Giant Cholesteatoma of the Temporal Bone

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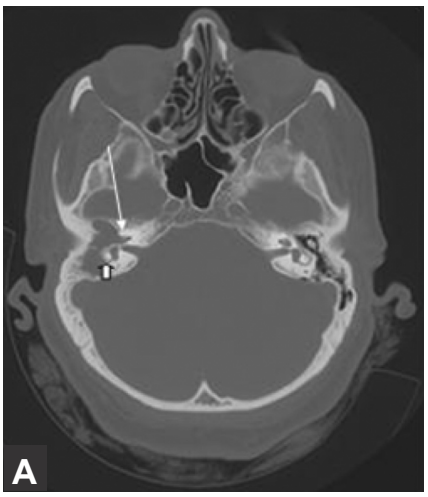
It is known that the destructive nature of cholesteatoma, its tendency to progress and erode the bone locally, and the fact that its growth, though benign, is practically non-self-limiting, can cause potentially severe intracranial complications. As a result of this awareness these complications now occur only rarely in developed countries, where in most cases established surgical techniques are used by well-trained otologic surgeons to treat the disease at an early stage. Nevertheless, some patients with cholesteatoma remain untreated to the point where life-threatening complications do occur. However, published descriptions of this entity are rare. In 1985 Arkin et al. [1] reported a case of an aggressive giant cholesteatoma that killed the patient by cerebellar invasion. In 1993, Borgstein et al. [2] reported a case of a giant congenital cholesteatoma of the temporal bone in a patient with longstanding unilateral hearing loss. Topaloglu and co-workers [3] in 1997 reported a case of a giant cholesteatoma extending through the temporal bone cortex and presenting as a postauricular mass. Rodriguez and team [4] in 2001 described a patient with a giant cholesteatoma of the temporal bone with an intact tympanic membrane. In 2003, Uygur et al. [5] described a patient with a giant middle ear cholesteatoma that had invaded the posterior cranial fossa leading to lateral sinus obliteration, while the clinical presentation was minimal.

We describe two patients, each with an unusually large and life-threatening cholesteatoma. Neither patient had ever been examined by an ENT specialist, although in both cases a cholesteatoma had clearly existed for many years before the patients were referred to us with obvious otologic signs. During that period they were seen, however, by general physicians who had apparently missed these signs.

Patient Descriptions

Patient 1

A 59 year old woman was referred to our department with right facial nerve palsy that had developed during the 3 months prior to the current presentation. There were no complaints of hearing loss, otalgia, or vertigo. The patient had a history of several episodes of otorrhea from the right ear during childhood, but in the last few years the ear had been dry. Upon referral, otoscopy revealed an intact tympanic membrane with a mass behind it. Audiometry showed right mixed (conductive and sensorineural) hearing loss. High resolution computed tomography of the temporal bone showed clouding of the right middle ear and mastoid with extensive bone destruction [Figure A]. The tegmen, roof of the temporomandibular joint, semicircular canals, cochlea, and lateral tip of the internal acoustic canal were eroded. The geniculate ganglion could not be identified because it was obscured by the disease. Magnetic resonance imaging showed a high-signal lesion on T2, a moderately low-signal lesion on T1 with rim enhancement after gadolinium infusion, consistent with the MRI diagnosis of cholesteatoma. The patient underwent right lateral temporal bone resection (radical mastoidectomy) with meatoplasty. At operation the dural plate was found to be largely eroded and there was extensive destruction of the semicircular canals by the cholesteatoma, which had penetrated the lumen of the lateral and posterior semicircular canals. The geniculate ganglion was exposed over 180 degrees of its circumference, and the tympanic segment of the facial nerve was completely exposed (360 degrees) and surrounded by the cholesteatoma, which had also penetrated medial to it, reaching the labyrinthine and intracanalicular segments of the facial nerve. The lateral end of the internal acoustic canal was eroded. The surgical findings corresponded closely to the preoperative HRCT findings. After complete removal of the cholesteatoma, leakage of cerebrospinal fluid was observed from an opening at the lateral part of the internal acoustic canal. The opening was obliterated with temporalis fascia, muscle tissue, and biologic glue. The postoperative period was uneventful. Follow-up examination 1 year after surgery showed a dry radical mastoidectomy cavity. Facial nerve function did not recover.

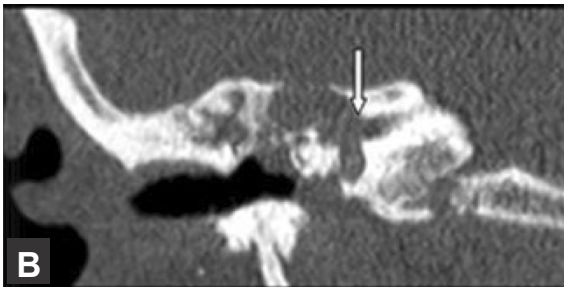


[A] Patient 1. Axial HRCT slice showing erosion of the right cochlea (long white arrow) and lateral semicircular canal (short white arrow).

Patient 2

A 68 year old woman was referred to us from the internal medicine department,

HRCT = high resolution computed tomography



[B] Patient 2. Coronal CT slice showing temporal bone erosion extending to the internal acoustic canal (arrow).

where she had been admitted with fever and urosepsis. During her hospitalization there she had complained of headaches and was examined by a neurologist, who noticed right-sided complete facial nerve palsy, which according to her history had existed for many years. Brain CT revealed a lesion of the right temporal bone. There were no complaints of otalgia or vertigo, but the patient mentioned a gradually progressive hearing loss over the last few years. There was no history of aural drainage. Otoscopy revealed right tympanic membrane perforation through which the middle ear cavity was seen to be completely occupied by soft tissue. On the left side there was a dry central perforation. Audiometry showed a profound mixed hearing loss bilaterally. HRCT of the temporal bone revealed a giant mass obliterating the right middle ear and mastoid with extensive bone destruction. The tegmen, semicircular canals, cochlea and fundus of the internal acoustic canal were eroded [Figure B]. The mass extended beyond the tympanomastoid space and involved the middle and posterior cranial fossa. MRI showed a high-signal lesion on T2 and a moderately low-signal lesion on T1 with rim enhancement after gadolinium infusion, typical of cholesteatoma. Despite repeated explanations about the life-threatening nature of her disease and the need for prompt surgical treatment, the patient refused surgical intervention.

Comment

The small number of published reports on the subject probably does not reflect the true prevalence of patients with life-threatening giant cholesteatomas seen by family physicians or pediatricians. This contention is supported by the present report. We describe two elderly women with giant cholesteatoma in whom disease had existed for some years, and who were first seen in our department only after facial nerve palsy or 'brain mass' appeared. In both patients HRCT and MRI of the temporal bone showed a giant cholesteatoma in the middle ear and mastoid, causing extensive destruction of the temporal bone including the cochlea, vestibular labyrinth, and internal acoustic canal.

In conclusion, cholesteatoma can exist for years as a non-aggressive state, with minimal symptoms that might seem mild to the family physician and therefore remain untreated until their potentially dangerous character becomes obvious. Otologists have a responsibility to heighten awareness among general practitioners and pediatricians of the potentially life-threatening nature of cholesteatoma and the importance of proper attention by the family physician to seemingly mild otologic symptoms.

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