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Surgical Strategy and Facial Nerve Outcomes in Petrous Bone Cholesteatoma

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Key Words

Petrous bone cholesteatoma · Sanna classification · Transotic approach · Modified transcochlear approach · Facial nerve · Petrous internal carotid artery

Abstract

Objective: To review the classification and management of petrous bone cholesteatomas (PBCs) at our center and the outcomes of facial nerve (FN) management in these lesions. ***Methods:*** This was a retrospective study. The setting was a quaternary referral center for skull base pathology in Italy. A total of 200 patients with 201 PBCs were included in the study. All patients diagnosed radiologically with PBCs were classified according to the Sanna classification. All patients were surgically treated and followed up with radiology. The main outcome measures – classification of PBCs, the surgical approach used, disease control, and FN outcomes – were analyzed. ***Results:*** Supralabyrinthine PBCs were the most common type with 92 cases (45.8%) followed by the massive PBCs with 72 cases (35.8%). Preservation of preoperative FN function was highest in the infralabyrinthine (72.2%) and infralabyrinthine-apical (73.3%) types. The transotic approach was used in 66 cases (32.8%) in this series. The modified transcochlear approach type A was applied in 55 cases (27.3%). Active management of the nerve (rerouting, anastomosis, or grafting) was required in 53 cases (26.4%). Postoperatively, of the 116 cases with FN House-Brackmann grade I and II, 107 cases (92.2%) retained the same grade or improved. Recurrence was seen in 7 cases (3.5%). The mean duration of follow-up was 6.3 years. ***Conclusions:*** Radical disease clearance must take precedence over hearing and FN preservation in PBCs. Active FN management, including rerouting, end-to-end anastomosis, and cable nerve grafting, routinely come to play in the surgical management of PBCs, and the postoperative FN results after such interventions can be satisfactory in most cases.

Sampath Chandra Prasad and Gianluca Piras contributed equally to this work.

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Introduction

Petrous bone cholesteatomas (PBCs) are slow-growing expansile epidermoid lesions arising in the petrous portion of the temporal bone with an incidence of 4–9% of all petrous pyramid lesions. These can be congenital, acquired, or iatrogenic. Congenital PBCs are most plausibly explained by the persistence of fetal epidermoid formation in the petrous bone or the middle ear from which it expands to the petrous bone [Persaud et al., 2007]. The acquired variety is due to the migration of squamous epithelium into the petrous bone secondary to a perforation in the tympanic membrane. The iatrogenic variety is due to the implantation of cholesteatoma after otological surgery. 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. 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 [Lin et al., 2009; Pandya et al., 2010; Rijuneeta et al., 2008; Sanna et al., 1993]. Also, the close proximity of the disease to the labyrinth and the facial nerve (FN) puts to risk both hearing and FN function, which is reflected in the high incidence of FN palsy (34.6–100%) seen in the important series reported in the literature [Kim et al., 2014; Magliulo, 2007; Moffat et al., 2008; Sanna et al., 2011; Yanagihara et al., 1992]. The classification proposed by Sanna et al. [Pandya et al., 2010; Sanna et al., 1993, 2011], which is now widely accepted, divides PBCs into five groups based on the relationship of the disease to the labyrinthine block. This radiological classification allows standardization in reporting and a clear planning of the surgical approach.

Surgery remains the mainstay of treatment of PBCs. We have come a long way since Gacek [1980] 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 [Yanagihara et al., 1992]. 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. 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. In this article we review our long-term results in the largest series of PBCs published in the literature to date, revisit the classification, and perform a comprehensive review of the literature.

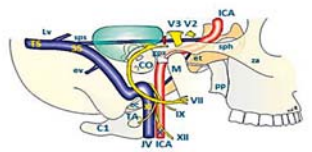
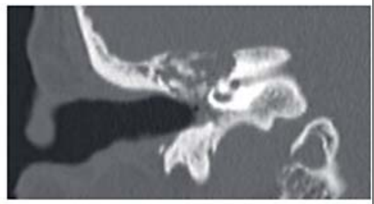
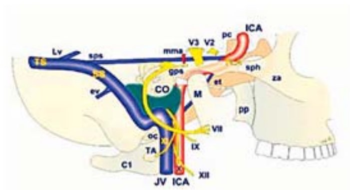
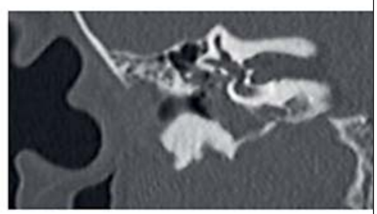
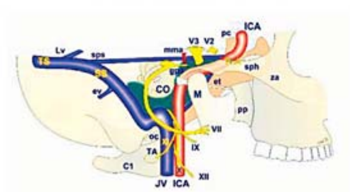
Materials and Methods

A total of 7,340 cases of cholesteatomas were surgically treated at the Gruppo Otologico, a quaternary referral neurotology and skull base center in Italy, from February 1979 to September 2015, of which the charts of 246 cases of PBCs were evaluated retrospectively; 45 cases with follow-up of less than 1 year or incomplete records were not included in the analysis. Hence, 201 cases of PBCs from 200 patients were included in the study, including 1 patient with bilateral cholesteatoma. The history of presenting illness, clinical examination findings, audiological and radiological examinations, and classification were documented. The type of approach, intraoperative findings, and postoperative complications were recorded. The pre- and postoperative FN function was graded according to the House-Brackmann (HB) grading system [House and Brackmann, 1985]. The follow-up period was analyzed for recurrences and late postoperative sequelae. Follow-up was defined as that period of time from surgery to the most recent office visit or patient contact.

At the Gruppo Otologico, pure-tone averages (PTAs) are noted for air conduction and bone conduction calculated before and after surgery as the mean of four thresholds at 500, 1,000, 2,000, and 4,000 Hz. Air-bone gaps were calculated using air and bone conduction values determined at the same time. At our center all patients with PBCs undergo preoperative high-resolution CT (cuts of 0.6–1 mm) and a cranial MRI with gadolinium enhancement. The PBCs were radiologically staged according to the Sanna classification for PBCs [Pandya et al., 2010; Sanna et al., 1993, 2011] that has been updated in this study (fig. 1). The surgical approaches and techniques of FN reconstruction have been comprehensively described elsewhere [Ozmen et al., 2011; Pandya et al., 2010; Sanna et al., 1993, 2006, 2008, 2011, 2012, 2013]. Since 1985, all the operated cavities have been obliterated with abdominal fat and the external auditory canal closed in a blind sac fashion. The postoperative follow-up imaging protocol includes a review of the operated area with CT and MRI (including diffusion-weighted imaging, DWI) at years 1, 2, 3, 5, and 10.

Descriptive statistics and comparison of proportions using χ^2 statistical analysis and Fisher's exact test were performed using IBM SPSS version 22 software (SPSS, Inc., Chicago, Ill., USA.). For Fisher's exact test requiring analysis of 2 × 5 contingency tables, Simple Interactive Statistical Analysis software was used. Statistical significance was accepted as $p < 0.05$ in all cases. This study was approved by the Ethics Committee of Casa Di Cura Hospital, Piacenza, Italy.

Color version available online

Class	Cholesteatoma location	Relations and features
Class I: Supralabyrinthine 	Centered on the geniculate ganglion area of the FN and the anterior epitympanum 	Superior: tegmen or dura Inferior: semicircular canals, apical turns of the cochlea Medial: limited extension beyond the otic capsule into the petrous apex Lateral: antrum, epitympanum, and further into middle ear Anterior: horizontal part of the pICA Posterior: posterior bony labyrinth Features: usually associated with fistula of the semicircular canals, erosion of tegmen, involvement of the FN
Class II: Infralabyrinthine 	Centered on the infracochlear, infralabyrinthine, and hypotympanic cells 	Superior: basal turn of the cochlea, vestibule Inferior: jugular bulb, lower cranial nerves, occipital condyle Medial: limited extension beyond the otic capsule into the petrous apex Lateral: hypotympanum and further into middle ear, retrofacial cells Anterior: vertical and horizontal part of pICA Posterior: posterior semicircular canal, IAC Features: fistula of the semicircular canals, erosion of the cochlea, jugular bulb, carotid canal, involvement of the lower cranial nerves
Class III: Infralabyrinthine-apical 	Involves infralabyrinthine cell tracts extending medially into the petrous apex 	Superior: basal turn of the cochlea, vestibule Inferior: jugular bulb, lower cranial nerves, occipital condyle Medial: extension into the petrous apex, lower clivus, along the greater wing of sphenoid into the foramen spinosum, foramen ovale, may extend up to sphenoid sinus Lateral: hypotympanum and further into middle ear, retrofacial cells Anterior: vertical and horizontal part of pICA Posterior: IAC, dura of the posterior cranial fossa (posterolaterally) Features: fistula of the semicircular canals, erosion of the cochlea, jugular bulb, involvement of the lower cranial nerves, extensive destruction of the carotid canal, involvement of the internal auditory canal

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(For rest of the figure and legend see next page.)

Results

A total of 246 cases of PBCs treated at the Gruppo Otologico were retrospectively analyzed. The incidence of PBCs among all cholesteatomas (7,340 cases) in our series was 3.3%. The age of the patients ranged from 9 to 83 years, with a mean age of 45 years; 141 patients were male and 59 were female, resulting in a male:female ratio of 2.4:1. The mean duration of follow-up was 6.3 years; 28 of the cases (14%) were purely congenital, 114 (57%) were acquired, and 59 (29%) were iatrogenic PBCs.

Classification

Supralabyrinthine PBCs were the most common type with 92 cases (45.8%) followed by massive PBCs with 72 cases (35.8%) (table 1). Apical PBCs were rare with just 4 cases (2%) in the series. Seven cases (7.6%) of supralabyrinthine PBCs had a limited apical extension. Eight cases had an extension to the clivus – 3 into the intradural areas, 2 into the sphenoid sinus, and 1 into the nasopharynx.

Clinical Features

A dead ear was seen in 98 of the cases (48.7%). There was no statistically significant difference in the rate of preoperative anacusis based on Sanna class ($p = 0.07$, Fisher's exact test). According to the American Academy of Otolaryngology-Head and Neck Surgery (AAO-HNS) classification, the mean preoperative PTA was 52 dB in the patients with supralabyrinthine, 45 dB in infralabyrinthine, 55 dB in infralabyrinthine-apical, 64 dB in massive, and 52 dB in apical PBCs. Two patients presented with disease in the only hearing ear. Hearing loss was most likely in massive PBCs ($p = 0.005$). Vertigo was seen in 32% of the cases. Tinnitus, headache, and trigeminal neuralgia were the other symptoms.

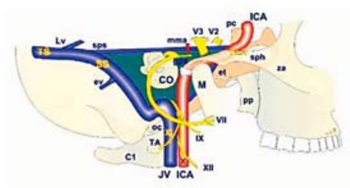
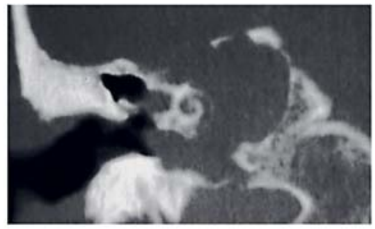
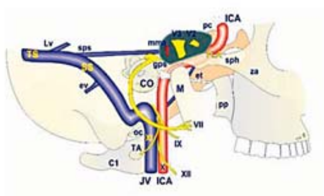
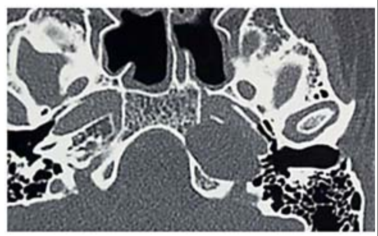
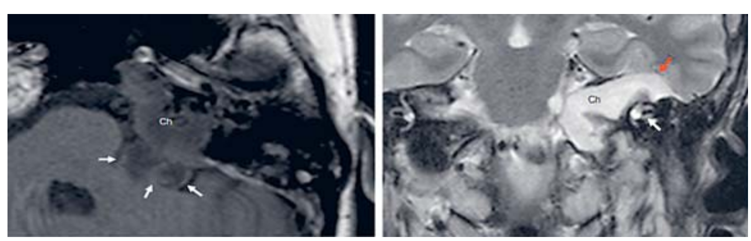
Class IV: Massive 	Centered on the otic capsule 	Superior: dura of the middle fossa, may extend intradurally Inferior: hypotympanic cells, infralabyrinthine cells, jugular bulb, lower cranial nerves Medial: extension into the petrous apex, along the greater wing of sphenoid into the foramen spinosum, foramen ovale, may extend up to sphenoid sinus Lateral: middle ear, antrum, retrofacial cells Anterior: vertical and horizontal part of pICA Posterior: IAC, dura of the posterior cranial fossa, may extend intradurally Features: various degrees of destruction of the otic capsule, involvement of FN
Class V: Apical 	Centered on the petrous apex 	Superior: dura of the middle fossa, Meckel's cave, may extend intradurally Inferior: hypotympanic cells, infralabyrinthine cells, jugular bulb, lower cranial nerves, infratemporal fossa Medial: extension into the spheno-petro-clival junction, midclivus, along the greater wing of sphenoid into the foramen spinosum, foramen ovale, may extend up to sphenoid sinus Lateral: otic capsule Anterior: horizontal part of pICA and foramen lacerum Posterior: IAC, dura of the posterior cranial fossa, may extend intradurally Features: otic capsule may be eroded medially, erosion of horizontal petrous carotid, clivus and intradural extensions into middle fossa or posterior fossa, extensions also possible into sphenoid, nasopharynx, or infratemporal fossa
Subclasses	**Relations and features**	
Clivus	Superior and midclival extensions are seen from massive, infralabyrinthine-apical and apical PBC, whereas the lower clival involvement is a feature of infralabyrinthine-apical PBC	
Sphenoid sinus	Sphenoid sinus involvement is seen from anteromedial extensions of massive, infralabyrinthine-apical, and apical PBC; it is a rare extension	
Nasopharynx	This is the rarest extension of the PBC; it is an extension of infralabyrinthine-apical or massive PBC, which may extend through the clivus beneath the sphenoid sinus into the nasopharynx	
Intradural	Intradural extensions may arise from the massive, infralabyrinthine-apical, and apical PBCs, usually into the posterior cranial fossa and rarely into the middle cranial fossa	
		

Fig. 1. Updated Sanna classification for PBCs. IAC = Internal auditory canal; CO = cochlea; M = mandible; PC = paraclival carotid; MMA = middle meningeal artery; SS = sigmoid sinus; TS = transverse sinus; EV = emissary vein; Lv = Labbe's vein; JV = jugular vein; TA = transverse process of the atlas; C1 = 1st cervical vertebra; OC = occipital condyle; PP = pterygoid plate; ET = Eustachian tube; SPH = sphenoid; ZA = zygomatic process; V3 = 3rd branch of the trigeminal nerve; V2 = 2nd branch of the trigeminal nerve; VII = FN; IX = glossopharyngeal nerve; XII = hypoglossal nerve; Ch = cholesteatoma.

Overall, 106 patients (52.7%) presented with preoperative FN palsy (HB grade II–VI) (table 2). In 35 (41.2%) of the 85 cases with definitive preoperative facial palsy (HB grade III or above), the duration of the paralysis was longer than 1 year. Preservation of preoperative FN function was highest in the infralabyrinthine (13/18, 72.2%) and infralabyrinthine-apical (11/15, 73.3%) types of PBCs.

Surgical Approaches

The TO approach, used in 66 cases (32.8%) in this series, was the preferred approach in all types of PBCs except the massive type. The modified transcochlear approach (MTCA) type A was applied in 55 of the cases (27.3%), mostly in massive PBCs, to achieve a better control over the horizontal petrous internal carotid artery (pICA). Subtotal petrosectomy was used in 26 cas-

Table 1. Sanna classification of PBCs vs. the approach used in this series

	Sanna classification of PBCs					
	SL	IL	IL-A	M	A	Total
Surgical approach						
TO	34	6	9	16	1	66 (32.8)
MTC						
Type A	17	2	2	32	2	55 (27.3)
Type B	0	0	1	3	0	4 (2)
TL	18	1	2	14	0	35 (18)
STP	17	6	0	3	0	26 (12.9)
TM	3	1	0	2	0	6 (3)
TM+MCF	2	0	0	2	0	4 (2)
RL+TM	1	2	0	0	0	3 (1)
IFTA-B	0	0	1	0	1	2 (1)
Total	92 (45.8)	18 (8.9)	15 (7.5)	72 (35.8)	4 (2)	201 (100)

Data are presented as n (%). SL = Supralabyrinthine; M = massive; IL = infralabyrinthine; IL-A = infralabyrinthine-apical; A = apical; TM+MCF = transmastoid + middle cranial fossa; TO = transotic approach; MTC = modified transcochlear; STP = subtotal petrosectomy; IFTA-B = ITFA type B; TL = translabyrinthine; RL+TM = retrolabyrinthine + transmastoid; TM = transmastoid.

Table 2. Pre- and postoperative FN function vs. class of PBCs

	Sanna classification of PBCs											
	SL		IL		IL-A		M		A		Total	
	pre	post	pre	post	pre	post	pre	post	pre	post	pre	post
HB grades												
I	45	40	13	12	11	10	25	25	1	0	95 (47.2)	87 (43.2)
II	8	11	1	1	2	2	9	5	1	1	21 (10.4)	20 (10)
III	15	24	3	3	0	1	10	12	0	0	28 (14)	40 (20)
IV	7	3	1	0	0	0	3	7	1	1	12 (6)	12 (6)
V	3	4	0	0	0	0	1	2	0	0	4 (2)	5 (2.4)
VI	14	10	0	2	2	2	24	21	1	2	41 (20.4)	37 (18.4)
Total	92		18		15		72		4		201 (100)	

Data are presented as n (%). SL = Supralabyrinthine; M = massive; IL = Infralabyrinthine; IL-A = infralabyrinthine-apical; A = apical; pre = preoperative; post = postoperative.

es (12.9%), mostly in supralabyrinthine PBCs. A translabyrinthine approach was used in 35 cases (18%) where the cholesteatoma was found to involve the internal auditory canal. The MTCA type B was the approach of choice in 4 cases of PBCs extending to the clivus or nasopharynx. In 2 patients undergoing a subtotal petrosectomy in the only hearing ear, an ipsilateral cochlear implantation was performed at the same time. None of the classes of PBCs were predictive of the individual approaches.

FN Management and Results

In total, 47.2% of the patients had a normal FN, 32.4% had various degrees of incomplete palsy, and 20.4% had complete FN palsy (table 3). All the FN palsies had a duration of less than a year, except in 8 cases where they were 1 year or longer. The most commonly involved segments of the FN were the tympanic (68%), geniculate ganglion (55%), and labyrinthine segments (50%). The FN was involved in multiple segments in 76% cases. Some form of management of the FN was required in all cases (table 3);

Table 3. Intraoperative FN management in PBCs

a Type of FN management

	Details	Cases	FN function	
			postoperative improvement[1]	postoperative worsening[2]
Skeletalization of the nerve	Drilling around various parts of the Fallopian canal but maintaining thin bone around it	46 (22.8)	2 (11.7)	2 (10)
Nerve decompression	Exposure of the nerve without incision of the epineurium; no mobilization	94 (46.8)	7 (41.2)	3 (15)
Active FN management				
Nerve sectioning and end-to-end anastomosis	In deficits less than 3 mm	6 (3)	1 (5.9)	1 (5)
Posterior rerouting				
Partial	Exposure and rerouting of the tympanic, labyrinthine, and/or IAC segments	2 (1)	–	2 (10)
Total	As part of MTCA	11 (5.5)	1 (5.9)	5 (25)
Posterior rerouting and end-to-end anastomosis	As part of MTCA	3 (1.5)	–	–
Nerve sectioning and cable sural nerve grafting	In cases where it was impossible to separate the nerve from the lesion	31 (15.4)	6 (35.3)	5 (25)
Facial hypoglossal anastomosis	Performed as a second stage in cases of longstanding FN paralysis in HB grade >IV (1 year or longer)	8 (4)	–	0 (0)
Total	All cases required some form of FN management	201 (100)	17 (8.5)	20 (9.9)

b Detailed analysis of cases with active intraoperative FN management

	Nerve sectioning and end-to-end anastomosis		Posterior rerouting		Posterior rerouting and end-to-end anastomosis		Nerve sectioning and cable sural nerve grafting		Facial hypoglossal anastomosis		Total	
	pre	post	pre	post	pre	post	pre	post	pre	post	pre	post
HB grades												
I	0	0	5	0	0	0	1	0	0	0	6	0
II	0	0	2	2	0	0	4	0	0	0	6	2
III	5	2	2	8	0	2	4	10	0	1	10	23
IV	0	2	0	1	2	0	3	4	2	3	8	10
V	0	1	1	0	0	0	1	3	3	2	5	6
VI	1	1	3	2	1	1	18	14	3	2	26	20
Total	6		13		3		31		8		61 (100)	

Data are presented as n (%). Pre = Preoperative; post = postoperative.
[1]Defined as a decrease in 2 HB grades or lower compared to that during presentation.
[2]Defined as an increase in 2 HB grades or higher compared to that during presentation.

however, active management of the nerve (rerouting, anastomosis, or grafting) was required in 53 cases (26.4%). In 8 cases with a preoperative FN function of HB IV or greater, the FN was restituted by facial-hypoglossal anastomosis. In all 8 cases the FN paralysis was present for 1 year or longer. There was a postoperative improvement in 3 cases.

Postoperatively, of the 116 cases with FN HB grades I and II (considered normal or near normal), 107 cases (92.2%) retained the same grade or improved postoperatively. An improvement of FN function (defined as a decrease in two HB grades and lower compared to that during presentation) was encountered in 17 cases. Conversely, a worsening of FN function (defined as an increase in two HB grades and higher compared to that during presentation) was encountered in 20 cases, including 2 cases where the FN was only skeletonized. Preoperative HB grading of the FN was found to have a significant effect on the outcome ($p = 0.002$).

A total of 53 of the 201 cases (26.4%) required active management of the FN due to the infiltration of the nerve by the lesion or the close proximity of the lesion over considerable lengths of the nerve. Active management included rerouting, sectioning, and end-to-end anastomosis or cable grafting of the FN. Only 12 of these patients (22.6%) had a preoperative FN function HB grade I–II. Of the 31 cases where the FN was reconstructed with a cable sural nerve graft or direct end-to-end anastomosis, the best postoperative result obtained was HB grade III. Of the 13 cases (58.5%) where the FN was rerouted posteriorly, 8 cases resulted in HB grade III, and 2 cases resulted in HB grade II. Of the 3 cases where a posterior rerouting of the FN was combined with end-to-end anastomosis due to an interruption of the FN, 2 recovered to HB grade III (table 3).

Of the 53 cases that required active management of the FN, 22 (41.5%) ended up with a HB grade V or VI after 1 year of follow-up. In 2 cases where the FN was not actively managed, the HB grade deteriorated to IV and V, respectively. Both patients were rehabilitated by facial-hypoglossal anastomosis. There was no statistically significant difference in pre- and postoperative HB grades between the classes of PBCs ($p > 0.05$ in all cases).

Hearing Results

Postoperatively, surgery led to destruction of hearing in 78.4% of cases; 88.3% of patients with massive and 74.4% of those with supralabyrinthine PBCs ended up with anacusis postoperatively. The mean postoperative PTA was 67 ± 20 dB in those patients with residual hearing. The details of hearing outcomes will be published separately in a subsequent article and are beyond the scope of this paper.

Complications and Recurrences

Postoperatively, 164 of the 198 ears (82.8%) that had various degrees of hearing loss ended up as dead ears. This was an obvious sequela of the surgical approaches chosen for the radical removal of the disease. One patient in this series developed a CSF leak leading to a brain abscess which was subsequently drained, and the patient recovered completely. Recurrence of PBC occurred in 7 cases (3.5%), of which 3 were in previous radical cavities. The recurrences in the 7 cases were noted between 1 and 3 years in 3 cases, between 3 and 5 years in 2 cases, and between 5 and 7 years in 2 cases. Disease recurrence in massive PBCs was statistically significant ($p = 0.005$).

Discussion

The main challenges of dealing with PBC are diagnostic and therapeutic: diagnostic due to a delay in identification of the disease owing to its silent growth patterns and therapeutic due to the difficulty in the complete surgical extirpation of the disease owing to its complex location. Diagnostic challenges have been partly surmounted by the introduction of techniques specific to cholesteatoma such as DWI, which has made it possible to detect lesions early and identify early recurrences in patients after surgery. The evolution of skull base surgery, both lateral microsurgical and anterior endoscopic, has made it possible to remove even the most centrally located lesions in the skull base with acceptable morbidity.

Terminology and Classification

Various nomenclatures have been used to describe this pathology. They include petrous apex cholesteatoma, petrosal cholesteatoma, congenital cholesteatoma of the temporal (or petrous) bone, middle ear cholesteatoma involving the petrous apex, or sometimes simply congenital cholesteatoma. According to us, the most appropriate terminology would be ‘petrous bone cholesteatoma’ as this is a lesion predominantly located in the petrous part of the temporal bone. Petrous apex cholesteatoma, which is the most commonly misused term, is inappropriate because, firstly, the apex by definition refers to the area of the tip of the petrous bone that meets the clivus whereto the disease extends infrequently, and secondly, the petrous apex excludes the middle ear, which is practically always involved by the disease. The use of the term congenital cholesteatoma is also erroneous as PBCs can be congenital, acquired, or iatrogenic.

The concept of classification is of paramount importance not only in surgical planning [Pareschi et al., 2001] but also in standardization in documentation and reporting. We use the Sanna classification proposed by the senior author (M.S.) to diagnose all cases of PBCs [Pandya et al., 2010; Sanna et al., 1993, 2011]. This classification has gained widespread acceptance and is used in most of the important series reported in recent times [Alvarez et al., 2011; Aubry et al., 2010a, b; Danesi, et al., 2016; Kim et al., 2014; Magliulo et al., 2007; Pareschi, et al., 2001; Senn et al., 2011; Tutar et al., 2013]. The updated classification (fig. 1) is an attempt to incorporate these complex but rare situations wherein the disease may extend into deeper areas like the clivus, sphenoid sinus, nasopharynx, and intradural areas.

Principles of Surgical Management

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 FN results and even hearing preservation. Hearing preservation surgery can be used in limited PBCs of the supralabyrinthine (combined middle cranial fossa-transmastoid approach) and infralabyrinthine (transmastoid-retrofacial approach) varieties. However, it must be noted that in most cases preservation of the otic capsule is impossible, as experienced by most authors [Alvarez et al., 2011; Aubry et al., 2010a, b; Axon et al., 1999; Danesi, et al., 2016; Magliulo, 2007; Pareschi et al., 2001], and this must be taken as the price to pay for achieving total disease clearance and in many cases also for the preservation of the FN.

The MTCA [Sanna et al., 1998, 2008], used in 59 cases (29.4%) in this series, provides excellent exposure of the clivus, petrous apex, dura of the posterior and middle fossa, sigmoid sinus, jugular bulb, and pICA. It also avoids cavity problems such as skin entrapment and disease recurrence, with minimal incidence of CSF leakage. Combining an infratemporal fossa approach (ITFA) type B and MTCA provides adequate exposure of the vertical and horizontal portions of the pICA, sphenoid sinus, and mid-clivus. The TO approach, used in 66 of the cases (32.8%) in our series, is used when the preoperative FN function is normal. Here the fallopian canal is kept intact, and disease is cleared all around it. On the other hand, the modified translabyrinthine approach with removal of the external auditory canal and blind sac closure, employed in 35 cases (17.4%) in this series, is used in extensive lesions without cochlear involvement [Omran et al., 2006]. This approach also allows simultaneous ipsilateral cochlear implantation if the cochlea and the cochlear nerve can be preserved. Subtotal petrosectomy with middle ear exclusion and blind closure of the external auditory canal, used in 28 cases (13.9%) in this series, is a useful procedure in infralabyrinthine or limited supralabyrinthine extensions. A simple transmastoid approach, used in only 6 cases (3%) in this series, is rarely sufficient to remove a PBC. A combination of a transmastoid with a middle cranial fossa approach, which was used in 4 cases (2%) in this series, is applied when the disease is in the ear wherein the sensorineural hearing is well preserved or in the only hearing ear. However, this also is a surgical option that rarely needs to be exercised in typical PBCs.

Management of the FN

The FN may be skeletonized, rerouted, cut with end-to-end anastomosis, or grafted with a cable motor nerve. The best possible functional outcome after FN grafting in our series was HB grade III. Also, in FN rerouting, the results were HB grade III or worse in most cases, except in 2 cases where it was grade II. The depreciation in FN function is because the surgical procedure usually involves disruption of the blood supply from the deep petrosal artery near the geniculate ganglion. In another study (in press), we analyzed the results of 213 cases where the FN was grafted for all pathologies of the skull base including vestibular schwannomas, meningiomas, FN tumors, and PBCs (follow-up longer than 1 year). The study revealed that PBCs had the worst outcome. The reason for this could be the fact that, preoperatively, most of the cases presented with varying grades (>HB III) of long-standing FN palsy. In patients with long duration of facial palsy (>12 months) facial-masseteric nerve grafting or facial-hypoglossal anastomosis is indicated.

Management of the Dura

Once the dura is exposed clearly all around the lesion, the matrix can be peeled off the dura using a flag or circular knife. In the case of adherence, the matrix can be neutralized by gently coagulating over it and the surrounding dura. There is a risk of opening the dura while removing the adherent matrix, causing an intraoperative CSF leak. CSF leaks resulting from dural tears do not need special repair but can be swiftly managed by inserting free muscle plugs into the subarachnoid space through the defect and by cavity obliteration with fat.

Management of the Jugular Bulb

Dealing with the jugular bulb in cases where it is involved is a well-thought-out strategy. Preoperative imaging must be carefully analyzed for two aspects: the relationship of the lesion with the jugular bulb and the patency of the contralateral venous drainage system by MR venography. In the presence of hypoplasia of the contralateral venous system, sacrifice of the bulb means occlusion of the main venous drainage of the brain, with the consequent risk of benign intracranial hypertension

Table 4. Comparison of results of important series of PBCs with the present series

Study	Classification used	According to Sanna classification for PBCs					Preoperative FN palsy	Surgical approaches	Postoperative FN results at 1-year follow-up following active intraoperative management of nerve[1]			Complications including FN palsy when no active management of FN was done	Recurrence
		SL	IL	IL-A	M	A			HB II and III	HB IV	>HB V		
Yanagihara et al., 1992 (n = 16)	Not classified	NA					16 (100)	RM+L: 9 (56), ICWM+L: 2 (13), STP: 3 (19), MCF+RM+L: 1 (6), MCF+ICWM+L: 1 (6)	5 cases; NA			Total FN palsy: 8 (50) 2nd cavity obliteration: 3 (19)	0 (0)
Axon et al., 1999 (n = 25)	Not classified	NA					14 (56)	TC: 17 (68), TL: 5 (20), TM+MCF: 3 (12)	4 (33), including 1 case of HB I	2 (17)	6 (50)	18 exteriorizations with 5 (20) chronic ear discharge	5 (20)
Steward et al., 2000 (n = 11)	Not classified	NA (all cases were called extensive anterior epitympanic cholesteatomas)					3 (27)	TM+MCF: 11 (100)	NA			–	2 (18)
Grayeli et al., 2000 (n = 19)	Fisch	3 (16)	3 (16)	4 TrL, 1 RtL (26)		8 (42)	12 (63)	MF/EMF: 6 (32), TM+MCF: 2 (11), TM+RM: 1 (5), STP: 1 (5), TO: 9 (47)	No active management, only decompression			FN palsy (HB V/VI): 3 (16) CSF leak: 2 (11)	2 (11)
Pareschi et al., 2001 (n = 25)	Pareschi	8 (32)	1 (4)	–	16 (64)	–	15 (60)	TC: 16 (64), TL: 6 (24), MCF: 2 (8), ITFA-A: 1 (4)	3 (100%)	–	–	Worsening FN: 11 (44)	1 (4)
Wu et al., 2004 (n = 12)	Not classified	NA					10 (83)	TL: 8 (67), MCF: 4 (33)	–	2 (67)	1 (33)	2nd cavity obliteration: 1 (8) Epidural hematoma: 1 (8) Infection: 1 (8)	1 (8)
Magliulo, 2006 (n = 52)	Sanna	19	15	4	12	2	28 (54)	STP: 24 (46), TT+SL: 5 (10), TO/TL: 18 (35), TC: 2 (4), ITFA-A/B: 3 (6)	<HB II, 0 (0), HB III–IV, 8 (57)		6 (43)	CSF leak: 3 (6)	4 (8)
Moffat et al., 2008 (n = 43)	Moffat-Smith	6 (14)	1 (2)	3 (7)	30 (70)	3 (7)	22 (51)	STP: 25 (58), TT+SL: 8 (19), TO: 4 (9), TC: 1 (2), ITFA-A: 3 (7), TT+SL+MCF: 1 (2), STP+MCF: 1 (2)	5 (12) primary anastomosis, 1 (2) GAN graft, 1 (2) SN graft			CSF leak: 4 (9) Deep vein thrombosis: 1 (2)	2 (5)
Aubry et al., 2010 (n = 28)	Sanna	2 (7)	6 (21)	4 (14)	10 (36)	6 (21)	10 (38)	TO: 14 (50), TL: 4 (14), TC: 1 (4), MCF+TC: 1 (4), MCF: 2 (7), ITFA-B: 2 (7), TS: 2 (7), IL: 1 (4), IC: 1 (4)	–			Transient hemiparesis and worsening VI nerve palsy: 1 (4)	8 (29)
Song et al., 2011 (n = 13)	Ishii	Anterior-superior labyrinthine (10, 77), posterior-superior labyrinthine (3, 23)					6 (46)	TL: 6 (46), STP: 3 (23), TM: 2 (15), TO: 1 (8), MCF: 1 (8)	2 (50), 1 case of HB I after rerouting	–	2 (50)	Infection: 1 (7)	0 (0)
Senn et al., 2011 (n = 21)	Sanna	14 (67)	2 (10)	2 (10)	3 (14)	–	6 (29)	SL/TL: 12 (57), TC: 3 (14), IL: 4 (19), STP+TCr: 2 (10), in 19 cases the cavity was left open after tympanoplasty	NA (in 2 cases FN rerouted)			XII–V anastomosis: 1 (5) CSF leak: 1 (5)	4 (19)
Alvarez et al., 2011 (n = 35)	Sanna	18 (52)	12 (34)	–	5 (14)	–	26 (74)	MCF: 6 (17), TL/TC: 15 (43), ITFA-B: 4 (12), ITFA-A: 2 (6), MRM: 4 (11), STP: 4 (11)	NA			NA	4 (11)
Tutar et al., 2013 (n = 34)	Sanna	13 (38)	3 (9)	4 (12)	13 (38)	1 (3)	20 (59)	NA	4 cases; NA			FN palsy: 2 (6) CSF leak: 3 (8) Meningitis: 1 (2) BA: 1 (2) SNHL: 1 (2)	2 (5)
Kim et al., 2014 (n = 31)	Sanna	16 (52)	–	1 (3)	13 (42)	1 (3)	11 (35)	TM: 15 (48), TL: 7 (23), TC: 5 (16), STP: 4 (13)	5 (100)	–	–	Infection: 4 (13) Dizziness: 1 (3)	1 (3)

Table 4 (continued)

Study	Classification used	According to Sanna classification for PBCs					Preoperative FN palsy	Surgical approaches	Postoperative FN results at 1-year follow-up following active intraoperative management of nerve[1]			Complications including FN palsy when no active management of FN was done	Recurrence
		SL	IL	IL-A	M	A			HB II and III	HB IV	>HB V		
Chen et al., 2015 (n = 38)	Not classified	NA					32 (84)	TO: 23 (61), MCF: 12 (32), ER: 3 (8)	NA			NA	NA
Danesi et al,, 2016 (n = 81)	Sanna	57	10	6	4	4	66 (81)	TLA: 27 (33), STP: 22 (27), LP+MCF: 14 (17), Others: 18 (23)	NA			–	21 (26)
Present Series, 2015 (n = 201)	Sanna	92 (46)	18 (9)	15 (8)	72 (36)	4 (2)	106 (53)	TC: 59 (29), TO: 66 (33), TL: 35 (18), STP: 26 (13), TM: 6 (3), TM+MCF: 4 (2), TM+RM: 3 (1), ITFA-B: 2 (1)	2 (4)	22 (42)	29 (55)	MEH: 2 (1) CSF leak+BA: 1 (0.5)	7 (4)

Data are presented as n (%). % = Percentages (not exact to decimals); NA = not available; GAN = great auricular nerve; SN = sural nerve; SL = supralabyrinthine; IL = infralabyrinthine; A = apical, IL-A = infralabyrinthine-apical; M = massive; TrL = translabyrinthine; RtL = retrolabyrinthine; IL = infralabyrinthine; IC = infracochlear; RM = radical mastoidectomy; L = labyrinthectomy; ICWM = intact canal wall mastoidectomy; MRM = modified radical mastoidectomy; MCF = middle cranial fossa; EMCF = extended middle cranial fossa; TCr = temporal craniotomy; TL = translabyrinthine; STP = subtotal petrosectomy; TM = transmastoid; ER = endoscopic resection; TS = transphenoidal; MEH = meningoencephalic herniation; BA = brain abscess; SNHL = sensorineural hearing loss.

[1] Active management includes mobilization, sectioning, and end-to-end anastomosis and grafting.

or venous infarction of the temporal lobe. In such cases damage to the jugular bulb or the sigmoid sinus must be avoided at all costs. However, in cases where the opposite venous drainage is patent, it is safe to occlude the jugular bulb or sigmoid sinus by intra- or extraluminal packing with Surgicel if it is damaged during dissection.

Management of Complex Cases

Radical clearance can be achieved even in such complex cases by the addition of an ITFA type B to a TO approach or one of the modified TC approaches. In the case of midline extensions, anterior endoscopic approaches can be used to good effect by combining them with the lateral approaches, either in a single or two-staged procedure. In our series, only 1 complex case had a recurrence near the paraclival ICA, which was removed using an anterior endoscopic approach. The details of management are described elsewhere and are beyond the scope of discussion in this article.

Review of the Literature

All authors reporting PBCs with a series of at least 10 cases since 1990 were reviewed. In the case of multiple publications from the same center, the latest publication was taken into account. A total of 16 series were analyzed for the classification system used, surgical approaches, FN results after active management (including end-to-end anastomosis, rerouting, and nerve grafting), complications, and recurrences (table 4). The present series emerged as the largest series in the world literature to date with 201 cases studied, the next biggest series being 52 cases. Almost all series after 2004 have used the Sanna classification for PBCs. Preoperative FN palsy was seen in 27–100% of cases. Lateral skull base approaches like TO, TC, and ITFAs were often used in series to deal with PBCs after 2000 with good outcomes, while most series before this used exteriorization procedures with poor outcomes. It was seen that most authors did not report FN outcomes after active management. In our opinion, for any meaningful results this must be analyzed separately from FN outcomes where no active management is used. For such a high number of cases, the complication and recurrence rates in our series were one of the lowest.

Conclusions

PBCs are rare and challenging lesions to treat, and they require thorough preoperative evaluation and surgical planning as well as training in lateral skull base approaches. Following a widely accepted classification allows standardization of reporting and, in turn, the formulation of treatment policy. Radical surgery is the treatment of choice, and hearing preservation must be a factor secondary to total disease clearance. Active FN management, including rerouting, end-to-end anastomosis, and cable nerve grafting consistently come to play in the surgical management of PBCs, and postoperative FN results can be satisfactory in most cases. Follow-up has been rendered safe and easy with the development of DWI and other improvements in imaging.

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Disclosure Statement

The authors have no conflicts of interest to declare.

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