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Case Report

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Cholesteatoma of petrous bone: Auditory improvement with surgical treatment novel application of the posterior retrosigmoid approach

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Abstract

Introduction: Petrous Bone Cholesteatomas (PBCs) are epidermoid cysts, which have developed in the petrous portion of the temporal bone and may be congenital or acquired. There are different types according to Sanna's classification. Our case represents a type IV surgically treated with a retrosigmosid approach, very infrequently used, with postoperative audiometry improvement.

Report: 68-year-old patient with a history of right ear accused of more than 10 years of evolution and right facial paralysis of 6 months without improvement with corticosteroid treatment. In the imaging tests, epidermoid cyst growth of the right petrous bone with extension to cistern of the pontocerebellar angle among others is visualized. The patient was operated on microsurgically by means of a retrosigmoid craniotomy and neurophysiological monitoring with exeresis of a tumor compatible with cholesteatoma. Pathological anatomy subsequently confirmed this diagnosis. In the long term, the patient surprisingly improved his sensorineural deafness objectively through audiometric studies.

Discussion: The literature reflects the large number of surgical approaches used for this pathology. Facial palsy and hearing loss, as well as tumor size and age of the patient, are the markers that define which approach to use. Recovery from sensorineural deafness is highly unlikely according to reports in the literature.

Conclusions: Petrous Bone Cholesteatoma (PBC) is a pathology associated with neurosurgeons but sometimes as this case and since they can compromise the cerebellopontine angle and cranial nerves of this region represent a neurosurgical challenge.

Introduction

Petrous Bone Cholesteatomas (PBCs) are epidermoid cysts, which have developed in the petrous portion of the temporal bone and may be congenital or acquired [1]. The incidence of PBCs was estimated as accounting for between 4 and 9% of all petrous bone lesions. A PBC gradually invades the bony labyrinth and erodes the petrous apex and the skull base around the Internal Auditory Canal (IAC) and may extend as far as the cerebellopontine angle. Furthermore, these lesions may affect other vital soft tissue structures within the temporal bone such as the sigmoid sinus or the jugular vein and carotid artery [2]. As regard the source and extension of cholesteatomas, Sanna et al [1] classified five types of PBC lesion: Supralabyrinthine, infralabyrinthine, massive labyrinthine, infralabyrinthine– apical, and apical. This radiological classification allows standardization

in reporting and a clear planning of the surgical approach Surgery remains the mainstay of treatment of PBCs. Lateral transtemporal or middle fossa approaches by microscopic surgery are usually employed for removing extensive PBCs.

Our case represents a type IV surgically treated with a retrosigmosid approach, very infrequently used, with postoperative audiometry improvement.

Clinical case

Came to our clinic a 68-year-old patient with right ear hearing for 10 years and and 4-month right facial paralysis (grade III according House-Brackmann scale). A subsequent nuclear Magnetic Resonance Imaging (MRI) confirmed structural alteration of the right petromastoid región and the presence of soft tissue which was hypointense on T1 and hyperintense on T2-weighted sequences, consistent with a diagnosis of cholesteatoma. The MRI confirmed the extension of the cholesteatoma to the jugular gulf and collapsed pontocerebellar cisterns (Figure 1) and preoperative pure-tone audiograms showed 105 db in the left ear (Figure 2).

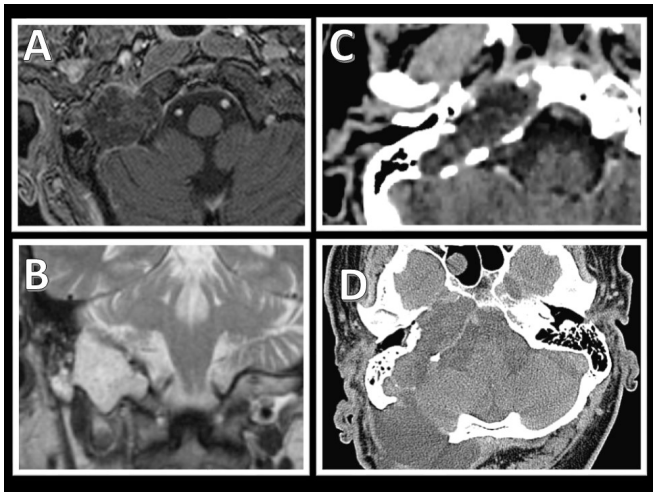

Figure 1: Pre-operative images of the lesion (MRI and CT).

Figures 1A and 1B are axial and coronal MRI slices of a lesion with extra-axial features inside the right rock, with extension to the jugular gulf that expands the bone and remodels it internally, occupying the internal auditory canal. It exerts a mass effect by partially collapsing the cisterna of the pontocerebellar angle, the prepontine cistern and the Meckel cavum. Figure 1C is a preoperative CT slices that show the same lytic lesion with expansive component and extra/intracranial hypodense soft tissue. In the postoperative figure 1D we can observe the complete resection of the lesion and the approach used.

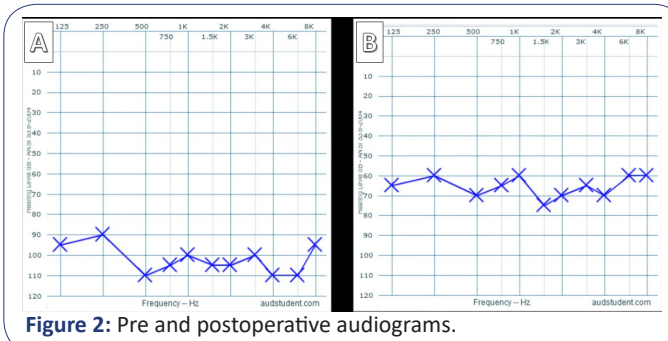

Figure 2: Pre and postoperative audiograms.

In this image we can observe the comparison between the audiograms of the left preoperative ear (Figure 2A) with an average of 105 db and the postoperative (Figure 2B) with an average of 60 db. A clear postoperative improvement was observed.

The aim of surgery was to remove the giant cholesteatoma completely. The surgery was performed under neurophysiological monitoring to maintain the integrity of the exposed pairs. A right laterocerebellar retrosigmoid approach was performed until the relief of the crag was seen, which presented a greater volume and bulge in its contour. A dural opening of the posterior border and exeresis of a white-scaly tumor was performed. In this manner, total removal of the lesion was achieved (Figure 3).

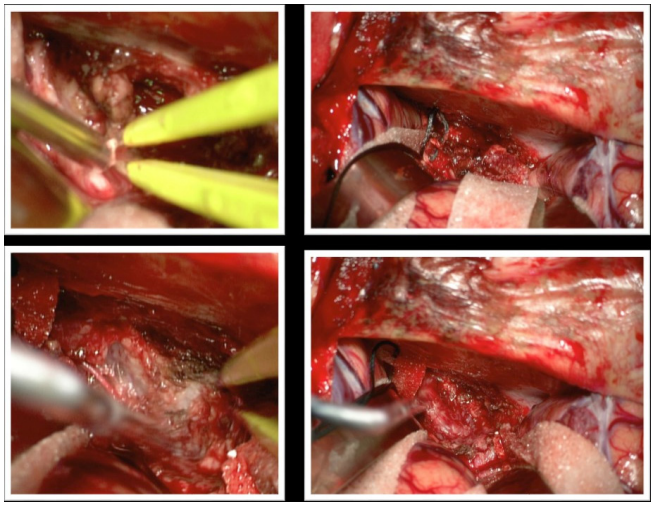

Figure 3: Intraoperative images.

In these images (Figure 3), it can be seen how a white-scaly lesion with a soft consistency that is completely resected appears through a right laterocerebellar retrosigmoid approach and an opening of the penile relief.

One month later, the patient went to the emergency department with rhinoliquorrhoea, which was successfully treated with lumbar drainage for 1 week without further complications. A control subsequent diffusion-weighted imaging sequence MRI showed no residual disease. The audiometry of control demonstrated the existence of a remarkable improvement in the right hearing with 60 db. The patient is disease-free after a 10 month follow-up.

Discussion

The rarity of these lesions, their slow and silent growth pattern, their complex location in the skull base, their proximity to vital neurovascular structures, and their tendency to recur make PBCs very challenging to diagnose and treat [3]. PBCs have been shown to be locally aggressive by involving the petrous bone and the areas surrounding it like the clivus, nasopharynx, sphenoid sinus, and infratemporal fossa and even extending intradurally [4]. Surgery with radical removal is the mainstay of treatment for PBCs, and there is no role for any form of expectant management. Early intervention could possibly result in better neurological results and even hearing preservation [5]. The choice of the best surgical approach is based on the location and extent of the lesion, hearing, preoperative facial nerve paralysis, and anatomic position of the internal carotid artery and jugular bulb. It must guarantee that the cholestea-

Table 1: Comparison of surgical approach used in relation to the class type in two different series (2006/2016).

	Massive		Supralabyrinthine		Infralabyrinthine		Infralab.-apical		Apical
	A	B	A	B	A	B	A	B	B
Modified transcoclear	43%	48.6%	19.51%	18.47%	28.6%	11.11%		23.4%	66.66%
Translaberintic	28.5%	19.4%	36.58%	19.56%		5.55%	33%	14.28%	
Middle fosa	4.8%		2.43%						
Transotic	9.5%	22.22%	19.51%	37%		33.33%	33%	62.28%	33.33%
Subtotal petrosectomy	7.14%	4.16%	17.07%	18.47%	42.85%	33.33%			
Radical mastoidectomy	7.14%		4.87%						
Retrolabyrinthine					28.6%				
Intratemporal							33%		
Transmastoid + others.	-	5.55%		6.5%		16.66%			
TOTAL	100		100%		100%		100%		

toma is visible in its entirety and ensure a sufficient exposure of the middle and posterior fossa dura, carotid artery, lateral sinus, jugular bulb, and facial nerve [6].

It has been come a long way since Gacek in 1980 [7] proposed permanent fistulization through the sphenoid or the middle ear for deep-seated PBCs due to the fact that such areas were not surgically accessible [8]. Advancements in neuroradiology and microscopic lateral skull base surgery have made it possible today to completely extirpate these lesions safely with minimal recurrences and perioperative morbidity. The primary objective in surgical approaches for PBCs today is to ensure macroscopic disease clearance along with complete control and safety of the surrounding important neurovascular structures [9]. The development of the Transotic (TO) and Transcochlear (TC) approaches, combined with various other skull base approaches, have helped achieve both these objectives and are considered the mainstay of surgery for PBCs [10] (Table 1).

This table (Table 1) reflects the comparison of the use of surgical approach performed according to class type of lesion by Mario Sanna in different years: A (2006) and B (2016) and visualized as there is no established standard.

Among the possible approaches, the Middle Cranial Fossa (MCF) approach is used when the pathology is localized to the supralabyrinthine region with a serviceable hearing.4 Posterior extension of this lesion necessitates a combined MCF-transmastoid approach. Transmastoid–retrolabyrinthine approach is appropriate for posterior lesions with limited superior extension. If hearing preservation is not aimed, Translabyrinthine Approach (TLA) ensures the adequate removal of the pathology. The retrosigmoid approach offers an excellent exposure of the pontocerebellar and cranial nerves IV through XII. This permits complete excision of small-to-medium-sized tumors when hearing preservation is the goal and safe resection of larger tumors that compress the brainstem and adjacent neurovascular structures. In our case, a grade massive, and because of the familiarity we have with this type of approach, we opted for a retrosigmoid approach to access the lesion in a simpler way.

Conclusion

Petrous bone cholesteatoma presents difficulties in its diagnosis and treatment. Adequate history-taking and high clinical suspicious with the advancement of imaging techniques made its diagnosis more feasible. Improvement of the lateral and posterior skull base approaches rendered safe, adequate, and complete removal of the tumor. The type of approach must take into account not only the tumor location but also the patient's facial paralysis and hearing loss. Although it is extremely uncommon, some patients can even better hear.

Declarations

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Conflict of interest: All authors certify that they have no affiliations with or involvement in any organization or entity with any financial interest (such as honoraria; educational grants; participation in speakers' bureaus; membership, employment, consultancies, stock ownership, or other equity interest; and expert testimony or patent-licensing arrangements), or non-financial interest (such as personal or professional relationships, affiliations, knowledge or beliefs) in the subject matter or materials discussed in this manuscript.

Ethical approval: All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee (name of institute/committee) and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent: Informed consent was obtained from all individual participants included in the study.

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