Giant Petrous Bone Cholesteatoma: Combined Microscopic Surgery and an Adjuvant Endoscopic Approach

Giannicola Iannella[1] Ersilia Savastano[1] Benedetta Pasquariello[1] Massimo Re[2] Giuseppe Magliulo[1]

[1] Organi di Senso Department, Sapienza University of Rome, Rome, Italy
[2] Department of Clinical Sciences, Polytechnic University of Marche, Ancona, Italy

Address for correspondence Giuseppe Magliulo, MD, Organi di Senso Department, Sapienza University of Rome, Via Gregorio VII n. 80, 00165 Rome, Italy (e-mail: giuseppemagliuloorl@yahoo.com).

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Abstract

Petrous bone cholesteatomas (PBCs) are epidermoid cysts, which have developed in the petrous portion of the temporal bone and may be congenital or acquired. Cholesteatomas arising in this region have a tendency to invade bone and functional structures and the middle and posterior fossae reaching an extensive size. Traditionally, surgery of a giant PBC contemplates lateral transtemporal or middle fossa microscopic surgery; however, in recent years, endoscopic surgical techniques (primary or complementary endoscopic approach) are starting to receive a greater consensus for middle ear and mastoid surgeries. We report the rare case of an 83-year-old Caucasian male affected by a giant cholesteatoma that eroded the labyrinth and the posterior fossa dura and extended to the infralabyrinthine region, going beyond the theca and reaching the first cervical vertebra. The giant cholesteatoma was managed through a combined approach (microscopic and, subsequently, complementary endoscopic approach). In this case report, we illustrate some advantages of this surgical choice.

Keywords
- petrous bone cholesteatoma
- giant cholesteatoma
- petrous bone lesions
- endoscopy

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.[1–3] The incidence of PBCs was estimated as accounting for between 4 and 9% of all petrous bone lesions.[1,2,4]

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.[1,2,5–7]

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.

Lateral transtemporal or middle fossa approaches by microscopic surgery are usually employed for removing extensive PBCs.[1,2,5–8]

In recent years, the instrumentation, techniques, and knowledge relating to middle ear endoscopic surgery have greatly improved. At present, the main application of endoscopic surgery is in the surgical treatment of middle ear cholesteatoma, but the natural evolution of the technique may provide an increasing number of applications in lateral skull base surgery.[9–11]

This case report illustrates the case of a giant cholesteatoma that eroded the labyrinth and the posterior fossa dura and extended to the infralabyrinthine region, thereby affecting the body of the first cervical vertebra. Few authors have dealt with the topic of a PBC, which presents this type of massive life-threatening infralabyrinthine extension, and

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there is apparently no literature documenting cases in which a cholesteatoma has extended to the extracranial area and cervical spine.

We performed an infralabyrinthine subtotal petrosectomy by means of a combination of microscopic surgery and an adjuvant endoscopic approach to remove the cholesteatoma matrix adherent to the vertebral body and the dura of the posterior fossa. No cases of endoscopic adjuvant management of giant infralabyrinthine cholesteatoma have previously been reported.

Case Report

An 83-year-old Caucasian male was referred to our department with otorrhea in his left ear which he had had for 3 months and a painful swelling behind the ear. Unspecified tympanoplasty surgery with mastoidectomy had been performed 60 years earlier. He also had a left facial palsy evaluated as V grade according to the House–Brackmann scale which had reportedly become progressively worse over a 10-year period.

Otomicroscopic evaluation revealed total perforation of the left hand tympanic membrane, through which the middle ear cavity appeared completely occupied by cholesteatomatous soft tissue. The external auditory canal drum appeared eroded. Audiological preoperative evaluation (pure-tone audiometry) showed left anacusis.

Temporal bone computed tomography revealed an extensive cholesteatoma that completely occupied the left middle ear cavity, the mastoid, and involved the petrous bones. Imaging showed erosion of the tegmen tympani, the bone around the geniculate ganglion, the semicircular canals, and the cochlea too. In the posterior section, the lesion was found to extend and destroy part of the occipital bone and skull base. It extended beyond the cranial theca and went distal to the body of the first cervical vertebra (atlas) which appeared eroded (►**Fig. 1**).

A subsequent nuclear magnetic resonance imaging (MRI) confirmed structural alteration of the petromastoid region and the presence of a 6 × 2 × 3 cm area 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 cerebrum to the first cervical vertebra (►**Fig. 2**).

The aim of surgery was to remove the giant cholesteatoma completely. Intraoperative facial nerve using NIM2 was planned.[12]

Left infratemporal and infralabyrinthine subtotal petrosectomy was performed using a combined approach microscopic and endoscopic surgery. First, the petrous bones and a massive cholesteatoma, which had occupied the lateral and posterior temporal areas on the left side and had caused widespread osteolisis of the mastoid cells, of the pyramid, and of the skull base theca, were removed through standard microscopic surgery (►**Figs. 3** and **4**). Furthermore, the remaining infralabyrinthine cholesteatoma matrix, which adhered to the body of the atlas was treated by means of delicate endoscopic dissection and bipolar coagulation (►**Fig. 5**). Rigid endoscopes

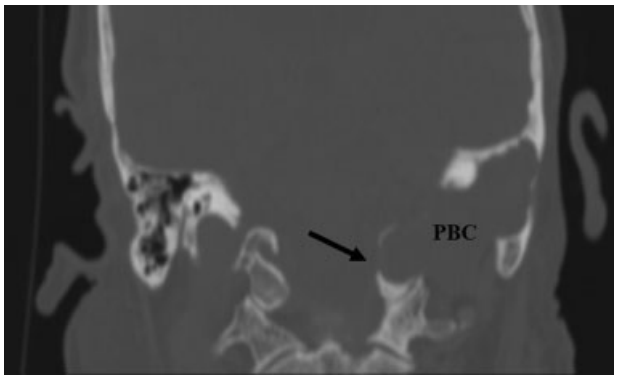

Fig. 1 Coronal CT; extensive cholesteatoma eroding part of skull base bone and of the first cervical vertebra (arrow). CT, computed tomography; PBC, petrous bone cholesteatoma.

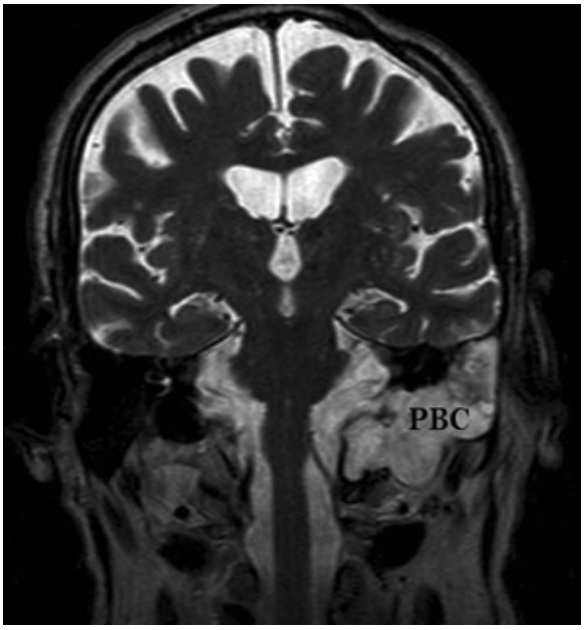

Fig. 2 Coronal MRI, T1-weighted sequence; presence of a hyperintense 6 × 2 × 3 cm soft tissue extended between the skull base and the first cervical vertebra. MRI, magnetic resonance imaging; PBC, petrous bone cholesteatoma.

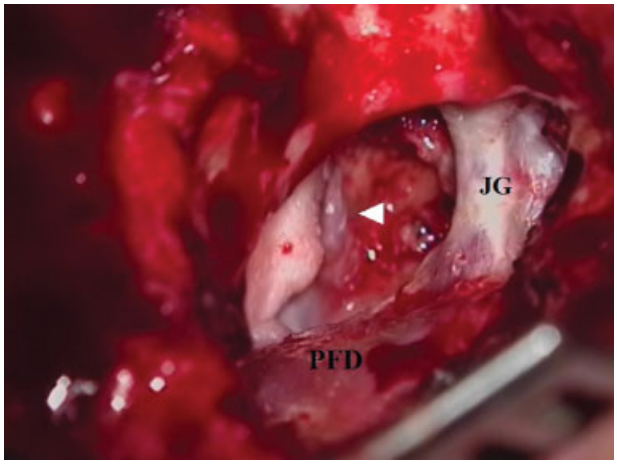

Fig. 3 Microscopic surgery; residual cholesteatoma adherent to the transverse apophysis of the first cervical vertebra (arrowhead). JG, jugular gulf; PFD, posterior fossa dura.

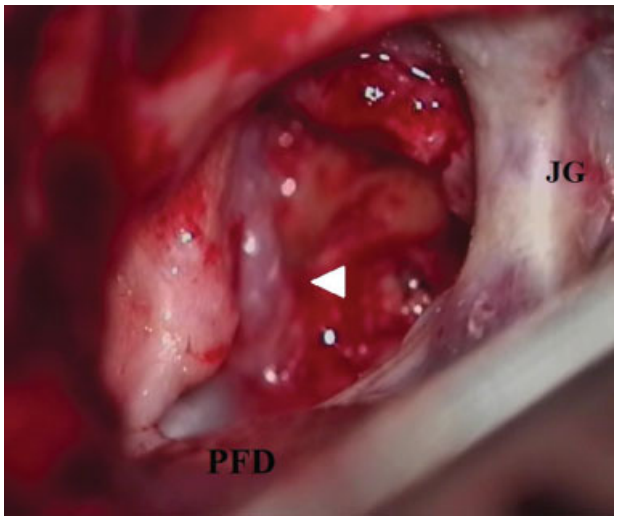

Fig. 4 Microscopic view (maximum magnification); residual cholesteatoma adherent to the transverse apophysis of the first cervical vertebra (arrowhead) not entirely visible.

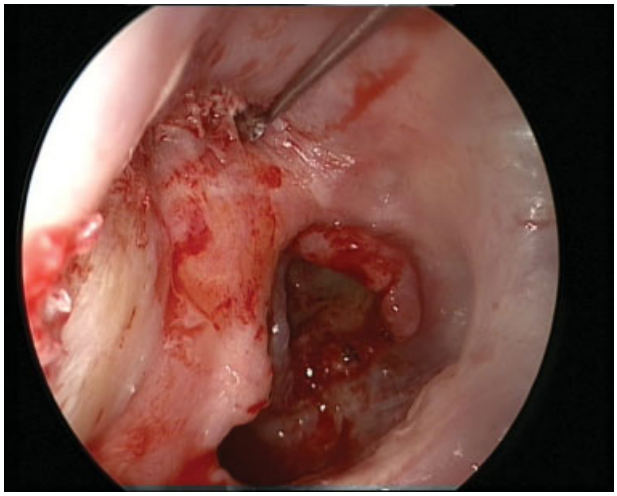

Fig. 5 Adjuvant endoscopic surgery; delicate dissection of the cholesteatoma matrix adhered to the transverse apophysis of the atlas clearly visible (round knife).

at 0- and 30-degree angled with an outer diameter of 4 mm (Storz, Germany) were used. Images were recorded through a full high-definition (HD) camera (Stroz, Germany), which was attached to the endoscope lens, and these images were displayed on a full HD monitor. In this manner, total removal of the lesion was achieved. No postoperative cerebrospinal fluid (CSF) leaks or other complications occurred. A subsequent diffusion-weighted imaging sequence MRI showed no residual disease.

The patient is disease-free after a 14-month follow-up.

Discussion

PBC is an uncommon pathology but may cause severe functional damage and negatively affect the quality of life. It has a known tendency to invade the labyrinth and Fallopian canal, thereby causing facial nerve paralysis and permanent hearing loss. It may also involve all the anatomical structures within the temporal bone and extend to the cerebellopontine angle or the infralabyrinthine region, which signifies a risk of damage to vital life structures (internal carotid artery, jugular vein, sigmoid sinus, dura).[1–5,8,11]

We have illustrated the case of a giant cholesteatoma which was classified as an infralabyrinthine–apical cholesteatoma according to Sanna et al[1] that eroded the labyrinth and the posterior fossa dura.

The case we have presented is a good example of a cholesteatoma which extends beyond the confines of the skull base and infralabyrinthine region. Indeed, although massive intracranial involvement of the bone, nerves, and vascular system of the posterior cranial fossa by such a huge cholesteatoma is not infrequent, this case is very unusual because the lesion went beyond the theca and reached the first cervical vertebra. To our knowledge, no previous cases of a cholesteatoma of this size have been documented.

Traditionally, surgery performed on giant cholesteatoma of the petrous bone has contemplated lateral transtemporal or middle fossa microscopic surgery. 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 cholesteatoma 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.[2,5–11,13–15]

The main problems regarding PBC removal are residual matrix, recurrence/relapse, and postoperative complications. Despite the illumination and magnification offered by the microscope, it has certain limitations. Insufficient primary resection of the epidermal matrix, caused by a recess which was not detected by the microscope, may lead to recurrence of the disease in the patient/relapse.[1–7]

In recent years, endoscopic surgical techniques, instrumentation, and knowledge have greatly improved due to an increased use of endoscopy in middle ear and mastoid surgeries.[9–11]

Marchioni et al[16] recently proposed the endoscopic transcanal surgical approach for the treatment of IAC and petrous bone lesions. Six of the 12 patients treated had a cholesteatoma of the tympanic cavity which affected the inner ear (vestibule, geniculate ganglion, middle cranial fossa, and IAC), although there was no massive infralabyrinthine extension.

Kanzara et al[4] reported the case of a petrous apex cholesteatoma which was removed in its entirety by means of an endoscopic approach. They stated that an endoscopic permeatal approach circumvents some of the problems encountered in microscopic surgery. It provides a better operative field and excellent vision of the important structures because, unlike the microscope, it bypasses the narrowest points and provides an excellent assessment of the surrounding structures. Moreover, they noted that the endoscopic approach provides more direct access to the apex.

In our case, a left-sided infratemporal and infralabyrinthine subtotal petrosectomy was performed using a

combined approach (microscopic and endoscopic). It resulted in total excision of the cholesteatomatous matrix.

Endoscopic petrous bone surgery offers some advantages as compared with the traditional microscopic technique, as it guarantees direct visual control of hidden areas such as the infralabyrinthine recess and its structures. In this way, the eradication of the cholesteatoma in its entirety may be possible, reducing the risk of residual persistence and recurrence (lower rate). Moreover, it may lower the risk of lesions of the dura and of the other functional and vital structures of the lateral skull base during cholesteatomatous matrix removal (postoperative CSF leaks).[8–10,14] As claimed by many authors, the main limitation of the endoscopic approach is that it is a one-hand surgical procedure that does not allow adequate drilling of the temporal bone to remove extensive lesions.[9–11,17]

In conclusion, in future, endoscopic surgery will gain increasing importance in surgery of PBCs. Initial microscopic surgery should be performed. Subsequently, a complementary endoscopic approach in hidden areas should be performed to remove the cholesteatomatous matrix and reduce possible postoperative complications and relapse.

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