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Miss the ear, and you may miss the diagnosis

She had experienced similar but unilateral ear pain months ago

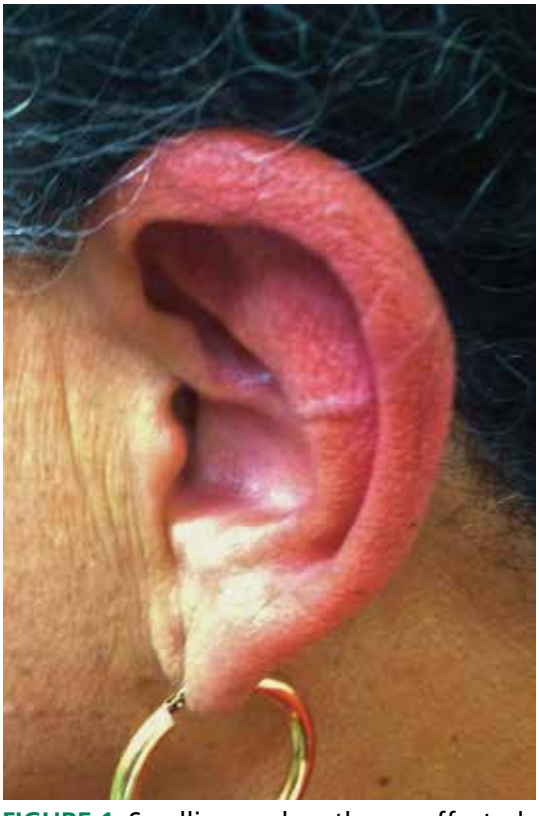

FIGURE 1. Swelling and erythema affected both ears, sparing the earlobes.

A 52-year-old woman presented with pain in both ears associated with redness and swelling. The symptoms appeared 3 weeks earlier. The pain had started on one side, then spread to the other over a period of 2 weeks. She denied fever, chills, rigor, rash, or upper respiratory symptoms. She had experienced similar but unilateral ear pain months before. Her medical history included bilateral knee pain and swelling (treated as osteoarthritis), hypertension, hyperlipidemia, and hypothyroidism. She also reported progressive bilateral hearing loss, for which she now uses hearing aids. She had no history of conjunctivitis or uveitis.

Physical examination showed swelling and erythema of both ears, sparing the earlobes (**FIGURE 1**), as well as bilateral knee-joint tenderness and restricted joint movement. The erythrocyte sedimentation rate was elevated at 52 mm/h (reference range 0–20); the complete blood cell count, creatinine, and liver enzyme levels were normal. An autoimmune panel was negative for antinuclear antibody, antineutrophil cytoplasmic antibody, and rheumatoid factor.

A clinical diagnosis of relapsing polychondritis was made based on the McAdam criteria.[1] The patient was initially started on steroids and then was maintained on methotrexate. Her symptoms improved dramatically by 3 weeks.

RELAPSING POLYCHONDRITIS

Relapsing polychondritis is a rare, chronic, and potentially multisystem disorder characterized by recurrent episodes of cartilaginous inflammation that often lead to progressive destruction of the cartilage.[2,3]

Auricular chondritis is the initial presentation in 43% of cases and eventually develops in 89% of patients.[2,4] The earlobes are spared, as they are devoid of cartilage, and this feature helps to differentiate the condition from an infection.

If the condition is not treated, recurrent attacks can result in irreversible cartilage damage and drooping of the pinna (ie, "cauliflower ear"). Biopsy is usually avoided, as it may further damage the ear. The diagnostic criteria for relapsing polychondritis formulated by McAdam et al[1] accommodate the different presentations in order to limit the need

doi:10.3949/ccjm.81a.13121

for biopsy. Systemic involvement may include external eye structures, vasculitis affecting the eighth cranial (vestibulocochlear) nerve, noninflammatory large-joint arthritis, and the trachea. There is also an association with myelodysplasia.

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