“Surgery of the lateral skull base: 50 years of endeavour”

Section 2. Advances and open issues

PETROUS BONE CHOLESTEATOMA

Diego Zanetti*, Giorgio Conte°, Laura Magnoni *, Federica Di Berardino*

*Unit of Audiology, Dept. of Clinical Sciences and Community Health, University of Milano; Dept. of Surgery, Fondazione IRCCS Cà Granda, Ospedale Maggiore Policlinico, Milano

°Dept. of Neuroradiology, University of Milano, Fondazione IRCCS Cà Granda, Ospedale Maggiore Policlinico, Milano

INTRODUCTION

Petrous bone cholesteatoma (PBC), also defined “invasive” or “medially extensive” cholesteatoma, is a benign lesion located medial to the otic capsule. It accounts for 10% of the lesions involving the petrous pyramid[1] [2].

Compared to middle ear cholesteatoma it is rare, representing less than 4% of all temporal bone cholesteatomas; its incidence has remained stable during the last 4 decades[2].

PBC can be congenital, arising from ectodermal embryonic remnants within the otic capsule, acquired (post-otitic), or iatrogenic, after failure of previous middle ear surgery[3]. Histologically, these 3 etiological variants are indistinguishable with a collection of desquamated epithelium, keratin, and cellular debris, sharing similar histologic features with odontogenic keratocysts and epidermal cysts.

After remaining silent for years, they manifest with either otorrhea, vertigo, otalgia or their combination, followed by facial nerve weakness and tinnitus[2] [4]; in Danesi’s[5] series no patient complained of vertigo pre-operatively, possibly due to long-standing vestibular areflexia related with the advanced extension of the disease at the time of surgery, often associated with multiple fistulas. In Álvarez’s [6] series of 25 patients, ¾ presented with facial paresis. Intracranial suppurative complications, such as recurrent aseptic meningitis [7] are frequently encountered in PBC, especially in populations living in lower socio-economic conditions.

A classification of the site and extension of PBC was initially proposed by Fisch, who separated supra- and infra-labyrinthine lesions. In 1993 Sanna et al categorized the PBC into 5 classes: Class I, supralabyrinthine; II, infralabyrinthine; III, infralabyrinthine-apical; IV, massive; and V, apical (Figure 1). Later on, the same Authors[4] added another stage for disease extending beyond the borders of the temporal bone.

Despite its slow-growing habit, the osteolythic capacity of PBC leads to exposure and adhesion to relevant anatomical structures such as the facial nerve, carotid artery and dura mater. In Danesi et al[5] series of 81 patients, a labyrinthine erosion / fistula was evident at surgery in 83% (infralabyrinthine/apical) to 100% (apical and massive) cholesteatomas. A cochlear erosion appeared again in all cases of massive PBC, while less frequently in other stages of the disease; erosion and infiltration of the internal auditory canal (IAC) was present in 100% of the massive and ¾ of the infralabyrinthine/apical. Conversely, an intracranial erosion and CSF leak occurred rarely.

Surgery is the only accepted treatment. Given the complex anatomy of the petrous bone, surgical removal of these lesions can be very challenging. A single best approach has not yet been identified.

Frequent post-operative complications or sequelae include cerebrospinal fluid (CSF) leaks, facial nerve palsies, anacusia[8]; recurrence is highly likely[6].

ADVANCES IN IMAGING

Multisection computed-tomography (MSCT) is the traditional technique of choice when cholesteatoma is clinically suspected, providing a spatial resolution of 0.4 x 0.4 mm in plane and 0.5 mm in slice thickness. In the last years the use of flat-panel CT has taken place in clinical practice, being preferred for its higher spatial resolution and lower radiation dose[9] [10]. Typically, PBC appears as an expansive soft-tissue mass in the apical portion of the petrous bone, with large areas of bone resorption (figure 2). CT yelds high negative predictive value in diagnosing cholesteatoma, ruling out the presence of cholesteatoma when when the tympanic cavity is well pneumatized. Conversely, CT has a low specificity because it can’t easily differentiate cholesteatoma from other soft-tissue masses[11]. Typical CT findings associated to a soft-tissue mass within the petrous apex that suggest a PBC (even if the tympanic membrane is normal at otomicroscopy) are the erosion of the tegmen tympani, of the posterior and inferior wall of the petrous pyramid, of the carotic canal and, sometimes, of the upper clivus. In the literature, bony erosion has been reported from 50 to 97% of CT scans of patients with histologically-proven cholesteatoma, thus resulting a reliable (but not perfect) imaging marker [11]. The density values of cholesteatoma are not significantly different from those of the other petrous bone lesions. Thus, CT is unable to differentiate cholesteatoma from inflammatory or scar tissue. Its value remains the high spatial resolution that allows defining the localization and extension of the PBC and the signs of erosion of relevant bony structures such as the tegmina, the Fallopian canal, the labyrinth and cochlea, the internal carotid artery (ICA), the jugular bulb (JB) and the sigmoid sinus.

Magnetic Resonance Imaging (MRI) usually provides complementary information compared to CT. It is superior in differentiating cholesteatoma from mimicking lesions but it is inferior in defining its localization and extension because of the lower spatial resolution. For this reasons, MRI is indicated after CT, when the latter and the clinical examination are not conclusive in the diagnosis or when a recurrence is suspected after surgery. MRI protocol for PBC includes multiplanar conventional T1-weighted and T2-weighted sequences, and diffusion-weighted (DW) sequence. (figure 3) Cholesteatomas show a high signal intensity on DW sequence attributed to impeded diffusion of water molecules and T2 shine-through effect [(12)]. In the literature, the DW sequence was associated with an excellent specificity and positive predictive values (up to 100%) in patients with or without history of a previous surgery [(12)]. Cholesterol granuloma and inflammatory/scar tissue do not show high signal on DW sequence. On pre-contrast T1-weighted sequence a PBC has a low signal intensity, easily differentiable from cholesterol granuloma which has an high signal. On T2-weighted sequence it appears with an intermediate signal, while inflammatory or scar tissue are characterized by a high signal intensity. After contrast-agent administration cholesteatoma does not show enhancement because it is a non-vascularised tissue; however, sometimes a thin peripheral rim of enhancement can be seen on T1-weighted images after intravenous administration of gadolinium because of the presence of epithelial (matrix) and granular (perimatrix) layers[(13)]. On early post-contrast T1-weighted slow-enhancing inflammatory/scar tissue can misdiagnosed as cholesteatoma, causing false positive results. The acquisition of late (45 min) post-contrast T1-weighted sequence can differentiate non-enhancing and avascular cholesteatoma from slow enhancing inflammatory/scar tissue [(14)]. Table I summarizes the MRI characteristics of cholesteatoma and mimicking lesions. It should be acknowledged that post-contrast sequences are not usually needed because pre-contrast sequences are sufficient for the diagnosis.

Table I: signals of cholesteatoma and mimicking lesions on MR sequences.

	T1-weighted	**T2-weighted**	**Diffusion-weighted**	**late post-contrast T1-weighted***
Cholesteatoma	low	intermediate	high	low
Inflammatory/scar tissue	low	high	low	high

Cholesterol Granuloma	high	high	low	high

* 45 minutes after intravenous contra-agent administration

APPROACHES AND SURGICAL ISSUES

The medial portion of the petrous bone and the petrous apex are difficult-to-access regions, owing to the anatomical boundaries represented by the ICA, JB, middle (MCF) and posterior cranial fossa (PCF) dura. Furthermore, all lateral skull base surgical trajectories come across the facial nerve, labyrinth and cochlea; despite wide bone removal and careful mobilization of the facial nerve and of the JB, some deep areas such as the medial surface of the ICA will remain a blind spot [15].

Traditional lateral skull base approaches, either transmastoid/transtemporal or through middle cranial fossa craniotomy, are usually performed under magnification by an operative microscope. They include the transcochlear, the modified transcochlear and the transotic approach[15].

The first two imply the mobilization of the facial nerve from the Fallopian canal (“re-routing”); in all the 3 techniques the destruction of the labyrinth and of the cochlea are inevitable surgical steps, followed by Eustachian tube plugging and blind-sac closure of the external auditory canal (EAC). Most surgeons prefer to fill the surgical cavity with adipose tissue harvested from the peri-umbilical region.

Most surgeons address PBC on the basis of their personal experience and surgical skills[16]; in tertiary referral Skull Base/Neuro-Otology centers these lesion are more often managed by a systematic approach [1] [4] [17]; nevertheless, an approach tailored on the location and extension of the disease observed with current imaging modalities and accounting for the pre-operative functional status is reasonable.

Van Dinther et al[18] developed a non-conventional transmastoid subarcuate approach in order to remove a small PBC through the center of the superior semicircular arch, preserving its integrity and sparing the hearing. Obviously enough, this peculiar approach is limited to small and very selected supralabyrinthine cholesteatomas.

Kojima et al[19] removed a supra- and infracochlear PBC through an extended middle cranial fossa approach with the assistance of endoscopy. Although acceptable in terms of functional sparing, in our opinion it should be reserved to cases with locations and extension hardly accessible through the more conventional routes, owing to possible higher morbidity.
As an alternative to the invasive lateral skull base procedures, a totally endoscopic trans-sphenoidal approach has been proposed by a few Authors with the purpose of avoiding a craniotomy, promoting faster recovery and reducing the postoperative morbidity[20][21]. Unfortunately, the experiences remain very limited and the outcomes uncertain.

Whatever the selected approach, a few basic surgical pitfalls have been derived from cumulative experiences, especially by leading Skull Base centers:

1) facial nerve

The main hurdle to the surgical access to the petrous apex is the Fallopian canal.
The facial nerve can be exposed, thinned and already damaged at the time of surgery in a relevant percentage of PBC. Danesi et al[5] (Danesi, 2016) found it interrupted in 35% of their patients. An interruption of the facial nerve occurred in ¾ of the apical lesions, half of the massive cases and 1/3 of the supra- and infralabyrinthine cholesteatomas. Half of their patients suffered a post-operative House-Brackmann grade V or VI facial palsy.
When the facial canal is intact, it represents an obstacle to complete visualization of deep and medial areas. The traditional solution to gain better access is represented by facial nerve “rerouting”, usually posterior position (Figure 4) by first skeletonizing and uncovering the bony shell of the canal, then sectioning the greater superficial petrosal nerve and translocating the nerve to the posterior edge of the petrosectomy cavity. Even delicate manipulations of the nerve lead to an inevitable transient palsy.
The transotic approach, which preserves the integrity of the bony Fallopian canal, combined with the use of endoscopes anterior and posterior to the canal to work “beyond the blind corner” (Figure 5) is an effective means to avoid a post-operative paresis. Care must be taken to prevent heating of the facial canal with the drill and with the endoscope.

2) ICA

Invasive cholesteatoma may erode the cortical bone of the carotic canal, exposing the adventitial layer of the intrapetrous carotid artery, without infiltrating it. Careful dissection of the adherent matrix from the carotid wall is essential to avoid relapses. The thickness of the adventitia is usually sufficient to prevent accidental tearing of the wall, which can result fatal to the patient; carotid stenting is usually not necessary, since no cases of severe hemorrhage are reported in the PBC literature [(1) (2) (17)].

For PBC extending beyond the ICA, its gentle mobilization may widen the surgical view of the medial surface under the microscope, but, again, it should only be performed cautiously.

3) dura mater

In PBC, the matrix is usually tightly adherent to the meninges and a loose perimatrix is missing. Bipolar coagulation of dural portions helps removing it or avoiding relapses[(1) (3) (4).] In our personal experience, the local application of solutions of mucolytic agents followed by brushing with neurosurgical cottonoids has proven helpful. In case of inadvertent dural breakdown with CSF leakage, immediate sealing with a dural patch is required, with careful avoidance of intracranial penetration of keratinized epithelium.

OUTCOMES

The main aim of surgery of PBC is radical removal of the disease with absence of recurrence. The latter should have a priority over an unpredictable preservation of hearing or even over the risk of facial palsy, although this aspect is still a subject of debate.

In agreement with other Authors [(16)] an individualized surgical treatment of PBC should strive to achieve the least recurrence rate as well as the facial functional outcome, at the price of sacrificing hearing.

1) Hearing and equilibrium

Post-operative vestibular impairment is rather unusual, which is not surprising, considering the very low incidence of pre-operative vestibular symptoms.

The chance of preserving the residual hearing is very low, although a few reports claim it is feasible[(22)], by means of modifications of the surgical techniques, such as partial (selective) ablation of the semicircular canals in the so-called modified translabyrinthine approach[(23)].

At the time of surgery, cholesteatoma has often already invaded the inner ear structures[24] and hearing is largely compromised. More than 90% of Danesi's[5] patients ended up with anacusis, even though half of them were already deaf ears before surgery.

Nevertheless, in those rare cases with preserved hearing and normal inner ear anatomy, a labyrinthectomy and a cochleostomy might be sidestepped and a more conservative approach considered.

Conversely, no question about sacrificing the hearing remnants should rise in case of massive or inaccessible cholesteatoma. [16] [23]

In the rare instances in which the PBC arises in the only hearing ear, the surgeon may elect either one of the following two solutions: 1) performing an "open" (canal wall down) technique in which the disease is simply exteriorized and its evolution can be easily followed-up without endangering the inner ear [6]; 2) the contralateral ear is rehabilitated with a cochlear implant (if feasible) and the PBC is addressed soon after [25].

In the era of cochlear implants, the opportunity of rehabilitating hearing has to be integrated in the pre-operative planning. Provided the contralateral hearing is at least partially compromised, a cochlear implant would certainly prove beneficial to the patient who will lose his/her hearing after PBC removal [25]. Quite obviously, the cochlea should be intact and free from disease in order to insert the array, and the implant would be buried under the final cavity obliteration. Then, the question of the timing of the implantation rises: simultaneous implantation would provide the greatest speech perception results and would spare the patient a second operation; however, it would also hamper the chance to follow-up the potential recurrence by means of MR imaging. On the other side, a delayed implantation might not be feasible because of potential ossification of the cochlear lumen after the surgical trauma.

2) facial nerve

Even if the nerve is intact, the surgical manipulations needed to access the most difficult to reach areas can endanger it.

Facial nerve function is more likely to be preserved in patients with preoperative facial palsy mild or moderate; an early diagnosis by current imaging methods is essential.

In Alvarez's[6] series of 25 patients, 80% had an immediate complete facial palsy (H-B grade VI) but 71% improved to H-B grades I to III at 1 year after surgery.

When preoperative facial palsy is present, a decompression of the Fallopian canal from the first genu to the silo-mastoid foramen is required. In the event of nerve interruption, its repair through either primary neurorrhaphy or nerve grafting, based on the length and location of the injured nerve segment, can be performed simultaneously. In case of long-standing palsies, a hypoglossal–facial nerve transfer is preferred [16].

In all procedures except for primary direct anastomosis, a recovery not exceeding House–Brackmann grade III can be expected[1] [4] [5] [24]. The facial nerve was surrounded of compressed by the cholesteatoma in 18 of the 52 patients with PBC described by Magliulo[24] and showed facial paralysis grade II to V. Fourteen were managed with cable grafts using sural or great auricular nerves, while the remaining recovered spontaneously after surgery.

1) Cholesteatoma recurrence

Recurrencies can develop from remnants of the cholesteatoma matrix adherent to the facial nerve, dura mater or ICA[4] and can also lead to postoperative complications.

The recurrence rate for the classical trans-mastoid routes ranges between 5 and 28% in the literature [1] [3] [4] [6] [17]; long-term observation is needed because relapses have been observed even 8 years after surgery.

“Exclusion” of the petrosectomy cavity by blind sac closure of the external auditory canal and plugging of the Eustachian tube orifice, combined with filling of the cavity with adipose tissue grafts and additional muscolo-periosteal flaps is the universally accepted surgical solution to minimize the risks of CSF leaks and infection, and to achieve adequate protection of the neurovascular structures. The drawback is the absence of control of the cavity for possible recidivism, which must then rely upon imaging. To partially overcome this issue fibrin glue can be used instead of fat and muscle to fill the operative field, in order to pneumatize the cavity and improve postoperative imaging.

Alternatively, an “open” surgical technique can be selected in order to allow periodical inspection of the petrosectomy cavity; actually it can expose the patient to a higher chance of post-operative complications and it is not clear if the recurrence rate is lowered [6]. In open techniques, Vashishth et al [2] recommended to reinforce the dura with full thickness cartilage palisades with attached perichondrium in an underlay fashion, covered by a posteriorly based periosteal flap.

2) Endoscopy vs microscopy

One recent advance in lateral skull base surgery has been the introduction of endoscopes. Although the microscope offers a wide field of view, different degrees of magnification and excellent illumination, deep recesses within the temporal bone may remain out of sight. In recent years, endoscopic surgical techniques applied to the ear have gained respect and widespread use, increasing knowledge and refining the outcomes(26) (27).

A variety of endoscopes that differ in size (4 mm vs 2.7mm), length (8-15-20mm) and angle of view (0°, 30°, or 70°) can assist in surgery for PBC, allowing access to areas such as the medial or anterior wall of the ICA, the infralabyrinthine compartment, the anterior part of the internal auditory canal and even the sphenoid sinus(15).

The sets of instruments specifically designed for endoscopic middle ear surgery can be too short for reaching the petrous apex, but modified sinus surgery (longer) tools or neurosurgical instruments can effectively replace them.

The main drawback of endoscopy is that one-hand surgical procedures somehow limit an adequate drilling of the temporal bone(15). The advantage of bimanual dissection under the microscope can be compensated by a second operator holding the endoscope, and, if needed, also a second suction tube (such as in endoscopic surgery of the anterior skull base).

Nowadays, endoscopes are commonly employed to assist conventional petrous bone microscopic procedures. as they allow inspection of hidden areas and guarantee a more reliable radical removal of the cholesteatoma.

Some Authors claim that an exclusively endoscopic approach can achieve complete eradication of the cholesteatoma in a less invasive fashion, reducing the risk of residuals, and lowering the risk of lesions of the dura, of the major vessels and of the facial nerve (15)(28).

Marchioni et al(29) recently treated 6 cholesteatomas of the middle ear affecting the inner ear structures (without significant infralabyrinthine extension) through an exclusive endoscopic trans-canal approach.

These approaches seem very promising in terms of functional preservation, but, up to now, the experiences are limited and long-term results are lacking, especially as far as the recurrence rate is concerned.

Indeed all otologic surgeons realize that the endoscopes are invaluable tools for assisting the dissection in the "blind spots" within the temporal bone, in combination with the otomicroscope.

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