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Picture Quiz

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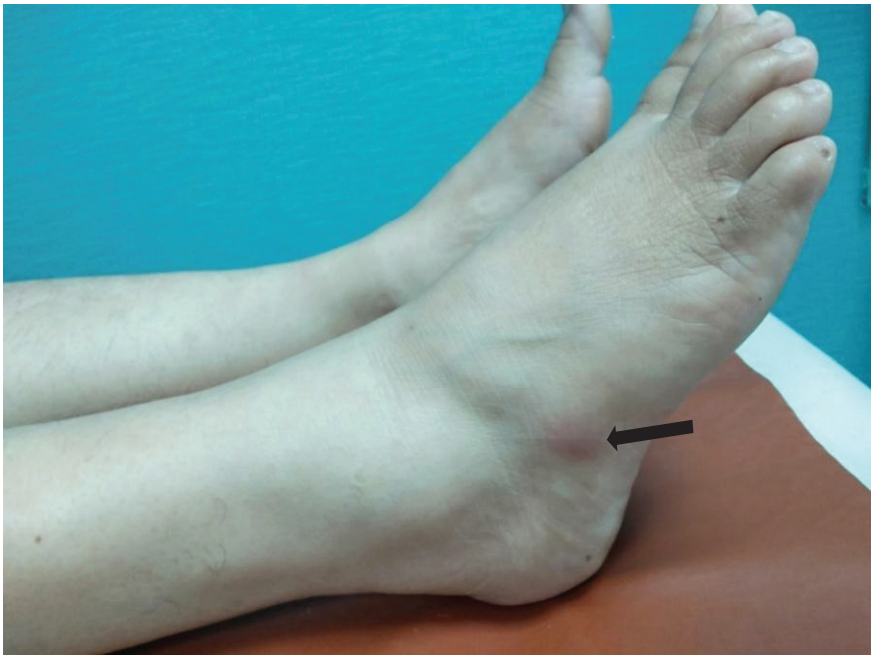
Question 1:

A 45-year-old female presented with a history of painful skin lesions on both legs, associated with bilateral ankle pain and swelling. She denied cough, fever, or lymphadenopathy, and no other joint involvement was noted.

On examination, multiple skin lesions similar to the one indicated by an arrow were observed, along with swollen ankle joints.



1.1. What is the most likely clinical diagnosis?
1.2. Name one investigation to assist the diagnosis.

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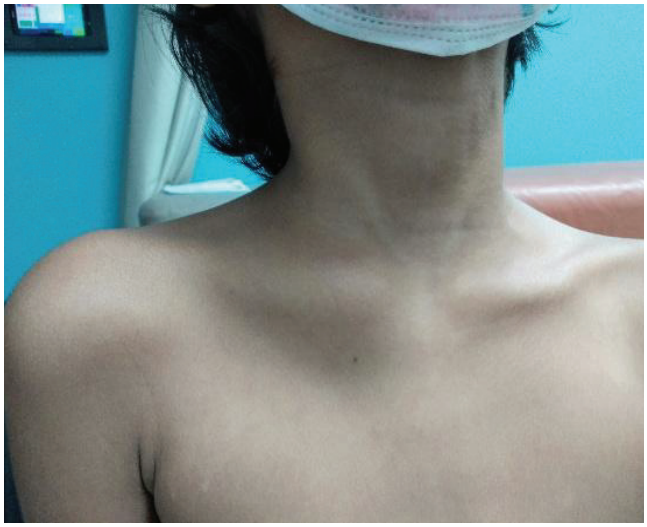
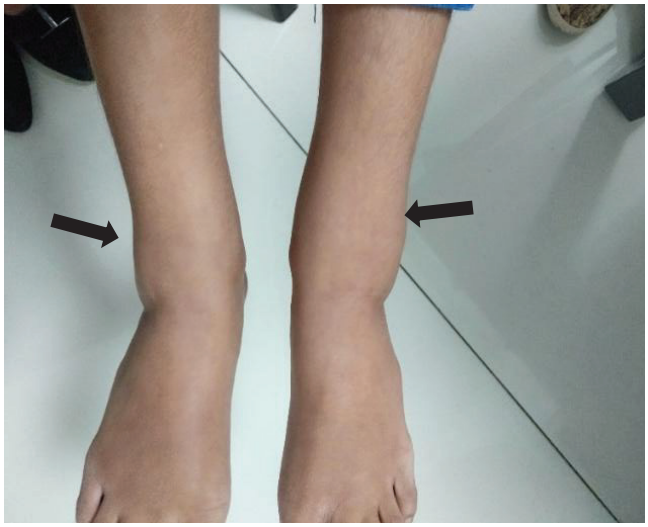
Question 2:

An 11-year-old girl presented with a 3-month history of pain and swelling of the right medial clavicle and of the bilateral ankles. Over the preceding month, she developed recurrent, deep, painful acne over her upper back, which healed with scarring. Examination revealed bony enlargement of the right medial clavicle. She also had tender enlargement of the distal tibiae bilaterally (arrow).





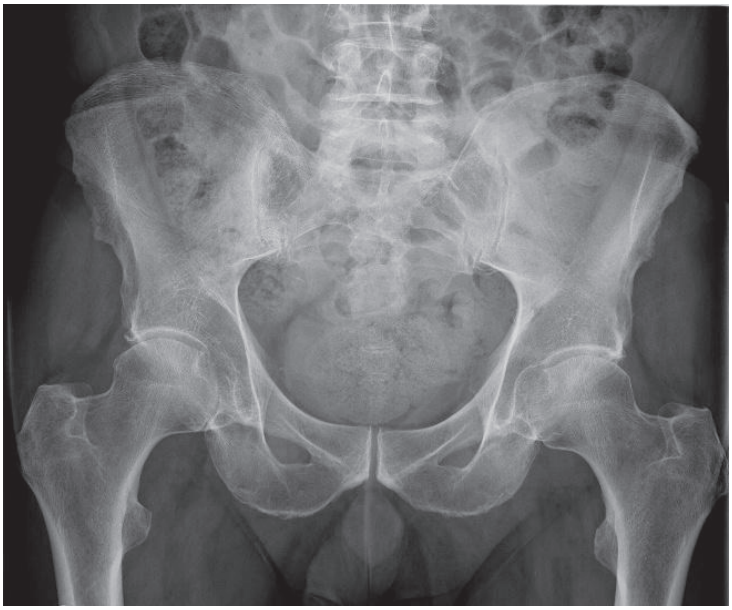
2.1. What is the most likely diagnosis?

Question 3:

A 52-year-old female presented with pain in her hands and chronic scarring skin lesions.

3.1. What is the complete diagnosis?
3.2. How do you confirm the dermatological diagnosis?

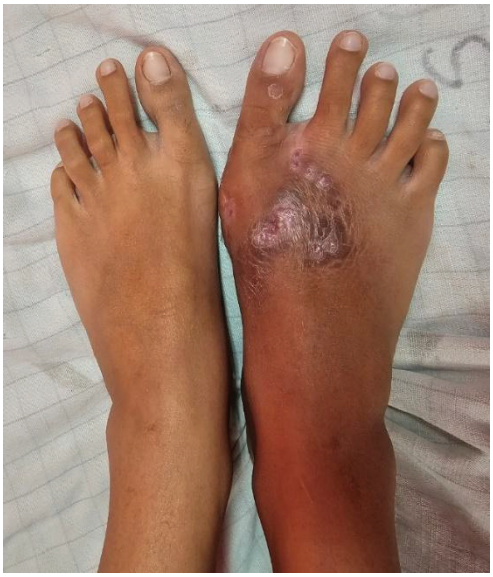
Question 4:

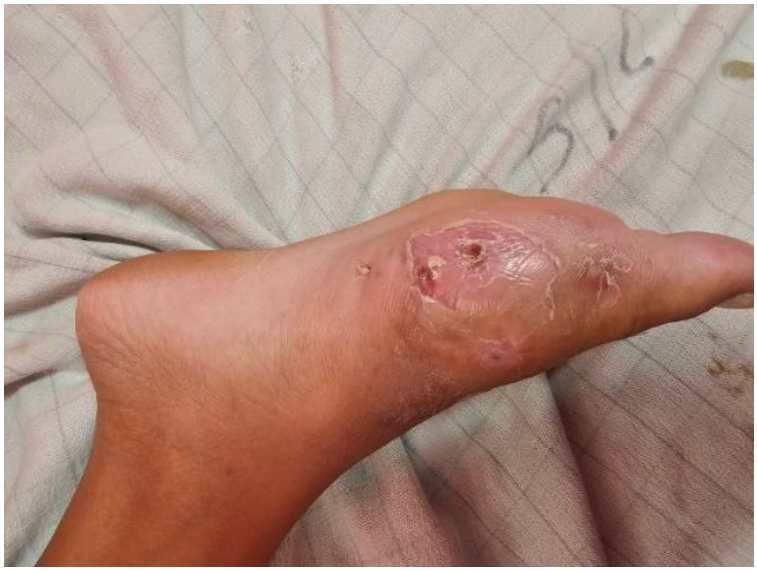


A pelvic X-ray of a 70-year-old man presenting with non-specific right-sided lower back pain is shown.

4.1 What is the most likely diagnosis? Give reasons.
4.2 Name one biochemical investigation that would support this diagnosis.
4.3 What is the first-line pharmacological treatment (gold-standard medication) for this condition?

Question 5:





A 23-year-old woman presented to the rheumatology clinic with recurrent episodes of metatarsophalangeal (MTP) joint pain and swelling, previously diagnosed as frequent gout flares. Her symptoms began at age 14, when she presented to a tertiary care hospital in the North Central Province of Sri Lanka with pain and redness of the right first metacarpophalangeal joint. Over the following 9 years, she had progressive swelling of the joint with recurrent flares leading to spontaneous ulceration of the overlying skin and discharge of a chalky white material.

5.1 What is the most likely diagnosis?
5.2 How would you confirm the diagnosis?

Answers

Answer 1:

1.1 Löfgren's Syndrome

1.2 Chest X-ray to identify bilateral hilar lymphadenopathy.
Löfgren's syndrome, an acute clinical variant of sarcoidosis, is characterised by a classic triad of erythema nodosum, bilateral hilar lymphadenopathy, and migratory polyarthralgia or periarthritis. While sarcoidosis often presents as a chronic interstitial lung disease, Löfgren's is an acute, often self-limiting phenotype with high diagnostic specificity (up to 95%) when the full triad is present. The ankle "arthritis" in this syndrome is typically periarthritis – inflammation of the subcutaneous fat and tendon sheaths – which explains the absence of significant intra-articular synovitis.[1]

Answer 2:

2.1 SAPHO Syndrome (Synovitis, Acne, Pustulosis, Hyperostosis, and Osteitis).

SAPHO syndrome is an auto-inflammatory disorder characterised by a constellation of bone, joint, and dermatological manifestations. The anterior chest wall, particularly the medial clavicle, sternum, and ribs, is the most frequently affected site with osteitis and hyperostosis. Skin manifestations such as pustulosis and/or acne are not present in all patients with SAPHO syndrome, which may make diagnosis more challenging.

Treatment typically begins with NSAIDs as first-line, with methotrexate or biologics for refractory cases.[2]

Answer 3:

3.1 Discoid Lupus Erythematosus (DLE) with deforming Rheumatoid arthritis.

Deforming joint disease of the right hand, with Boutonniere deformities of the 3rd to 5th fingers and a swollen right wrist, suggests rheumatoid arthritis.

The prominent, well-demarcated atrophic plaques with hypopigmented centres and hyperpigmented borders on the forearms are consistent with discoid lupus erythematosus.

In addition, her left hand is clenched in a fist. There was hypertonia with flexion deformities of the rest of the left upper limb. Further investigation revealed systemic features of systemic lupus erythematosus (SLE) and co-existing antiphospholipid syndrome (APLS), with multiple cerebral ischaemic infarcts.

3.2 Full-thickness skin biopsy from the edge of a lesion.

Answer 4:

4.1 Paget's disease of bone.

The marked thickening of the iliopectineal line, cortical thickening, and a coarse trabecular pattern involving the right hemipelvis are characteristic of Paget's disease. The "pelvic brim sign" (thickening of the iliopectineal line) and enlargement of the hemipelvis are pathognomonic radiographic features of Paget's disease. While often asymptomatic, Paget's disease can lead to bone pain, secondary osteoarthritis, or, rarely, malignant transformation into osteosarcoma.[3]

4.2 Serum alkaline phosphatase (ALP) level. This is elevated due to increased bone turnover and is frequently used as a marker to monitor treatment response.

4.3 Intravenous zoledronic acid is considered the gold standard. Oral alendronate and risedronate (not available in Sri Lanka), are less expensive alternatives.

Answer 5:

5.1 Madura Foot (Actinomycetoma /eumycetoma).

Examination of the foot reveals multiple draining sinuses at various stages of healing. Active lesions typically discharge serosanguinous fluid containing 'grains', which in this case were misidentified as gouty tophi. Significant architectural distortion of the midfoot is also evident.

Another learning point in this case is that gout is extremely uncommon in menstruating female adolescents and adults in the absence of other precipitating factors, such as cyanotic congenital heart disease or chronic kidney disease. The presentation of Madura foot is highly variable, with an indolent, protracted disease course.

5.2 Deep tissue biopsy and culture.

Mycetoma is a chronic granulomatous infection caused by fungi (eumycetoma) or bacteria (actinomycetoma). It is characterised by the clinical triad of localised swelling, underlying bone destruction, and sinus tracts that discharge grains containing the causative organisms. The organisms are saprophytic and can be isolated in deep tissue culture. Affected individuals have a history of exposure to contaminated soil. Widespread barefoot walking and the agricultural economy make Sri Lankans vulnerable to the condition, especially in the rural farming communities of the north-central, northern, and eastern provinces.[4]

Author declaration

Conflicts of interest

The authors declare that they have no known competing interests.

Consent for publication

Informed consent was obtained from the patients (or the patient's guardian) for the publication of any images or clinical details included in this article.

References

1. Tejera Segura B, Holgado S, Mateo L, Pego-Reigosa J M, Carnicero Iglesias M, Olivé A. Síndrome de Löfgren: estudio de 80 casos [Löfgren syndrome: a study of 80 cases]. *Medicina Clinica,* 2014;143(4):166–169. doi:10.1016/j.medcli.2014.02.029
2. Demirci Yildirim T, Sari İ. SAPHO syndrome: current clinical, diagnostic and treatment approaches. *Rheumatol Int.* 2024;44(11): 2301–2313. doi:10.1007/s00296-023-05491-3
3. Ralston S H, Corral-Gudino L, Coope, C, et al. Diagnosis and Management of Paget's Disease of Bone in Adults: A Clinical Guideline. *J Bone Miner Res.* 2019; 34(4): 579–604. doi:10.1002/jbmr.3657
4. Zijlstra E E, van de Sande W W J, Welsh O, Mahgoub E S, Goodfellow M, Fahal A H. Mycetoma: a unique neglected tropical disease. *Lancet Infect Dis*. 2016;16(1), 100–112. doi:10.1016/S1473-3099(15)00359-X