Our Experience in the Management of Petrous Bone Cholesteatoma

Karine Aubry, M.D., Ph.D.,¹ Lana Kovac, M.D.,¹ Elisabeth Sauvaget, M.D.,¹ Patrice Tran Ba Huy, M.D.,¹ and Philippe Herman, M.D., Ph.D.¹

ABSTRACT

We present the management of a series of petrous bone cholesteatomas. We performed a retrospective analysis on 28 patients with petrous bone cholesteatoma who underwent surgery between 1991 and 2008 at Lariboisière Hospital, Paris, France. Main outcome measures included age of patients, surgical approaches, complications, and recurrence. The mean age was 47 years. Five were congenital cholesteatomas and 23 were acquired ones. Seventeen patients had undergone previous mastoid surgery elsewhere. Ninety-six percent of patients presented with hearing loss and 37.5% with facial nerve palsy. The surgical approaches varied according to the classification. Postoperatively, four patients developed facial nerve palsy; two patients, XII nerve paresis; one patient, X nerve paresis; and eight patients, deafness. The mean follow-up was 5 years. Twenty patients had no long-term recurrence. Two cases of petrous apex cholesteatoma presenting with double vision were removed by an endoscopic transsphenoidal approach. Control of the lesion was satisfactory in both cases. However, a minor pontic stroke resulted in transient hemiparesis in the case with dehiscent dura around the petrous bone cholesteatoma. Petrous bone cholesteatoma surgery is difficult. Lateral transtemporal and middle fossa approaches are classically used to remove petrous bone cholesteatoma. Recent progresses in endoscopic surgery, using image guidance system, allow removal of the petrous apex cholesteatoma by an endoscopic transsphenoidal approach with minimal morbidity.

KEYWORDS: Petrous bone cholesteatoma, petrous apex, endoscopic transsphenoidal approach

Petrous bone cholesteatoma (PBC) is an epidermoid cyst developed in the petrous portion of the temporal bone. PBC gradually invades the bony labyrinth and erodes the petrous apex and the skull base around the internal auditory canal. It may invade the auditory canal and grow into the cerebellopontine angle. It may also involve all the anatomic structures within the temporal bone with the risk of damaging structures vital for life (internal carotid artery [ICA], jugular vein,

sigmoid sinus).¹ PBC surgery is challenging because complete removal of the cholesteatoma matrix from the dura, sigmoid or superior petrosal sinus, jugular bulb, ICA, and facial and lower nerves is cumbersome. Improvements in skull base surgery and recent advances in radiological imaging allow refining the management of PBC. Indeed, high-resolution computed tomography (CT) and magnetic resonance imaging (MRI) allow determining the size, morphology, and anatomic site of

¹ENT Department, Assistance Publique des Hôpitaux de Paris (AP-HP), Lariboisière Hospital (AP-HP), Paris, France.

Address for correspondence and reprint requests: Karine Aubry, M.D., Ph.D., ENT Department, University Hospital Center, 2 Avenue Martin Luther-King, 87000 Limoges, Paris, France (e-mail: aubryorl@aol.com).

Skull Base 2010;20:163–167. Copyright © 2010 by Thieme

Medical Publishers, Inc., 333 Seventh Avenue, New York, NY 10001, USA. Tel: +1(212) 584-4662.

Received: September 24, 2009. Accepted: October 10, 2009.

Published online: January 5, 2010.

DOI: <http://dx.doi.org/10.1055/s-0029-1246228>.

ISSN 1531-5010.

Table 1 Sanna's Classification²

Classification	Definition
Supralabyrinthine	It involves the anterior epitympanum and extends medially toward the internal auditory canal and anteriorly toward the carotid artery. The basal turn of the cochlea may be involved. Posteriorly, the cholesteatoma may spread toward the posterior aspect of the bony labyrinth and the retrolabyrinthine mastoid cells.
Infralabyrinthine	It involves the hypotympanic and infralabyrinthine regions and extends anteriorly toward the internal carotid artery and posteriorly toward the posterior cranial fossa.
Massive labyrinthine	It involves the entire anterior and posterior labyrinth.
Infralabyrinthine-apical	It extends either anteriorly into the petrous apex and may involve the sphenoid sinus and the horizontal portion of the internal carotid artery, or arises in the apical compartment and extends superiorly to the sphenoid sinus and inferoposteriorly to the infralabyrinthine compartment.
Apical	It involves only the apical compartment of the temporal bone; it can cause erosion of the internal auditory canal and extend toward the posterior cranial fossa or anteriorly to the trigeminal nerve.

the cholesteatoma preoperatively. Based on a series of 28 cases with PBC, the aim of this work is to present our management and to define the indications for endoscopic transsphenoidal approach.

PATIENTS AND METHODS

Twenty-eight patients (15 men, 13 women) with PBC referred between 1991 and 2008 to a tertiary care center were reviewed retrospectively. All patients underwent thorough clinical otoneurological examination. Audiological examination (air conduction measured by pure tone audiometry using the modified American Academy of Ophthalmology and Otolaryngology) was performed pre- and postoperatively. Cranial nerve deficits were evaluated. Facial nerve function was graded according to the House-Brackmann grading system. The presenting signs and symptoms of the patients were documented. CT scans (axial and coronal bone window images) and MRI (T1- and T2-weighted images) were analyzed to evaluate the extent of the disease. Patients were grouped according to Sanna's classification (Table 1).² Postoperative morbidity (hearing loss; cranial nerve deficits; cerebrospinal fluid [CSF] leak) and residual or recurrent cholesteatoma were reported. Postoperatively, patients were regularly followed both clinically and radiologically by high-definition CT, MRI, and diffusion-weighted scans.

RESULTS

The average age was 47 years (range from 33 to 70). Seventeen patients had undergone previous mastoid surgery elsewhere, from one to six previous procedures. Five cholesteatomas were assumed to be congenital and 23 were acquired. We observed two supralabyrinthine, six infralabyrinthine, 10 massive labyrinthine, four infralabyrinthine-apical, and six apical cholesteatomas. The symptoms are reported in Table 2. Twenty-seven

patients presented more than two symptoms. The main symptom was hearing loss (27 patients, 96%). Mixed hearing loss was present in four cases (14%), conductive hearing loss in one case (3.5%), sensorineural hearing loss in seven cases (25%), and total deafness in 15 cases (54%). Hearing was normal in just one case (3.5%). Facial nerve palsy was observed in 10 cases (37.5%), otorrhea in eight cases (28%), and vertigo in four cases (14%). Meningitis, tinnitus, otalgia, headache, and (V, VI, and XII) nerve paresis or palsy occurred less frequently.

The surgical approach versus localization of cholesteatoma is reported Table 3. Endoscopes (0-degree, 30-degree, 70-degree) were used nine times as an adjunct to the lateral approach. Monitoring of the facial nerve was performed for each case. Computer-assisted navigation (Digipointer, Collin-ORL, Cachan, France) was used for the two patients with petrous apex cholesteatoma removed by an endoscopic transsphenoidal approach.

Postoperatively, four patients developed a transient facial nerve palsy; two patients, a XII nerve paresis; one patient, a X nerve paresis; eight patients, deafness;

Table 2 Clinical Presentation in 28 Patients with PBC

Symptoms	No. of patients (%)
Hearing loss	27 (96)
Facial palsy	10 (37.5)
Otorrhea	8 (28)
Vertigo	4 (14)
VI palsy	3 (11)
Tinnitus	2 (7)
Meningitis	2 (7)
Otalgia	1 (3.5)
Headache	1 (3.5)
V dysesthesia	1 (3.5)
Facial myoclonia	1 (3.5)
XII palsy	1 (3.5)

Table 3 Surgical Approaches Used for 28 Patients Grouped According to Sanna's Classification

Classification	No. of Patients	Surgical Approach/No. of Patients
Supralabyrinthine	2	Middle cranial fossa/2
Infralabyrinthine	6	Translabyrinthine/4 Transcochlear/1 Transotic/1
Infralabyrinthine-apical	4	Transotic/3 Middle cranial fossa-transcochlear/1
Massive labyrinthine	10	Transotic/10
Apical	6	Infratemporal fossa type B/2 Transsphenoidal/2 Infralabyrinthine/1 Infracochlear/1

and one patient, a transient hemiparesis with worsening of the VI nerve palsy.

Four of 18 patients with a preoperative normal facial nerve function developed a facial palsy postoperatively (two grade II, two grade III), which recovered completely. Preoperatively, 10 patients presented a facial palsy (seven grade III and three grade V). During surgery, the facial nerve was preserved and decompressed in eight cases, and in two cases it had to be resected because of profound atrophy. Postoperatively, six patients improved to grade II, one kept a grade III, and patients with grade V did not recover. Facial rehabilitation with hypoglossal-facial anastomosis had to be performed in these three cases. Botulism toxin injection was performed for one patient with facial hemispasm.

Postoperative hearing was normal in two patients. Two patients had a sensorineural hearing loss, one patient had a mixed hearing loss, and 23 patients had deafness.

The mean follow-up was 5 years (range, 4 months to 10 years). All patients remained symptom-free after surgery. However, the follow-up evidenced recurrent disease in eight cases. Revision surgery was performed in five cases because of substantial growth during follow-up. In the three other cases, residual disease is still looked after. Two cases of petrous apex cholesteatoma were removed by an endoscopic transsphenoidal approach without recurrence.

DISCUSSION

How Extensive Should the Removal of PBC Be?

PBC is to be treated surgically. The goal is to remove completely PBC, a challenging issue when dealing with soft tissues as dura, jugular bulb, and nerves. Choice of approach is based on the location and extent of the lesion, hearing on the affected side, and anatomic position of the ICA and jugular bulb. It must provide safe and adequate exposure of the middle and posterior fossa dura, carotid artery, lateral sinus and jugular bulb,

and facial nerve.²⁻⁴ Whether surgery should peel off the matrix or be aggressive and remove all involved anatomic structures is still the matter of ongoing debate, for facing a benign and slowly progressive lesion.⁵⁻⁷ As regards dural involvement, bipolar coagulation of suspected dural portions has been advocated for devitalization of the cholesteatoma matrix.^{4,8} There is no report of systematic removal of dura, an option that might expose intractable CSF leak. Whether the walls of jugular bulb and sigmoid sinus should be removed seems controversial. Some have advocated complete removal of their lateral wall after luminal packing of Surgicel (Ethicon, Issy les Moulineaux, France) and ligation of the jugular vein in the neck. However, sacrifice of a permeable sigmoid sinus or jugular bulb might expose the patient to major complications such as headache, loss of vision, VI nerve palsy, cerebral and cerebellar edema, or infarctus.^{9,10} Concerning the ICA wall involvement, there is no rationale for the removal of a major artery. Therefore, the adequate exposure of the involved area should be allowed either by infratemporal fossa approach type B, modified transcochlear approach,^{2,4} or endoscopic transsphenoidal approach. As in the case of this series, complementary use of endoscopes in the course of lateral approaches allows to expose the anteromedial wall of the ICA for an adequate dissection of the matrix.

Functional and Dural Outcome

As in our series, hearing loss is often the main symptom.⁵⁻⁷ For most authors, removal of the otic capsule, when it is involved, is usually performed without hesitation to achieve the goal of radical removal of the pathology, but the status of the contralateral ear has to be considered to adjust the therapeutic approach. In fact, PBC in the only hearing ear is managed conservatively with regular radiological follow-up (CT scan and MRI are advised) and watchful waiting, and insertion of a cochlear implant in the contralateral ear may be considered.^{2,4}

Facial prognosis is of utmost importance for the patient. When the facial function is preoperatively normal, the prognosis is excellent, as underlined by our results, as far as matrix dissection on the nerve is performed gently under monitoring. In the contrary, preoperative facial nerve palsy, which is present in more than 50% of cases in the literature^{6,11} and was the second symptom in our series (37.5%), deserves a poor prognosis. Grade III and over palsies do not recover after conservative surgery. Therefore, aggressive management may be considered in those cases instead of nerve decompression, with rehabilitation by faciofacial end-to-end anastomosis or cable graft interposition if the proximal portion of the nerve is still available. In case of long-standing facial palsy (more than 1 year), most authors agree to recommend a hypoglossal-facial nerve anastomosis.^{2-4,8}

CSF leakage may result from dural tears occurring during matrix removal. Those are usually related to surgery rather than to the disease, and the case of patient 1 is unusual. A small dural tear is sealed by a muscle plug introduced through the defect in the subarachnoid space, and large defects require muscle plug, fascia, and fibrin glue.^{2,4} In the case of endoscopic transsphenoidal approach, various techniques are reliable, such as fascia lata tobacco pouch or use of a nasoseptal flap eventually combined with a CSF lumbar drainage when tight closure of the dural defect cannot be achieved.

Approach to the Petrous Apex

The petrous apex is a complex area due to the close proximity of vital structures. The thin matrix membrane of cholesteatoma often adheres tightly to the dura, the ICA, or the jugular bulb.^{8,12} Surgical approaches to the petrous apex include lateral transtemporal (translabyrinthine, transcochlear, infralabyrinthine, infracochlear) and middle fossa approaches, as opposed to the anterior transsphenoidal approach.^{13,14} By lateral transtemporal approaches, direct exposure of petrous apex cholesteatoma is difficult because of overlying structures including the facial nerve, ICA, jugular bulb, and otic capsule. Recent advances in minimally invasive surgery with the use of intraoperative navigational systems allow transnasal endoscopic access to petrous apex lesions such as cholesterol granuloma, petrous apicitis, and chondrosarcoma with minimal morbidity.^{13,15,16} To our knowledge, we are the first to use this approach for removing petrous apex cholesteatoma. Potential advantages of endoscopy include avoidance of a craniotomy, faster recovery, minimal postoperative symptoms, and shorter hospitalization. Facial weakness and hearing-vestibular loss are common complications of the lateral temporal and middle fossa approaches, which are not described in the endoscopic transsphenoidal approach. Potential major complications of this surgery include nerve and vascular injuries that can

be avoided by knowledge of anatomic relationships, recognition of anatomic variations, and use of image guidance system. If—as in most cases—the lesion is surrounded by an intact dura, morbidity seems minimal.

A combination of two surgical approaches can be of interest according to the initial PBC classification.

Long-term follow-up of PBC should be based on long-term MRI assessment, at least 5 years,⁸ as well as on clinical evaluation. Indeed, most recurrences are asymptomatic and detected only on MRI. Follow-up of an asymptomatic recurrence is proposed.

CONCLUSION

PBC surgery remains difficult, and functional deficits are frequent. Lateral transtemporal and middle fossa approaches are classically used but recent progresses in endoscopic surgery, using image guidance system, allow removing PBC in the petrous apex by an endoscopic transsphenoidal approach with minimal morbidity. Combination of an endoscopic transsphenoidal approach with a transtemporal or middle fossa approach may be considered for complete removal of a PBC reaching the petrous apex.¹⁴

REFERENCES

1. Magliulo G. Petrous bone cholesteatoma: clinical longitudinal study. *Eur Arch Otorhinolaryngol* 2007;264:115–120
2. Sanna M, Zini C, Gamoletti R, et al. Petrous bone cholesteatoma. *Skull Base Surg* 1993;3:201–213
3. Moffat D, Jones S, Smith W. Petrous temporal bone cholesteatoma: a new classification and long-term surgical outcomes. *Skull Base* 2008;18:107–115
4. Omran A, De Denato G, Piccirillo E, Leone O, Sanna M. Petrous bone cholesteatoma: management and outcomes. *Laryngoscope* 2006;116:619–626
5. Boko E, Morinière S, Lescanne E, Robier A. [Intrapetrous cholesteatoma. A retrospective study of seven operated cases]. [in French]. *Rev Laryngol Otol Rhinol (Bord)* 2006;127:211–216
6. Atlas MD, Moffat DA, Hardy DG. Petrous apex cholesteatoma: diagnostic and treatment dilemmas. *Laryngoscope* 1992;102(12 Pt 1):1363–1368
7. Magliulo G, Terranova G, Sepe C, Cordeschi S, Cristofar P. Petrous bone cholesteatoma and facial paralysis. *Clin Otolaryngol Allied Sci* 1998;23:253–258
8. Yanagihara N, Nakamura K, Hatakeyama T. Surgical management of petrous apex cholesteatoma: a therapeutic scheme. *Skull Base Surg* 1992;2:22–27
9. Keiper GL Jr, Sherman JD, Tomsick TA, Tew JM Jr. Dural sinus thrombosis and pseudotumor cerebri: unexpected complications of suboccipital craniotomy and translabyrinthine craniectomy. *J Neurosurg* 1999;91:192–197
10. Stramandinoli E, Lot G, Herman P, Corre-Gaubert H, Tran Ba Huy P. [Interruption of the lateral sinus in surgery of paraganglioma and acoustic neurinoma. Apropos of 2 complications]. [in French]. *Ann Otolaryngol Chir Cervicofac* 1998;115:362–366

11. Gyo K, Jyokou H, Komori M, Zenke K. A case of acquired petrous cholesteatoma associated with insidious middle ear infection treated by staging the surgical procedures. *Auris Nasus Larynx* 1995;22:192–196
12. Komune S, Nakagawa T, Haruta A, Matsuda K, Tono T. Management of cholesteatoma in the petrous apex. *Skull Base Surg* 2000;10:47–51
13. Zanation AM, Snyderman CH, Carrau RL, Gardner PA, Prevedello DM, Kassam AB. Endoscopic endonasal surgery for petrous apex lesions. *Laryngoscope* 2009;119:19–25
14. Mattox DE. Endoscopy-assisted surgery of the petrous apex. *Otolaryngol Head Neck Surg* 2004;130:229–241
15. Georgalas C, Kania R, Guichard JP, Sauvaget E, Tran Ba Huy P, Herman P. Endoscopic transsphenoidal surgery for cholesterol granulomas involving the petrous apex. *Clin Otolaryngol* 2008;33:38–42
16. Chatrath P, Nouraei SA, De Cordova J, Patel M, Saleh HA. Endonasal endoscopic approach to the petrous apex: an image-guided quantitative anatomical study. *Clin Otolaryngol* 2007;32:255–260