Images in Rheumatology

### 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,  $-\lambda$  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. - $\lambda$  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. - $\lambda$  shows the compensator axis (b).

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#### Reference

1. Zuckner J, Uddin J, Gantner Ge Jr, Dorner Rw. Cholesterol crystals in synovial fluid. Ann Intern Med 1964; 60: 436-46. [CrossRef]

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

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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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Figure 1. a, b. Multiple angiofibroma on the left side of the face and in the back

#### Azami and Habibzadeh. Unilateral short third MCP in tuberous sclerosis



Figure 2. a, b. Third metacarpal of the left hand during physical examination and radiography

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- Leung AK, Robson WL. Tuberous sclerosis complex: a review. J Pediatr Health Care 2007; 21: 108-14. [CrossRef]
- Tzaveas A, Paraskevas G, Gekas C, Vrettakos A, Antoniou K, Spyridakis I. Anatomical variation of co-existence of 4th and 5th short metacarpal bones, sesamoid ossicles and exostoses of ulna and radius in the same hand: a case report. Cases J 2008; 1: 281. [CrossRef]
- Lodh M, Mukhopadhyay R. A Case of Primary Hypogonadism with Features of Albright's Syndrome. J Reprod Infertil 2016; 17: 188-90.
- Suresh SS, Abraham R, Ravi P. Isolated symmetrical brachymetacarpia of the thumb-case report. Hand (NY) 2009; 4: 424-6. [CrossRef]

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

Nurşen Düzgün<sup>1</sup> , Haldun Umudum<sup>2</sup>

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 rhinnore/ discharge (Figure 1) since 5 years. Her ophtalmic and ear nose, and throat (ENT) examination after admission showed episcleritis and nasal purulant crusting. There was no evidence of other organ/tissue involvement.

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Figure 1. Large nasal discharge with bloody purulent appearance

Her laboratory findings revealed systemic inflammation. Blood parameters were as follows: white blood cell count  $11 \times 103/\mu$ L, 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-

#### Düzgün and Umudum. Nasal bloody crusting and recurrent episcleritis in limited GPA

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**Figure 2.** Scanning magnification of biopsy material. Respiratory mucosa is present on upper part of biopsy. A prominent inflammatory infiltration is seen on full thickness of biopsy material (X2, Hematoxylen&Eosin).



Figure 3. Medium magnification of biopsy material shows a section of vascular structure (X20, Hematoxylen&Eosin). Fibrinoid necrosis is present on intimal part of vessel.



**Figure 4.** Higher magnification of inflammatory infiltration shows multinucleated giant cells (40X, Hematoxylen&Eosin).

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 remission, followed by a less toxic immunosuppressant, such as azathioprine (8).

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

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- Jenette JC, Folk RJ, Bacon K, Basu N, Cid MC, Ferrario F, et al. 2012 revised International Chapel Hill Consensus Conference Nomenclature of vasculitides. Arthritis Rheum 2013; 65: 1-11. [CrossRef]
- Schilder AM. Wegener's granulomatosis vasculitis and granuloma. Autoimmun Rev 2010; 9: 483-7. [CrossRef]
- Hoffman GS, Kerr GS, Leavitt RY, Hallahan CW, Lebovics RS, Travis WD, et al. Wegener granulomatosis: an analysis of 158 patients. Ann Intern Med 1992; 116: 488-98. [CrossRef]
- Leavitt RY, Fauci AS, Bloch DA, Michel BA, Hunder GG, Arend WP, et al. The American College of Rheumatology 1990 Criteria for the Wegener granulomatosis. Arthritis Rheum 1990; 33: 101-7.
- Cannady SB, Batra PS, Koening C, Lorenz RR Citardi MJ, Longford C, et al. Sinonasal Wegener granulomatosis: a single-institution experience with 120 cases. Laryngoscope 2009; 119: 757-61. [CrossRef]
- Srouji IA, Andrews P, Edwards C, Lund VJ. Patterns of presentation and diagnosis of patients with Wegener's granulomatosis: ENT aspects. J Laryngol Otol 2007; 121: 653-8. [CrossRef]
- Yoo J, Kim H, Ahn S, Jung S, Song J, Park Y, et al. Clinical and prognostic features of Korean patients with MPO-ANCA, PR3-ANCA and ANCA-negative vasculitis. Clin Exp Rheumatol 2017; 35 (Suppl 103): 111-8.
- Jayne D. How to induce remission in primary systemic vasculitis. Best Pract Res Clin Rheumatol 2005; 19: 293-305. [CrossRef]

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

Wolfgang Jungraithmayr<sup>1</sup> , Njanja Enz<sup>1</sup> , Frank Lippek<sup>2</sup>

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 and complained of pain and swelling in the feet and knees. Treatment with prednisolone (35 mg/d), lodotrate (5 mg/d), and methothrexate (15 mg/week) was initiated. Upon deterioration of symptoms, the patient received an additional therapy with a monoclonal antibody against the interleukin (IL)-6 receptor. Computed tomography of the thorax revealed multiple round-shaped nodules of different size that were peripherally and centrally located within the 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 of nodules (Figure 2a) revealed a 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 an IL-6R antagonist and is free of symptoms until now.

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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) (40× magnification)

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

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- Truong MT, Ko JP, Rossi SE, Viswanathan C, Bruzzi JF, Marom Em, et al. Update in the evaluation of the solitary pulmonary nodule. Radiographics 2014; 34: 1658-79. [CrossRef]
- 2. Sagdeo P, Gattimallanahali Y, Kakade G, Canchi B. Rheumatoid lung nodule. BMJ Case Rep 2015; 2015.
- Andres M, Vela P, Romera C. Marked improvement of lung rheumatoid nodules after treatment with tocilizumab. Rheumatology (Oxford) 2012; 51: 1132-4. [CrossRef]

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### Transient osteoporosis of the hip

Joe Thomas<sup>1</sup> , Kurian Ninan<sup>2</sup>

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)

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#### References

- 1. Curtiss PH Jr., Kincaid WE. Transitory demineralization of the hip in pregnancy. A report of three cases. J Bone Joint Surg Am 1959; 41: 1327-33. [CrossRef]
- 2. Hadidy AM, Al Ryalat NT, Hadidi ST, Tarawneh ES, Hadidi MT, Samara OA, et al. Male transient hip osteoporosis: Are physicians at a higher risk? Arch Osteoporos 2009; 4: 41-5.
- 3. Harrington S. Idiopathic TOH. Phys Sportsmed 2000; 28: 34.
- 4. Bloem JL. Transient osteoporosis of the hip: MR imaging. Radiology 1988; 167: 753-5. [CrossRef]

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### Acro-osteolysis

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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

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**Figure 2.** Radiography of the resorption of the distal phalangeal tufts (acro-osteolysis) and soft tissue calcifications (calcinosis cutis)

#### Thomas and Jose. Acro-osteolysis

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#### References

- Avouac J, Guerini H, Wipff J, Assous N, Chevrot A, Kahan A, et al. Radiological hand involvement in systemic sclerosis. Ann Rheum Dis 2006; 65: 1088-92. [CrossRef]
- Koutaissoff S, Vanthuyne M, Smith V, De Langhe E, Depresseux G, Westhovens R, et al. Hand radiological damage in systemic sclerosis: comparison with a control group and clinical and functional correlations. Semin Arthritis Rheum 2011; 40: 455-60. [CrossRef]

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## Nasal septal perforation in systemic lupus erythematosus

Joe Thomas<sup>1</sup> , Praveen Gopinath<sup>2</sup>

A 40-year-old female with systemic lupus erythematosus was admitted with typical characteristics of active lupus in the form of palatal ulcers, alopecia, bicytopenia, lupus nephritis (class 4), necrotizing lymphadenopathy, high-titer anti-double-stranded deoxyribonucleic acid, and low complements. P-ANCA and C-AN-CA tests performed using ELISA method were negative. Otorhinolaryngology opinion was taken for nasal block, and a diagnostic nasal endoscopy showed a large septal perforation involving the anteroinferior and anterosuperior parts of the cartilaginous nasal septum with severe crusting over the edges of the septal perforation. The crusts were endoscopically cleared, edges of the perforation were smeared with an antibiotic cream, and the patient started the use of saline nasal douches. She was treated with pulse methylprednisolone, mycophenolate mofetil, hydroxychloroquine, and other supportive medications. The patient attained remission over time, and periodic endoscopic examination showed the perforation to be stable in size and free of major crusting (Figure 1). Nasal septal perforation is an underdiagnosed complication of lupus because it is asymptomatic and the patients are often not aware of their nasal problem (1). Nasal septal perforation in lupus may be secondary to vasculitis or to ischemia with subsequent chondrolysis (2). Treatment should primarily be directed to control disease activity.



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Figure 1. Large septal perforation involving the anteroinferor and anterosuperior parts of the cartilaginous nasal septum

#### Thomas and Gopinath. Nasal septal perforation in SLE

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- Reiter D., Myers A.R. Asymptomatic nasal septum perforation in Systemic Lupus Erythematosus. Ann Otol 1980; 89: 78-80.
- Rahmam P, Gladman D, Urowitz M. Nasal septum perforation in Systemic Lupus Erythematosus - Time for a closer look. J Rheumathol 1999; 26: 1854-5.

### Chronic postrheumatic fever arthropathy

Nagaraja Moorthy 🗅, Rajiv Ananthakrishna 🗅

A 36-year-old female presented with deformities of the hands and feet for the last 10 years. She had a history of acute rheumatic fever at the age of 15 years. Clinical examination revealed painless reducible deformities of her hands and feet, characteristic of Jaccoud's arthropathy (Figure 1a, Video 1). Laboratory investigations were negative for acute phase reactants, rheumatoid factor, and antinuclear antibodies. Radiography of both hands showed ulnar deviation of the fifth digit and absence of bony erosions (Figure 1b). Echocardiography was consistent with rheumatic mitral stenosis (Figure 1c-d, Video 2). The mitral valve orifice area measured 1.4 cm<sup>2</sup>. Jaccoud's arthropathy is typically observed in rheumatic fever, but it has also been described in systemic lupus erythematosus and other connective tissue disorders (1, 2). The deformities are reducible and are primarily caused by soft tissue abnormalities rather than destruction of the joints. Jaccoud's arthropathy is benign and reversible and should be differentiated from fixed deformities of rheumatoid arthritis. The mainstay of treatment includes physiotherapy and the use of orthotic devices. Our patient was referred to physical therapy for muscle strengthening and is on medical therapy for mitral stenosis.



**Figure 1. a-d.** Photograph of both hands showing flexion deformities at the proximal interphalangeal joints and extension deformities at the distal interphalangeal joints (a). Radiograph showing ulnar deviation of the fifth digit and absence of bony erosions (b). Two-dimensional echocardiography illustrating the "hockey stick" appearance of anterior mitral leaflet in parasternal long axis view (arrowhead) (AO: aorta, LA: left atrium, LV: left ventricle, RV: right ventricle) (c). Three-dimensional echocardiography demonstrating the thickened mitral valve leaflets with fused commissures (fish-mouth appearance) in parasternal short axis view, suggesting rheumatic mitral stenosis. The mitral valve orifice area measured 1.4 cm<sup>2</sup> (d)

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**Video 1.** Video illustrating the hand deformities that are reversible. These reducible deformities are characteristic of Jaccoud's arthropathy

**Video 2.** Three-dimensional echocardiography demonstrating the thickened mitral valve leaflets

with fused commissures (fish-mouth appearance) in parasternal short axis view, suggesting rheumatic mitral stenosis. The mitral valve orifice area measured 1.4 cm2

- 1. Santiago M, Machicado V. Jaccoud's Arthropathy. N Engl J Med 2015; 373: e1.
- Chaurasia AS, Nawale JM, Patil SN, Yemul MA, Mukhedkar SM, Singh D, et al. Jaccoud's arthropathy. Lancet 2013; 381: 2108. [CrossRef]

Images in Rheumatology

## Osseous tuberculosis mimicking Kienböck's disease of the wrist

### Ganesh Singh Dharmshaktu 💿

Wrist is an uncommon site for tuberculosis and its description is limited to few reports or small series in the literature. Synovial soft tissue is usually involved more and before the skeletal pathology, making radio-



**Figure 1.** Radiograph of the wrist showing abnormal shape, irregularity, flattening, and decreased height of lunate with sclerosis (arrow) in anteroposterior and oblique views. Rest of the carpal bones appear normal

logical presentation a late feature (1). The involvement of single focus may ultimately spread to multiple carpal joints, which can lead to serious morbidity. In endemic regions, tuberculosis can masquerade other disorders, and it thus warrants cautious approach to diagnose it early.

A 65-year-old female presented to us with mild atraumatic right wrist pain that was insidious at onset but had increased in severity in the last six months. The pain was located to the dorsal midline of wrist and she noticed mild swelling of the wrist for the past three weeks. There was limitation of terminal range of motion but she could still perform activities of daily living. There was no constitutional feature or relevant medical history suggestive of any chronic systemic disease. The radiograph of the wrist revealed abnormal shape of lunate bone with decrease height, radio-opacity, and flattening (Figure 1). She was diagnosed with osteochondritis of lunate, also known as Kienböck's disease, and was managed elsewhere for one month leading to no relief. Magnetic resonance imaging (MRI) was advised and showed abnormal signal



Figure 2. Sagittal MRI scans showing hyper-intensities in lunate and distal radius suggesting edema and fluid collection in volar space (asterisk)

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#### Dharmshaktu G.S. Tuberculosis as Kienböck disease



**Figure 3.** Coronal images showing multiple proximal carpal bone and distal radioulnar region edema and adjacent collection. The volar collection along with extensive flexor tendon tenosynovitis is also present

intensities in lunate and distal radius with soft tissue collection more in volar space (Figure 2). The non-contrast MRI revealed multiple bony erosions and edema was noted to be severe on proximal carpal row along with extensive tenosynovitis of flexor tendons (Figure 3). Provisional diagnosis of tuberculosis of the wrist was made. The synovial fluid that aspirated from the wrist showed the presence of mycobacterium tuberculosis in polymerase chain reaction (PCR) test. Appropriate anti-tubercular therapy was initiated leading to gradual improvement in clinical-radiological profile in 6 weeks and the same was continued for a total of 18 months as per the institution protocol. No recurrence or further complication was noted regarding the course of disease and pharmacotherapy.

Osteonecrosis of lunate bone is termed Kienböck's disease and is a rare disease with no distinct etiology. Lunatomalacia and ischemic ne-

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crosis of lunate are other terms for describing this disease. Progressive debilitation, however, is noted as it has a negative impact on wrist biomechanics. Trauma or stress injuries are etiological risk factors associated with this disease (2). Modalities such as MRI have made it readily identifiable in dubious cases. It usually affects men in the adult age group. There is a need to form well-defined diagnostic and intervention criteria for the disorder (3). Initial presentation of a sinister disorder like tuberculosis as Kienböck's disease is an uncommon event and has only been reported once to our knowledge (4). This short case snippet highlights the importance of careful assessment of radiographs and judicious use of MRI to confirm the diagnosis or to rule out other variables.

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- Kotwal PP, Khan SA. Tuberculosis of the hand: clinical presentation and functional outcome in 32 patients. Bone Joint J 2009; 91: 1054-7.
- Bain Gl, MacLean SP, Yeo CJ, Perilli E, Lichtmann DM. The etiology and pathogenesis of Kienbock's disease. J Wrist Surg 2016; 5: 248-54. [CrossRef]
- Bain GI, Yeo CJ, Morse LP. Kienbock'S disease: recent advances in the basic science, assessment and treatment. Hand Surg 2015; 20: 352-65. [CrossRef]
- Jaiswal Y, Agnihotri A. Tuberculosis wrist mimicking Kienbock's disease. J Clin Orthop Trauma 2015; 6: 78. [CrossRef]

# Nuclear medicine imaging in idiopathic inflammatory myopathies

Thomas Nadin<sup>1</sup> , Mohammed Akil<sup>1</sup>, Manu Shastry<sup>2</sup>, Michael Hughes<sup>1</sup>

A 76-year-old female presented with a three-month history of progressive proximal muscle weakness and systemic disturbances, including fatigue and malaise. There was a contemporaneous onset of a widespread rash (including the trunk and limbs) in keeping with dermatomyositis. Before the onset of her illness, she was fully mobile and independent. Physical examination revealed marked reduction in power in the proximal musculature and neck. Investigations revealed a raised creatine kinase (4018 IU/L). Magnetic resonance imaging of the femurs demonstrated florid muscle edema. Computerised tomography - positron emission tomography (CT-PET) (Figure 1) (Top and bottom left) imaging demonstrated increased uptake of fluorodeoxyglucose tracer in the skeletal muscle of the proximal upper limbs and neck. Electromyography findings were consistent with diagnosis of myositis, including profuse fibrillation and positive sharp waves in the deltoid muscle group. Immunology was negative, including myositis-specific antibodies. A diagnosis of dermatomyositis was made based on the clinical findings and investigations. CT-PET (Figure 1) (Bottom right) revealed an incidental facial squamous cell carcinoma (arrow) which was subsequently completely excised at an early stage, avoiding lymph node/other spread. Increased colonic uptake from diverticular disease was also noted and subsequent colonoscopy was negative for malignancy. Our case highlights the potential clinical utility of nuclear medicine imaging to screen for underlying malignancy and to assess muscle disease activity (Figure 1).



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Figure 1. Nuclear medicine imaging in idiopathic inflammatory myopathies

#### Nadin et al. Nuclear medicine imaging in idiopathic inflammatory myopathies

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## Periungual dryness without resultant desquamation in a child with kawasaki disease: A new clinical finding?

Aman Gupta 💿, Surjit Singh 💿

Kawasaki disease (KD) is a multisystem vasculitis with predominant muco-cutaneous manifestations. Desquamation of the skin is usually observed 10-14 days after the onset of the disease (1). Periungual desquamation may precede by dryness at the fingertips (2). Dryness may rarely persist without resultant desquamation.

A 4.5-year-old-girl presented with fever for 15 days along with redness of the eyes, cracking of the lips, and swelling over the dorsum of the hands and feet for 7 days. No rash, redness of the tongue, or preceding history of upper respiratory tract infection was observed. The patient had received oral amoxicillin–clavulanate for 7 days without any improvement. Examination revealed a febrile child with dry, cracked, and reddened lips. Dryness of skin was observed in the periungual location on both the toes without any desquamation. Systemic examination was unremarkable. The patient had hemoglobin 89 g/L, total leukocyte count 13.9×10<sup>9</sup>/L, platelet count 674×10<sup>9</sup>/L, erythrocyte sedimentation rate 58 mm in the first hour, total serum protein 7.1 g/dL with albumin of 4.2 g/dL. No coronary artery abnormalities were observed on 2-dimensional transthoracic echo-

cardiography. The patient developed periungual desquamation on the right toe, whereas dryness persisted on the left toe. A diagnosis of incomplete Kawasaki disease (KD) was made, and the patient received 2 g/kg intravenous immunoglobulin (IVIG).

Kawasaki disease classically presents with muco-cutaneous manifestations that sequentially appear over time (3). Desquamation occurs during the subacute phase of the disease and is typically periungual and sheet-like (1). However, flaky desquamation can also occur (1). We have recently reported dryness of the fingertips as a premonitory sign of skin peeling in KD (2). However, in the present case, the dryness progressed to desquamation in one toe (Figure 1a), whereas it continued to persist in the other toe without resulting in any desquamation (Figure 1b). Recognition of this finding may facilitate the early diagnosis and treatment of KD.

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**Figure 1. a, b.** Periungual desquamation over big toe of right foot (a); Periungual dryness over big toe of left foot (b)

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#### Gupta and Singh. Periungual dryness without desquamation in KD

2.

#### References

1. Son MB, Sundel RP. Kawasaki disease. In: Petty RE, Laxer RM, Lindsley CB, Wedderburn LR, Eds.

Textbook of Pediatric Rheumatology. 7th ed. Philadelphia: Elsevier Saunders; 2015. p. 467-83. Jindal AK, Singh S. Dryness at Fingertips: Is it a Premonitory Sign of Skin Peeling in Kawasaki Disease. Eur J Rheumatol 2019; 6(4): 231-2

J Clin Rheumatol 2017; 23: 286. [CrossRef] Gupta A, Singh S. Kawasaki disease for Dermatologists. Indian Dermatol Online J 2016; 7: 461-70. [CrossRef]

3.