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Petrous Bone Cholesteatoma: Classification, Management and Review of the Literature

Mario Sanna[a] Yash Pandya[b] Fernando Mancini[a] Giuliano Sequino[a] Enrico Piccirillo[a]

[a]Gruppo Otologico, Casa di Cura, Piacenza, Italia; [b]Pandya's ENT Clinic, Rajkot, India

Key Words

Skull base surgery · Petrous bone cholesteatoma · Facial nerve · Internal carotid artery · Jugular bulb · Lower cranial nerves · Dura and hearing preservation

Abstract

Objective: To discuss the classification of petrous bone cholesteatoma (PBC) and add a subclassification; to review the existing literature and to propose the ideal surgical management of PBC based upon the experience of the largest series published in the literature until now. ***Study Design:*** Retrospective analysis. ***Setting:*** Quaternary referral neuro-otologic private practice. ***Materials and Methods:*** The data of 129 patients who underwent surgery for PBC between 1979 and 2008 were analyzed with respect to the classification, type of the approach used, facial nerve lesion and its management, recurrences and outcome. ***Results:*** Out of the 129 PBC cases 64 were supralabyrinthine, 9 infralabyrinthine, 7 infralabyrinthine-apical, 48 massive and 1 apical. The facial nerve was involved in 95% of the cases. Hearing could not be preserved in 82% of the cases due to the extent of the lesions and the surgical approaches used. The internal carotid artery, jugular bulb and the lower cranial nerves were infrequently involved, but demanded careful identification and meticulous care to avoid complications. Obliteration of the cavities provided a safe solution for protection of the exposed dura and the vital neurovascular structures. Recurrences were observed in 5 cases. ***Conclusion:*** The classification of PBC is fundamental to choose the appropriate surgical approach; the facial nerve is involved in almost all the cases, radical removal takes priority over hearing preservation and cavity obliteration is important to protect the vital neurovascular structures which may be exposed.

Introduction

The term petrous bone cholesteatoma (PBC) is used to define an epidermoid cyst of the petrous portion of the temporal bone. It is a rare pathologic entity with a reported incidence of 4–9% of all petrous pyramid lesions [Omran et al., 2006].

The central position of the otic capsule and the complex anatomical relationship of this part of the bone with vital intracranial structures (facial nerve, internal carotid artery, sigmoid sinus, jugular bulb, lower cranial nerves, middle and posterior fossa dura, temporal lobe and cerebellum) render PBC a challenging pathology even for

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Dr. Yash Pandya, MS
Dr. Pandya's ENT Hospital
Dr. Rajendra Prasad Road
Rajkot 360001 (India)
Tel. ■■■, Fax ■■■, E-Mail yash_pandya81@hotmail.com

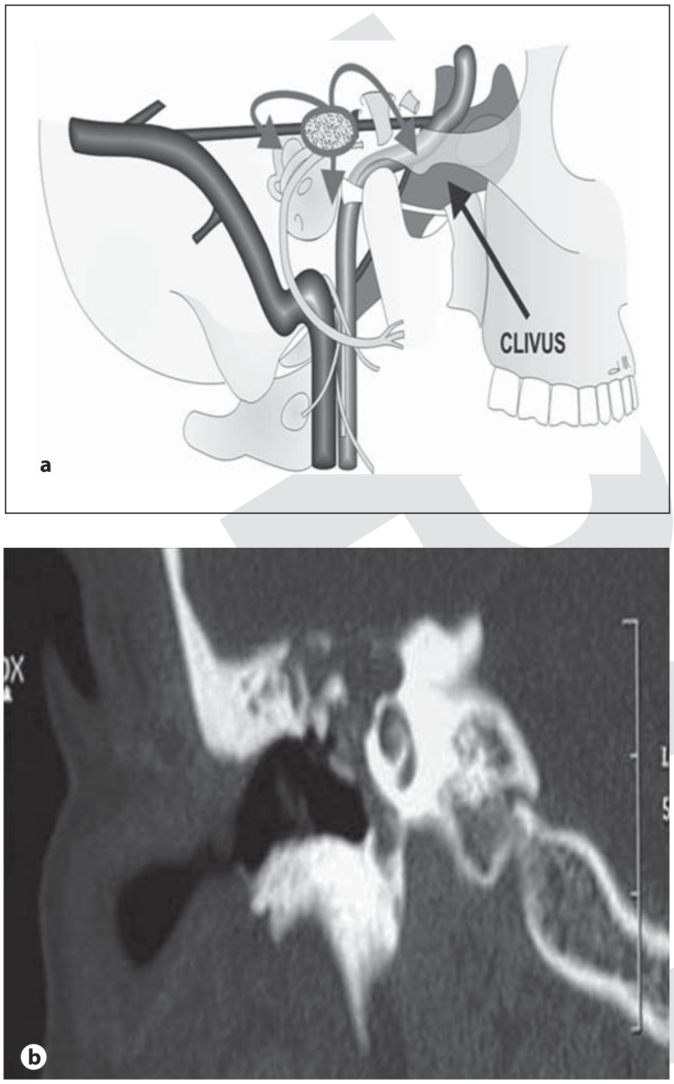


Fig. 1. a Diagrammatic representation of a supralabyrinthine PBC as viewed from the lateral aspect. The dotted ovoid area represents the site of the lesion and the arrows represent the route of spread. Directions for the route of spread are given in table 1. **b** Coronal HRCT image of the temporal bone showing a supralabyrinthine PBC. The dural plate is eroded. The cochlea is intact.

the most experienced surgeons. Extension of the PBC to the clivus, sphenoid sinus or rhinopharynx, even if rare, can be extremely difficult to treat.

According to Sanna et al. [1993], PBCs can be classified into five groups: supralabyrinthine, infralabyrinthine, massive, infralabyrinthine-apical, and apical (fig. 1–8). These terms describe both the location and the extent of the lesion.

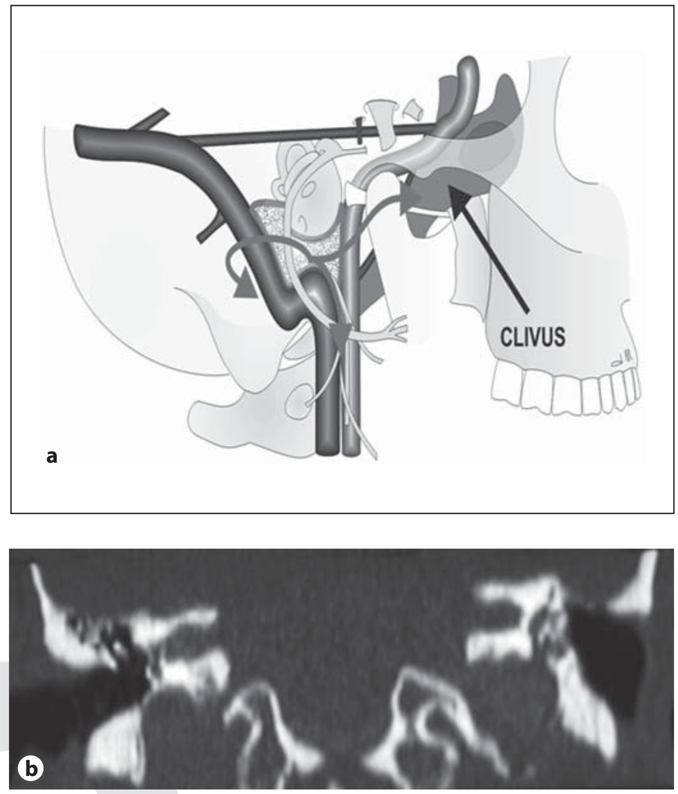


Fig. 2. a Diagrammatic representation of an infralabyrinthine PBC as viewed from the lateral aspect. The dotted area represents the site of the lesion and the arrows represent the route of spread. Directions for the route of spread are given in table 1. **b** Coronal HRCT image of the temporal bone showing an infralabyrinthine PBC.

The advances in lateral skull base approaches over the past few decades have greatly influenced the management of PBC and minimized morbidity. In spite of the excellent imaging techniques and the advanced surgical approaches, postoperative morbidity still remains a disparaging issue.

The present article aims at proposing an ideal surgical management for various classes of PBC based on the experience of 129 cases, introducing a subclassification for the extension of PBC to the existing Sanna classification (table 1) along with a review of the literature on PBC.

Materials and Methods

A retrospective case study was performed on 128 patients diagnosed and treated for PBC out of 4,500 cholesteatoma cases from 30/02/79 till 30/11/08 by the Gruppo Otologico, Piacenza and Rome, Italy. All the patients were operated on by the senior

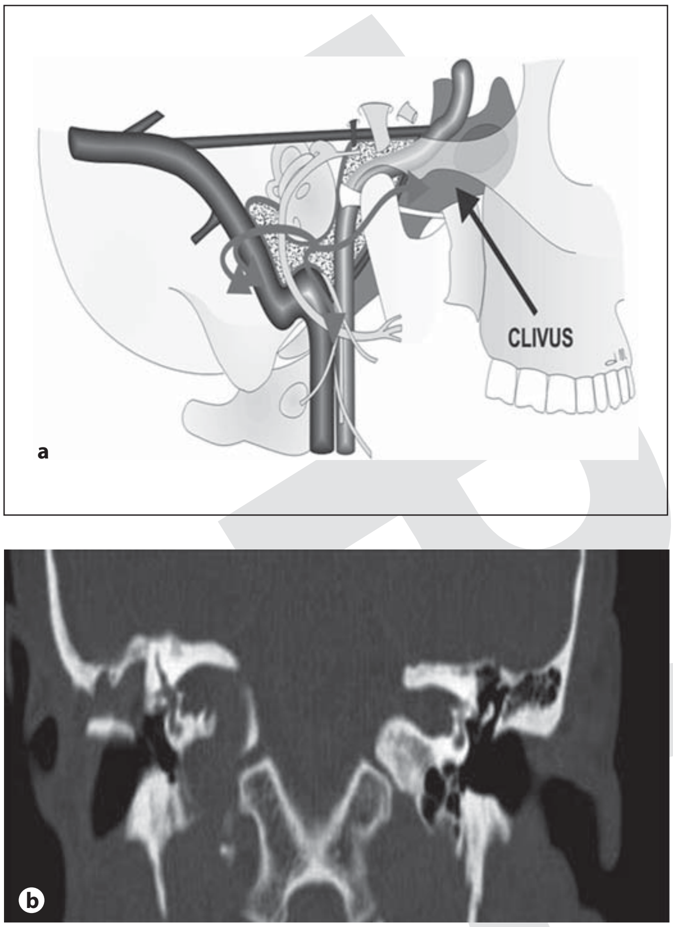


Fig. 3. a Diagrammatic representation of an infralabyrinthine-apical PBC as viewed from the lateral aspect. The dotted area represents the site of the lesion and the arrow represents the route of spread. Directions for the route of spread are given in table 1. **b** Coronal HRCT image of the temporal bone showing an infralabyrinthine-apical PBC. Cholesteatoma involves the inferior aspect of the internal auditory canal.

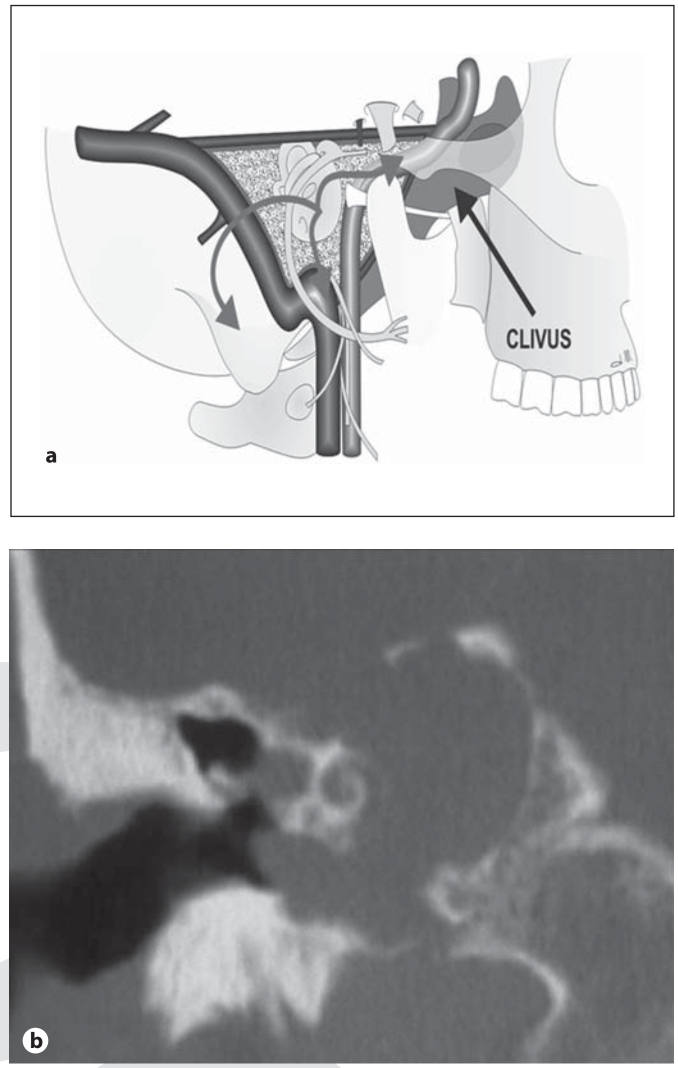


Fig. 4. a Diagrammatic representation of a massive PBC as viewed from the lateral aspect. The dotted area represents the site of the lesion and the arrows represent the route of spread. Directions for the route of spread are given in table 1. **b** Coronal HRCT image of the temporal bone showing a massive PBC. The cholesteatoma has eroded the cochlea extending into the petrous apex reaching up to the clivus. Cholesteatoma is eroding the dural plate.

author. There was 1 patient with bilateral PBC, hence the total number of cases was 129. There were 93 males and 35 females. The right side was affected in 74 patients whereas 55 patients had a left-sided lesion. All these patients underwent a thorough otoneurologic evaluation followed by pure-tone audiometric evaluation.

A high-resolution CT (HRCT) scan of the temporal bone (coronal and axial scans; bone window images with a width of 1–2 mm) was indicated in all patients with a history of chronic otorrhea, anacusis, facial palsy, vertigo, or lower cranial nerve paralysis. A history of previous ear surgery was another indication for a CT scan. A cerebral MRI (T_1, T_2-weighted images with gadolinium enhancement) was obtained in cases suspected of extratemporal spread on the CT scan till 1999, after which we got MRI in all cases.

All the lesions were classified according to Sanna et al. [1993] based on the topographic location and the extent of the cholesteatoma on the CT scan. Depending on the class of the PBC, the management was planned. Between 1978 and 1985 the surgical approach evolved from open techniques to closed techniques adopted in most of the patients since then. Facial nerve function was graded preoperatively, immediately postoperatively and at 1 year postoperatively according to the House-Brackmann grading system.

Minimum follow-up was 1 year in 126 patients. Subsequently the patients underwent an annual radiological follow-up (CT scan and MRI) for detection of any recurrence for at least 5 years.

Table 1. Sanna classification of PBC

Class	Location	Spread
Class I: supralabyrinthine (fig. 1a, b)	geniculate ganglion of facial nerve	anterior: horizontal part of ICA posterior: posterior bony labyrinth medial: IAC, petrous apex inferior: basal turn of the cochlea
Class II: infralabyrinthine (fig. 2a, b)	hypotympanic and infralabyrinthine cells	anterior: ICA vertical part, petrous apex, clivus posterior: dura of the posterior cranial fossa and sigmoid sinus medial: IAC, lower clivus, occipital condyle inferior: jugular bulb, lower cranial nerves
Class III: infralabyrinthine-apical (fig. 3a, b)	infralabyrinthine compartment, ICA reaching up to petrous apex	anterior: ICA vertical ± horizontal parts posterior: posterior fossa through the retrofacial air cells medial: petrous apex, clivus, sphenoid sinus, rhinopharynx inferior: jugular bulb, lower cranial nerves
Class IV: massive (fig. 4a, b)	entire otic capsule	anterior: ICA vertical ± horizontal parts posterior: posterior fossa dura and IAC medial: petrous apex, superior and mid clivus, sphenoid sinus inferior: infralabyrinthine compartment
Class V: apical (fig. 5a, b)	petrous apex	anterior: Meckel's cave area and may involve the V nerve posterior: IAC and posterior cranial fossa medial: superior or mid clivus, sphenoid sinus inferior: infralabyrinthine compartment
Subclass	**Features**	
Clivus (C) (fig. 6a, b, c, d)	superior and mid clival extensions are seen from massive, infralabyrinthine-apical and apical PBC whereas the lower clival involvement is a feature of infralabyrinthine-apical PBC	
Sphenoid sinus (S) (fig. 7a, b)	sphenoid sinus involvement is seen from anteromedial extensions of massive, infralabyrinthine-apical and apical PBC; it is a rare extension	
Rhinopharynx (R) (fig. 8a, b, c)	it 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 rhinopharynx	

IAC = Internal auditory canal; ICA = internal carotid artery.

Table 2. Classification of PBC versus pathologic type

Class/pathology	Congenital	Acquired	Recurrent/iatrogenic	Total
Supralabyrinthine	6	41	18	64 (49%)
Infralabyrinthine	2	6	1	9 (7%)
Infralabyrinthine-apical	1	4	2	7 (5%)
Massive	5	26	17	48 (38%)
Apical	1	0	0	1 (1%)
Total	15 (12%)	76 (58%)	38 (30%)	129 (100%)

Table 3. Surgical approach and class of PBC

Surgical approach	SL	IL	ILA	M	A	Total
MTCA	12	2	1	19	0	34
MTCB	0	0	0	2	1	3
ETLA	17	0	2	14	0	33
TO	19	0	3	7	0	29
SP	11	5	0	6	0	22
MF + TM	4	0	0	0	0	4
TM + RL	1	2	0	0	0	3
IFTB	0	0	1	0	0	1
Total	64	9	7	48	1	129

SL = Supralabyrinthine; IL = infralabyrinthine; ILA = infralabyrinthine-apical; M = massive; A = apical; MTCA = modified transcochlear approach type A; MTCB = modified transcochlear approach type B; ETLA = enlarged translabyrinthine approach; TO = transotic approach; SP = subtotal petrosectomy; MF + TM = middle fossa + transmastoid; TM + RL = transmastoid + retrolabyrinthine; IFTB = infratemporal fossa approach type B.

Table 4. Preoperative facial nerve (FN) function for various classes of PBC according to HB grading system

FN grading	SL	IL	ILA	M	A	Cases, n
Grade I	32	7	6	15	1	61 (46%)
Grade II	5	0	0	8	0	13 (10%)
Grade III	10	1	0	6	0	17 (13%)
Grade IV	4	1	0	2	0	7 (6%)
Grade V	2	0	0	0	0	2 (2%)
Grade VI	11	0	1	17	0	29 (23%)
Total	64	9	7	48	1	129 (100%)

SL = Supralabyrinthine; IL = infralabyrinthine; ILA = infralabyrinthine-apical; M = massive; A = apical.

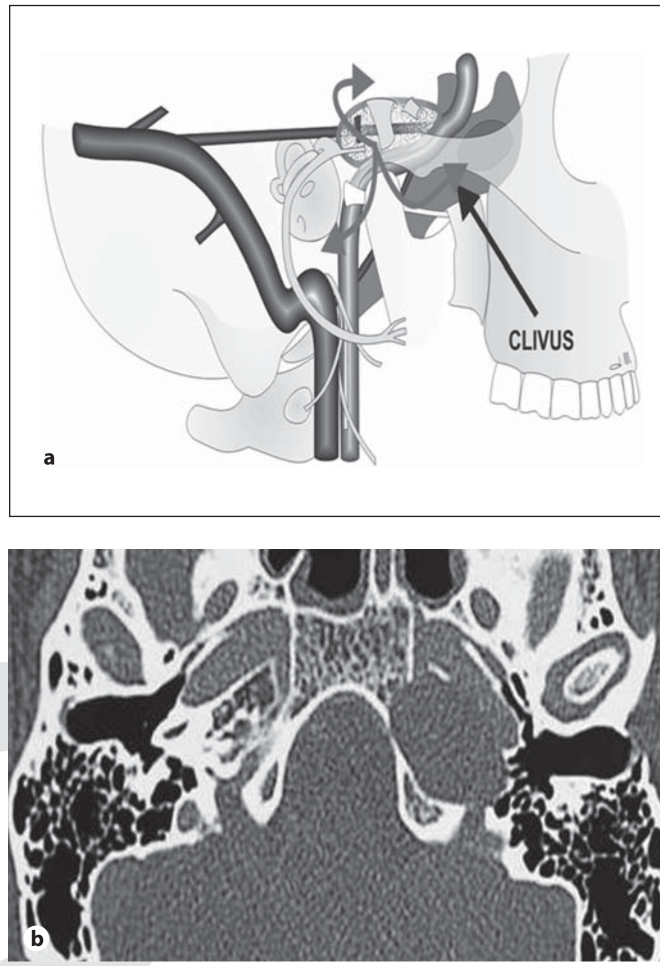


Fig. 5. a Diagrammatic representation of an apical PBC as viewed from the lateral aspect. The dotted area represents the site of the lesion and the arrows represent the route of spread. Directions for the route of spread are given in table 1. **b** An axial HRCT image of the temporal bone showing an apical PBC. There is a smooth osteolytic lesion occupying the petrous apex reaching up to the clivus, lying medial to the horizontal portion of the internal carotid artery. Note that the bony canal of the horizontal portion of the internal carotid artery is eroded.

Results

The incidence of PBC was 2.9% (129/4500) in our series out of all the patients diagnosed and treated for cholesteatomas.

The age range in our series was from 9 to 83 years with a mean age of 45 years. Hearing loss was the commonest symptom in 69% followed by facial palsy in 53% of patients.

Table 2 illustrates PBC classes versus pathological type. Thirty percent of the cases were recurrent or iatrogenic in origin. The supralabyrinthine type of PBC was the commonest type, followed by massive, infralabyrinthine, infralabyrinthine-apical and apical.

Infratemporal fossa type B approach was performed for 1 case with extension to the sphenoid sinus. Modified transcochlear approach type B was used in 3 cases with extension to the clivus and rhinopharynx, respectively. The different surgical approaches used are summarized in table 3.

The commonest sites for the involvement of the facial nerve in the present series were the tympanic part (94%),

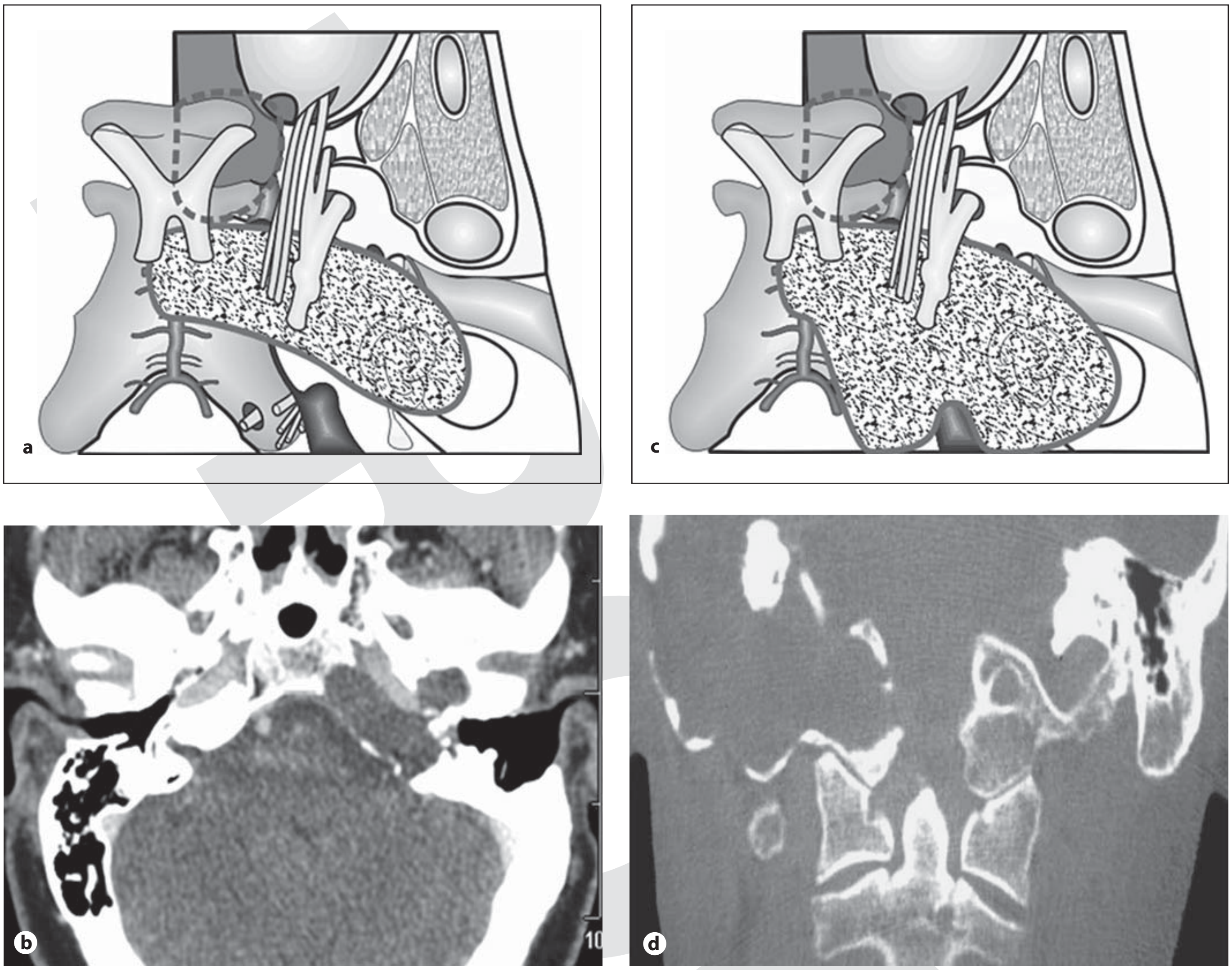


Fig. 6. a Diagrammatic representation of massive PBC with clival extension as seen from the superior aspect (see text). The cholesteatoma involves the petrous apex and superior and mid clivus. **b** Axial CT scan showing clival extension of a massive PBC. Note the radical mastoid cavity; this patient had previous surgery 15 years prior to presentation. **c** Diagrammatic representation of extension of massive PBC to clivus and occipital condyle as viewed from the superior aspect. **d** Coronal CT scan showing extension of massive PBC to clivus and occipital condyle.

geniculate ganglion (84%) and the labyrinthine portion (69%). The facial nerve was involved in multiple segments in most of the patients. Table 4 presents the preoperative distribution of facial nerve function in various types of PBC. Facial nerve decompression was performed in 55 cases, which was the commonest form of management of the facial nerve. It had to be frequently performed even in cases with grade I preoperative facial nerve function. 6/29 patients with preoperative grade VI palsy did not undergo any treatment for the facial nerve because of the long duration of facial palsy of >3 years. Sural nerve grafting was the next common form of treatment for facial nerve lesions (22 cases). End-to-end anastomosis was done in 7 cases and facial-hypoglossal anastomosis was performed in 2 cases. A total number of 10/61 cases with grade I preoperative function developed various grades of facial paresis. Out of these 10 cases 5 had grade II and 4 cases had grade III facial nerve function, and 1 patient with an apical PBC had postoperative grade VI facial palsy at 1-year follow-up. Later this patient was operated for facial-hypo-

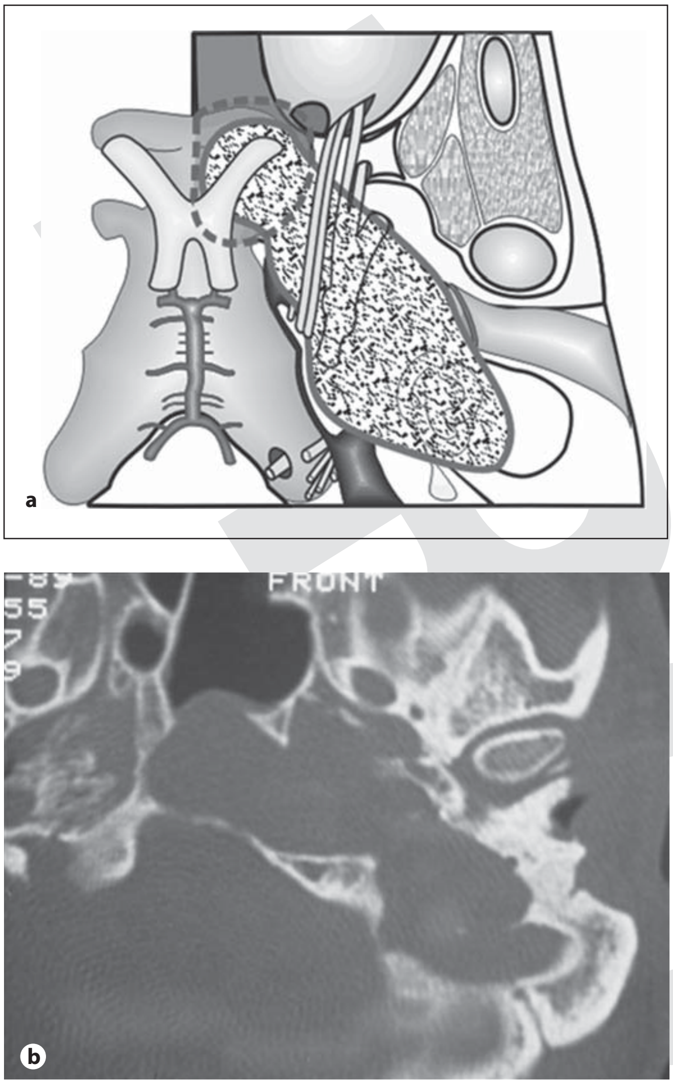


Fig. 7. a Diagrammatic representation of an infralabyrinthine-apical PBC with extension to the sphenoid sinus as viewed from the superior aspect. **b** Axial CT scan showing an infralabyrinthine-apical PBC with extension to the sphenoid sinus.

Table 5. Postoperative facial nerve (FN) grades in different classes of PBC according to HB grading system

FN grading	SL	IL	ILA	M	A	Cases, n
Grade I	25	7	4	15	0	51 (41%)
Grade II	8	0	2	5	0	15 (11%)
Grade III	17	0	0	9	0	26 (21%)
Grade IV	3	0	0	5	0	8 (6%)
Grade V	5	0	0	1	0	6 (5%)
Grade VI	4	2	1	12	1	20 (16%)
Total	62	9	7	47	1	126 (100%)

Two patients were lost to follow-up including the 1 case of bilateral PBC. SL = Supralabyrinthine; IL = infralabyrinthine; ILA = infralabyrinthine-apical; M = massive; A = apical.

Table 6. Postoperative complications

Postoperative complications	Cases	
	n	%
Facial palsy	10/61	16
Cavity infection	2/129	2
Meningeal herniation	1/129	<1
CSF leak	1[a]/129	<1
Brain abscess	1[a]/129	<1

Facial palsy was noted as a complication when a grade I preoperative facial nerve function deteriorated postoperatively.
[a] Same patient.

glossal anastomosis, following which facial nerve function improved to grade IV (table 5). Out of 128 patients 1 patient died of carcinoma of the colon, and 1 case of bilateral PBC was lost to follow-up.

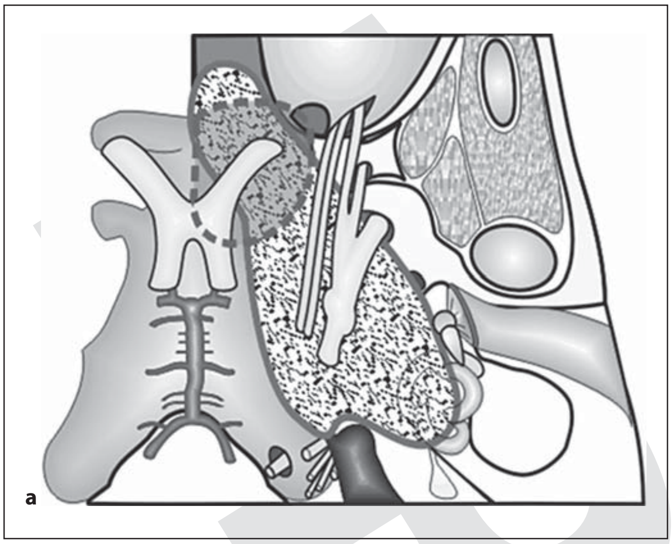
Intraoperative complications included cerebrospinal fluid (CSF) leak in 24 cases (19%), accidental opening up of the jugular bulb and sigmoid sinus in 1 patient each (<1%). There were no perioperative deaths.

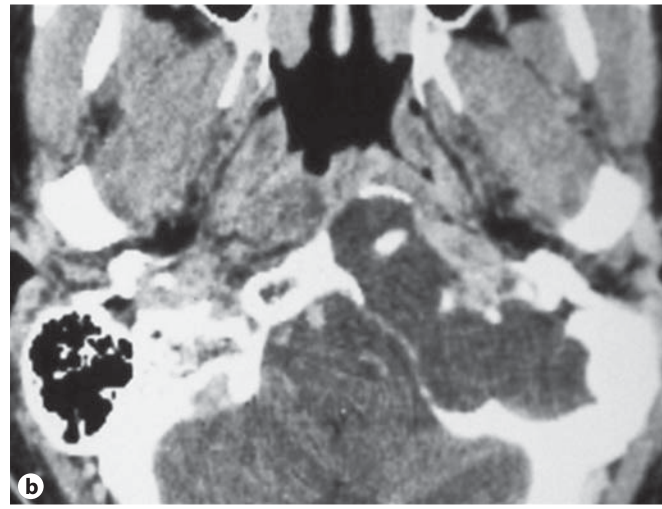
Postoperatively, 82% of the patients had a dead ear (106/129 patients), which was a sequela to the surgical approaches chosen for the radical removal of the disease. Postoperative complications are listed in table 6. Facial palsy was noted in 10/61 (18%) cases who had been classified as grade I preoperatively.

Out of the 129 cases 126 had a minimum follow-up of 1 year. HRCT of the temporal bone and MRI with fat suppression were performed on follow-up. Recurrence was observed in 5 cases (table 7).

Review of the Literature

A review of the literature on PBC was done using a PubMed database search. Inclusion criteria comprised case series describing cholesteatomas involving the petrous bone that had more than 10 cases. The following





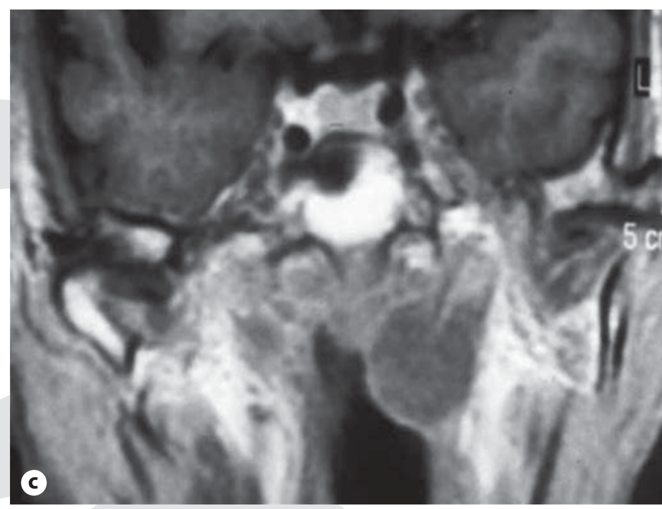


Fig. 8. a Diagrammatic representation of a massive PBC with extension to the rhinopharynx. The cholesteatoma passes beneath the sphenoid sinus to reach the rhinopharynx (see text). **b** Axial CT scan showing a massive PBC with extension to the rhinopharynx. **c** Coronal T_1-weighted MRI showing extension of PBC into the rhinopharynx.

Table 7. Recurrences and their management

Class	Primary surgery	Revision surgery
Supralabyrinthine	RPME	SP with obliteration
Infralabyrinthine	SP	SP with obliteration
Infralabyrinthine	SP	SP with obliteration
Supralabyrinthine	MF + TM	MTCA
Massive	SP	MTCA

RPME = Radical petromastoid exenteration; SP = subtotal petrosectomy; MF + TM = middle fossa + transmastoid; MTCA = modified transcochlear approach type A.

aspects were analyzed: nomenclature, classification, management protocols, preoperative and postoperative facial nerve function, surgical approaches used, hearing preservation, complications and recurrences. There were seven case series in the English literature that met our inclusion criteria, with a total of 221 cases (table 8). Including our own series of 129 cases, the total number of cases was 350. Bartels [1991] presented a review of 80 cases that were mentioned in the literature till 1991; we tried to compare the review of this case series and the one done by him. Part of this series has already been published by the senior author [Omran et al., 2006; Sanna et al., 1993]; we included those cases in our series.

Hearing loss was present in almost 70% of these patients on presentation. Facial palsy was the presenting

Table 8. Review of the literature

Series and year	Classi-fication	Cases n	Preoperative		Recur-rences	Complications (number of patients)
			hearing loss	facial palsy		
Charachon et al. [1989]	not given	42	35	15	6	not given
Hawthorne and Fisch [1989]	Fisch	33	NA	22	none	meningitis (2 patients) cardiac infarct (1 patient)
Pyle and Wiet [1991]	Schuknecht's anatomical	10	8	5	3	CSF leak (1 patient) wound infection (1 patient) recurrent cavity infection (4 patients)
Yanagihara et al. [1992]	not given	16	11	16	none	not given
Axon et al. [1999]	Sanna	25	16	14	5	chronic discharge (3/6 patients) stenosis of the meatus (2 patients)
Magliulo [2007]	Sanna	52	36	28	4	not given
Moffat et al. [2008]	modified Sanna	43	26	22	2	CSF leak (4 patients)
Present series 2009[a]	Sanna	129	82	68	5	meningeal herniation (1 patient) cavity infection (2 patients) CSF leak and brain abscess (same patient)

Sorted according to publication date, the table presents the various series along with references. Classification, number of cases, along with preoperative hearing loss and facial paralysis are given as well as recurrences and complications.
[a] The present series includes the case series published by the senior author [Sanna et al., 1993; Omran et al., 2006].

symptom in 54% of the patients in all the series. In the review by Bartels et al. [1991], hearing loss was reported in 55% of patients and facial palsy was seen in 46% of the patients.

The transtemporal middle fossa approach was only used by few authors [Hawthorne and Fisch, 1989; Moffat et al., 2008] for hearing preservation in supralabyrinthine lesions. In our experience, with this approach hearing could not be preserved with a fistula in the basal turn of the cochlea [Omran et al., 2006]. Hawthorne and Fisch [1989] in their series had a similar experience in such cases. Therefore these patients should have a preoperative CT scan evaluation to identify a fistula in the basal turn of the cochlea. In the absence of a fistula this approach should be performed, otherwise hearing preservation is not possible. Detailed statistical analysis of the postoperative hearing status is not mentioned in most of these series; hence it could not be evaluated.

Hawthorne and Fisch [1989] observed that the facial nerve was involved in almost all of their cases of PBC. Various theories have been put forward regarding the pathogenesis of facial palsy in PBC, such as the theory of perigeniculate ischemia [Axon et al., 1999] and the theory of facial nerve impairment caused by direct compression or by raised intrafunicular pressure or by the direct toxic effects of bacteria and enzymes [Atlas et al., 1992]. We believe that it is the combination of direct compression and repeated inflammation leading to fibrous tissue interposition which results in facial nerve dysfunction, depending upon the site involved [Sanna et al., 2006].

Preoperative facial nerve status and postoperative results were not published in four of these series [Charachon et al., 1989; Hawthorne and Fisch, 1989; Pyle and Wiet, 1991; Yanagihara et al., 1992]. Available cumulative statistics from the remaining series revealed that 89% of the patients who had preoperative grade I facial nerve function maintained it in the postoperative period. Omran et al. [2006] observed that cases with onset of facial nerve dysfunction <12 months preoperatively had a better outcome than patients who presented later. Axon et al. [1999] introduced the theory of geniculate ganglion ischemia; they reported complete facial paralysis with bony erosion around the geniculate ganglion and thought that the ischemia was the main factor. They resected the ischemic nerve segment in 6 patients and performed an end-to-end anastomosis over the posterior fossa dura. But only 2 of these patients showed improvement. The other 4 underwent facial-hypoglossal anastomosis. The duration of paralysis was the important cause in all these pa-

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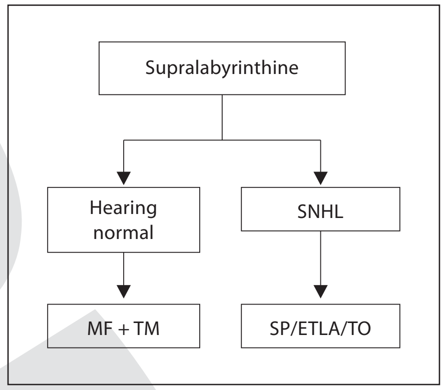

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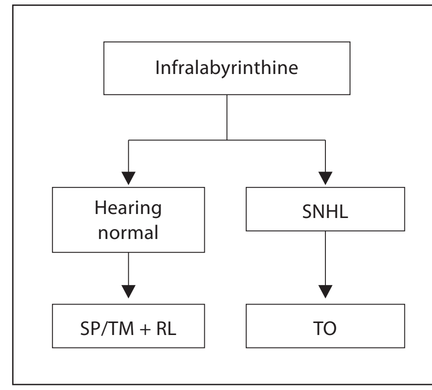

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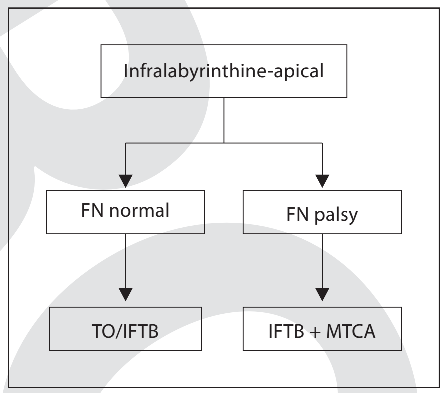

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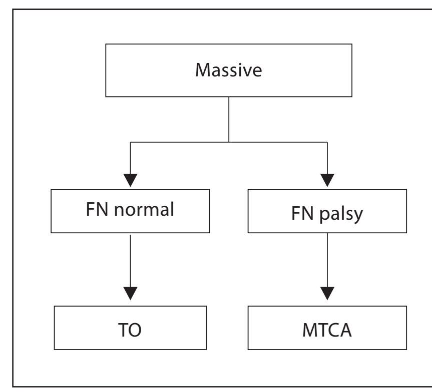

Fig. 9. Algorithm for the management of class I supralabyrinthine PBC. SNHL = Sensorineural hearing loss; FN = facial nerve; SP = subtotal petrosectomy; MTCA = modified transcochlear approach type A; MF + TM = middle fossa + transmastoid approach; ETLA = enlarged translabyrinthine approach; TO = transotic approach; RL + TM = retrolabyrinthine + transmastoid approach; IFTB = infratemporal fossa approach type B; MTCB = modified transcochlear approach type B.
Fig. 10. Algorithm for the management of class II infralabyrinthine PBC.
Fig. 11. Algorithm for the management of class III infralabyrinthine-apical PBC.
Fig. 12. Algorithm for the management of class IV massive PBC.

tients; all these patients had paralysis of >12 months [Omran et al., 2006].

The cumulative incidence of intracranial complications was 2%. It included 6 patients with CSF leak, 2 cases of meningitis and 1 case of brain abscess. Bartels [1991] in his review reported a 6% rate of intracranial infections. Meningitis was reported in 2 cases by Hawthorne and Fisch [1989], and 1 of their patients died of otogenic meningitis in an open cavity. Pyle and Wiet [1991] published a comparison of an open technique and cavity obliteration. Cavity obliteration had the advantage of obviating the need for postoperative draining and of protecting vital intracranial structures (internal carotid artery, dura mater, facial nerve, jugular bulb, etc.) that could be exposed.

There was no perioperative or immediately postoperative mortality as compared to the 3% death rate in the review by Bartels [1991]. There is no mention of cholesteatomas with extension into the petrous bone, particularly into the sphenoid sinus and the rhinopharynx, and their management. No definite management protocols are mentioned.

Our series of 129 PBC cases (as far as we know) is the largest series published until now in the English literature. The present article focuses on the nomenclature and presents management guidelines based on the classification of PBC.

Discussion

The classification of PBC is of paramount importance as it gives information regarding the anatomical position and the extent of the disease. The subclassification proposed by us in this article aims at preoperatively diagnosing the extension of PBC beyond the temporal bone (usually clivus, sphenoid sinus and rhinopharynx). This helps to plan the surgical approach, which is important to clear the disease from these areas.

The choice of surgical approach has evolved from radical petromastoid exenteration with marsupialization of the cavity to closed and obliterative techniques following complete eradication. Decision making is the crucial aspect of surgical management. It depends on several factors, the most significant of which are the extent of the

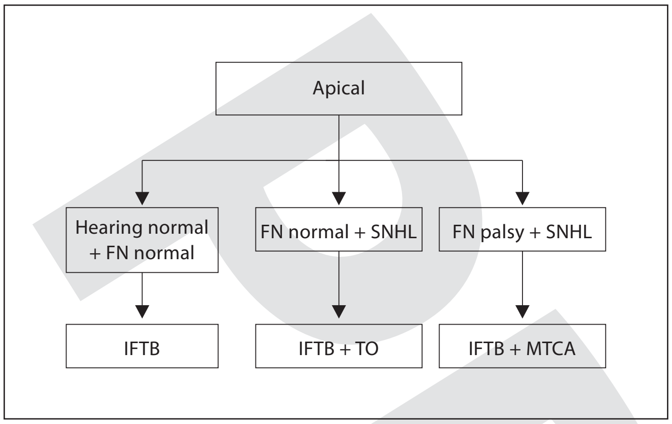

Fig. 13. Algorithm for the management of class V apical PBC.

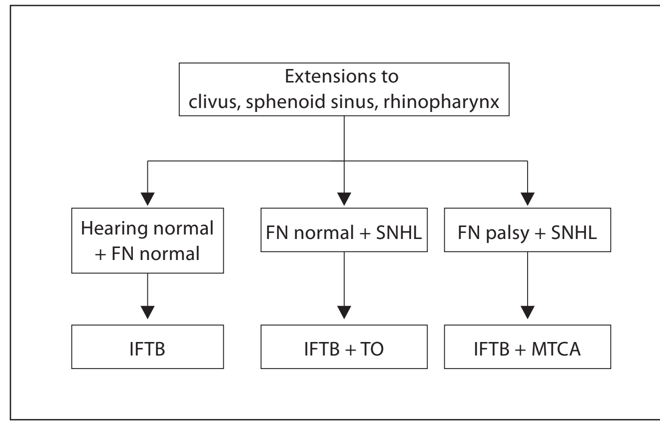

Fig. 14. Algorithm for the management of the extensions of PBC to clivus, sphenoid sinus and rhinopharynx.

disease and preoperative facial nerve function. The approach is chosen depending upon the type of PBC and its extent, which should be determined according to the CT scan and MRI findings (fig. 9–14, algorithms 1–6).

Algorithms of PBC Management

The main factors to be taken into consideration while treating these lesions are as follows: (1) complete eradication of the disease, (2) preservation of facial nerve function, (3) prevention of CSF leak and meningitis, (4) cavity obliteration, and (5) hearing preservation whenever feasible.

In supralabyrinthine PBC (fig. 9), if hearing is normal without any evidence of a fistula in the basal turn of the cochlea, we prefer a middle fossa approach which may be combined with a transmastoid approach depending on the extension of the disease. In the presence of sensorineural hearing loss or CT evidence of a fistula in the basal turn of the cochlea we prefer a radical approach (subtotal petrosectomy/enlarged translabyrinthine approach/transotic approach) with cavity obliteration (fig. 15). In infralabyrinthine PBC (fig. 10) bone conduction can be preserved with subtotal petrosectomy and blind sac closure of the external auditory canal with cavity obliteration. In very rare cases a transmastoid approach can be combined with a retrolabyrinthine approach to preserve bone conduction.

Hearing preservation is usually not possible in infralabyrinthine-apical (fig. 11) and massive PBC (fig. 12), hence we use a transotic approach or a modified transcochlear approach type A depending on preoperative facial nerve function. Modified transcochlear approaches provide excellent access to the petrous apex, clivus, sphenoid sinus and rhinopharynx depending upon the type used [Sanna et al., 1993]. The posterior rerouting of the facial nerve carries the disadvantage of postoperative facial paresis, which is attributed to the loss of blood supply from the deep petrosal artery near the geniculate ganglion; this can recover up to grade III of the HB grading system. Therefore we prefer a transotic approach when facial nerve function is normal preoperatively and a modified transcochlear approach when the patient presents with facial paresis. Facial nerve involvement carries the maximum risk of morbidity as it not only has functional impairment but also a psychological effect.

Whenever the cholesteatoma involves the apical portion (fig. 13) of the temporal bone or when it extends further to the clivus, sphenoid sinus or rhinopharynx (fig. 14), an infratemporal fossa approach type B is incorporated into the transotic approach or modified transcochlear approach type A depending upon the preoperative status of the facial nerve. In selected cases with preoperatively normal hearing, if the otic capsule is not involved, bone conduction can be preserved with infratemporal fossa approach type B.

Problems and Solutions

Hearing Preservation

Hearing preservation would be important in exceptional cases, i.e. bilateral PBCs or in a PBC of the only hearing ear. However, in the era of BAHA, cochlear implants and Vibrant Soundbridge even these cases can be treated successfully with hearing rehabilitation. Bartels [1991] pointed out the possibility of a cochlear implant in

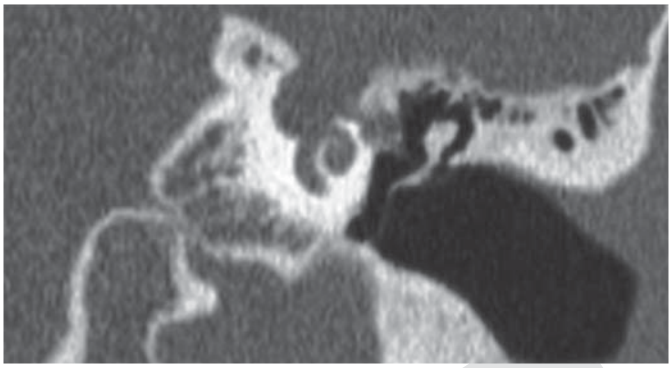

Fig. 15. Coronal HRCT suggestive of a fistula in the superior basal turn of the cochlea. In this case it is not possible to preserve hearing.

ears with an accessible cochlear lumen even after labyrinthectomy and hence the cochlea should not be destroyed beyond what is essential for the clearance of the disease. Another option is a BAHA if bone conduction is preserved in the ipsilateral ear in cases of PBC in the only hearing ear. Linder et al. [2008] published results on Vibrant Soundbridge in patients having undergone subtotal petrosectomy; this promises to be a good option for selective patients with mixed hearing loss.

Facial Nerve

Facial nerve lesions may vary from simple erosion of the fallopian canal to total interruption of the nerve with fibrous tissue interposition. Management depends on three principal factors: preoperative status, degree of facial nerve involvement and the extent of the lesion.

Decompression. If there is a compression of the nerve with preserved anatomical integrity, then decompression of the nerve should be performed.

Rerouting. When the lesion is present medial to the facial nerve and complete control over the lesion is hampered due to the position of the nerve, then rerouting of the facial nerve is undertaken, which could be partial or complete.

End-to-End Anastomosis. If there is a discontinuity of the nerve or a fibrous tissue interposition, the affected segment should be excised and a tension-free end-to-end anastomosis should be performed.

Nerve Grafting. Whenever the nerve segment lost is long and a tension-free end-to-end anastomosis is not possible, continuity of the nerve can be restored using a nerve graft. We prefer a sural nerve for grafting purposes.

Facial-Hypoglossal Anastomosis. In patients with long duration of facial palsy (>12 months) facial-hypoglossal anastomosis is indicated [Falcioni et al., 2003].

In cases where the preoperative facial nerve function is good, a surgeon can be optimistic about a favorable facial nerve outcome depending upon the extension of the pathology.

Internal Carotid Artery

The PBC may involve the internal carotid artery in the vertical and/or the horizontal parts. In this situation a complete control over the artery is important prior to attempting its removal. A modified transcochlear approach type A/transotic approach is used for involvement of the vessel in the vertical part, whereas an infratemporal fossa type B/modified transcochlear approach type B is used for involvement of vertical and horizontal parts. In case of a lesion extending into the petrous apex, clivus, sphenoid sinus and into the rhinopharynx it is important to perform a complete control of the internal carotid artery in order to mobilize the artery if necessary. PBC are less aggressive in terms of arterial involvement [Charachon et al., 1989; Hawthorne and Fisch, 1989; Bartels, 1991; Magliulo, 2007] and are easier to dissect as compared to other tumors (i.e. glomus jugulare). The internal carotid artery has a thick adventitia which resents the dissection of the matrix but it requires extreme caution and surgical skill to clear it.

Sigmoid Sinus and Jugular Bulb

The involvement of the sigmoid sinus and the jugular bulb presents a problem in matrix removal due to the thin wall and the fragility of these structures. In such cases it is important to control the internal jugular vein in the neck prior to the dissection of the matrix. The ligation of the internal jugular vein in the neck and the sigmoid sinus packing (extraluminal and intraluminal) enables removal of the lateral wall of the dome of the jugular bulb and the sigmoid sinus to clear the matrix in the rare cases of accidental opening of the bulb. This maneuver also helps in preserving the IX, X, and XI cranial nerves. During this procedure bleeding from the inferior petrosal sinus is controlled with Surgicel packing. In cases where its involvement is suspected it is always advisable to ensure preoperatively the patency of the contralateral cerebral vein.

Dura

The matrix is often adherent to the dura of the middle and posterior fossa. Bipolar coagulation of all the suspected portions of the dura mater can be performed to destroy all the possible remnants of the matrix. We have been using bipolar coagulation in all cases to devitalize the epithelium, and also other authors [Axon et al., 1999] who have used the same technique agree with us. Bipolarizing large areas of the dura does not lead to any dural necrosis if carefully performed. Long-term follow-up has shown that this maneuver is safe and adequate for complete control. There is a risk of opening the dura while removing the adherent matrix causing an intraoperative CSF leak.

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 cavity obliteration with fat. CSF leak from the internal auditory canal can be treated by adopting a translabyrinthine approach.

Examination of Hidden Areas

Once the removal of the disease has been achieved it is useful to carry out an endoscopic examination of the cavity with a 30-degree rigid endoscope to visualize the hidden areas that might not be accessible to the microscope. In some cases epithelium missed by the conventional technique can be found on endoscopic examination [Axon et al., 1999]. In our experience, if the approach is correct this technique is rarely required.

Residual Lesions or Recurrences

After complete eradication of the disease, it is mandatory to obliterate the cavities with autologous abdominal fat. The major disadvantage of cavity obliteration is that the recurrence cannot be directly visualized and detected. Therefore it is mandatory to follow up these patients radiologically. We perform a high-resolution CT scan and a cerebral MRI (T_1, T_2-weighted images with fat suppression) and gadolinium enhancement every year for at least 5 years.

Recurrent/iatrogenic cholesteatomas can be avoided if the surgeries for middle ear and mastoid cholesteatomas are meticulously performed. To achieve complete clearance we advocate that in the canal wall up (CWU) technique for cholesteatoma the operation has to be staged: second stage surgery is mandatory (after 1 year) to avoid the risk of recurrence and future development of PBC, especially when we are dealing with children or young patients, whereas in the canal wall down (CWD) technique it is mandatory to perform a good mastoidectomy with saucerized margins, adequately exposed anterior and posterior epitympanum and with the facial ridge lowered till the level of the facial nerve. The cholesteatoma matrix should be accurately removed from the middle ear cleft after it has been adequately exposed to prevent any recurrences. Following these principles a second stage is performed in cases in which cholesteatomas occupy the oval window or the anterior epitympanum or infiltrate deeply into the hypotympanic cells. In cases with small cholesteatomas an alternative to the second stage can be a CT scan of the middle ear and temporal bone. In all cases of middle ear cholesteatomas treated either with the CWD or CWU technique, long-term follow-up is strongly recommended.

Conclusions

- The management of PBC requires thorough pretreatment evaluation, planning, execution and a meticulous radiological follow-up.
- Classification is fundamental in order to apply the appropriate surgical approach.
- The facial nerve requires special consideration as it gets involved in almost all the cases and is a cornerstone of management.
- Radical removal takes priority over hearing preservation even in only hearing ears thanks to the possibility of hearing rehabilitation (CI, BAHA and Soundbridge).
- Cavity obliteration holds the key for a problem-free cavity.
- Regular follow-up is mandatory.

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References

Atlas M, Moffat D, Hardy D: Petrous apex cholesteatoma: diagnostic and treatment dilemmas. Laryngoscope 1992;102:1363–1368.

Axon P, Fergie N, Saeed S, Temple RH, Ramsden R: Petrosal cholesteatoma: management considerations for minimizing morbidity. Am J Otol 1999;20:505–510.

Bacciu A, Falcioni M, Pasanisi E, Di Lella F, Lauda L, Flanagan S, Sanna M: Intracranial facial nerve grafting after removal of vestibular schwannoma. Am J Otolaryngol 2009;30:83–88.

Bartels LJ: Facial nerve and medially invasive petrous bone cholesteatomas. Ann Otol Rhinol Laryngol 1991;100:308–316.
Charachon R, Martin Ch, Gratacap B, Perron X: Temporal bone cholesteatoma; in Fisch U, Valavanis A, Yasargil MG (eds): Neurological Surgery of the Ear and the Skull Base. Berkeley, Kugler, 1989, pp 21–28.
De Donato G, Caylan R, Falconi M, Landolfi M, Titliz A, Russo A, Taibah A, Sanna M: Facial nerve management and results in petrous bone cholesteatomas surgery; in Nakano Y (ed): Cholesteatoma and Mastoid Surgery. Proc 4th Int Conf, Niigata, Sept 1992. Amsterdam, Kugler, 1993, pp 500–506.
Falcioni M, Taibah A, Russo A, Piccirillo E, Sanna M: Facial nerve grafting. Otol Neurotol 2003;24:486–489.
Glasscock ME 3rd: Middle fossa approach to the temporal bone: an otologic frontier. Arch Otolaryngol 1969;90:15–27.
Hawthorne M, Fisch U: The surgical management of supralabyrinthine and infralabyrinthine-apical cholesteatoma of the temporal bone; in Fisch U, Valavanis A, Yasargil MG (eds): Neurological Surgery of the Ear and the Skull Base. Berkeley, Kugler, 1989, pp 11–19.
House W, Histelberger W: The transcochlear approach to the skull base. Arch Otolaryngol 1976;102:334–342.
Jackler R, Parker D: Radiographic differential diagnosis of petrous apex lesions. Am J Otol 1992;13:561–574.
Linder T, Schlegel C, Demin N, van der Westhuizen S: Active middle ear implant in patients undergoing subtotal petrosectomy: new application for Vibrant Soundbridge device and its implication for lateral cranium base surgery. Otol Neurotol 2008;30:41–47.
Magliulo G: Petrous bone cholesteatoma: clinical longitudinal study. Eur Arch Otorhinolaryngol 2007;264:115–120.
Moffat D, Jones S, Smith W: Petrous temporal bone cholesteatoma: a new classification and long-term surgical outcomes. Skull Base 2008;18:107–115.
Omran A, De Donato G, Piccirillo E, Leone O, Sanna M: Petrous bone cholesteatoma: management and outcomes. Laryngoscope 2006;116:619–626.
Pyle GM, Wiet R: Petrous apex cholesteatoma: exteriorization versus subtotal petrosectomy with obliteration. Skull Base Surg 1991;1:97–105.
Rosenberg R, Hammerschlag P, Cohen N, Bergeron RT, Reede D: Cholesteatoma versus cholesterol granuloma of the petrous apex. Otolaryngol Head Neck Surg 1986;94:322–327.
Sanna M, Khrais T, Mancini F, Russo A, Taibah A: The Facial Nerve in Temporal Bone and Lateral Skull Base Microsurgery. Stuttgart, Thieme, 2006.
Sanna M, Mazzioni A, Saleh E, Taibah AK, Russo A: Lateral approaches to the median skull base through the petrous bone: the system of the modified transcochlear approach. J Layngol Otol 1994;108:1036–1044.
Sanna M, Saleh E, Khrais T, Mancini F, Piazza P, Russo A, Taibah A: Atlas of Microsurgery of the Lateral Skull Base, ed 2. Stuttgart, Thieme, 2008.
Sanna M, Zini C, Gamoletti R, Frau N, Taibah A, Russo A, Pasanici E: Petrous bone cholesteatoma. Skull Base Surg 1993;3:201–213.
Selesnick S, Abraham M, Carew JF: Rerouting of the intratemporal facial nerve: an analysis of the literature. Am J Otol 1996;17:793–805.
Yanagihara N, Nakamura K, Hatakeyama T: Surgical management of petrous apex cholesteatoma: a therapeutic scheme. Skull Base Surg 1992;2:22–27.