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CASE REPORT

Hearing Improvement after Partial Labyrinthectomy

Resection of petrous apex cholesterol granuloma

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تحسن السمع بعد استئصال الورم الحبيبي الكولسترولي من قمة العظم الصخري عن طريق استئصال جزئي للتيه

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الملخص: نقدم هنا حالة رجل عمره 57 عاما كان يعاني من ورم حبيبي كولسترولي في قمة العظم الصخري مع أعراض ضعف الأعصاب القحفية وصمم الأذن اليسرى. تم إختيار التدخل الجراحي لإستئصال الورم عبر التيه. نتج عن هذا تحسن وظيفة الأعصاب القحفية ومنها عمل الوجه. كما تحسن السمع في الأذن المصابة في غضون ثلاثة أشهر بعد الجراحة. لم يشهد المريض أية مضاعفات بعد الجراحة. نستنتج أنه يمكن استخدام التدخل الجراحي لإستئصال الورم الحبيبي الكولسترولي عبر التيه مع إحتمال تحسن السمع.

كلمات البحث: التيه ؛ سمع ؛ كولسترول ؛ ورم حبيبي ؛ تقرير حالة ؛ المملكة العربية السعودية.

ABSTRACT: We present the case of a 57 year-old male presenting with symptomatic petrous apex cholesterol granuloma, multiple cranial nerve weaknesses and deafness of the left ear. The chosen intervention was a cholesterol granuloma resection via the translabyrinthine approach. This resulted in cranial nerve recovery and improved facial functionality. Hearing in the operated ear improved within 3 months after surgery. The patient experienced no postoperative complications. We conclude that a translabyrinthine approach can be used for drainage of petrous apex cholesterol granulomas with a chance of hearing loss recovery

Keywords: Labyrinth; Hearing; Cholesterol; Granuloma; Case report; Saudi Arabia

Cholesterol granulomas (CGs) are also known as cholesterol cysts, unicameral cysts, xanthomas, or chocolate cysts of the temporal bone. They are benign, expansile, round, or ovoid cysts containing cholesterol crystals surrounded by foreign body giant cells and chronic inflammation, all contained within a thick fibrous capsule.

The petrous apex is the region of the petrous pyramid bounded medially by the clivus, laterally by the inner ear, anteriorly by the carotid, and posteriorly by the dura of the posterior fossa. The petrous apex CG was not recognized as a distinct entity until the mid-1980s.[1] CGs are ten times more common than cholesteatomas and forty times more common than petrous apex mucoceles.[2] Its occurrence in the petrous portion of the temporal bones is of particular interest because of its effect on a variety of cranial nerves and its surgically challenging location. The exact cause of CGs is unknown, but it is thought to be cause by blocked air cell tracts in well-pneumatised petrous apex spaces. As the mucosa absorbs air negative pressure, hypoxia develops, which leads to mucosal oedema, blood vessel rupture, and haemorrhage. Catabolism of haemoglobin leads to the formation and accumulation of blood products such as cholesterol, fibrin, and haemosiderin. These materials subsequently induce a foreign body reaction, resulting in fibrosis, neovascularisation, and CG formation. The granuloma continues to enlarge as new vessels haemorrhage and the cycle repeats itself.

Mass effects of petrous apex CGs can result in a variety of symptoms such as: unilateral hearing loss, vertigo, headaches, tinnitus, facial numbness,

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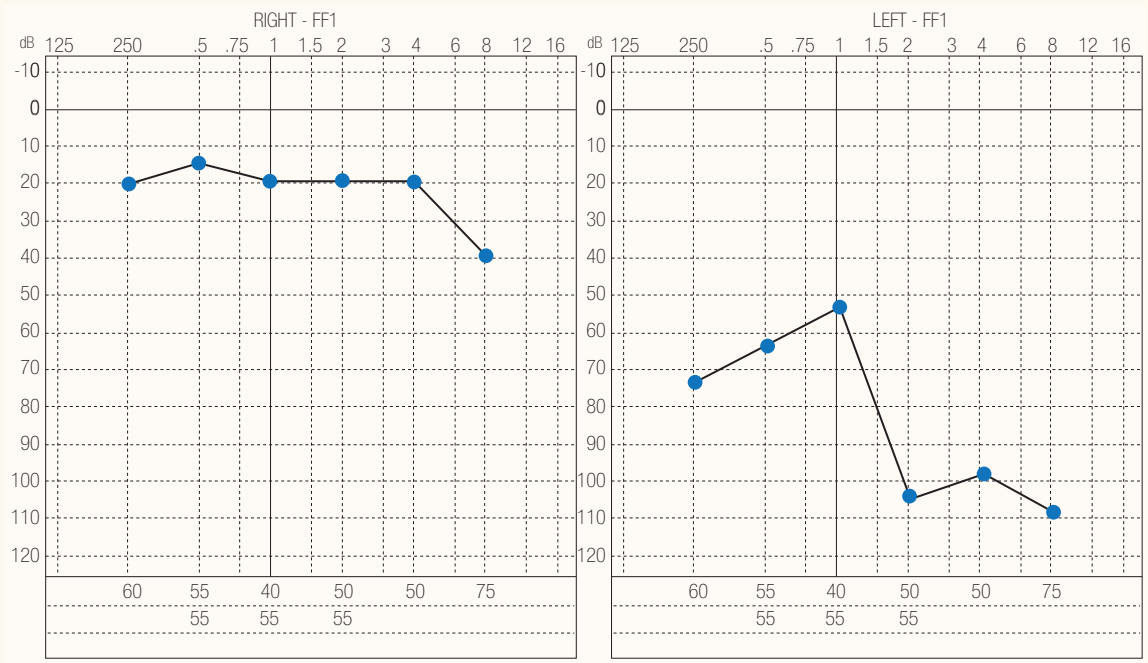


Figure 1: Preoperative audiogram documenting sensorineural hearing loss in the patient's left ear.

diplopia, and cranial nerve deficits. As the lesions enlarge, signs of cerebellopontine compression can be seen.

Differential diagnoses of a petrous apex CG include: cholesteatoma, mucocele, chordoma, petrous apicitis, metastasis, and epidermoid cysts. Thus, accurate preoperative diagnosis of petrous apex lesions is critical, because the surgical approaches used for this region vary depending on the specific disease process involved. It is best diagnosed by magnetic resonance imaging (MRI). Partial labyrinthectomy, where only the semicircular canals are drilled without opening of the vestibule, is one option for draining large CG at the petrous apex with hearing preservation. To our knowledge, this case is the first report on improvement in hearing after translabyrinthine surgery for a CG.

Case Report

A 57 year-old man presented at King Abdulaziz University Hospital, Riyadh, Saudi Arabia, with a five-month history of progressive left-sided hearing loss, facial weakness, and numbness. Seven months before presentation to our institution, he had undergone subtotal excision and decompression of the CG via a middle fossa approach, which was performed by another surgeon. He had no history of tinnitus, vestibular dysfunction, or middle ear disease. On examination, both ears appeared normal, but left-side grade VI facial weakness (according to the House-Brackmann scale) was observed.

An audiogram showed severe sensorineural hearing loss in the left ear [Figure 1]. A computed tomography (CT) scan revealed extensive erosion of the petrous bone [Figure 2]. Magnetic resonance imaging (MRI) showed that the lesion was hyperintense on both pre-contrast and post-

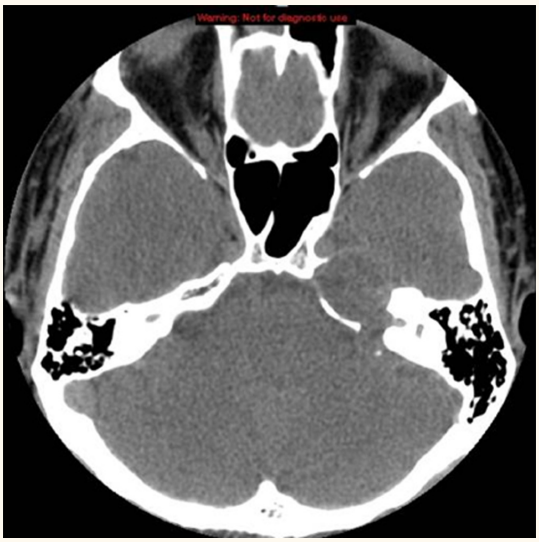
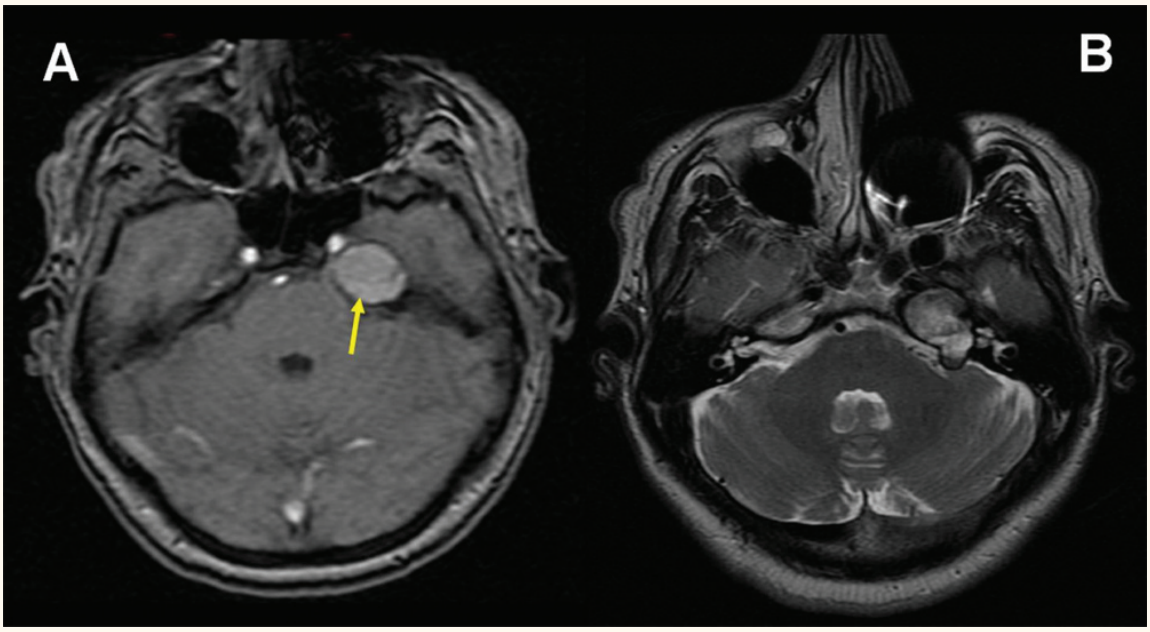


Figure 2: Axial computed tomography scan of the temporal bone demonstrating a well-defined expansile lesion in the left temporal bone isodense to brain tissue



Figures 3A & 3B: A: Magnetic resonance imaging (MRI) (T1-weighted) image showing the mass (arrows) to be hyperintense in signal, which is characteristic for cholesterol granulomas (CGs). **B**: T2-weighted MRI scan showing high-signal lobulated masses in the petrous apex compatible with CGs.

contrast T1- as well as T2-weighted images [Figure 3].

We planned a translabyrinthine as well as transcochlear approach for wide excision and drainage of the lesion in view of the hearing loss. A facial nerve monitor was set in place, and a partial labyrinthectomy approach was done where the lateral, posterior and superior semicircular canals are drilled, but the vestibule was left undisturbed. Drilling was continued until the gelatinous blue-brown mass was identified. When the mass was opened, it oozed the typical thick, viscous, brownish fluid, which had a characteristic sheen to it. We had wide access to the petrous apex and were able to excise the lesion. Thus, the transcochlear approach that was part of the intervention plan was not necessary. The window in the labyrinth was left open for drainage of the petrous apex.

No complications or adverse consequences of the petrous apex surgery (e.g. postoperative headache, cerebrospinal fluid leak, and meningitis) were noted. In fact, immediately after the surgery, the patient noticed improved facial functionality, which further improved over time. Three months after the operation, the patient reported improved hearing in the operated ear. Ear examination revealed an intact tympanic membrane and the Weber response lateralised to the operated ear. His audiogram, with masking of air and bone, showed only moderate mixed hearing loss in the operated ear [Figure 4].

Discussion

Cholesterol granulomas cannot usually be detected by otoscopic evaluation, and it is difficult to diagnose them before the operation without appropriate imaging. While CT and MRI scans have improved the accuracy of preoperative diagnosis of petrous apex pathology, these imaging techniques are most helpful when used in combination. When systematically applied, the combination of contrast CT and MRI (with or without gadolinium) permits a narrowing of the extensive differential diagnosis.

CT scans of CGs typically show a well-defined, sharply marginated, non-enhancing expansile mass that breaks down the septate structure of the pneumatised petrous apex. It has a density similar to that of brain tissue and it is frequently limited to the petrous apex. Remodeling and erosion of the bone can occur.

While CT scanning can provide superior evaluation of the bony structures of the temporal bone, MRI scanning can aid in the detection of intracranial extension and the involvement of adjacent structures, such as the carotid artery and sigmoid sinus. CG is often unique and diagnostic on

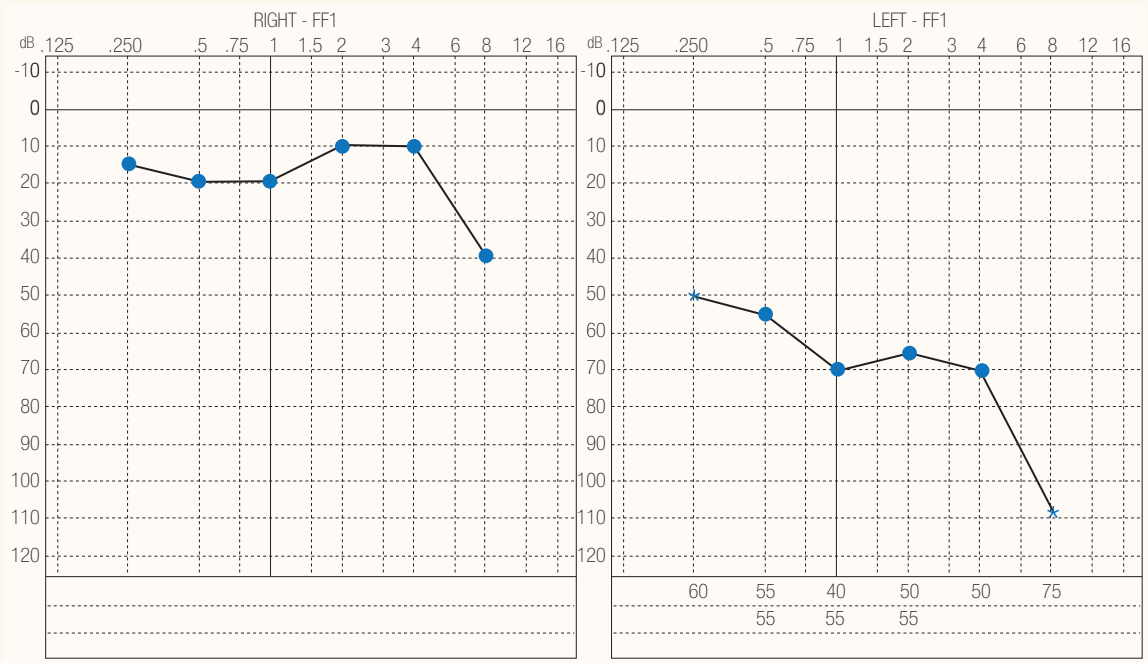


Figure 4: Postoperative audiogram demonstrating mixed hearing loss on the patient's operative side.

MRI, showing hyperintense T1- and T2-weighted images and lacking enhancement with gadolinium infusion, as seen in our patient. Moreover, postoperative MRI can help in the verification of complete removal of the CG and in the evaluation of complications, as well as recurrence or formation of complicating granulation tissue.

In our case, the high-signal characteristics of the CG within the petrous apex allowed reliable distinction of this lesion from other deferential diagnoses. In general, management of CGs includes surgical draining and permanent aeration of the air cell. It is believed that total removal is not necessary in all cases due to the lack of epithelial lining. Therefore, the management of CGs is dictated by the location of the lesion relative to adjacent neurovascular structures in the temporal bone, complications, and recurrence, as well as the status of the patient's hearing.

Since CGs usually arise in extensively pneumatised temporal bones, several well-developed possible access routes usually exist, and many approaches have been used successfully. If hearing is to be preserved, the infra-labyrinthine, infra-cochlear trans-sphenoidal, and middle cranial fossa approaches are the main non-invasive routes. Most commonly, the infra-labyrinthine and infra-cochlear approaches are used by otologists. However, recurrences are not uncommon, and may occur in up to 60% of cases, which may require further revision.[3] These recurrences may be related to closure of the surgically created tract with new bone formation, granulation tissue, or fibrosis, or may be secondary to obstruction of a surgically placed drainage catheter.

Although the translabyrinthine approach provides the most direct route and wide exposure for aeration from the mastoid through the surgical defect to the petrous apex, it has previously been considered to be incompatible with hearing preservation and was only used in cases where hearing and vestibular function were absent. However, most patients with CGs have excellent hearing. In this case, a partial labyrinthectomy approach with hearing preservation was used; it is interesting to note that this is led to improvement in hearing after this surgical procedure.

In patients with large and multi-loculated petrous apex CGs, there may be extensive critical structure involvement, including the cranial nerve, brain, jugular foramen, carotid canal, and internal auditory canal. In this subset of patients, it may be impossible to remove the CG without wide drainage from multiple access routes, which was the planned approach in our patient, with translabrynthine, as well as transcochlear, approaches. This approach was used because our patient did not meet the well-accepted Gardner-Robertson preoperative

criteria of the "50/50 rule" for useful hearing, in which subjects with pure-tone audiometry > 50 dB and speech discrimination score < 50% are not candidates for hearing preservation.

In a previous report in 1991, McElveen *et al.*[4] modified the traditional translabyrinthine approach for removal of an intracanalicular acoustic schwannoma by sealing the vestibule with bone wax, which allowed hearing function to be preserved in one patient. Two years later, the authors reported two cases in which the same technique was used,[5] but few other authors have managed to obtain similar results in case reports.[6,7] Moreover, case series were reported with hearing preservation in all patients[8,9] or in some patients.[10,11] In these reports, partial labyrinthectomy approaches were used for different lesions at the petrous apex or the cerebellopontine angle. Our case is unique because there was marked improvement in inner ear function after partial labyrinthectomy. Although our patient was followed up for a short time after surgery, the partial labyrinthectomy approach has been shown to maintain serviceable hearing for a long time.[12,13,14]

We propose two theories to explain the recovery of hearing observed in our patient, namely, neurovascular compression and ototoxicity. First, it is possible that our patient experienced compression of the arterial or nerve supply to the cochlear, which resulted in transient ischemia or neuropraxia, respectively; this had not yet progressed to irreversible Wallerian degeneration. Second, toxins from the CG are another theoretical cause of this transient hearing loss, as similar recovery of hearing was also reported with acoustic neuroma after middle fossa tumor resection.[15]

This case report is instructive in that many otologists would consider hearing to be irretrievably lost, on the basis of the preoperative audiogram, and may thus plan a wide translabyrinthine or transcochlear approach. This case illustrates that a partial labyrinthectomy approach may actually result in recovery of hearing, which is a useful functional result. We therefore believe that this case report is an important contribution to the literature.

Conclusion

Partial labyrinthectomy is an option for petrous apex lesion and we recommend consideration of this approach for extensive petrous apex lesions, or for access to the cerebellopontine angle, even in the presence of moderate to severe hearing loss.

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