

Gouty Tophi on the Ear: A Review

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Practice Points

- Consider gout in the differential of papulonodular lesions on the ear.
- If biopsying to rule out gout, fix the biopsy specimen in ethanol or Carnoy fixative to preserve birefringence of urate crystals.

Although the classic location of gouty tophi is the great toe (podagra), gouty tophi of the ear also is common and is worth including in the differential diagnosis in patients presenting with ear lesions. Other entities presenting as papules or nodules on the ear include chondrodermatitis nodularis helices (CNH), actinic keratosis, basal cell carcinoma, squamous cell carcinoma, verruca vulgaris, amyloids, rheumatoid nodules, and elastotic nodules. If tophaceous gout is suspected, alcohol fixation of the biopsy specimen is preferable, as it enables visualization of characteristic needle-shaped urate crystals.

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Gout is the most common cause of inflammatory arthritis in men and classically presents as painful acute monoarthritis of the great toe (podagra) or the knee. In women, gout usually occurs after menopause.¹ Hyperuricemia is the primary risk factor for gout; other risk factors include hypertension, renal insufficiency, diuretic use, alcohol consumption, a purine-rich diet, and obesity.² Due to high cell turnover, leukemic patients and solid-organ transplant recipients who are taking immunosuppressive agents, especially cyclosporine, also have an increased propensity to develop hyperuricemia

and gout. Presentation of gout in younger patients without these risk factors should raise suspicion of renal disease or an underlying enzyme deficiency, such as hypoxanthine guanine phosphoribosyltransferase deficiency in X-linked recessive Lesch-Nyhan syndrome.³

Clinical Features

Gouty tophi classically present as hard, yellow-white papules and nodules of a chalky consistency on the joints of the hands and feet. Less common locations for gouty tophi include the ears, elbows (olecranon bursae), and the Achilles tendon. Decreased temperature and reduced blood flow in these areas may explain the predilection for these locations. The ears are the most common location for gouty tophi in the head and neck region and may be a heralding sign of gout.⁴ Auricular tophi usually are located on the helical rims but also may be located on the anti-helix (Figure 1). Lesions usually are painless and well circumscribed without any surrounding erythema, bleeding, or eschar formation. Larger tophaceous nodules may perforate through the overlying skin and extrude their chalky content.

Histopathology

Histopathologic diagnosis of auricular gouty tophi is greatly aided by selection of the correct fixative. Inclusion of gout in the differential diagnosis should alert the clinician to use ethanol or Carnoy fixative, an ethanol-based fixative, for a portion of the biopsy specimen because the aqueous nature of formalin dissolves urate crystals.⁵ Alcohol-fixed tophi show characteristic needle-shaped urate crystals packed in radially arranged bundles surrounded by a granulomatous infiltrate; however, after formalin fixation,

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Figure 1. Gouty tophus on the right helical rim. Photograph courtesy of Joseph C. English III, MD, Pittsburgh, Pennsylvania.

only remnant, amorphous, slightly eosinophilic, nonbirefringent material with surrounding histiocytes (CD68⁺) and foreign body giant cells are seen (Figure 2). Via the traditional de Galantha technique⁶ for preserving gout crystals, tissue is fixed in absolute ethyl alcohol, and staining yields black urate crystals in a yellow background.

Polarizing microscopy is useful in distinguishing gout from pseudogout and tumoral calcinosis. The monosodium urate crystals that are characteristic of gout are negatively birefringent, while the calcium pyrophosphate crystals of pseudogout are positively birefringent. Urate crystals are yellow when parallel and blue when perpendicular to the direction of the compensator. Calcium-detecting alizarin red and phosphate-detecting von Kossa stains also can be helpful in ruling out calcium pyrophosphate deposits seen in pseudogout.⁷ In contrast to gout and pseudogout, calcium hydroxylapatite deposition in tumoral calcinosis lacks birefringence.

To preserve the birefringent property of gout crystals, different staining and nonstaining techniques have been developed. Staining of formalin-fixed specimens (more than 6 hours but fewer than 12 hours) with a nonaqueous alcoholic eosin staining method, which avoids the use of hematoxylin, preserves the birefringence of gout and pseudogout crystals.⁸ Additionally, a nonstaining method for detecting birefringent urate crystals in a formalin-fixed specimen using a thick, unstained, cover-slipped microscopy slide has been described.⁹

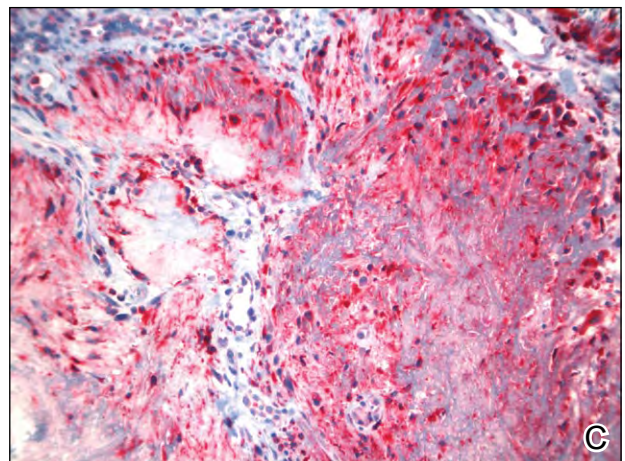
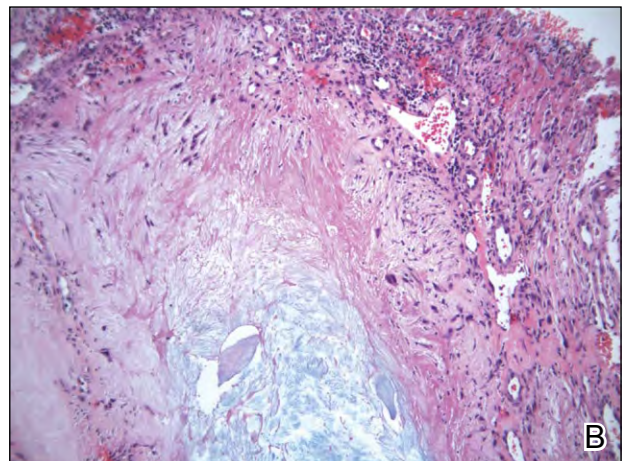
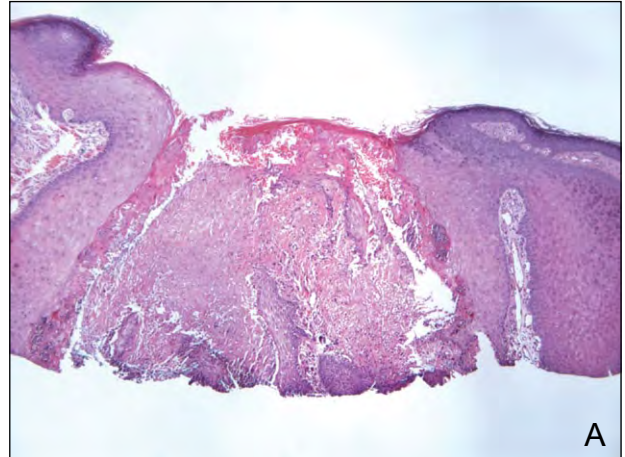


Figure 2. Formalin-fixed auricular tophi biopsy showing a perforating aggregate with a mixed inflammatory infiltrate (A and B)(H&E; original magnifications $\times 100$ and $\times 200$, respectively). Numerous histiocytes also were appreciated (C)(anti-CD68 antibody, original magnification $\times 200$).

Differential Diagnosis

In addition to gout, the differential diagnosis for papules on the helix and antihelix includes

chondrodermatitis nodularis helices (CNH), actinic keratosis, basal cell carcinoma, squamous cell carcinoma, verruca vulgaris, amyloids, rheumatoid nodules, and elastotic nodules.¹⁰ Chondrodermatitis nodularis helices presents as consistently painful lesions unlike gouty tophi, which usually are painless. Transdermal elimination of destroyed collagen as seen in CNH may explain the clinical resemblance between CNH and gout, especially perforating tophaceous gout nodules (Figure 2A). Auricular elastotic nodules are rare lesions resulting from chronic actinic damage that also may resemble gouty tophi; however, unlike gouty tophi and CNH, elastotic nodules on the ear usually are bilateral and typically present on the anterior crus of the antihelix and less often on the helix.¹¹ Histologically, these dome-shaped papules and nodules show clumps of irregular eosinophilic elastotic fibers on a background of remarkable solar elastosis. Amyloids, specifically primary localized cutaneous amyloidosis of the lichen variant, have been reported in the literature as presenting as papules on the ears; however, unlike tophaceous gout, these papules usually appear on the concha and are intensely pruritic.¹² Rheumatoid nodules, which also are painless, are rare but should be considered if nodules are found on the elbows and other extensor joints.¹³ Rheumatoid nodules histologically differ because the pathology lies in the deep dermis and subcutis, but they can perforate. Fibrinoid degeneration of collagen is seen with surrounding palisading histiocytes and foreign body giant cells.¹⁴

Treatment

Acute gouty tophi are managed with colchicine, nonsteroidal anti-inflammatory drugs, and corticosteroids. Drugs that lower uric acid levels in the blood (eg, allopurinol, probenecid) are not started or are discontinued during short-term episodes because of the possibility of triggering further episodes.¹⁵ Tophi usually resolve with urate-lowering therapy, but it may take more than 30 months for any perceptible reduction in size.¹⁶ Larger nodules may require excision.

Conclusion

Gout should be considered as part of the differential diagnosis in patients who present with ear lesions, especially on the helix and antihelix, particularly older men, postmenopausal women, organ transplant recipients, and patients with myeloproliferative disease. In cases of suspected gout, biopsy specimens should be fixed in alcohol to facilitate visualization of needle-shaped urate crystals or should be processed using newer staining and nonstaining

methods that preserve the birefringence of formalin-fixed urate crystals.

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