Eosinophilic Granuloma of the Temporal Bone in Children

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Abstract: Eosinophilic granuloma (EG) is a bony destructive disease that frequently occurs in children; it is a subtype of Langerhans cell histiocytosis. The aims of this study were to detect the presenting features of temporal bone lesions in children and to evaluate the efficacy of surgery combined with radiotherapy in treatment of the disease. A retrospective study on 12 children with EG of the temporal bone was done. Computed tomography and hearing assessment were performed for all patients. All patients were treated with cortical mastoidectomy followed by postoperative radiotherapy. Follow-up was carried out for at least 2 years. The patients' presenting symptoms were external ear canal mass in 10 patients (83.3%), postauricular swelling in 8 patients (66.7%), and persistent otorrhea in 4 patients (33.3%). Ten patients (83.3%) showed conductive hearing loss, whereas 2 patients (16.7%) showed mixed hearing loss on the affected side. Computed tomography showed osteolytic defects without sclerotic margins filled with soft tissue masses involving the mastoid bone. Histopathologic examination showed eosinophils and Langerhans cells that were immune reactive for CD1 antigen and S-100 protein. Postoperative follow-up showed complete cure of the disease in 10 children (83.3%), with recurrence detected in 2 patients (16.7%) who needed second surgical intervention. We concluded that temporal bone EG in children may present with features that mimic the features of chronic suppurative otitis media. However, computed tomography and histopathologic examination are diagnostic. Cortical mastoidectomy together with postoperative radiotherapy is an achievable treatment in most cases.

Key Words: Eosinophilic granuloma, temporal bone, cortical mastoidectomy, otorrhea

Langerhans cell histiocytosis (LCH) refers to a group of diseases whose primary pathogenesis is an abnormal polyclonal proliferation of Langerhans cells.[1] These cells, which are macrophages that are normally present only in the dermis, are the hallmark of this disease.[2] It is a disease that possesses 3 overlapping states called eosinophilic granuloma (EG), Hand-Schuller-Christian disease, and Letterer-Siwe disease.[3]

The common lesion is a destructive inflammatory granuloma of the skeletal and extraskeletal soft tissues and organs. Males are more affected than females. The disease is not hereditary or familial and has no racial predominance. eosinophilic granuloma frequently occurs in the flat bones such as the ribs, the pelvis, the scapula, and the skull. Temporal bone involvement is rare, whereas only 25% of them present bilateral occurrence.[4,5]

Eosinophilic granuloma commonly presents with otorrhea, mastoid swelling, deafness, and aural polyps eroding the posterosuperior ear canal wall.[5] These otologic findings can mimic common diseases, including simple otitis externa, otitis media, aural polyps, acute mastoiditis, and metastatic lesions.[6] Involvement of the inner ear is rare because of compact otic capsule.[7–9]

Treatment of EG is not specific and depends on the extent of disease at the time of diagnosis. Single lesions can be successfully treated with either surgery or radiotherapy alone. Although controversial, some authors report good results with local injection of steroids, avoiding irradiation in children.[10] For systemic disease or recurrence after irradiation in addition to surgery and radiotherapy, chemotherapy is required. Systemic or intratumoral steroids have also been used effectively. However, for treatment of unifocal bone lesions distant from vital structures, surgical curettage of tumor can provide local control of the disease, avoiding side effects of irradiation and steroid.[11]

The aims of this study were to detect the presenting features of temporal bone EG in children and also to evaluate the efficacy of surgery combined with radiotherapy in the treatment of this disease.

MATERIALS AND METHODS

This study included 12 children with EG. The patients have been diagnosed and treated at the departments of otolaryngology of 3 university hospitals in Egypt (Cairo, Beni Suef, and Suez Canal) in the period from June 2007 to July 2011, with their age ranging from 2 to 7 years, with a mean age of 3.5 years. They were 10 boys and 2 girls.

A computed tomography (CT) of the temporal bone was performed for all patients as a part of the workup to delineate the cranial extension of the lesion. In addition, chest x-ray and abdominal ultrasound were done to all patients.

Hearing was assessed in all patients, either by pure tone audiometry or auditory brain-stem response audiometry according to the tolerability of the patients.

Cortical mastoidectomy through a postauricular incision was performed for all children, and removal of soft tissues and bony lesions was carried out. All surgical specimens were sent for histopathologic examination to confirm the diagnosis. The lesions showed abundant infiltration with eosinophils and histiocytes. However, the diagnosis of EG was confirmed by using surface protein antigens S-100 and CD1 immunohistochemical staining.

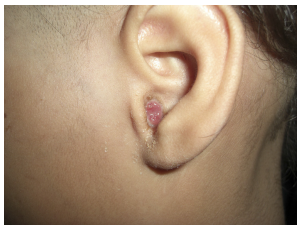

FIGURE 1. A 4-year-old child presenting with a mass on the left external auditory canal.

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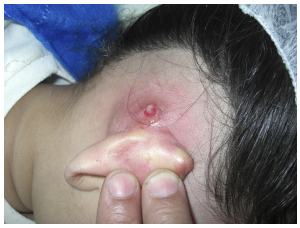

FIGURE 2. A 5-year-old child presenting with soft tissue swelling on the right mastoid region.

After confirmation of the diagnosis of EG, all patients received postoperative radiotherapy.

All patients were followed for at least 2 years after completion of the course of radiotherapy with monthly regular visits. A CT of the temporal bone was requested on suspicion of recurrence.

Informed consents were obtained from the parents of the patients, and the principles outlined in the Declaration of Helsinki were followed.

RESULTS

Twelve children were enrolled in this study, and all were diagnosed to have EG of the temporal bone. The presenting symptom was external ear canal mass or polyps in 10 patients (83.3%) (Fig. 1); 2 of them showed a normal tympanic membrane that was hardly seen on otoscopic examination, 8 patients (66.7%) had postauricular swelling (Fig. 2), and 4 patients (33.3%) had persistent otorrhea. No patients presented with bilateral lesions.

Hearing was assessed with pure tone audiometry in 8 patients and with auditory brain-stem response audiometry in 4 patients; 10 patients (83.3%) showed conductive hearing loss, whereas 2 patients (16.7%) showed mixed hearing loss on the affected side (Table 1).

Computed tomography of the temporal bone showed osteolytic defects without sclerotic margins filled with soft tissue masses involving the mastoid in all cases. The middle ear cavity and the external auditory canal were involved in 10 patients (83.3%), whereas the otic capsule was not involved in any case (Fig. 3).

Chest x-ray and abdominal ultrasonography showed no systemic lesions in any of our patients.

Intraoperatively, a soft tissue mass was detected filling the mastoid region with variable extension in all patients (Fig. 4); the middle ear cavity and the external auditory canal were involved in 10 patients.

Histopathologic examination of the specimens showed Langerhans cells and eosinophils.

The Langerhans cells were immune reactive for CD1 antigen and S-100 protein.

Follow-up showed complete cure of the disease in 10 children, whereas recurrence was detected in 2 patients. One patient developed a small aural polyp in the external auditory canal and the other developed postauricular fistula, 6 and 8 months, respectively, after completion of the treatment course. Computed tomography showed superficial soft tissue lesion without deep invasion. The first patient was treated with curettage of the lesion under a microscope, whereas

TABLE 1. Presentations of EG

Presentation	No. Patients	Percentage
External ear canal mass or polyp	10	83.3
Persistent otorrhea	4	33.3
Postauricular swelling	8	66.6
Conductive hearing loss	10	83.3
Mixed hearing loss	2	16.7

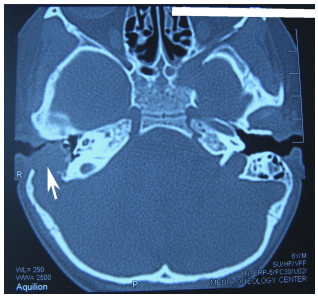

FIGURE 3. Computed tomographic scan (axial cuts). The arrow points to a soft tissue mass occupying the right mastoid process and the external auditory canal.

the fistula of the second patient was excised via postauricular approach. Fortunately, both patients showed no recurrence until the end of the follow-up period.

DISCUSSION

Eosinophilic granuloma is a single or multiple bony disease that frequently occurs in children.[12] It is a subtype of LCH that results from clonal proliferation of Langerhans cells. Although its etiology is still unknown, the hypothesis of neoplastic, inflammatory, or immunologic processes has been widely discussed in the literature, but no conclusive proof has ever been provided.[13]

A high index of suspicion is required to recognize the otologic manifestations of histiocytosis.[10] In addition, despite the fact that it is not easy to diagnose an EG isolated to temporal bone, which can mimic other more common otologic disorders, there are some clues that can suggest the EG. A young child or even an infant with otitis media or swelling in the mastoid region that is refractory to medical treatment should be suspected as having EG.[14] In particular, a negative aspiration result of mastoid swelling should suggest the EG. However, in the case of temporal bone involvement, a patient may present with otitis media alone or a large mass eroding the bone and extending to the adjacent neurovascular structures. Eosinophilic granuloma may involve the temporal bone bilaterally as well.[12]

Saliba et al[15] found that the most common presentation of LCH was a temporal bone mass (70%), followed by otitis media and/or externa (60%). they reported hearing loss in 28.5% of their patients. Nicollas et al[16] reviewed 42 patients with LCH, 7 of whom had temporal bone affection and some degree of hearing loss. In our study, the most common presenting feature is a mass or a polyp in the external canal (83.3%), with hearing affection in all patients; 10 patients had conductive hearing loss and 2 patients had sensorineural hearing loss.

Most authors believe that the method of choice in the primary diagnosis of LCH is the conventional radiographic survey, whereas others believe that the radionuclide scintigraphy is more accurate.[17] In our study, we used CT for radiologic evaluation; it showed osteolytic defects without sclerotic margins filled with soft tissue masses involving the mastoid in all children. The middle ear and the external

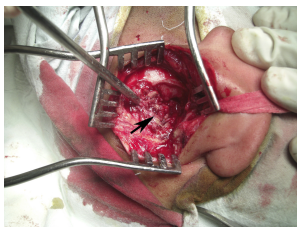

FIGURE 4. An intraoperative finding, with the arrow pointing to a soft tissue mass occupying the right mastoid region and the external auditory canal.

canal were involved in 10 patients (83.3%), whereas the otic capsule was not involved in any patient.

The diagnosis of EG was established after histopathologic examination. Histologically, the lesion was characterized by expanding, erosive accumulations of Langerhans cells within the medullar cavity of the bone. Histiocytes were variably admixed with eosinophils, lymphocytes, plasma cells, and neutrophils. The cells were immunoreactive for CD1 antigen and S-100 protein in all our patients, whereas Saliba et al[15] found positivity for these immunohistochemical stains in only 80% of their patients.

The LCH has an unpredictable course varying from a rapidly fatal progressive disease to spontaneous resolution.[2] However, the usual prognosis of the disease affecting the temporal bone is good in patients having limited organ involvement, with a survival rate of more than 90%.[1] Age at presentation, multisystem involvement, and vital organ dysfunction are the most relevant prognostic factors, with bad prognosis in children younger than 2 years.[2]

There are several treatment modalities for histiocytosis, including surgery, radiotherapy, and chemotherapy. These modalities can be used either alone or in combinations, depending upon the extent and the severity of the disease. Different treatment protocols were used in different clinical settings.[18]

In our institute, the treatment protocol of EG is to perform cortical mastoidectomy followed by postoperative radiotherapy; in our study, there was no recurrence in 83.3% of our patients. Even the 2 patients who developed recurrence were treated successfully, with limited surgical excision because the recurrent lesions were superficial.

Because of the fact that there are no large series in the literature, the optimal treatment protocol of histiocytosis is not well established. This is particularly true for EG because most published cases are sporadic without a standard protocol for treatment. With accumulation of data in the literature, the treatment of temporal bone EG may be standardized in the future.[3]

Bayazit et al[3] stated that the most preferable method for treatment of temporal bone EG is surgical excision, and radiotherapy can be used for residual tissues when complete excision is not achieved. However, some authors preferred the use of corticosteroids, either local or systemic; others used chemotherapy especially in systemic lesions.[19,20]

del Río et al[21] proposed a treatment of temporal bone histiocytosis with radiosurgery, through isodoses of 10 Gy, with a good therapeutic response after 2 years of follow-up.

Saliba et al[15] treated a case of EG in the temporal bone only with steroid injections (Triamcinolone) in the mastoid, external auditory canal, and orbital region; they achieved complete eradication of the lesion after 1 year. In addition, Rodríguez Rivera et al[22] treated EG with bone curettage and intralesional corticosteroid therapy; they obtained good clinical and radiologic response, without local and systemic relapse after a 3 month follow-up.

Finally, we conclude that children with temporal bone EG may present with features that mimic the features of chronic suppurative otitis media. However, CT shows characteristic lesions, and histopathologic examination is diagnostic. Cortical mastoidectomy together with postoperative radiotherapy is a successful method for treatment of EG of the temporal bone; however, a long follow-up is essential to detect any recurrence.

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Functional Outcomes of Preauricular Underparotid Retrograde Approach for Mandibular Condyle Fractures

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Abstract: Management of condyle fractures includes a wide spectrum of alternatives including analgesia alone, physiotherapy, intermaxillary fixation, and open reduction and internal fixation.