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CASE REPORT

Eosinophilic granuloma of the temporal bone with extensive bilateral otic capsule involvements: Incomplete reossification despite theraphy

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Summary Bilateral temporal bone involvement of eosinophilic granuloma is extremely rare. It has been estimated that approximately 22 cases have been reported so far in the world literature. In many cases, the symptoms are misinterpreted as otitis media, cholesteatoma, or mastoiditis. We report a case of bilateral temporal eosinophilic granuloma with otic capsule involvement, who was initially misdiagnosed and treated for non-specific otitis media. After mastoidectomy and chemotherapy with vinblastine and prednisone the patient's bony lesions of the temporal bone incompletely reossified and labyrinthine function was not satisfactory. Children with otitis media or mastoiditis with no response to medical treatment should be evaluated with Computerized tomography (CT) scanning of the temporal bone.

1. Introduction

Langerhans' cell histiocytosis (LCH), previously known as histiocytosis X, is a rare, proliferative disorder in which the accumulation of pathologic Langerhans' cells lead to local tissue infiltration and destruction [1]. LCH principally affects children younger than 4 years of age, with an incidence of 0.5—5 cases per million children per year [1].

Based on the sites of involvement and extent of the disease, LCH shows various clinical manifestations [2]. The incidence of otologic manifestations in children ranges from 11% to 61%. The most common symptoms are otorrhea, mastoid swelling, deafness, and aural polyps eroding the posterosuperior canal wall [3]. These otologic findings can mimic more common diseases, including simple otitis externa, otitis media, aural polyps, acute mastoiditis, and metastatic lesions [4]. Involvement of the inner ear is rare due to compact otic capsule [5—7].

Eosinophilic granuloma (EG) or benign focal histiocytosis X is one of the three variants of LCH. The

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other two are Letterer-Siwe's and Hand-Schüller-Christian's diseases [8]. We report a case of bilateral temporal eosinophilic granuloma with otic capsule involvement, who was initially misdiagnosed and treated for non-specific otitis media. The diagnosis, radiologic findings and the management of this relatively rare disease are discussed.

2. Case report

A four-year-old boy who had intermittant otorrhea in both ears during the last year attended the Ear Nose Throat Clinic. Clinical examination showed a polypoid mass which had totally filled up external auditory canal. Bilateral profound sensorineural hearing loss was diagnosed with audiological test battery. At the age of 2 years, he had earache and otorrhea in the right ear and applied to Pediatric Clinic. Examination demonstrated purulent aural discharge in the right ear and normal findings in the left ear, and the systemic medical treatment was given. The patient was lost to follow-up after that initial diagnosis of right otitis media and was treated for chronic otitis media by other institutions.

Computerized tomography (CT) scanning was performed with a 64-detector (multi-slice) CT scanner (Aquillion 64, Toshiba, Tokyo, Japan). CT showed bilateral osteolitic defects without sclerotic margins filled with soft tissue masses involving the middle ear, mastoid, squama and petrous part of the temporal bone. The involvement was worse on the right side. Bilateral bony labyrinth was involved (Fig. 1). For the differential diagnosis of cholesteatoma and LCH, Magnetic Resonance Imaging (MRI) was performed. MRI of the temporal bone revealed bilateral involvement of inner ear with internal auditory canal involvement on the right. The sof t-tissue mass was relatively isointense to hyperintense on T1-weighted images (Fig. 2a), iso-hyperintense on T2-weighted images (Fig. 2b) and showed marked enhancement after administration of contrast material (Fig. 2c). For differentiation of LCH, osteomiyelitis and malignant tumors bilateral punch biopsy was performed under general anesthesia.

Microscopically, the tissue fragments contained a dense cellular infiltrate consisting of Langerhans' cells, eosinophils and scattered lymphocytes (Fig. 3a). There were small foci of necrosis. Langerhans' cells were immunoreactive for CD1a (Fig. 3b) and s-100 protein. The histopatological and immunohistochemical findings were consistent with LCH.

Mastoidectomy in the right ear was performed under general anesthesia. All mastoid cells completely blocked with the mass and lesion extended into the cochlea although the outer layer of the cochlea was intact. Postoperative pathology confirmed preoperative pathology.

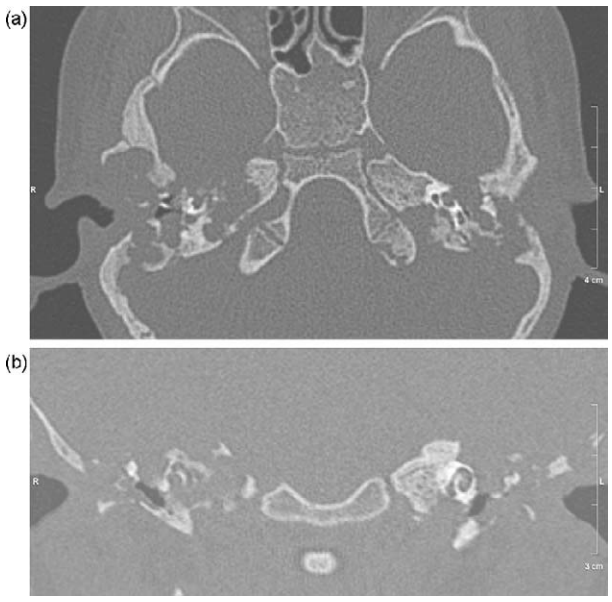

Figure 1 (a, b) Axial and coronal bone window computed tomography reveals destructive lesions of bilateral mastoid with bilateral otic capsule involvement.

Bone scintigraphy revealed no other lesions. As recommended by the French Langerhans' cell study group [9], he was treated with vinblastine and corticosteroid by pediatric hematologist for a year because of the multifocal bone involvement.

Fourteen months later, follow-up temporal bone CT revealed no soft tissue masses on both inner ears, and the bony lesions of the temporal bone were reossified and remodeled (Fig. 4a). The right lateral (Fig. 4b) and posterior (Fig. 4c) semicircular canals were dehissent and these canals were tortious and did not complete their normal configuration. Cochlea and superior semicircular canal were normal. On the left posterior semicircular canal was irregular and dehissent (Fig. 4d). Cochlea and other semicircular canals were normal on the left.

During the follow-up temporal bone CT, bilateral profound sensorineural hearing loss was present and therapy options such as cochlear implantation were discussed with the parents and the final decision was expected.

3. Discussion

Involvement of the temporal bone has been described in 11—61% of all cases of LCH [3]. Most authors agree that otologic involvement usually occurs during the course of multisystemic disease, although it may be the only symptom in 5—25% of patients [10]. Bilateral involvement of EG as