Eosinophilic granuloma: Bilateral temporal bone involvement

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Abstract
Eosinophilic granuloma is an uncommon condition that is characterized by unifocal or multifocal osteolytic lesions that often affect the skull. Unilateral lesions of the temporal bone are not uncommon, but bilateral temporal bone lesions are rare. In fact, to the best of our knowledge, fewer than 20 such cases have been reported during the past 40 years. We report a new case of bilateral temporal bone eosinophilic granuloma, and we review the disease process and its treatment.

Introduction
Eosinophilic granuloma is an uncommon condition that primarily affects children and adolescents. It is characterized by unifocal or multifocal osteolytic lesions that often affect the skull. Temporal bone lesions are not an uncommon manifestation, but bilateral temporal bone lesions are rare. Our review of the literature revealed that fewer than 20 such cases have been reported worldwide during the past 40 years.

Patients with eosinophilic granuloma usually present with otorrhea, conductive hearing loss, and symptoms mimicking those of persistent otitis media or mastoiditis. Further investigation by computed tomography (CT) generally reveals periauricular soft-tissue swelling and osteolytic lesions. A biopsy will provide the diagnosis, but bilateral temporal bone eosinophilic granuloma may not be included in the differential diagnosis because it is so rare.

We report a new case of bilateral temporal bone eosinophilic granuloma. We also review the disease process and its treatment in order to remind clinicians to consider this disease process when evaluating patients with similar signs and symptoms.

Case report
A 14-month-old boy was referred to the ENT clinic by the pediatrics unit, where he had been unsuccessfully treated for otitis media over the preceding 2 months. Despite treatment that included oral and topical antibiotics, the patient continued to exude left-sided bloody otorrhea. Just prior to the referral, a polypoid mass had developed in the patient’s left auditory canal; a biopsy of the mass identified only granulation tissue. In addition, a soft-tissue swelling had begun to develop in the patient’s left preauricular area (figure 1). The child had no systemic symptoms, and findings on the physical examination were otherwise unremarkable.

In the ENT clinic, we ordered a CT scan of the temporal bones. CT revealed the presence of an osteolytic lesion of the squamous and mastoid portions of the left temporal bone with erosion into the middle cranial fossa (figure 2, A). The facial nerve and otic capsule were spared. CT also unexpectedly showed involvement of the right temporal bone (figure 2, B). At this point, concern was raised for a systemic and/or malignant process, and we consulted with pediatrics and hematology/oncology staff. Further radiologic studies revealed no other organ system involvement. A bone marrow aspirate was obtained to look for hematologic malignancy; it revealed a normal cell population.

The child was taken to the operating room, and a biopsy of the temporal bone lesion was performed. The pathology report indicated that it was a benign-appearing lesion with a proliferation of mononuclear histiocytes that were positive for S-100 and CD1a immunohistochemical stains. These findings are consistent with Langerhans’ cell histiocytosis. Given the histopathology and physical examination findings, the child was diagnosed with eosinophilic granuloma. He was treated with a steroid (prednisolone) and a single-drug (vinblastine) chemotherapy regimen for 6 weeks. Four months after the cessation of therapy, an excellent response was noted.
**Discussion**

Langerhans’ cell histiocytosis (formerly known as histiocytosis X) is a histopathologic diagnosis characterized by a monoclonal proliferation of histiocytes. The disease is manifested in three different clinical conditions: eosinophilic granuloma, Hand-Schüller-Christian disease, and Letterer-Siwe disease. Eosinophilic granuloma is the most benign of the three conditions. Hand-Schüller-Christian disease is the chronic systemic variant, and Letterer-Siwe disease is an acute, fulminant, systemic condition.

Eosinophilic granuloma manifests clinically as osteolytic lesions of the skull, long bones, ribs, and/or vertebrae; there is a notable lack of systemic involvement. Its incidence is slightly less than 1 per 200,000 population. Approximately 75% of affected patients are younger than 20 years, and the male-to-female ratio is 2:1. The disease has no predilection to any particular race. Skull lesions are present in approximately 40% of all cases; some 20 to 30% of these lesions involve the temporal bone.

Patients with temporal bone lesions usually present with symptoms similar to those of persistent otitis media or mastoiditis. Otorrhea, soft-tissue swelling, and conductive hearing loss are common; sensorineural hearing loss and facial nerve paresis are less common. The definitive diagnosis is made via biopsy. Histopathology results show an abundance of histiocytes with grooved nuclei intermingled with large numbers of eosinophils. The tissue stains positively for S-100 protein and CD1a immunoreactivity. Although it is not usually performed, electron microscopy will reveal tennis-racket–shaped organelles known as Birbeck’s granules. Their function is unknown.

Treatment is almost always curative. Regimens vary, but most include surgical curettage. Radiation therapy is often considered, and it is sometimes used in conjunction with surgery, especially for lesions that are close to vital structures. Administration of intralesional steroids, with or without surgery, is often successful. Chemotherapy regimens that include prednisone, vinblastine, or methotrexate are usually reserved for cases of Hand-Schüller-Christian or Letterer-Siwe disease, in which multiple organ systems are involved. These agents have also been used in cases such as ours, in which multifocal bone lesions are present.

**References**