presented with arthritis of the right knee and both ankles since 2 weeks. On performing an examination, apart from arthritis of the affected joints, dactylitis of the left thumb was seen. On questioning, he admitted to having dysuria, and a genital examination revealed multiple erythematous shallow ulcers with serpiginous margins on the glans penis (Figure 1). He did not have a history of inflammatory back pain, heel pain, or pain at other entheseal sites. He never had pain or redness of the eyes, prolonged diarrhea, or rashes elsewhere. He did not have a family history of arthritis, uveitis, or psoriasis.

Circinate balanitis, which is seen in up to 40% of men with reactive arthritis (ReA), is often painless; hence, its presence is not voluntarily mentioned by patients, unless they are specifically asked about it. In almost half of the patients with ReA, the causative pathogen of preceding infection cannot be identified (1, 2). In such cases, the presence of circinate balanitis can serve to differentiate ReA from other infections associated acute arthritides, such as poststreptococcal ReA, rheumatic fever, or viral arthritis.

In patients with arthritis but with no history of preceding infection, undifferentiated spondyloarthritis (UspA) is an important differential. UspA is a chronic disease marked with asymmetric oligoarthritis and enthesitis. The presence of Circinate balanitis in such a setting indicates ReA rather than UspA, signifying a shorter disease duration and better prognosis. It is now believed that most cases of UspA are forme fruste of ReA itself in the absence of identified infectious triggers (3). As classic ReA with balanitis circinata has been described in the setting of Human Immunodeficiency virus (HIV) infection, it is important to test for the same in all these cases (4). Hence, the presence of circinate balanitis is not specific but is an important clue to the etiology of arthritis. The presence of long-standing psoriasiform lesions with destructive arthritis or prominent distal interphalangeal joint involvement suggests a diagnosis of psoriatic arthritis. On the other hand, a preceding history of infection in a healthy adult with a short history of arthritis is best labelled as of ReA.

Although circinate balanitis by itself usually does not require treatment, it can serve as a salient marker of underlying genitourinary infections, which warrant treatment to prevent relapses (3). Most often seen with genitourinary chlamydia induced reA, circinate balanitis can also occur with gastrointestinal infections. In refractory patients, topical salicylates usually suffice, though mild topical glucocorticoids and calcineurin inhibitors have also been successfully used (1).

Figure 1. Picture of the uncircumcised penis showing multiple shallow ulcers with a white base and serpiginous edges on the glans; a few lesions have coalesced to give a polycyclic appearance.
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Looking for a needle in a haystack

Paloma Vela, Laura Ranieri

A 56-year-old man with long-standing chronic seronegative polyarthritis was first seen in our clinic. Treatment with synthetic DMARD and anti-TNF drugs was ineffective in controlling inflammation, and structural damage was obvious. A clinical exam showed prominent indurated nodules in both elbows. For an accurate diagnosis, aspiration of a nodule for microscopic examination was performed. The small sample obtained showed abundant cholesterol crystals (Figure 1a, compensated polarized light), but a careful examination allowed identifying typical acicular crystals with a strong negative birefringence characteristic of monosodium urate (Figure 1b, black arrow, -λ shows the compensator axis), confirming the diagnosis of gout. Cholesterol crystals are commonly found in chronic processes, but they are not specific (1). However, the finding of monosodium urate crystals allows the accurate diagnosis of gout. Joint, bursa, or nodule aspiration is a simple and easy procedure, and it could result in findings as interesting and useful as these.

Figure 1. a, b. Cholesterol crystals seen by compensated polarized light microscope to 200X. -λ shows the compensator axis (a); Black arrow showing acicular crystals with a strong negative birefringence characteristic of monosodium urate, seen by compensated polarized light microscope to 400X. -λ shows the compensator axis (b).

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Reference
Chronic large nasal bloody crusting and recurrent episcleritis: Limited granulomatosis with polyangiitis

Nurşen Düzgün1, Haldun Umudum2

Granulomatosis with polyangiitis (GPA) (formerly named Wegener’s granulomatosis) is an uncommon kind of systemic vasculitis involving small-to-medium sized vessels, and categorized as ANCA-associated vasculitis with the presence of anti-neutrophil cytoplasm antibodies (1). It is characterized by the formation of necrotizing granuloma in the upper and/or lower respiratory tract and glomerulonephritis. Subsequently, it affects almost any organ or tissue. GPA is influenced by genetic, immunologic, and environmental factors (2). GPA affects people at any age, usually between 60 and 70 year of age in both sexes. Two forms of GPA are systemic and diffuse forms; systemic GPA typically includes renal and pulmonary manifestations and/or vital organ involvement and systemic symptoms, such as fever, anorexia, or weight loss, and as localized/limited forms that predominantly affect the upper respiratory tract, but they are recurrent (3). The diagnosis of disease is based on the classification criteria for granulomatosis with polyangiitis provided by the American College of Rheumatology (4). Currently, ANCA is used for diagnosis in clinical practice.

A 69-year-old female with painless, redness in the left eye, which persisted for 2 weeks, and chronic weakness and general malaise, was referred to the rheumatology clinic for further evaluation of her condition. She had ophtalmic history, including an episcleritis requiring systemic corticosteroid treatment 2 years ago. Medical history revealed that she has been suffering from chronic nasal dryness and nasal bloody crusting rhinorrhea/discharge (Figure 1) since 5 years. Her ophtalmic and ear nose, and throat (ENT) examination after admission showed episcleritis and nasal purulent crusting. There was no evidence of other organ/tissue involvement.

Her laboratory findings revealed systemic inflammation. Blood parameters were as follows: white blood cell count 11x10^3/µL, neutrophil 81%, C-reactive protein 84 mg/dl, erythrocyte sedimentation rate 89 mm/h. Urine analysis result was normal. Anti-PR3, anti-MPO, and antinuclear antibodies were negative. Rheumatoid factor was 75.6 IU (normal value: <20 IU).

Sinusal tomography showed increased amount of soft tissue in the paranasal sinuses. Chest graphy and computed tomography results were normal. Biopsy of the nasal mucosa and the histopathological findings were consistent with GPA (Figure 2-4).

Nasal-sinus involvement occurs in approximately 85% of patients with GPA, such as bloody nasal discharge or crusts, chronic sinusitis, bone, and/or cartilage destruction (5, 6). The ocular manifestations, such as conjunctivitis, episcleritis, keratitis, scleritis, uveitis, and retinal vasculitis, can occur in approximately half of the patients. Here, the patient presented with chronic nasal symptoms and recurrent episcleritis, and histopathological granulomatous manifestations of nasal mucosa without evidence of lung and kidney disease, and demonstrated the presence of limited form GPA with absence of ANCA. Histopathological examination is essential for diagnosing, particularly in ANCA-neg-
ative patients; however, the absence of ANCA does not exclude this diagnosis. ANCA may not be present in cases of limited form of GPA (7). Current treatment comprised corticosteroid and cyclophosphamide until disease remis-
sion, followed by a less toxic immunosuppres-
sant, such as azathioprine (8).

Clinical presentation of primary vasculitis can
be variable; therefore, careful attention needs
to be paid to patient’s anamnesis, clinical ex-
amination, and laboratory findings. Our aim was
to show the importance of nasal bloody crusting in the diagnosis of vasculitis.

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Transient osteoporosis of the hip

Joe Thomas¹, Kurian Ninan²

A 31-year-old male presented with pain in the right hip on activity which had persisted for the last 2 weeks. He denied any other joint pain and did not have any other co-morbid illness. On examination, range of hip movements was found to be painful in all directions. His investigation revealed normal acute phase reactants. Magnetic resonance imaging (MRI) revealed diffuse T2w-hyperintense signal in the head and neck of the right femur which is consistent with marrow edema (Figure 1a). No fracture or collapse of the femoral head or joint effusion was observed. These changes were consistent with transient osteoporosis of hip joint. Patient was advised conservative treatment and his symptoms completely subsided within 4 weeks. Follow-up MRI performed 2 months later showed complete resolution of the marrow edema in the right femoral head and no residual subarticular bone changes were observed (Figure 1b). The transient osteoporosis of hip (TOH) is an idiopathic and self-limiting disorder which is characterized by unexplained hip pain and was first reported by Ravault (1947) followed by Curtiss and Kincaid in 1959 (1). The TOH has been reported more frequently in healthy middle-aged males with a male:female ratio of 3:1 (2). The etiopathogenesis of TOH may include microvascular injury, nontraumatic reflex sympathetic dystrophy, metabolic factors, viral infection, neurological factors, and endocrine factors (3). An MRI is a sensitive test for diagnosing TOH and was described first in the radiology literature by Bloem (4). TOH is a self-limiting disease, a symptomatic and supportive treatment is recommended, and TOH should be included in the list of differential diagnoses of acute onset of hip pain.

Figure 1. a, b. Magnetic resonance imaging (MRI) showing diffuse T2w hyperintense signal involving the head and neck of the right femur which is in keeping with marrow edema (a); complete resolution of the marrow edema in the right femoral head and no residual subarticular bone changes (b)

Informed Consent: Written informed consent was obtained from the patient who participated in this study.

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References
Unilateral shortening of third metacarpal bone in a patient with tuberous sclerosis

Ahad Azami, Afshin Habibzadeh

A 33-year-old woman with a history of hypothyroidism and celiac disease presented with symmetric inflammatory polyarthritis. She had no history of seizure. She presented with multiple angiofibroma on the left side of the face and in the back (Figure 1a, b) with bone cysts in phalanxes, which were indicative of tuberous sclerosis with normal neurologic findings. Kidney ultrasonography showed cortical nephrocalcinosis. We observed shortening of the third metacarpal of the left hand during physical examination and radiography (Figure 2a, b). Inflammatory tests showed positive ANA=39.1 IU/mL (using ELISA with normal range <20), CRP: 3+, and ESR: 52. She had no history of hand trauma or surgery. After full evaluations and treatment of arthritis, the patient was discharged symptom free with normal inflammatory markers.

Although different bone manifestations, including bone cysts in the phalanxes of the hands and feet, sclerotic lesions, and periosteal new bone formation (1), are reported for tuberous sclerosis, the shortening of the third metacarpal has not been mentioned. The shortening of MCP may be a part of a syndrome acquired due to a disease during the childhood or idiopathic and is usually reported in pseudohypoparathyroidism (2, 3). In unilateral short MCP cases, we should evaluate the possible childhood injury, osteomyelitis, and infections of epiphysis, which we could not exclude in our case (4). Short third MCP alone has not been reported previously. It is possible for short third MCP to be an incidental finding or another form of presentation in tuberous sclerosis.
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References

Figure 2. a, b. Third metacarpal of the left hand during physical examination and radiography
Acro-osteolysis
Joe Thomas, Jewel Jose

A 45-year-old woman presented with long-term Raynaud’s phenomenon, esophageal dysmotility, sclerodactyly, telangiectasia, and shortened fingertips (Figure 1). Examination revealed positive anticentromere antibody (ACA), with no evidence of pulmonary hypertension and interstitial lung disease. Radiography of her hands revealed the resorption of the distal phalangeal tufts (acro-osteolysis) and soft tissue calcifications (calcinosis cutis) (Figure 2). Acro-osteolysis is a characteristic of systemic sclerosis (SSc) and has been estimated to occur in approximately 20%-25% patients (1, 2). The pathogenesis of acroosteolysis in SSc is not well understood, and presumed mechanisms include a reduction of vascular supply, compression from skin tightening, and impaired angiogenesis, among many others.

Figure 1. Long-term Raynaud’s phenomenon, esophageal dysmotility, sclerodactyly, telangiectasia, and shortened fingertips

Figure 2. Radiography of the resorption of the distal phalangeal tufts (acro-osteolysis) and soft tissue calcifications (calcinosis cutis)
Informed Consent: Written informed consent was obtained from the patient who participated in this study.

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References
Disseminated hollow and solid lung nodules as a unique pulmonary manifestation of rheumatoid arthritis

Wolfgang Jungraithmayr¹, Njanja Enz¹, Frank Lippek²

Pulmonary round nodules can represent various diseases (1). Moreover, rheumatoid arthritis can cause lung infiltrations that usually present as irregularly shaped formations (2). Here, we report a unique case of disseminated hollow pulmonary nodules in the context of rheumatoid arthritis.

The patient was diagnosed with seropositive rheumatoid arthritis in 2016. The patient complained of pain and swelling in the feet and knees. Treatment with prednisolone (35 mg/d), lodostrate (5 mg/d), and methotrexate (15 mg/week) was initiated. Upon deterioration of symptoms, the patient was administered with an additional therapy with the monoclonal antibody against interleukin (IL)-6 receptor. Computed tomography of the thorax revealed multiple round-shaped nodules of different size that were peripherally and centrally located within lung parenchyma (Figure 1a, b). Some nodules were solid, some hollow, representing a "ring shape," and some were in between these morphologies (Figure 1a, arrows). Respiratory symptoms were absent. Thoracoscopic resection of the three types nodules (Figure 2a) revealed necrotizing, granulomatous inflammation with central necrosis and margins containing epithelioid cells, fibroblasts, lymphocytes, and histiocytes (Figure 2b). Malignancy, tuberculosis, or other infections, such as fungus and bacteria, were ruled out by Grocott and Ziehl-Neelsen and Gram staining along with PCR. The patient continued to receive weekly treatment with IL-6R antagonist and is free of symptoms until now.

In conclusion, the unusual presentation of solid and hollow pulmonary nodules, normally highly suspicious of metastases of cancer, may be induced by rheumatoid arthritis and may be resolved by treatment with IL-6 inhibitor (3). Nevertheless, these nodules must undergo a work-up, including nodule resection, particularly in the presence of a positive smoking history.

Figure 1. a, b. Lung nodules in preoperative CT. Some nodules were solid (a,*), some hollow (b), and some nodules changed from solid to hollow (white arrow)

Figure 2. a, b. Thoracoscopic view and section (a) of a solid nodule in Segment 6 on the right side, corresponding to the nodule shown in Figure 1a; histology (H&E) revealed necrotizing, granulomatous inflammation with central necrosis and margins containing epithelioid cells, fibroblasts, lymphocytes, and histiocytes (b) (40x magnification)
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