Giant Congenital Cholesteatoma of the Petrous Bone: Personal Experience and Literature Review

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Abstract

Congenital cholesteatoma of the petrous bone, though originating in childhood, has often a delayed diagnosis due to its poor symptoms and can grow achieving great extension. When asymptomatic or pauci-symptomatic, the diagnosis is incidental and purely radiological. Leaving the lesion untreated implies an inexorable progression leading to extensive erosion of the petrous bone and cranial nerves involvement. First choice treatment is radical surgical excision. An aggressive treatment is recommended, a combination of approaches to the petrous bone and skull base may be used to obtain complete removal. Surgical morbidity is a balance between the choice of the approach and the goal of radical excision. Prevention of relapse is essential not to expose the patient to further morbidity some years later. Long term follow-up is essential.

ABBREVIATIONS

CC: Congenital Cholesteatoma; MRI: Magnetic Resonance Imaging; CT: Computed Tomography; DWI: Diffusion Weighted Imaging; EPI: Echo-Planar Image

INTRODUCTION

Congenital cholesteatoma (CC) of the middle ear was first described by Howard House in 1953. CC is thought to be caused by inadequate folding of the epidermoid formation inside the middle ear cleft. Stratified squamous epithelium accumulates by inadequate folding of the epidermoid formation inside the middle ear cleft. Stratified squamous epithelium accumulates during the development of the middle ear mucosa through the 3rd and 5th weeks of embryonic life [1]. The incidence of CC is estimated from 1% to 5% of all cholesteatomas in most important series [2-4], although some authors esteemed they rise up till the 24% of all cholesteatomas in children [5]; diagnosis in adults is quite rare. CC is often asymptomatic for years, and may manifest in advanced stage, it may be revealed incidentally as a retro-tympanic mass during a routine clinical evaluation, or a myringotomy [3]. Poor symptoms often delay diagnosis. Common symptoms include otalgia, hearing impairment, vertigo and facial nerve palsy [6,7]. Extradural sites of involvement include middle ear cavity, external meatus, mastoid, squamous and petrous portions of the temporal bone [8]. Advanced disease could involve intradural tract of 7th, 9th, 10th, 11th, 12th cranial nerves extending through the auditory canal, the jugular foramen and the condylar canal respectively. A posterior intracranial extension may cause mass effect and obstructive hydrocephalus, or may involve inner ear structures [9]. Such destructive disease, with the mentioned neurologic symptoms, was observed in older children and in adults [1,7,10,11]. Higher incidence of recurrence was found to be associated with advanced disease and incomplete removal at 1st surgery; to date, the specific recurrence rate of giant cholesteatoma in adult is not reported, due to the rarity of the disease. Lesion growth and disease progression are straightly associated with age [1,2,13].

The first choice treatment for congenital cholesteatoma is radical surgical excision, since any residual could become a new growing disease. The attempt to preserve a useful or serviceable hearing is mandatory, when feasible. We present a rare case of petrosal giant CC extended to the middle ear, the mastoid, and the jugular foramen and along the mastoid, tympanic tract of the facial nerve, the geniculate ganglion, the supra labyrinthe area as well as the dura of the posterior cranial fossa.

CASE PRESENTATION

A 43 year old man presented to our tertiary referral centre for a left massive petrous bone lesion, detected incidentally by mean of a cerebral CT scan, performed in occasion of a syncope episode. The patient had history of left progressive hearing loss in the previous 4 years, suspected to be noise-induced and never investigated. No other symptoms were reported. At admission, high resolution petrous bone CT scan and cerebral contrast-enhanced MRI were performed. The first showed an extensive soft tissue mass involving the left petrous bone (Figure 1). Magnetic resonance imaging (MRI) suggested specific signal-intensity characteristics for cholesteatoma: high signal intensity on T2-weighted images, low signal intensity on unenhanced and
post contrast T1-weighted images with a thin rim enhancement on the late gadolinium-enhanced images and a very high signal intensity on DWI (Figure 2, Figure 4A). To date, no clear-cut definition of “giant” cholesteatoma is reported in literature. We refer to a lesion extending beyond the temporal bone to adjacent structures such as jugular foramen, internal carotid artery, and dura and intradural sites. A mass effect on the cerebello pontine angle was also observed.

A pure tone audiometry showed a left mixed hearing loss, with PTA of 70 db. Facial nerve function was clinically normal; an electromyographic study of the 7th cranial nerve was however required, showing signs of left neurogenic partial suffering with concomitant impairment of myelin fibers. The planned treatment was radical surgical excision. The patient underwent a sub-total petrosectomy combined with a sub-temporal access (Figure 3).

The approach involved the resection of the mass, with soft and not infiltrating margins from the area of erosion in the skull base and was dissected from the facial nerve, without evidence of intra nervous infiltration. The facial nerve was completely uncovered in all its portions. The subtotal petrosectomy was combined with a temporal craniectomy to expose and remove the supralabyrinthine cholesteatoma and to dissect it from
the geniculate ganglion. An area of dura of the posterior fossa was resected since the matrix was there firmly adherent. A fat obliteration prevented any cerebrospinal leakage, the external auditory canal was sutured and the Eustachian tube was closed.

The postoperative was regular, the patient reported no major complications and was discharged with a normal facial nerve function (I grade, House Brackmann). Antibiotic therapy was administered during the recovery.

The follow up was planned with yearly contrast enhanced and DWI MRI. The first one was asked after six months as baseline exam. 18 months after surgery the patient showed no signs of relapse (Figure 4B).

**DISCUSSION**

Pathogenesis of CC is difficult to establish since the incidence of the disease is low and a proper direct examination of the whole middle ear in children or asymptomatic infants is not always feasible [14].

A debated question is still present in literature, whether this disease is truly congenital or acquired. It was suggested that ectodermal tissue from the external acoustic meatus may migrate into the middle ear cavity due to failure of the inhibitory function of the tympanic ring [15]. The most amenable theory is based on the presence of residual embryonic epithelial tissue in the middle ear [16]. There is increased evidence of CC on affected family [17] and on identical twins [18].

CC may be completely asymptomatic, especially when arising in silent regions such as the mastoid, and the diagnosis occurs incidentally by radiological imaging. As a result, the diagnosis is often delayed until mid to late adulthood. It could manifest itself with progressive hearing loss, dizziness, otalgia, tinnitus, temporal bone swelling, facial nerve palsy, cerebrospinal fluid leak [6,19,20].

When diagnosis is delayed in adulthood as in our case (4x4x3cm), the lesion may be much extended and is referred to as giant CC. In the adult and the elderly, the disease may involve unusual sites, and mixed hearing loss is the most common symptom [8]. Facial nerve palsy could also be present although less frequently then in acquired cholesteatoma [10]. In the case of a delayed diagnosis of a giant lesion, radical surgery is mandatory, with complete removal which implies extended or combined surgical approaches to the petrous bone. Surgery may be affected by higher morbidity such as labyrinthine fistula, facial paralysis, CFS leak, cranial nerves impairment (9th to 12th according to the higher morbidity such as labyrinthine fistula, facial paralysis, CFS leak, cranial nerves impairment (9th to 12th according to the

The lesion extends to the jugular foramen. Lesion size increases linearly with age; this finding emphasizes the importance of early detection and management. This is to prevent complications and reduce the risk of relapse. A combination of approaches to the petrous bone and skull base may be used [21] and an aggressive treatment is recommended to obtain complete removal. Surgical morbidity is a balance between the choice of the approach and the goal of radical excision. Prevention of relapse is essential not to expose the patient to further morbidity some years later. Long term follow-up is essential.

**REFERENCES**


2. Darrouzet V, Duclos JY, Portmann D, Bebear JP. Congenital middle ear

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