

# A cervical rib presenting as a hard, immobile lump in the neck

Angel Checa 💿

A cervical rib, also called as an accessory or a supernumerary rib, is a congenital anomaly that usually develops from the seventh vertebra and extremely rarely from the fifth or sixth vertebra. It may blindly end up in the soft tissues of the neck, articulate with the first thoracic rib and sternum, or connect to a fibrous band. Cervical ribs can bilaterally appear in approximately 50% of patients. The presence of two ribs on the same side is an unusual finding (1).

A 53-year-old Hispanic woman with a history of fibromyalgia visited our rheumatology office for a second opinion about a lump in the neck. Although she detected the mass several years ago, it did not cause any symptoms. Two months ago, the patient visited her primary doctor for medical advice because her family was concerned about any malignancy. At the time of presentation to the rheumatology office, the patient denied fatigue, weight lost, fever, and local manifestations including pain or difficulty to swallow. Symptoms related to regional conditions such as upper limb pain, paresthesia, coldness, or color changes in the fingertips were not experienced.

On physical examination, a firm, non-pulsatile, non-tender, and immobile rounding mass of approximately 1.5 cm2 was noted at the anterior triangle of the right side of the neck. No regional enlarged lymph nodes or satellite nodules were observed. Phalen's and Tinel's test were both normal. Adson's maneuver test and the Upper Limb Tension Test (Elvey's) were both negative. Several studies were prescribed at a previous visit by the primary doctor including hemoglobin level, white cell count, erythrocyte sedimentation rate, thyroid panel (TSH, free T4, and T3), vascular carotid Doppler, and thyroid ultrasound, which showed no abnormality.

Ultrasonography of the neck protuberance at the rheumatology office revealed a hyperechoic enhancement with acoustic shadowing, consistent with a bony pattern. This was immediately followed by a simple anteroposterior radiograph of the chest, which confirmed the bony structure at the seventh cervical rib on the right side (Figure 1).

Based on results of radiographies, the incidence of supernumerary ribs is approximately 0.05%-3% (2). These ribs are observed more commonly in females than in males. In a report that included 5000 chest X-rays, these ribs were found in 70% females and 50% males (3, 4).

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Regarding clinical manifestation, cervical ribs are often asymptomatic throughout life. In patients who required surgery due to thoracic outlet syndrome, only 8%-10% had a supernumerary rib (3, 5). Leong and Karkos (6) described a woman in Singapore with a hard lump in the neck due to a protruding rib without any other symptoms. A teenage male with a similar clinical feature was recently reported (2). The occurrence of a palpable mass has been overlocked, perhaps as a result of physical examination based on neurovascular clinical maneuvers. However, among 19 patients with a thoracic outlet syndrome due to a cervical rib, Brannon (7) found that 68% had a firm mass in the neck.



**Figure 1.** A supernumerary rib (arrow) is emerging from the last cervical vertebral body on the right side of the cervical spine.

The presence of an indolent firm mass in the neck

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should alert clinicians about the possibility of a cervical rib in the lower cervical spine, even in cases without any symptoms of neurovascular compression. Ultrasound, in addition to physical examination, can be helpful as a first diagnostic step to confirm the bony prominence in the neck region.

Omitting or postponing a simple diagnostic test using modern tests, such as in this case, can cause a tortuous and delayed diagnosis process. Despite the extreme usefulness of novel imaging modalities in the assessment of symptoms due to a supernumerary rib (8, 9), a simple chest radiograph is still the first step in the diagnosis of a cervical rib. Peer-review: Externally peer-reviewed.

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# Symmetrical cutaneous ulcers: Are they associated with severe disease in children with juvenile dermatomyositis?

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A 4-year-old girl presented with difficulty in climbing stairs, getting up from supine position, and raising arms above the shoulder for 1.5 years. She also reported rash over face and dorsum of hands. There was worsening of symptoms in the last 1 month with fever and difficulty in holding neck. Physical examination revealed a heliotrope rash (Figure 1a); healed Gottron papules over bilateral metacarpo-phalangeal and proximal interphalangeal joints; weakness of the proximal limb, trunk, abdomen, and neck flexor muscles; and weak gag reflex. Symmetrical punched-out ulcers overlying bilateral scapula were also noted (Figure 1b). However, no calcinosis was noted. Laboratory investigations revealed the following: hemoglobin, 105 g/L; white blood cell count,  $12.9 \times 10^{9}$ /L; platelet count,  $352 \times 10^{9}$ /L; C-reactive protein, 3.4 mg/L; erythrocyte sedimentation rate, 68 mm in first hour; alanine aminotransferase, 70 IU/L; aspartate aminotransferase, 64 IU/L; lactate dehydrogenase, 561 U/L; and creatine kinase, 73.8 IU/L. Antinuclear antibody by indirect



Figure 1. a, b. (a) Heliotrope rash and Symmetrical punched-out ulcers overlying bilateral scapula in a girl with juvenile dermatomyositis (b).

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immunofluorescence was negative. Magnetic resonance imaging revealed diffuse hyperintensities involving bilateral thigh muscles. She was diagnosed with juvenile dermatomyositis (JDM) and received five doses of intravenous methylprednisolone followed by tapering doses of oral prednisolone and subcutaneous methotrexate. Her symptoms persisted, and she received three doses of monthly intravenous immunoglobulin (1 g/kg body weight), intravenous cyclophosphamide (500 mg/m<sup>2</sup> body surface area/month) for six doses, and mycophenolate mofetil (1000 mg/m2 body surface area/day). Informed written consent was obtained from the caregiver.

JDM is the most common childhood inflammatory myopathy. Cutaneous ulcers occur in 5%-30% of children with JDM and are often associated with severe disease (1). These are associated with significant pain and can develop secondary bacterial infection. In children with JDM and cutaneous ulcers, the clinical course is marked by frequent muscle and skin relapses (2). These children show unresponsiveness to routine medical management and often require multiple immunosuppressants for disease control (1, 3). Development of cutaneous ulcers in patients with JDM should make the treating physician alert regarding a severe disease course in these children.

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## Groove sign: Heeding clue to eosinophilic fasciitis

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**Figure 1.** Groove sign (white arrows): Linear depressions along the course of the veins on the forearm, better visualized with the forearm elevated.

A 34-year-old male farmer presented with polyarthralgia involving the small and large joints of the upper and lower limbs with early morning stiffness lasting 20-30 min for the past 2 months. He complained of bounding of skin involving the limbs and trunk along with difficulty in rising from a sitting posture for the last 1 month. He had no history of fever, Raynaud's phenomenon, dyspepsia, dyspnea, symptoms of sicca, or pentazocine abuse. Examination revealed pitting edema of the legs and skin thickening in the arms and legs up to the elbow and knee joints, respectively, with trunk involvement. Skin thickening was absent on the face and digits. Both forearms exhibited linear depression along the superficial veins, which was more prominent upon elevation of the arms, suggestive of groove sign (Figure 1). Laboratory test results revealed peripheral eosinophilia (E=12%)

on differential leucocyte count, with a total leukocyte count of 9000 cells/mm<sup>3</sup>. Skin biopsy revealed mild eosinophilic infiltrate of the vessels with few macrophages infiltrating the fascia, suggestive of eosinophilic fasciitis (EF). He was treated with prednisolone (40 mg/day) and methotrexate (15 mg/week). After 1 month of treatment, he showed improvements in joints pain, muscle weakness, and skin thickening.

In 1974, Schulman described the first case of EF (also known as Shulman Syndrome), which closely mimicked scleroderma (1). EF is a rare fibrosing condition of fascia characterized by skin indurations caused by predominant eosinophilic infiltration, peripheral eosinophilia, edema, and progressive muscle weakness involving the extremities. Groove sign is a classical and characteristic feature of EF and can be observed as a depression along superficial veins, best visualized when the limbs are elevated. Limb elevation results in decreased peripheral venous pressure, and the superficial skin is tethered inward, accentuating the depressions along the course of the veins, which is caused by sparing of epidermis and upper-dermis layers by a fibrotic process. EF is often misdiagnosed, leading to delays in management. Identification of the groove sign, which is characteristic of EF, can help establish diagnosis (2).

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Images in Rheumatology

# Tuberculosis monoarthritis of the wrist mimicking rheumatoid arthritis

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A 78-year-old man presented with a 10-month history of pain and swelling in his left wrist. Medical history was negative for preceding infections or trauma. Physical examination revealed a swollen and tender wrist joint. Erythrocyte sedimentation rate (32 mm/h) and C-reactive protein level (16 mg/L) were increased. Tests for rheumatoid factor, anti-cyclic citrullinated peptide, and anti-nuclear antibody gave negative results. Synovial aspiration showed increased leukocyte count (8200/mm<sup>3</sup>) without crystals. X-ray showed juxta-articular osteoporosis, erosions, and narrowing of the intercarpal joint space (Phemister's triad; Figure 1a). Computed tomography showed multiple osteolytic lesions in the carpal bones and calcifications in soft tissue (Figure 1b). Radiocarpal dislocation was also observed (Figure 1c). Synovial biopsy showed granulomatous inflammation, and tissue culture revealed the presence of Mycobacterium tuberculosis. Chest radiograph and sputum culture findings were normal. Tuberculosis (TB) monoarthritis was diagnosed, and a four-drug anti-TB regimen was started for the patient.

Skeletal TB accounts for 5%-15% of extra-pulmonary TB cases. Articular involvement is generally monoarticular and involves weight-bearing joints such as hip and knee. Wrist joint involvement is quite uncommon and accounts only for 1% of the skeletal TB cases (1). Joint findings were non-specific; however, Phemister's triad, including periarticular osteoporosis, joint space narrowing, and erosions, could be seen (2). Our case is a good example of TB monoarthritis. The striking radiographic features that helped us in diagnosis were Phemister's triad and cold abscess formation in the soft tissue. In our patient, 9 months of TB treatment resulted in symptomatic control of pain and swelling. However, due to the extensive erosions, wrist joint movement was restricted with some functional impairment.

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**Figure 1. a-c.** X-ray of the wrist, showing juxta-articular osteoporosis (white arrow), erosions (arrowhead), and narrowing of the intercarpal joint space (black arrow) (a); computed tomography revealed multiple osteolytic lesions in the carpal bones (arrowheads) and calcifications in the soft tissue, suggesting cold abscess formation (highlighted in ellipse) (b); radiocarpal dislocation was also observed (black arrow) (c).

### Uslu et al. Wrist monoarthritis of tuberculosis

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### Raccoon eyes in cutaneous neonatal lupus syndrome

Kavitha Mohanasundaram<sup>1</sup> , Ashok Govindaraj<sup>2</sup>

Neonatal lupus syndrome occurs in babies of mothers who are anti-SSA and/or anti-SSB positive; most mothers are often asymptomatic. It can also occur in neonates born to mothers with Sjogren's syndrome, lupus, or undifferentiated connective tissue disease (1). The child can present with erythematous rashes in the peri-orbital region called as raccoon eyes, congenital heart block, thrombocytopenia, and transaminitis. It is often termed as cutaneous neonatal lupus syndrome if the presentation is limited to skin alone.

A 4-week infant was referred from a pediatrician to a rheumatologist for ruling out lupus considering skin rashes on the face. The infant was examined and found to have erythematous rashes over the face; she was active, and her milestones were appropriate for the age (Figure 1). She was delivered through a normal vaginal delivery. Her mother was asymptomatic and denied treatment for any rheumatological illness; this was her first pregnancy. The child was diagnosed with raccoon eyes, suggestive of neonatal lupus syndrome.

Investigations done on the infant before referral showed a normal blood count; indirect immunofluorescence showed antinuclear antibodies (ANA) positive at 1:80 dilution with a speckled pattern. ECHO and ECG performed on the infant showed normal results; her C3 and C4 levels were 115 mg/dL and 25 mg/ dL, respectively. We advised the mother to undergo immunological workup; mother had ANA positivity at 1:160 dilution with a speckled pattern. On further workup, she had a high positive anti-SSA (Ro 52 and Ro 60) and anti-SSB. The infant was followed up regularly and advised mild low-potency steroid application along with limitation of sun exposure. At the end of 9 months, we observed complete resolution of the erythematous lesions with mild hypopigmentation, and her repeat ANA was negative (Figure 2).

This infant presented only with cutaneous manifestation and did not have any internal organ involvement. It is ideal to look for anti-SSA/SSB antibodies in mothers since fetal monitoring during the next pregnancy

becomes an absolute necessity. ANA, since it is of Ig G type, can be transferred from mother to fetus, and ANA positivity in this infant could be attributed to such a passive transmission. Repeat ANA testing is recommended at 6-9 months of age.

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**Figure 1.** Erythematous lesions in the periorbital region - Raccoon eyes.



**Figure 2.** Complete resolution of cutaneous lesions leaving behind mild atrophic scars.

### Mohanasundaram and Govindaraj. Raccoon eyes in NLS

Most infants born to anti-SSA positive mothers are born without any major abnormalities. Complete heart block, the most dreaded complication of neonatal lupus syndrome happens in 2% of anti-Ro-positive pregnancies; the recurrence of which rises to 20% in subsequent pregnancies. Fetal monitoring starting at week 16 becomes mandatory to detect early conduction abnormalities, some of which may be reversible (2).

The cutaneous manifestations of neonatal lupus syndrome are usually reversible, occur more frequently, and resemble those of subacute cutaneous lupus erythematosus lesions. They occur in the first 8 weeks of birth, though rarely they can be observed at birth itself. They resolve without scarring at around 6 months, often coinciding with the disappearance of maternal anti-Ro and -La antibodies from the infant (3).

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Images in Rheumatology

## Lupus with "thumb-prints," "targets," and "combs"

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A 28-year-old woman was undergoing evaluation for proteinuria (urine protein/creatinine ratio=9.6 [normal<0.3]). Two days following kidney biopsy, she presented to the emergency department with abdominal pain, vomiting, and obstipation. In the previous 3 months, she had experienced frequent episodes of abdominal pain, which used to subside spontaneously. She also had a history of intermittent fever and passage of frothy urine. Examination revealed distended abdomen with rebound tenderness and absence of bowel sounds. Reduced breath sounds were noted over the infrascapular area on both sides. Laboratory test results were as follows: hemoglobin, 9 g/dL; total leukocyte count, 9779 cells/mm<sup>3</sup>; erythrocyte sedimentation rate (ESR), 76 mm in the first hour; positive anti-nuclear antibody (1:640, homogeneous); C3, 61 mg/dL; and C4, 8 mg/dL. An erect radiograph of the abdomen excluded any bowel perforation. Computed tomography of the chest and abdomen revealed pleural effusion on both sides with diffuse thickening and edema of the bowel wall giving rise to "thumb-printing" (Figure 1) and "target" sign (Figure 2). The mesenteric vasculature ("comb" sign) and ascites were accentuated (Figure 2). Kidney biopsy revealed Class IV lupus nephritis. The patient was diagnosed with systemic lupus erythematosus (SLE) with nephritis and mesenteric vasculitis and was treated with high-dose pulsed methylprednisolone followed by monthly pulses of intravenous cyclophosphamide and tapering doses of oral prednisolone. Her bowel symptoms improved remarkably after 4 days of treatment. At the 6-month follow-up, she was relieved of abdominal symptoms and her ESR and proteinuria had normalized. She was started on maintenance therapy with azathioprine (2 mg/kg).

Mesenteric vasculitis is a rare manifestation of lupus, which may present with abdominal pain, intestinal obstruction, abdominal distension, or gastrointestinal bleeding. In patients with SLE, mesenteric vasculitis is the most common cause of severe abdominal pain and requires admission (1). Signs of peritonitis can be observed in some cases, which mandates ruling out intestinal perforation. It is commonly associated with evidence of active SLE in other organ systems (2). It can be fatal if not diagnosed and treated promptly with glucocorticoids, to which good response is frequently noted (1, 3). Cyclophosphamide has been successfully administered in steroid refractory cases (3).

Small vessels of the bowel wall, which are involved in lupus mesenteric vasculitis, cannot be visualized using mesenteric angiography. However, angiography can be performed on suspicion of involvement of larger arteries, which occurs in polyarteritis nodosa, atherosclerosis, or thrombosis. In addition, the most common site of involvement is the small bowel, which is not easily amenable to biopsy. Because of these limitations, computed tomography of the abdomen is the most useful modality for diagnosis. It reveals characteristic features such as edema of the bowel wall, giving rise to the "target" sign on cross-sections (4). Thumb-like

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**Figure 1.** Contrast-enhanced CT of the abdomen showing thumb-like projections of the intestinal mucosa into the lumen: "thumb-print" sign (aster-isk).



Figure 2. Contrast-enhanced CT of the abdomen showing edema of the bowel wall: "target" sign (black asterisks); prominence of mesenteric vasculature: "comb" sign (white asterisk) and ascites (star).

### Kumar et al. Lupus with mesenteric vasculitis

projections of the intestinal mucosa into the lumen appear as "thumb-printing" on longitudinal sections, which indicate bowel wall ischemia (4). Prominence of mesenteric vessels resembles the teeth of a comb ("comb" sign) (4). Prompt treatment is necessary to prevent complications such as pneumatosis intestinalis and bowel perforation.

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## Hypercholesterolemic arthritis in a young female

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A 30-year-old female presented with arthritis of the small joints with swellings on the proximal inter phalangeal joints since 18 months (Figure 1). She with normal Erythrocyte Sedimentation Rate (ESR) and C-reactive protein levels; her rheumatoid factor, anti-cyclic citrullinated peptide, and antinuclear antibody (ANA) tests were negative. Ocular examination revealed a grayish-white ring at the periphery of cornea, which indicated corneal arcus (Figure 2). Lipid examination showed the following: total cholesterol, 325 mg/dL (normal value <200 mg/dL) and low density lipoprotein cholesterol, 271 mg/dL (normal value <130 mg/dL), with normal serum triglyceride and high density lipoprotein levels. The biopsy sample obtained from skin nodules revealed numerous foam cells in the dermis, and this was consistent with the findings of xanthomas (Figure 3). Plain radiographs of the hands did not show erosions (Figure 4). Her family history was negative for similar illnesses. She received atorvastatin 20 mg/day and paracetamol; 2 months later, her lipid levels returned to normal



**Figure 1.** Swelling over the proximal inter phalangeal (PIP) joints.



Figure 2. Gray white ring at periphery of cornea: corneal arcus.

levels with significant improvement in arthritis, but corneal arcus and xanthomas persisted. She was diagnosed with arthropathy related to dyslipidemia. Other rheumatologic features include recurrent achilles pain or tendinitis, acute mono/oligoarthritis, and migratory (rheumatic fever-like) polyarthritis (1). The symptoms might resemble those of rheumatoid



Figure 3. Photomicrograph of lower dermis with numerous foam cells (lipid-laden macrophages) within the dermal collagen (H&E, 400×).



Figure 4. Plain radiographs of hands with soft tissue swelling on PIP joints.

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### Padhan et al. Hypercholesterolemic arthritis

arthritis with rheumatoid nodules, indicative of tophaceous gout. Unlike xanthomas, rheumatoid nodules are often found over the extensor surfaces, but they are almost invariably associated with circulating RF and a characteristic histological appearance (2). Hyperlipidemia should be considered in a patient presenting with articular symptoms with normal ESR and CRP.

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### Eur J Rheumatol 2020; 7(2): 96-7



## A case of parotid sarcoidosis

Sajal Ajmani 🕩

A 51-year-old female presented with right parotid enlargement for 2 months. She had no other complaints such as dry eyes, dry mouth, shortness of breath, or skin lesions or any other lump in the body. She was a known diabetic and also had history of Bell's palsy 3 years ago. On examination, she had a firm, diffuse, and nonnodular right parotid gland swelling (Figure 1). Her serum angiotensin-converting enzyme (ACE) levels were elevated 198 (66-114) U/L, ESR (Erythrocyte Sedimentation Rate) was 43 mm/hr, Mantoux test was negative, and serum calcium level was 9.6 (8.8-10.6) mg/dL. A fine needle aspiration cytology from the parotid gland showed granulomatous parotitis. MRI showed (Figure 1) enlarged right parotid gland with multiple ill-marginated enhancing lesions. Focal similar enhancing lesions were also seen in the left parotid gland along with enlarged mediastinal lymph nodes. A diagnosis of sarcoidosis was made, and she was started on oral prednisolone 40 mg, which was subsequently tapered over the next 6 months. With treatment, the parotid enlargement resolved and the serum levels normalized. In a population-based cohort, only 2% (7 out of 345) patients with sarcoidosis had parotid gland involvement (1), wherein parotid gland disease was usually painless, unilateral, and associated with intrathoracic disease. It was the initial presentation in 4 patients. The ACE level was elevated in 25% of patients, while none had hypercalcemia. Gland swelling regressed after steroid treatment in all patients, although one patient had relapse. Written informed consent and publication consent was obtained from the patient.



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**Figure 1. a, b.** (a) clinical image (b) T2 weighted coronal sections showing enlargement of bilateral parotid glands (right>left) with focal areas of T2 hyperintensities within the substance of the gland (arrow).

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## Immune-mediated necrotizing myopathy associated with antibodies to the signal recognition particle: A rare cause of hyperCKaemia

Elsa Meireles<sup>1</sup>, Joana Malheiro<sup>1</sup>, Ricardo Taipa<sup>2</sup>, Manuela Alves<sup>1</sup>

A 73-year-old Caucasian male patient presented to the emergency department with 3 months evolution of severe progressive proximal weakness and atrophy. Weakness was characterized by difficulty in standing up from sitting position, combing hair, and changing clothes. No history of dysphagia, dyspnea, skin rashes, or weight loss was noted. Our patient was not taking any myotoxic drugs. Clinical examinations revealed right upper and lower limb proximal and distal muscle weakness and atrophy, with a clinical power grade of 4/5. Laboratory investigation revealed an elevated sedimentation rate of 61 mm/h, with markedly elevated serum creatine kinase (CK) levels, 9562 (normal range 30-200) units/L. Antisignal recognition particle (SRP) was positive. Electrophysiologic studies evidenced diffuse myopathy with fibrillations. Necrotic fibers and regeneration, in the absence of inflammatory infiltrates, were seen in the muscle biopsy (Figure 1. a-d). The patient was initially started on prednisolone 1 mg/kg/ day. His weakness improved, with a CK level of 2198 U/L at 1 month after beginning the treatment. Steroid-sparing agent was initiated with azathioprine 2 mg/kg/day, and 1 month after, he achieved a steady functional recovery.

Immune-mediated necrotizing myopathy (IMNM) is a rare clinicopathologic entity that is composed of three serologically subtypes: antihydroxy-3-methylglutaryl-CoA reductase (HMGCR) myopathy, anti-SRP myopathy,



**Figure 1. a-d.** Left deltoid muscle biopsy showed scattered necrotic fibers without associated inflammation (a, b). There was regenerating fibers (c) and upregulation of sarcolemmal MHC class I (d). a) H&E, b) Gomori trichrome, c) Fetal myosin; d) MHC class I

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### Meireles et al. Immune-mediated necrotizing myopathy

and autoantibody-negative IMNM (1). However, only 10% of the cases have positive antibodies (1). Unlike the other subtypes, anti-SRP myopathy is not associated with cancer (2, 3) and has characteristically poor responsiveness to steroid monotherapy and others immunosuppressive therapies (4). In conclusion, we present a case of a patient with an anti-SRP antibodies-associated inflammatory myopathy, a rare diagnosis that should be considered in cases of elevated CK levels that do not resolve with appropriate management and exclusion of other causes of rhabdomyolysis.

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## Bilateral retinal detachments presenting as manifestations of Vogt-Koyanagi-Harada syndrome

Sidney Ariel Schechet D, Rahul Komati, Seenu M. Hariprasad D

A 38-year-old African-American female patient with a past medical history of hypertension presented with blurry vision, headache, and neck stiffness for 1 week. Her vision was decreased at 20/40 and 20/200 in the right and left eyes, respectively. She denied recent illness, skin changes, trauma, travel, sick contacts, or family history of diseases. Dilated fundus examination revealed bilateral panuveitis with serous retinal detachments (Figure a, b; white tracing outlines the subretinal fluid). Laboratory and imaging findings were unremarkable, and the patient was diagnosed with Vogt-Koyanagi-Harada (VKH) syndrome. She was treated with a slow taper of ophthalmic and systemic steroids and subsequently exhibited complete recovery of ocular (Figure c, d) and systemic findings. VKH syndrome is a systemic disease that can present with hearing loss, meningismus, vitiligo/poliosis, and typically bilateral uveitis and serous retinal detachments. High-dose systemic corticosteroid therapy is the gold standard for treating this syndrome; however, other immunomodulatory agents may be needed in refractory cases (1, 2).



**Figure 1. a-d.** Images of the fundus in the right (a) and left (b) eyes at presentation outlining the significant serous retinal detachments. After 1 month of initiating corticosteroid therapy, the retinal detachments resolved in both the right (c) and left (d) eyes.

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### Ulcerated tophaceous gout

Jaclyn Rosenthal<sup>1</sup>, Robert J. Smith<sup>2</sup>, Megan H. Noe<sup>2</sup>

A 51-year-old male presented with ulcerated nodules over the bilateral elbows. These nodules had been present for >10 years; however, 3 weeks prior to presentation, they became more painful and chalky material started to drain from them. At that time, the patient was prescribed a short course of prednisone, which led to mild subjective improvement in swelling and pain. On presentation, physical examination of the right elbow revealed a tender, deep-red, exophytic nodule with central ulceration surrounding deposits of solid, yellow-white chalky material (Figure 1). On the left elbow, there was a similar tender, deep-red firm plaque that distally extended to the forearm with central ulceration and chalky debris. The patient exhibited limited extension and flexion of the left elbow. Additionally, there were several yellow-orange nodules overlying the small joints of the bilateral hands and wrists. An X-ray of the elbows revealed significant swelling with foci of mineralization. An arthrocentesis and culture of the left elbow identified uric acid crystals without bacteria. In this patient with a history of tophaceous gout, the clinical presentation was most consistent with ulcerated tophaceous gout. The patient was instructed to continue the dose of allopurinol and surgical excision and debridement of the gouty tophi on the left elbow was performed.

Tophaceous gout is a rare condition characterized by the subcutaneous accumulation of monosodium urate crystals in a matrix of lipids, proteins, and mucopolysaccharides, surrounded by giant cell inflammation (1, 2). Ulceration is rare and occurs when tophi break through the skin. The most commonly reported sites of ulcerations are over the first metatarsophalangeal joints as well as on the joints of fingers, wrists, elbows, and ankles. Comorbidities that impair wound healing, such as diabetes mellitus, peripheral vascular disease, and hypertension, are commonly noted in patients with ulcerated tophaceous gout (1). A Significant morbidity is associated with this condition considering the extent of pain, impaired mobility, and risk of secondary infection (3).

Although tophaceous gout is often clinically diagnosed, when ulceration occurs, arthrocentesis of synovial fluid is necessary to rule out a secondary septic arthritis. Histopathology of a gouty tophus demonstrates palisaded granulomas surrounding amorphous, gray-blue material with a feathery appearance. Because the ulceration of a tophus is rare, there are no standard guidelines for its treatment. In case reports and small case series, several medical and surgical treatment modalities have been demonstrated to expedite the resolu-

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tion of pre-existing ulcers as well as prevent superimposed infections of pre-existing ulcers and future ulcer formation (1). Medical management includes long-term antihyperuricemic agents (i.e., allopurinol and probenecid) and topical wound care aimed at preventing secondary infection and promoting healing (i.e., mupirocin, 3% citric acid in petroleum jelly, allogeneic culture dermal substitute, silver-containing dressing, and heterologous lyophilized collagen) (1). Surgical management should be considered for patients with infection of tophi, mechanical impairments, uncontrollable pain, and cosmetic disfigurement (3). Intralesional shaving, hydrosurgery, and skin grafting are the most commonly performed surgical interventions (4). Curettage and debridement are used less frequently due to high rates of delayed wound healing and skin necrosis (5).

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**Figure 1.** Ulcerated tophaceous gout involving the right elbow joint.

An elbow with a deep-red, exophytic nodule with central ulceration surrounding a deposition of solid, yellow-white chalky material is shown.

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### Rosenthal et al. Ulcerated tophaceous gout

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# Multiple embolic strokes in primary antiphospholipid syndrome

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A 58-year-old female was brought to our institution with an ischemic stroke. A holosystolic murmur in the fifth intercostal space in the mid-clavicular line was heard on cardiac auscultation. Cranial magnetic resonance imaging (MRI) revealed a sub-acute stroke in the posterior cerebral artery and multiple images related to previous embolic events (Figure 1). Treatment with atorvastatin and rivaroxaban was initiated for ischemic stroke. A transthoracic echocardiogram and a transesophageal echocardiogram (TEE) showed nodular thickening in both mitral leaflets and the sub-valvular apparatus with moderate mitral regurgitation. Moreover, small nodules were observed on the vascular side of the aortic valve. Mild tricuspid regurgitation was observed, and the pulmonary artery systolic pressure was normal (32 mm Hg). The patient had normal biventricular systolic function with a left ventricular ejection fraction of 65% and tricuspid annular plane systolic excursion of 19 mm (Figures 2 and 3). The laboratory examination revealed anemia (hemoglobin: 9.9 g/dL, hematocrit: 29.1%); no leukocytosis; and concentration of D-dimer: 10,000 ng/mL, protein C: 150 mg/dL, protein S: 80%, C3: 145.8 mg/dL, C4: 29.7 mg/dL, anti-cardiolipin IgA: 20 A phospholipids (APL), anti-cardiolipin IgM: 15 M phospholipids (MPL), anti-cardiolipin IgG: 19 G phospholipids (GPL), anti-B2GP1: positive, anti-dsDNA: 10 IU/mL, anti-nuclear positive and increased IgA: 505.5 mg/dL, and a heterozygous mutation in MTHFT (WT/C677T) was observed. At 12 weeks, increased levels of anti-cardiolipin IgG (50 GPL) and anti-cardiolipin IgM (29 MPL) were observed. A diagnosis of antiphospholipid syndrome (APS) and mitral valvulopathy was established. The New York Heart Association classification placed the patient in functional class II; she is currently receiving medical treatment (warfarin, atorvastatin, and aspirin).

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It is observed that 70% of patients with APS have at least 1 valvular lesion diagnosed through echocardiography (valvular thickening, stenosis, regurgitation, or non-infective vegetation). However, the most important manifestation is Libman-Sacks endocarditis (LSE), primarily in the mitral and aortic valves (1). TEE detects cardiac involvement in 75.9% to 82% of patients (2). The diagnosis requires the presence of non-infective vegetation on the echocardiogram and the exclusion of infective endocarditis (3). The prevalence of stroke in a patient with APS is 19.8% (2). Patients with APS and stroke should be evaluated using



**Figure 1. a-c.** Cranial MRI. Hyperintense lesions in the right posterior cerebral artery territory that compromises the ipsilateral temporal region. T2-weighted MRI (a). FLAIR (b). Multiple axial hyperintense lesions in FLAIR within the cerebellum, and right occipital-cortical and frontoparietal midline thalamic regions (white arrows) (c).

FLAIR: fluid-attenuated inversion recovery; MRI: magnetic resonance imaging.



### Garcia-Diaz et al. Stroke in primary antiphospholipid syndrome



**Figure 2. a-d.** Bidimensional and color-flow transthoracic echocardiogram in apical views. Four-chamber view showing nodular thickening on the tip of both mitral leaflets (arrows) (a). Mild to moderate mitral regurgitation and mild tricuspid regurgitation (arrows) (b). Two- (c) and three-chamber (d) views showing nodular thickening of the mitral leaflets (arrows). Ao: aortic valve; LA: left atrium; LV: left ventricle; RA: right atrium; RV: right ventricle.



**Figure 3. a-d.** Two- and three-dimensional transesophageal echocardiograms in 12° (a) and 27° (b) showing nodules in the auricular side of mitral leaflets (arrows). The image at 147° (c) with nodules in mitral leaflets (arrows) and on the vascular side of the aortic valve (arrow). Doppler color with moderate mitral regurgitation (d).

Ao: aortic valve; LA: left atrium; LV: left ventricle; RA: right atrium; RV: right ventricle.

### Eur J Rheumatol 2020; 7(4): 199-200

MRI, and intra- and extracranial vascular imaging (angiotomography). It is recommended to perform TEE in all patients with APS and stroke (4). Neuroimaging abnormalities have been reported in 35% to 90% of patients with APS. The most frequent findings were heart attacks, reported in 22% to 45.7% of patients, and white matter hyperintensities, found in 17% to 45% of patients with APS. Ischemic lesions can be small or large and involve superficial and deeper areas of the brain (2, 5).

This case is of great interest because in the sixth decade of life presented a cardioembolic stroke as the first manifestation of APS. The echocardiogram demonstrated non-infective vegetation that suggested the diagnosis of APS, which was confirmed by her immune profile.

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# Digital artery vasospasm in primary Raynaud's phenomenon

Steven Rogers<sup>1, 2</sup> , Michael Hughes<sup>3</sup>

A 60-year-old female with known primary Raynaud's phenomenon (RP) presented with an episodic unilateral attack of digital pallor change in the left index finger (Figure 1a) associated with paresthesia. Ultra-high-frequency (48 MHz) ultrasound of the affected digit revealed vasospasm of the proper digital arteries (Figure 1b), where color flow imaging shown in the longitudinal view was absent (Figure 1b). Contralaterally, the unaffected respective digit (Figure 1c) had normal color flow (Figure 1d). The attack lasted approximately 15 minutes and self-aborted.

Primary RP is a common vasospastic condition primarily affecting the digital artery. Typical attacks of the RP affect the extremities, presenting as episodic digital color changes in response to cold and/or emotional stress (1). Secondary RP can occur because of a broad range of medical and drug causes and is often the earliest manifestation seen in patients with systemic sclerosis (SSc) (1). Significant abnormalities (e.g., intimal hyperplasia and occlusion) of the digital arteries have been reported in SSc (2). Irrespective of the underlying cause, RP can have a significant impact on quality of life and function (3).

First-line treatment is patient education and behavioral adaptations (e.g., cold avoidance) (1, 4, 5). Pharmacological therapy (e.g., oral vasodilators) is indicated after the failure of general measures/behavioral adaptions. Calcium channel blockers are often used as the first-line drug treatment; however, clinicians are increasingly using phosphodiesterase type 5 inhibitors earlier for the pharmacological treatment of SSc-associated digital vasculopathy (1, 4, 5).

Patients with RP should seek emergency medical attention if they develop a permanently discolored digit and/or develop signs of tissue ulceration or gangrene.



**Figure 1. a-d.** Unilateral attack (a) of primary Raynaud's phenomenon and digital artery vasospasm as assessed by ultra-high-frequency ultrasound (b). The contralateral digit was unaffected (c) and had normal color flow (d).

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# Tenosynovial giant cell tumor mimicking an ankle monoarthritis

Francisco Vílchez-Oya D, Ana Pros D

A 24-year-old woman was referred to our clinic for suspected monoarthritis located in her left ankle (Figure 1). She denied any previous trauma and personal or familiar history of rheumatologic or infectious diseases.

Examination of the affected foot revealed swelling overlying the ankle joint, with limited of the range of motion and without redness or warmth in the examined area. There was no swelling in other locations.

Initial blood test revealed normal levels of C-reactive protein and erythrocyte sedimentation rate, negative antinuclear antibody, rheumatoid factor, anticitrullinated protein antibodies, and HLA-B27. Besides, an osteoarticular ultrasound was performed, which showed a marked thickening of the tibiotalar synovial membrane, which widely distends the anterior recess of the joint, showing a pseudotumor (Figure 2). After the clinical and initial imaging findings, nuclear magnetic resonance imaging (MRI) of left foot and ankle was performed (Figure 3). Because of the broad differential diagnosis of the detected mass, a biopsy was needed to confirm the diagnosis and rule out malignancy. The histopathological findings were compatible with a tenosynovial giant cell tumor (TGCT). Subsequently, because of the extension and the bone involvement, maximal resection of pathological tissue was done through open surgery.



Figure 1. Swelling overlying the left anterolateral ankle joint. Longitudinal scar (\*) secondary to biopsy.



**Figure 2.** Marked thickening of the tibiotalar synovial membrane, with heterogeneous and hypoechogenic appearance and diffuse increase in vascularization pattern with Doppler study. The synovial membrane widely distends the anterior recess of the joint, leading a mass effect on the extensor tendons.

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Follow-up was necessary because of the risk of recurrence in this case.

TGCT is a rare (1) but well-recognized proliferative lesion that involves the synovium, bursae, and tendon sheath. The pathogenesis is not well understood, although it has been observed that a chromosomal translocation involving 1p13 chromosome causes overexpression of the CSF1 (macrophage colony stimulating factor 1), which binds to the receptor (CSFR1) on the tumoral cell surface, leading to the expression of cells of mononuclear phagocyte lineage that constitute the tumor mass (2-4).

MRI is quite useful and shows a characteristic pattern that helps differentiate the neoplasm

from other masses. Nevertheless, a definitive diagnosis should be possible through biopsy. Regarding the treatment, classically, a surgical approach with resection has been the preferred treatment, but nowadays, medical treatment is also proposed with monoclonal antibodies inhibiting CSF1 receptors overexpressed in TGCT (2, 4, 5).

The advance in the knowledge of the etiopathogenesis of the TGCT has opened up a greater range of possibilities for a therapeutic approach beyond the classic surgical approach.

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## The view of joints in the Wilson's disease

Bengisu Aslan<sup>1</sup>, Hatice Erdoğan<sup>2</sup>, Veli Yazısız<sup>1</sup>

A 57-year-old female presented with pain in the metacarpophalangeal (MCP) joints of the wrist and knee. She was diagnosed with Wilson's disease 25 years ago. She had undergone liver transplantation, with a liver from a living donor two years ago because of cirrhosis associated with Wilson's disease. In addition, she had undergone hip replacement surgery for femoral neck fracture one and a half years ago. The patient had had muscle-joint pains for a long time. Rheumatoid factor and anticyclic citrullinated peptide tests were negative. Erythrocyte sedimentation rate and C-reactive protein levels were normal. Plain radiographs of the joints showed erosions and osteoarthritic changes. Although the view of distal and proximal interphalangeal joints was normal, irregularity of articular surfaces and joint space narrowing was observed in the MCP joints of both hands, especially in the right 3 and 4 joints. Chondrocalcinosis, bone fragmentations, changes of articular surfaces, and osteophyte protrusions were seen in the wrist joints (Figure 1). Classical osteoarthritic changes such as joint space narrowing in the medial compartments, subchondral sclerosis, and marginal osteophytes were visible in the radiograph of the knees (Figure 2a). Spine radiographs had shown spinal osteoarthrosis with Schmorl's nodes, narrowing of the intervertebral disc spaces, and osteophytes at the edges of the vertebral body (Figure 2b). Degenerative changes and irregularity of trochanters were seen on the right hip joint and the left hip joint was replaced with a prosthesis because of traumatic fracture (Figure 2c). Although some radiographic features were resembling osteoarthritis, the patient was diagnosed with arthropathy related to Wilson's disease.

Wilson's disease is a recessively inherited autosomal disease caused by mutations in the ATP7B gene, which leads to impaired copper excretion into the bile and causes a combination of hepatic, neurologic, and psychi-



![](_page_25_Figure_8.jpeg)

atric symptoms. It has a disabling and fatal course if the diagnosis is overlooked and treatment is not initiated. The estimate prevalence ratio for this disease is 1:30,000 - 1:50,000 in the USA, Europe, and Asia (1). Joint involvements and radiological abnormalities have been described in clinical case studies. Early osteoarthritic changes were reported at the joints, especially at the knee, hip, and wrist joints (2). In addition, Wilson's disease is associated with premature osteoarthritis at the joints that are unaffected in patients with classic osteoarthritis, such as the MCP joints (3). Other abnormalities include joint space narrowing in the medial compartments, subchondral sclerosis, osteophytic protrusions at bone ends, and bunches of tongue-like osteophytes at the joint margin of the knees; irregularity of femoral trochanters; and osteochondritis, reduction of intervertebral joint spaces, osteoar-

![](_page_25_Figure_10.jpeg)

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![](_page_25_Picture_20.jpeg)

### Aslan et al. Joints in the Wilson's disease

throsis, and a tendency of squaring of vertebral bodies at the spine. Although the exact pathogenic mechanisms responsible for joint changes are unknown, high levels of copper were found in synovial biopsies, and copper and sulfur deposits were found in the cartilage biopsies (4, 5).

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