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Endoscopic-assisted exenteration of massive petrous apex cholesteatoma misdiagnosed as Bell's Palsy

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Abstract. *Problem/objective:* Multiple differential pathologies can occur at the petrous apex. Clinicians must be highly suspicious of temporal bone pathology to diagnose lower motor neuron facial nerve palsy, particularly in patients with hearing impairments. The clinical presentation of a petrous apex lesion is variable, depending on the involvement of intimately adjacent structures. It cannot be diagnosed accurately from clinical findings alone. Hence, computed tomography and magnetic resonance imaging provide complementary data for more definitive characterization. Here, we described a case of massive petrous apex cholesteatoma in a young adult female that had been misdiagnosed with Bell's palsy for years. We aimed to highlight a safe treatment approach that could preserve hearing.
Methodology: We used a supralabyrinthine approach for an endoscopic-assisted transmastoid removal of the petrous apex lesion.
Results: Post-operative residual hearing was preserved and healing was good. **Conclusion**: Endoscopy approach could be a less invasive and effective way in treating PBC without compromising the ultimate aim of complete cholesteatoma removal.

Introduction

Petrous bone cholesteatoma (PBC) is defined as a cholesteatoma that forms medial to the otic capsule. PBC accounts for 4–9% of all petrous pyramid lesions and for 3% of all chronic otitis media with cholesteatoma.[1] PBC lesions are classified as either congenital or, more frequently, acquired, but they are histologically indistinguishable. Congenital, or primary, petrosal cholesteatoma is believed to arise from embryonic cells that reside within the petrous temporal bone, due to abnormal development of the first branchial groove. On the other hand, acquired, or secondary, PBC is believed to arise from the medial extension of an acquired middle ear cholesteatoma.

Case report

A 29-year-old woman had a 7-year history of progressive right-sided facial weakness. She also complained of reduced hearing and an occasional right temporal headache with otalgia. Nonetheless, there was no ear discharge, dizziness, vomiting, or body weakness. There was no history of head trauma. Seven years ago, her condition was diagnosed as Bell's palsy and treated at the district hospital. She had been an excellent student pursuing her final semester in a local university, but she had to drop out 4 years ago, after developing schizophrenia. She was living alone, due to a poor socioeconomic background. Her psychiatrist referred her to our clinic, after her condition was deemed treatment resistant. Upon examination, she demonstrated good orientation and obeyed commands. However, further testing indicated a right-sided House Brackmann grade V facial nerve palsy with right abducens nerve palsy. Other cranial nerves were normal, and cerebellar signs were absent. An otoscopic examination produced normal findings. A pure tone audiogram showed moderately mixed hearing loss in the right ear and normal hearing in the left ear. High resolution computed tomography (CT) and magnetic resonance imaging (MRI) revealed an expansile lesion, suggestive of a cholesteatoma, within the right petrous apex, which

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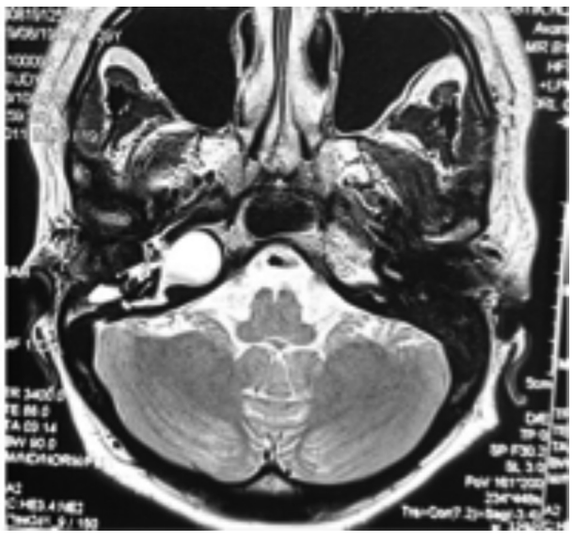

Figure 1

Hyperintense expansile lesion within the right petrous apex bone on MRI T2 sequence.

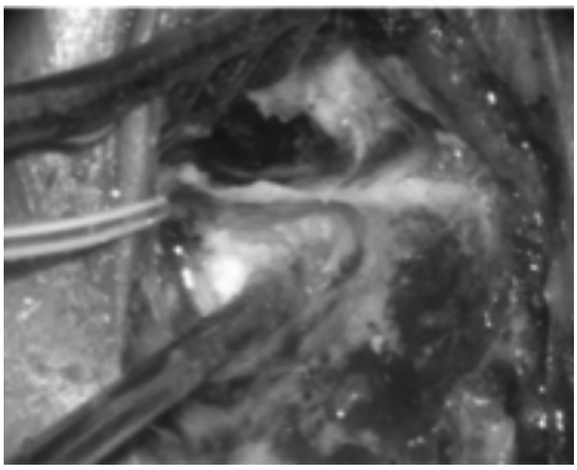

Figure 3a

Cortical mastoidectomy (transmastoid) as an initial approach to the petrous apex.

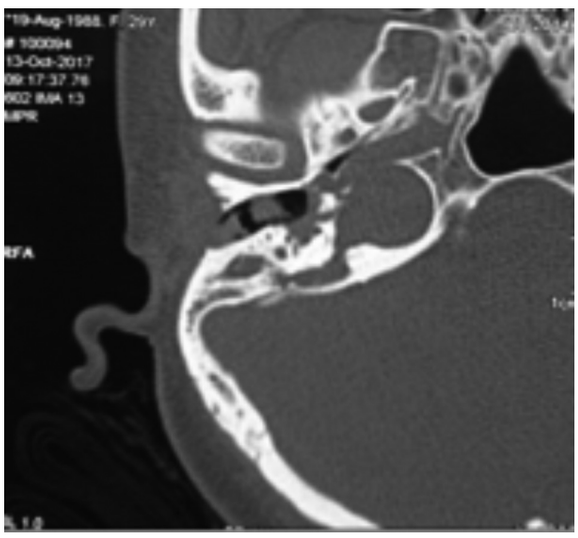

Figure 2

Heterogenous soft tissue lesion occupying the right petrous apex extending into middle ear, aditus ad antrum and Prussak's space.

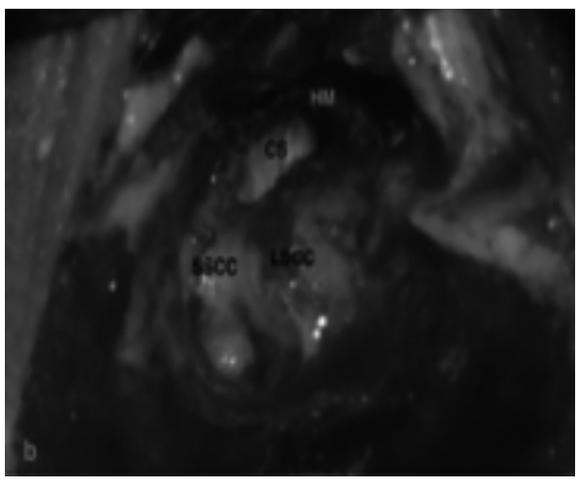

Figure 3b

Microscopic view of the mastoid bowl after CWD. It showed a huge cholesteatoma sac extending to petrous apex from junction between SSCC and LSCC. [CWD = canal wall down; CS = cholesteatoma sac; HM = handle of malleus; LSCC = lateral semicircular canal; SSCC = superior semicircular canal]

caused obliteration of the right internal auditory meatus (Figures 1,2). It extended to the middle ear, the aditus ad antrum, and Prussak's space.

We performed a right modified radical mastoidectomy, followed by a supralabyrinthine approach combined with an endoscopic technique to remove most of the cholesteatoma matrix; a small flake remained, which was adherent to the exposed carotid artery (Figure 3). We did not perform any reconstruction, in view of the small size of residual lesion over the carotid artery. Post-operatively, both the facial nerve function and the hearing threshold remained unchanged. However, subjectively, both the frequent headache and otalgia had resolved. There has been no sign of recurrence noted during the 12-month follow up after surgery (Figure 4).

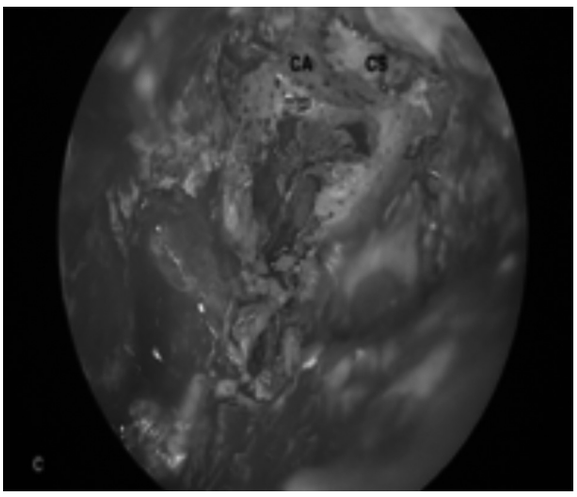

Figure 3c

Endoscopic view using 300, by following the cholesteatoma sac showed in figure 3b. Near complete removal was done with small flakes left on exposed dehiscenced carotid artery.

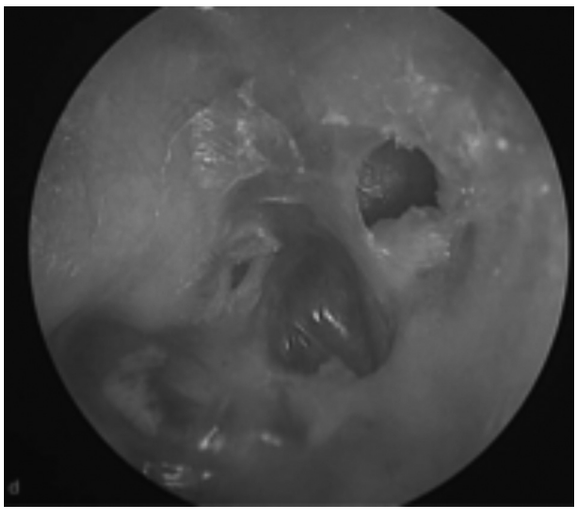

Figure 4

Post-operative view 3 weeks, showed a well healed cavity with minimal crusting

Discussion

The main presenting symptoms of PBC are hearing loss and facial weakness, due to the invasion of a cholesteatoma into the labyrinth or fallopian canal. Vertigo/imbalance and ear discharge also affect nearly a third of patients. Tinnitus, otalgia, and headache are less common manifestations. PBC must also be considered in patients that present with recurrent bacterial meningitis, lateral rectus palsy, due to sixth cranial nerve involvement, or trigeminal neuralgia.

Clinical findings from patient symptoms and signs, together with a preoperative hearing test, are frequently insufficiently specific to determine a conclusive diagnosis of PBC. Imaging techniques, like high resolution CT and MRI, are more reliable methods for narrowing down a differential diagnosis of the lesion. Preoperatively, the size, morphology, anatomical site, and extent of the lesion should be assessed, because these features are critical for surgical planning. High resolution CT provides details of bony destruction and additional features like elevation of the petrous ridge associated with PBC.[2] With MRIs, clinicians can differentiate cholesteatomas from other lesions, such as cholesterol granulomas. Indeed, cholesteatomas are only hyperintense on a T2-weighted MRI, and cholesterol granulomas are hyperintense on both T1 and T2-weighted MRIs.[2] Cholesteatomas can also be distinguished from mucoceles based on the presence of restricted diffusion with diffusion-weighted imaging, and they can be distinguished from benign tumors based on the absence of post-contrast enhancement.[3]or may be detected during an investigation for an unrelated disease. CT and MRI are complementary in providing an appropriate differential diagnosis and in aiding surgical planning. This pictorial review appraises the anatomy and contents of the petrous apex. Benign (e.g. cholesterol granuloma, cholesteatoma

PBC classifications have evolved since Fisch proposed two classes (supralabyrinthine and infra-labyrinthine-apical). An additional translaby-rinthine category was proposed by Bartels. Then, five classes were proposed by Sanna et al (supralabyrinthine, infralabyrinthine, massive laby-rinthine, infralabyrinthine-apical, and apical). Recently, Moffat et al. proposed seven classes. (supralabyrinthine, infralabyrinthine, massive laby-rinthine, supralabyrinthine apical, infralabyrinthine apical, massive labyrinthine apical, and apical).[4]

PBC surgery is challenging, because many difficulties confront otoneurosurgeons during the complete removal of a cholesteatoma matrix. This matrix often adheres to or invades the surrounding vital structures, including the dura, internal carotid artery, jugular bulb, sigmoid sinus, labyrinth, and the facial and lower cranial nerves. Moreover, it is often difficult to obtain an adequate visual field during surgery. Nonetheless, the mainstay of treatment is surgery, and the goal is complete removal to prevent recurrence.

The choice of surgical approach depends on the location and extent of the lesion, the hearing status on the affected side, and the anatomic positions of the internal carotid artery and jugular bulb. The selected approach should provide safe, adequate exposure to allow the surgeon to identify and preserve vital structures. When there is no evidence of inner ear involvement, inner ear sparing approaches are adopted, such as transmastoid or middle cranial fossa approaches. On the other hand, when there is definite evidence of inner ear involvement, a translabyrinthine, transotic, or subtotal petrosectomy with labyrinthectomy approach is performed to obtain wider exposure in addressing the cholesteatoma. An endoscopic trans-sphenoidal approach has also been introduced. This approach provides several benefits: the patient recovers more rapidly, a craniotomy is avoided, and vital structures can be preserved. Despite literature that might advocate one or more approaches over another, depending on the PBC classification, a randomized controlled trial for comparing these approaches has never been conducted. Even the most experienced surgeons could be biased toward a few approaches, based on their surgical skills and preferences.

In this case study, we elected a transmastoid supralabyrinthine approach, because the PBC had predominantly grown along the supralabyrinthine area from the petrous apex and had extended posteriorly, toward the superior part of the mastoid. The advantage of the transtemporal/transmastoid approach was that it provided the most direct route to the pathology for bone removal, and it avoided the potential risk involved in a dural retraction. Moreover, the supralabyrinthine approach to PBC allowed minimal disruption to labyrinthine structures; thus, hearing could be preserved. However, an extensive cholesteatoma might require combining the supralabyrinthine approach with a classical middle fossa craniotomy.

A variety of approaches can be used in patients with massive labyrinthine cholesteatomas. A subtotal petrosectomy is appropriate for infralabyrinthine lesions, with or without an apical extension. The transotic (translabyrinthine-transcochlear) and transcochlear approaches enable access to the petrous apex and clivus.[4] Fisch described several infratemporal fossa approaches that enabled access to more anterior structures, including the internal carotid artery.[4] We performed a "minimally invasive" supralabyrinthine approach, which allowed us to avoid disrupting the labyrinthine structures. We followed the cholesteatoma sac between the superior semicircular canal and the lateral semicircular canal to the petrous apex. We used rigid endoscopes with angulations of 0^0 and 30^0, a length of 14 cm, and an outer diameter of 4 mm (Storz, Germany). Figure 3c clearly shows that, without endoscopic assistance, it would not have been possible for the senior surgeons to reach the lesion at the petrous apex. The endoscope allowed visualisation of the deeper structures situated at angles away from the microscopic line of sight. In addition, deeper hidden corners could be explored to enable complete removal of the lesions and to avoid critical neurovascular structures.[5]

In this case, our senior surgeons decided to leave some small keratin flakes, which were attached to the exposed, pulsating carotid artery (Figure 3c). Complete removal of these flakes carried the risk of rupturing the artery. Moreover, this patient was unlucky to have schizophrenia and a poor socioeconomic background. Morbidity or mortality from a carotid artery injury would have done her more harm than running the risk that the remnants might develop into a recurrence. We believed that the tract that we had opened to approach the petrous apex was adequate for ventilating the petrous apex, which could reduce the risk of disease recurrence.

After surgery, the patient showed no complications, her residual hearing was preserved, and some of her presenting symptoms had resolved. This outcome should improve her quality of life and facilitate the management of schizophrenia. Once her psychiatric symptoms have been well controlled, we plan to prescribe a conventional bone conduction hearing aid for hearing rehabilitation.

Joan et al. reported that the recurrence rate of cholesteatoma after the combined endoscopic and microscopic technique was similar to the rate after a conventional canal-wall-down surgery.[6] Giuseppe et. al also showed that hearing preservation and healing time were better when endoscopy was employed in cholesteatoma surgery.[7] With improvements in instrumentation, techniques, and knowledge, endoscopic surgery has become a valuable adjunct to the operating microscope.[8] However, despite the usefulness of endoscopy, the possibility of leaving inspissated keratin or injuring the critical neurovascular structures remains a valid concern.[8]

Conclusion

Endoscopy provides superior visibility in areas of the petrous apex that are difficult to access. Naturally, it is changing conventional surgical approaches for treating petrous apex cholesteatoma. Patients must be carefully selected before offering this "minimally invasive" supralabyrinthine approach, rather than conventional radical surgeries. This study further demonstrated that an endoscopy approach could be effective in treating PBC without compromising the ultimate aim of complete cholesteatoma removal.

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