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Bilateral primitive cholesteatoma of external auditory canal with congenital stenosis

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1. Introduction

Cholesteatoma of external auditory canal (EAC) is a very rare disease, representing 0.1–0.5% of otologic disease. It is defined as an accumulation of epidermis associated with bone erosion osteitis, which is often ignored at an early stage.

Its diagnosis is primarily clinical, it has significantly benefited from the advances in imaging, which helped to differentiate it from other inflammatory diseases or tumor of the ear canal.

We describe an unusual case of bilateral congenital cholesteatoma EAC with stenosis, through its clinicoradiological features and offering clinicians our therapeutic attitude vis-à-vis this particular otologic entity.

2. Case report

A 5 year old, boy, without specific medical history, who consulted for bilateral stenosis of the external auditory meatus seen at birth by his parent.

Abbreviations: EAC, external auditory canal; CT, computed tomography.

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The evolution was marked by the installation of a bilateral hearing loss associated with delayed language acquisition. Clinical examination revealed a complete, bilateral stenosis of the external auditory canal without fistula or other malformation of both ears (Fig. 1).

The objective audiometry, showed a hearing threshold of 80 dB in the right side and 90 dB on the other. CT of the temporal bone objectived a polypoid tissue filling the right canal bulging in the middle ear. The same aspect in the left EAC, associated with enlargement and smoothing of the walls, a discreet ossicular lysis, eroded scutum and Tegman tympani dehiscence, we noted also bilateral fibrous stenosis of the external auditory meatus (Fig. 2).

At surgical exploration; on the left: presence of cholesteatoma in the EAC with lysis of its walls, extending to the middle ear, which imposed a modified mastoidectomy with Canaloplasty.

On the right side (in a second time): we noted the presence of the cholesteatoma in the right ear canal, lysing the posterior wall and depressing eardrum without invading it, thing that facilitated its dissection until its wholly ablation. Then we performed a reconstruction of the attic and the posterior wall by cartilage, also we conducted a canaloplasty with thin skin graft (Fig. 3).

The postoperative course was uneventful. During follow-up was not recorded recurrence. Post-operative hearing gain is 30 dB and 5 dB right to left, and anatomical results are very satisfactory (Fig. 4).

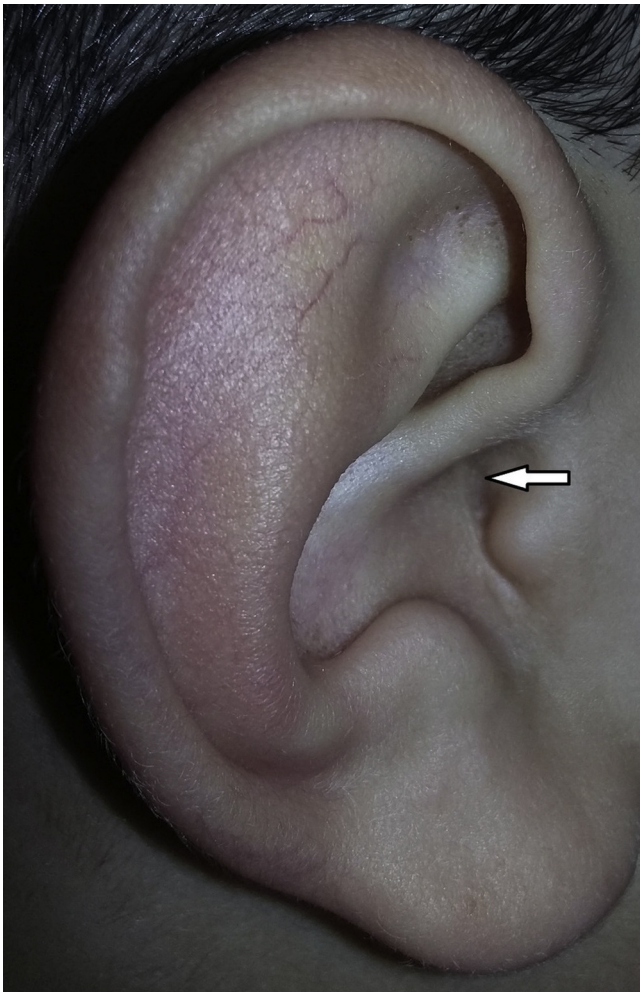


Fig. 1. Entire stenosis of the right external auditory canal.

3. Discussion

Cholesteatoma of EAC is very rare with an incidence of 0.1–0.5% of otologic disease [1,2]. It is gradually developing in the duct floor from epithelial dander that can invade the structures of the middle ear. The exact etiology is not well known [6–8]. In most cases, it is either spontaneous or it occurs after surgery or trauma to the ear canal, however obstruction or stenosis of the EAC was also described as a causative factor [9]. This accords with our patient, who had a bilateral congenital stenosis.

The diagnosis is made by physical examination and CT of the temporal bone [3]. Generally, the tympanic membrane is intact, except in the case of extensive cholesteatoma [4,5]. In our case the damage was severe at diagnosis especially the left side, and we were not able to visualize the characteristics of the mass by endoscopy due to bilateral stenosis. The CT of temporal bone was important to determine the site, severity and extension into adjacent structures. It remains the method of choice to study the EAC cholesteatoma, middle ear and mastoid.

Its differential diagnosis includes neoplasms of EAC, epidermal cap, epidermal cysts and external necrotic otitis [4,9].

The principle of surgical treatment involves the excision of the mass with its capsule and the necrotic bone. The chosen surgical technique depends on the site and extent of bone destruction. The conservative approach is indicated for limited lesions such as what was done to the right side with skin grafting. Modified radical mastoidectomy is indicated if the mastoid is invaded.



Fig. 2. CT scan of the temporal bone; axial section.

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