A 51-year-old man presented with numerous mobile, skin-colored, tender, subcutaneous nodules over the extensor aspects of his superior and inferior extremities for the past 2 years. These nodules measured between 1 and 5 cm in diameter. Some of the right pretibial lesions were ulcerated and oozed chalky substances. The associated small joints of hands and feet were disfigured [Figure 1]. Their mobility was partially compromised. However, the skin overlying these distorted peripheral joints lacked any inflammatory changes. On further interrogation, he also recalled few episodes of asymmetric arthritis involving hands and feet during the past 15 years. Family history of gout as well as personal history of diabetes, hypertension, alcoholism, purine-rich diet, and exposure to chemicals/heavy metals was negative.

Plain skiagram detected only soft tissue swellings [Figure 2]. Serum biochemical investigation revealed ↑uric acid (13.8 mg/dl, normal: 4–7 mg/dl), ↑urea (92 mg/dl, normal: 10–50 mg/dl), and ↑creatinine (1.8 mg/dl, normal: 0.5–1.2 mg/dl). Liver function test, urinalysis, and serum electrolytes remained within normal reference ranges. Rheumatoid factor was negative. Fine-needle aspiration cytology from the nodules and also the cytological preparation with chalky discharges from the ulcers expressed amorphous fluffy masses of brown-colored crystals with foreign body reaction. Rare polymorphs and histiocytes were present. The morphology of these crystals was best visualized under higher magnification at the periphery of crystallized clumps as well as in dispersed crystals lying singly. Structurally, these crystals appeared long and extremely fine, sharply pointed at both ends, resembling the shape of a needle [Figure 3]. No other cellular elements were found within any of the smears.

**Question**

What is your diagnosis?
Answer

Diagnosis: Chronic tophaceous gout (CTG).

Discussion

Gout is a chronic disease resulting from deranged purine metabolism and is mostly characterized by hyperuricemia. It leads to the accumulation of monosodium urate crystals within the synovial limb joints, extra-articular soft tissue, and kidneys. Patients exacerbate with recurrent episodes of acute arthritis, which is precipitated by the crystallized deposits within the joints. The first metatarsophalangeal joint is most frequently involved. Gout is relatively common among Westerners, with an overall lifetime risk of 1%–2% whereas among Indians, its prevalence has been reported between 0.12% and 0.19%.\(^1\)\(^-\)\(^3\)

In long-standing untreated cases of gout, the urate crystals conglomerate to form an amorphous mass, namely “gouty tophus”. The pseudotumoral process that ensues in this way, is known as “tophaceous gout”. Clinically, visible tophi are usually preceded by repeated attacks of acute arthritis for about 12 years on an average.\(^3\)\(^,\)\(^4\)

Under rare circumstances, “CTG” is initialized. Here, generalized tophi deposition occurs in the tendons and soft tissue surrounding the wrist, elbow, ankle, knee, and small joints of hand and foot. Infrequently, so extensor surfaces of limbs, ear helices, and eyelids also become affected.\(^3\)\(^\)\(^,\)\(^5\) Such a rare manifestation of gout is evidenced in only 3%–14% patients.\(^3\)\(^,\)\(^4\) Wang et al. opined that in long-standing cases, tissue damage and venous stasis caused by subcutaneous urate deposits eventually result in panniculitis and subsequent cutaneous ulceration.\(^5\) In the currently reported patient, decades-long negligence to his condition led to extensive tophi deposition all over the extensors and limb joints with joint distortion. Some of his right shin tophi were ulcerated with discharging chalky substances as well.

In case of the discussed patient, his clinical and biochemical profile was indicative of “tophaceous gout.” However, still some multifocal coexistent cutaneous cysts or tumors and soft tissue tumors such as lipoma or neurofibroma were subject to be excluded from the study.\(^5\) A possibility of dual pathology was kept on the mind. The fine-needle aspirate was granular chalky white in appearance, which reminisced about gouty tophi but also about few relatively uncommon multifocal cutaneous conditions such as pilomatrixoma, calcified infundibular cysts, and calcinosis universalis.\(^6\) In such situation, a confident diagnosis of gout requires demonstration of urate crystals in synovial fluid and/or cytological/histopathological examination of the tophus.\(^3\)\(^-\)\(^5\) On aspirated smears, gouty tophi appear as amorphous dark brown clumps of crystalline substances under low power objectives. Few inflammatory cells including histiocytes and foreign body-type multinucleated histiocytic giant cells are often interspersed within the debris. On magnification, these amorphous masses are comprised by tangled stacks of innumerable, bipolar pointed tip, slender, needle-shaped crystals of monosodium urate. These crystals measure approximately 5–20 \(\mu\) in length. Their morphology is appreciated best for individually dissociated crystals or at the periphery of those clumps.\(^7\)\(^,\)\(^8\) Similar cytological features were also found in the present case. The characteristic crystals were well visualized on aspirated smears from tophi as well as within the chalky discharge from pretibial ulcers. When such a distinctive cytological finding was corroborated with serum uric acid level plus the typical location and presentation of the lesions – it virtually clinched the diagnosis of CTG. Needless to mention that diagnostic excisional biopsy was averted thereby.

Risk factors of gout include chronic diuretic therapy, alcoholism, and cyclosporine use.\(^4\) However, none of
such risk factors could be validated in the reported case. Treatment modalities of CTG encompass diet modification, medical and surgical interventions. Medical options include nonsteroidal anti-inflammatory drugs (NSAIDs), hypouricemic (allopurinol), and uricosuric drugs (probenecid). Surgery is usually preserved for critical joint deformities.\(^2,4\) Therapeutic strategies for gouty panniculitis with ulceration are debated. Anti-inflammatories and surgical excision have been practiced with some success by researchers.\(^1\) In this regard, the discussed patient was symptomatically relieved with NSAIDs and allopurinol. However, before implementing any serious consideration toward his ulcerated tophi or distorted joints, he was missed out on further follow-up.

**Learning points**

- Uncontrolled hyperuricemia from undertreated or untreated gout precipitates into widespread tophaceous deposits over the limbs: a condition known as chronic tophaceous gout (CTG)
- In CTG, the pseudotumoral lesions are predominantly juxta-articular in location, but sometimes, these may also evolve at extensor surfaces of limbs, away from joint proximity
- Definitive diagnosis of this condition can be rendered by demonstration of classic urate crystals on cytological smears from these nodules and discharging ulcers. Thereby, the more hazardous excision procedure is averted
- Conjugal chemotherapy with uric acid-lowering drugs and anti-inflammatory drugs has the best patient outcome
- Surgery is reserved for debilitating joint deformity.

**Financial support and sponsorship**
Nil.

**Conflicts of interest**
There are no conflicts of interest.

**References**
1. Wang L, Rose C, Mellen P, Branam G, Picken MM. Gouty panniculitis with ulcerations in a patient with multiple organ dysfunctions. Case Rep Rheumatol 2014;2014:320940.
2. Mahajan A, Jasrotia DS, Manhas AS, Jamwal SS. Prevalence of major rheumatic disorders in Jammu. JK Sci 2003;5:63-6.
3. Bones HA. Joints and soft tissue tumors. In: Kumar V, Abbas AK, Aster JC, editors. Robbins and Cotran Pathologic Basis of Disease. 9th ed. Philadelphia: Elsevier; 2014. p. 1214-7.
4. Kelley WN, Schumacher HR Jr. Crystal-associated synovitis. In: Kelley WN, editor. Textbook of Rheumatology. 4th ed. Philadelphia: Saunders; 1993.
5. Elder DE, editor. Lever's Histopathology of the Skin. 10th ed. Philadelphia: Lippincott Williams & Wilkins; 2008. p. 438-9, 791-810, 1057-99.
6. Koss LG, Melamed MR, editors. Koss' Diagnostic Cytology and Its Histopathologic Bases. 5th ed. Philadelphia: Lippincott Williams & Wilkins; 2006. p. 1290-1.
7. Gupta A, Rai S, Sinha R, Achar C. Tophi as an initial manifestation of gout. J Cytol 2009;26:165-6.
8. Walke V, Ramraje S, Jadhao V. Cytodiagnosis of gouty tophus. Cytojournal 2013;10:11.