The Laryngoscope
Lippincott Williams & Wilkins, Inc.

Petrous Bone Cholesteatoma: Management and Outcomes

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Objective: **Cholesteatoma of the petrous bone is a rare pathology that grows slowly and is often asymptomatic. High clinical suspect is recommended for its diagnosis. With the advancement of imaging, clinical diagnosis is confirmed and follow up is assured. The present study is presented to state our experience in management of petrous bone cholesteatoma (PBC) and an overview of the outcome results.** ***Methods:*** **This study was a retrospective case review conducted at a private quaternary referral center. From January 1984 to October 2004, 93 of 2,739 patients with chronic otitis media with cholesteatoma were treated surgically for PBC with varying location types.** ***Results:*** **The mean age in this study was 46 years. Ninety-four percent of patients presented with hearing loss. Facial paralysis constituted 55% of the presented manifestations and grade VI palsy was the most common. A massive type of PBC was found to affect 45% of patients and it was mainly the acquired pathology. The surgical approaches varied according to the class type of PBC. Postoperative complications were minimal, and follow up was carried out after 1 year from the surgery date.** ***Conclusion:*** **Petrous bone cholesteatoma presents difficulties in its diagnosis and treatment. Adequate history-taking, high clinical suspicious, with the advancement of imaging techniques made its diagnosis more feasible. Improvement of the lateral skull base approaches rendered possible safe, adequate, and complete removal of the pathology.** ***Key Words:*** **Petrous bone cholesteatoma, facial nerve palsy, facial nerve reconstruction, internal carotid artery.**

Laryngoscope, **116:619–626, 2006**

INTRODUCTION

Petrous bone cholesteatoma (PBC) is used to define an epidermoid cyst of the petrous portion of the temporal bone. This pathology is generally a slow-growing lesion that may remain asymptomatic for years that produces sometimes misleading symptoms.[1] Congenital and acquired etiologies are the main attributers in the pathology of cholesteatoma of the petrous bone. Congenital cholesteatoma is suggested to arise from epithelial cell rest within the temporal bone.[2] Acquired type is the result of medially invasive cholesteatoma of the tympanomastoid region.[3]

According to Sanna's classification, PBC is subdivided into five classes in relation to the labyrinth: supralabyrinthine, infralabyrinthine, massive labyrinthine, infralabyrinthine apical, and apical cholesteatoma to facilitate surgical planning and management. With improvement of lateral skull base and transpetrous approaches and recent advances in radiologic imaging, the attitude in management of PBC has been changed with a high percentage of safety to assure radical removal of the pathology. The current study aim is to present our management experience of 93 cases.

Classification of Petrous Bone Cholesteatoma

We have classified PBC into five classes according to the relationship of the lesion to the labyrinth. Class I: Supralabyrinthine (Fig 1A, B); Class II: Infralabyrinthine (Fig 2A, B); Class III: Massive (Fig 3A, B); Class IV: Infralabyrinthine Apical (Fig 4A, B) with or without sphenoid sinus extension (Fig 4C, D) or with clival extension (Fig 4E, F); and Class V: Apical (Fig 5A, B).

In addition to its functional importance, the labyrinthine block was chosen as the reference base of this classification because of its central position within the petrous bone.

Supralabyrinthine cholesteatoma is characteristically congenital or may arise from deep extension of acquired epitympanic cholesteatoma. It involves the anterior epitympanum and extends medially toward the internal auditory canal and anteriorly toward the internal carotid artery and the cochlea. Rarely, it extends posteriorly to involve the posterior labyrinth and retrolabyrinthine air cells. This type of cholesteatoma is usually centered on the geniculate ganglion (Fig 1A, B).

Infralabyrinthine cholesteatoma is an acquired type that arises in the hypotympanic and the infralabyrinthine cells with anterior extension toward the petrous apex, the

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Editor's Note: This Manuscript was accepted for publication January 20, 2006.

This study was supported by a grant from Associazione Studio Aggiornamento Basicranio.

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DOI: 10.1097/01.mlg.0000208367.03963.ca

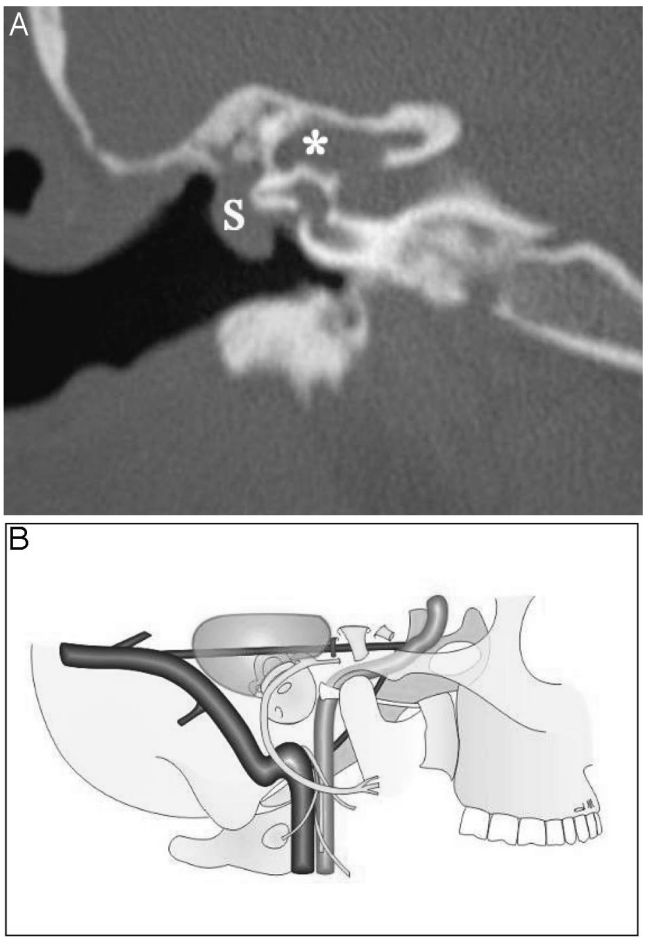

Fig. 1. (A) Supralabyrinthine cholesteatoma (SLC); S = granulation tissue in middle ear. (B) Schematic illustration of SLC.

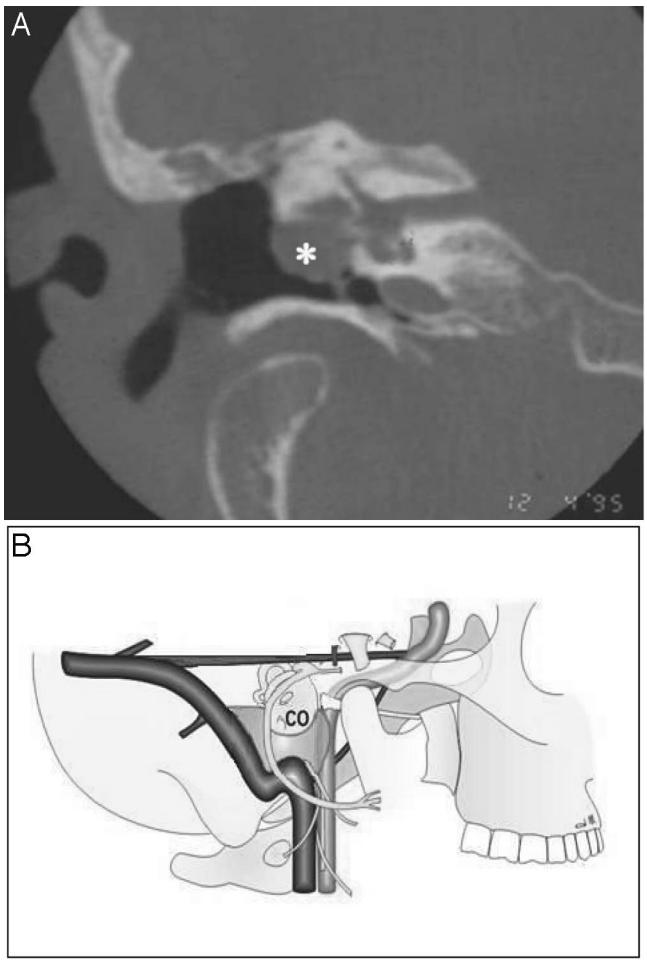

Fig. 2. (A) Infralabyrinthine cholesteatoma (ILC). (B) Schematic illustration of ILC. CO = cochlea.

clivus, and may involve the internal carotid artery. Posteriorly, it extends toward the dura of the posterior cranial fossa (Fig 2A, B).

Massive cholesteatoma is a diffuse type involving the otic capsule and may progress toward the internal carotid artery, the internal auditory canal, the posterior fossa dura, or the infralabyrinthine compartment. The origin of this type of cholesteatoma is difficult to define. It may be congenital or acquired (Fig 3A, B).

Infralabyrinthine apical cholesteatoma originates from the infralabyrinthine or apical compartments. If it arises from the former, it extends along the internal carotid canal toward the clivus and the sphenoid sinus, which may be involved or arises from the latter, the apical cells, and extends posteriorly toward the infralabyrinthine cells. It may be congenital in origin when it arises from the apical compartment or acquired when it arises from the infralabyrinthine cells (Fig 4A–F).

Apical cholesteatoma is typically congenital and rare type. It arises from the petrous apex and may extend toward the horizontal portion of the carotid artery or the trigeminal nerve or the dura of the posterior and middle fossa. Its diagnosis is suspected when associated with facial paralysis, vertigo, or deafness and rarely with trigeminal symptoms (Fig 5A, B).

Surgical Problems

In cholesteatoma surgery, dissecting the cholesteatomatous matrix from its bony attachment is simple and ensures radical removal of the pathology provided that the surgeon needs to check all the suspected hidden sites. The problem arises when cholesteatoma is attached to soft tissue like dura, sigmoid sinus, jugular bulb, internal carotid artery, or the facial nerve in which its radical removal is a challenge to the surgeon.

Dural Involvement

Dural involvement with cholesteatoma recommends wide bony removal. If any doubt exists about leaving cholesteatoma matrix over the dura, a bipolar coagulation of the suspected dural portions is the safest solution for devitalization of the cholesteatoma matrix and avoids dural tear with subsequent cerebrospinal fluid leakage. Other options are either to remove the affected dura and close the defect with fascia, which we do not recommend for the risk of meningitis, or to use 90% ethyl alcohol as proposed by Fisch.[4] The latter two options are not used in our center. The former option is simple, safe, and less time-consuming with no evidence of recurrence in the present study.

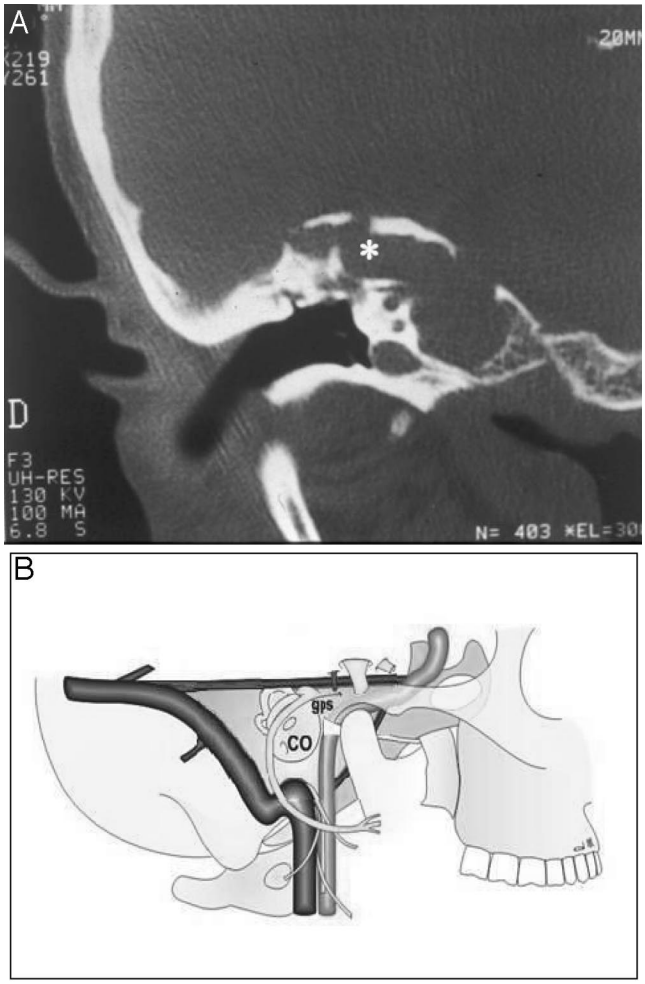

Fig. 3. (A) Massive cholesteatoma (MC). (B) Schematic illustration of MC. CO = cochlea; gps = greater superficial petrosal nerve.

Sigmoid Sinus and Jugular Bulb Involvement

These structures have a very delicate wall that might rupture during cholesteatoma dissection. Thus, it might be necessary to decompress them with intraluminal and extraluminal packing of the sigmoid sinus using Surgicel and to ligate the jugular vein in the neck. This aids in the removal of the lateral wall of the sigmoid sinus and the dome of the jugular bulb covered by the cholesteatoma matrix. Bleeding from the inferior petrosal sinus is controlled by Surgicel packing. Performance of such a step should be wise enough not to produce intracranial complication in case of a dominant ipsilateral sigmoid sinus. Therefore, if there is radiologic evidence of involvement of these structures, the contralateral sigmoid sinus must be assessed.

Carotid Artery Involvement

Dissection of cholesteatoma matrix from the internal carotid artery is relatively easy as a result of its thicker wall providing safe dissection in experienced hands. Infratemporal fossa type B and modified transcochlear approaches render safe, adequate exposure and control of the vertical and horizontal portions of the intrapetrous carotid artery.

Facial Nerve Involvement

Facial nerve paresis or palsy is one of the common presentations of PBC. When the facial nerve is preoperatively involved, the surgeon must suspect the presence of facial nerve compression, interruption, or being replaced by fibrous tissue. In the former condition, simple decompression of the nerve is the treatment of choice by removal of cholesteatoma matrix. If complete removal is not possible, resection of the involved segment is the only sure alternative for complete removal of the pathology. In the latter two, facial nerve endings should be refreshed or the fibrous segment be excised followed by facial nerve repair according to the size of the defect. If the defect is small, the nerve could be approximated after rerouting to perform tension-free primary anastomosis. Otherwise, sural nerve cable grafting is performed. In cases presented with facial paralysis after more than 1 year, hypoglossal facial nerve anastomosis should be considered.

Inner Ear Involvement and Hearing Status

In PBC surgery, we usually aim for radical removal of the pathology. We do not hesitate to remove the cochlea if there is evidence of its invasion by cholesteatoma matrix provided that the contralateral ear is normal or possesses acceptable hearing status. The problem arises when we deal with the only hearing ear. We used to follow up these cases with regular hearing assessment, high-resolution computed tomography, and magnetic resonance imaging if indicated. If there is progressive hearing loss, surgical removal of the pathology is required. Before removal of the pathology, we insert a cochlear implant in the contralateral ear if it is feasible. Otherwise, a planned surgical removal of the pathology with simultaneous insertion of a cochlear implant is performed. If the lesion is small and not eroding the otic capsule, the pathology is removed and the inner ear is saved. This condition seldom exists because involvement of the otic capsule is a frequent finding.

Cerebrospinal Fluid Leaks

Cerebrospinal fluid leaks may result during dissecting the cholesteatoma matrix from the dura. A small dural tear is sealed by a muscle plug introduced through the defect in the subarachnoid space. On the other hand, in large defects, the edges of the dura are sutured over the muscle plug and secured with fibrin glue.

MATERIALS AND METHODS

From January 1984 to October 2004, the senior author (M.S.) had operated 93 patients with PBC out of 2,739 (3.4%) cases presented with chronic ear discharge and cholesteatoma formation. Different types of cholesteatomas were encountered in the study of 93 ears, with 52 right and 41 left. All patients underwent thorough clinical otoneurologic examination. Audiologic examination was performed preoperatively and in follow-up visits. High-resolution computed tomography with an axial and coronal bone window images was indicated in patients presenting with facial paralysis, vertigo, anacusis, and cranial nerve paralysis in a discharging ear. A history of middle ear surgery was another indication for computed tomography scan. In some cases,

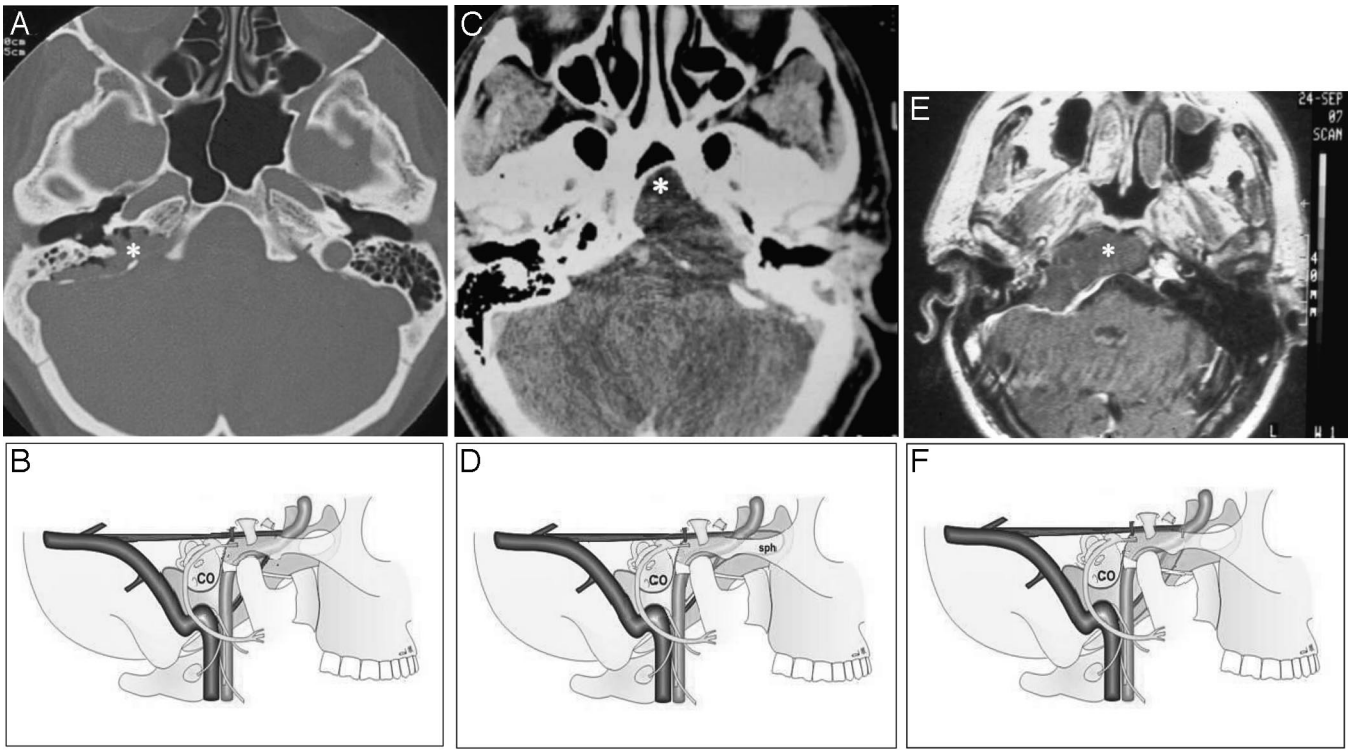

Fig. 4. (A) Infralabyrinthine apical cholesteatoma (ILAC). (B) Schematic illustration of ILAC. CO = cochlea. (C) Sphenoid extension of ILAC. (D) Schematic illustration of ILAC with sphenoid extension. CO = cochlea; sph = sphenoid sinus. (E) Clival extension of ILAC. (F) Schematic illustration of ILAC with clival extension. CO = cochlea.

the study was completed with imaging T_1- and T_2-weighted images.

RESULTS

The age in this series ranged from 9 to 83 years with a mean age of 46 years. Males were 66, whereas females were 27 with a ratio 2:1. Hearing loss was the most common presentation (94%) and facial nerve dysfunction was the second. Different symptoms are illustrated in Table I.

Massive cholesteatoma affected 42 ears. Supralabyrinthine and infralabyrinthine cholesteatoma were found in 41 and seven ears, respectively. Meanwhile, only three had infralabyrinthine–apical. Acquired cholesteatoma was the dominating pathologic type. Thirteen percent were recurrent cases as a sequence of a previous operation performed elsewhere. These cases are considered as iatrogenic cholesteatoma (Table II).

Table III represents the distribution of the total number of patients with facial nerve dysfunction preoperatively versus different types of PBC. Grade VI facial nerve dysfunction was common with massive cholesteatoma.

The most common treatment used for facial nerve paralysis was facial nerve decompression. Sural nerve grafting was used in 18.3% of cases. End to end anastomosis and facial nerve rerouting were different modalities of management that were not constantly used. Forty-one percent of cases did not need management of the facial nerve; this was the result of several factors because the duration of total paralysis (grade IV) was longer than 2 years or the pathology did not involve the facial nerve (Table IV).

The minimum duration of paralysis was 2 months and the maximum was 37 years. Of 42 patients with a preoperative normal facial nerve function, only six patients developed facial palsy (four patients had grade II, two patients had grade III). Forty-six of 51 patients presented with preoperative facial nerve dysfunction and underwent follow up for more than 1 year by means of House-Brackmann's grading system. The duration of preoperative facial nerve paralysis in relation to postoperative recovery revealed 60% improvement in patients with a preoperative facial nerve dysfunction less than 1 year.[5] On the other hand, 19.2% was the improvement in cases presented with a more than 1-year facial nerve paralysis (Tables V and VI). Those patients who did not accomplish 1-year follow up of facial nerve recovery were excluded (five of 51 cases).

The modified transcochlear approach (MTCA) was more frequently used for massive and supralabyrinthine types. Enlarged translabyrinthine approach (ETLA) with blind closure of the external canal was applied to 14 ears. Radical petromastoidectomy and middle cranial fossa approach were used in five and three ears, respectively (Table VII).

Follow Up

Follow up was carried out with a minimal 1-year postoperative period. Twelve patients who were operated in 2003 and 2004 were excluded from the follow-up period. Computed tomography and magnetic resonance imaging with fat suppression test, either used separately or in

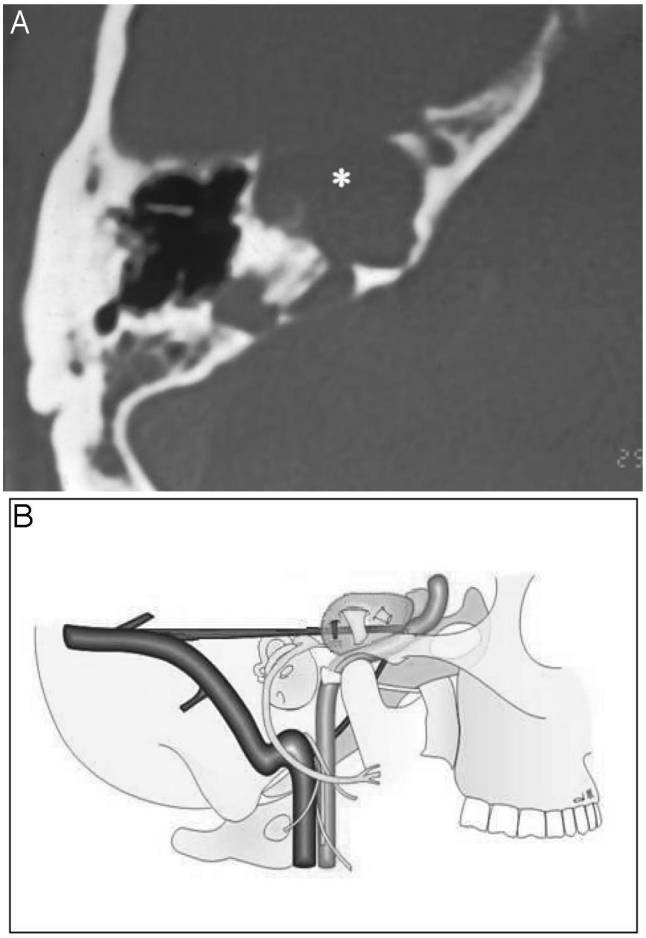

Fig. 5. (A) Apical cholesteatoma (AC). (B) Schematic illustration of AC.

combination, were the major tools for follow up. Recurrence was observed in two cases; the first was primarily treated with petrosectomy operation in which a revision surgery was performed and the cavity was left exteriorized. The second had residual cholesteatoma after previous radical mastoidectomy, and revision mastoidectomy with cavity obliteration was sufficient.

Inner ear involvement in some cases and the use of the modified transcochlear, enlarged translabyrinthine, and transotic approaches in others resulted in total hearing loss (HL) in 83 cases. Postoperative audiogram showed four cases with conductive HL and six cases affected with mixed HL. The only patient presented with a preoperative normal hearing in this series was found to have cholesteatoma eroding the cochlea and labyrinth that was managed through translabyrinthine approach (Table VIII).

Intraoperative complications were documented in four patients. Two patients had injury of sigmoid sinus and jugular bulb. Lower cranial nerve injury manifested in one case and encephalomeningocele was encountered in the last case. Postoperative complications are tabulated in Table IX. No single mortality was recorded in this series.

TABLE I.
Different Presentations and Their Percentages.*

Symptom	No. of Patients	Percent
Hearing loss	87	94
Facial palsy	51	55
Otorrhea	46	49
Vertigo	27	29
Instability	7	8
Other	19	20

*Patients usually presented with more than one symptom. Other presentations were tinnitus, headache, and otalgia.

TABLE II.
Classification of Petrous Bone Cholesteatoma versus Pathologic Type.

Class/Pathology	Congenital 15%	Acquired 72%	Recurrent/ Iatrogenic 13%	Total
Massive (45%)	6	32	4	42
Supralabyrinthine (44%)	5	30	6	41
Infralabyrinthine (8%)	2	4	1	7
Infralabyrinthine–apical (3%)	1	1	1	3

DISCUSSION

Petrous bone cholesteatoma represents a clinical challenge in both its diagnosis and surgical management. It may arise from an acquired middle ear cholesteatoma or embryonic epithelial remnant. It should be underlined that failure of the middle ear surgery may result in iatrogenic PBC. The availability and the advances of imaging techniques using high-resolution computed tomography scanning and magnetic resonance imaging renders petrous bone cholesteatoma feasible diagnosis and management.[6,7] The concept of classification is of paramount importance in surgical planning. The introduction and advancement of lateral skull base approaches made radical removal of the disease more assured. The ideal surgical approach of this type of lesion must provide an adequately wide route for its extirpation and safe control of the dura, facial nerve, carotid artery, sigmoid sinus, and

TABLE III.
Distribution of Preoperative Facial Nerve Dysfunction: Grades of Different Types of Petrous Bone Cholesteatoma.

Preoperative FN Grading	Classes of Petrous Bone Cholesteatoma				Number of Cases and Percents
	Massive	SL	IL	ILA	
Grade I	12	22	5	3	42 (45%)
Grade II	9	3	0	0	12 (13%)
Grade III	4	5	1	0	10 (11%)
Grade IV	2	3	1	0	6 (6%)
Grade V	15	8	0	0	23 (25%)
Total	42	41	7	3	93 (100%)

FN = facial nerve; SL = supralarabyrinthine cholesteatoma; IL = infralabyrinthine cholesteatoma; ILA = infralabyrinthine apical cholesteatoma.

TABLE IV.
Management of the Facial Nerve in Petrous Bone Cholesteatoma.

Technique	No. of Patients	Percentage
No treatment	38	41%
Decompression	28	30%
Sural nerve grafting	17	18.3%
Rerouting	5	5.4%
End to end anastomosis	4	4.2%
XII–VII anastomosis	1	1.1%

the jugular bulb. Hearing preservation is secondary in respect to radical removal of the pathology. Our strategy for surgical removal was changed over the years from opened exteriorized radical cavity to MTCA with cavity obliteration. The widely and commonly used approaches, which provided radical removal of the pathology and safe management of important structures were MTCA or ETLA.

In 1969, Glasscock[8] described the middle cranial fossa (MCF) approach for various petrous bone lesions. This approach is used when the pathology is localized to the supralabyrinthine region with a serviceable hearing.[9] Posterior extension of this lesion necessitates a combined MCF–transmastoid approach. Transmastoid–retrolabyrinthine approach is appropriate for posterior lesions with limited superior extension. If hearing preservation is not aimed, translabyrinthine approach (TLA) ensures the adequate removal of the pathology. Yanagihara et al.[10] in 1992 believed that the translabyrinthine approach with or without transcochlear approach is the basic technique for removal petrous apex cholesteatoma and middle fossa approach should be conducted when the cholesteatoma is deeply situated in the petrous apex and its matrix adheres to the middle fossa dura in such a way that it can be removed safely with this approach.

Based on experience with 93 cases, we did not consider a radical cavity as the treatment of choice for PBC and believe that the obliteration technique is a recommended procedure. We could rarely preserve the hearing without compromising total removal of cholesteatoma; therefore, removal of the otic capsule, when it is involved, is usually performed without hesitation to achieve our primary goal of radical removal of the pathology, especially if the contralateral ear has serviceable hearing. In cases of the only hearing ear, regular follow up is the rule. If there is continuous progression and deterioration of hearing, surgical intervention is planned. A trial to insert a cochlear implant in the contralateral ear before removal of cholesteatoma could be performed if feasible or a simultaneous insertion of a cochlear implant in the same ear is carried out.

We combined and modified the transcochlear approach introduced by House and Hitselberger[11] in 1976 with the infratemporal fossa approach described by Fisch[12] in 1977 together with obliteration as suggested by Coker et al.[13] This modified transcochlear approach[14] provides an excellent exposure of the clivus, the petrous apex, the dura of the posterior and middle fossa, sigmoid sinus, jugular bulb, and the internal carotid artery. It also avoids cavity problems as skin entrapment and disease recurrence with minimal incidence of cerebrospinal fluid leakage. Disadvantages of this technique are removal of the otic capsule and the risk of residual pathology that can not be detected clinically and requires follow up with computed tomography and magnetic resonance imaging with the fat-suppression technique.

We use MTCA for a massive type of PBC. A transotic approach is also used for massive type provided that preoperative facial nerve function is normal. On the other hand, modified translabyrinthine approach with removal of the external auditory canal and blind sac closure is used in extensive lesions without cochlear involvement. Fat obliteration of a radical petromastoid cavity with blind closure of the external auditory canal and the eustachian tube is preferred in infralabyrinthine extension. Combined infratemporal fossa type B approach and modified transcochlear approaches provide adequate control of the vertical and horizontal portions of the internal carotid artery and the sphenoid sinus and the midclivus.

Dealing with the cholesteatoma matrix represented some difficulties. Cholesteatoma matrix adhered to the dura is managed with a bipolar coagulation. Cases with cerebrospinal fluid leakage intraoperatively were controlled with adequate cavity obliteration. Thin delicate

TABLE V.
Improvement of Facial Nerve Dysfunction in Relation to Duration (less than 12 months).

		Postoperative Facial Grading						
Group	Preoperative Facial Grading	I	II	III	IV	V	VI	Total
Patients with <12 months facial nerve dysfunction	II	3	1	2	1			7
	III			1			1	2
	IV		1					1
	VI			6	1	2	1	10
	Total	3	2	9	2	2	2	20

TABLE VI.
Improvement of Facial Nerve Dysfunction in Relation to Duration (more than 12 months).

Group	Preoperative Facial Grading	Postoperative Facial Grading						Total
		I	II	III	IV	V	VI	
Patients with >12 months facial nerve dysfunction	II		1	3				4
	III		2	3			2	7
	IV			1	1	1	2	5
	VI				2		8	10
	Total		3	7	3	1	12	26

wall of the sigmoid sinus and the jugular bulb rendered difficulty in dissection of cholesteatoma matrix adherent to their walls. The use of bipolar coagulation and/or removal of their lateral wall after extraluminal and intraluminal packing of Surgicel solved the problem.

Facial nerve showed a spectrum of different pathologies, and for each, a management technique was chosen. Decompression of the facial nerve was the frequently performed technique when the nerve was found to be compressed but anatomically intact. Fibrotic segment was excised and a tension-free anastomosis was performed after facial nerve rerouting or using a cable nerve graft (sural nerve). Axon et al.[15] stated that the prime factor for facial paralysis is perigeniculate ischemia. Accordingly, they adopted an aggressive policy whereby the presumed ischemic perigeniculate segment is resected, including the adherent cholesteatoma matrix, followed by end to end anastomosis over the posterior fossa dura. This technique hopes to promote axon regeneration along a presumed healthy and well-vascularized nerve.

Early diagnosis and management of patients who developed facial nerve dysfunction for a period less than 1 year revealed a significant improvement of the outcome (60%). On the contrary, only 19.2% improvement of facial nerve paralysis was observed in a patient with late presentation and management of their facial nerve deficit for a duration more than 1 year. In the literature, it is reported that the duration of the preoperative facial nerve deficit is the most important influencing factor for the final outcome of facial nerve results, and the 1-year period after the occurrence of the preoperative clinical deficit seemed to be the cutoff point in achieving a high rate of good postoperative recovery.[5]

TABLE VII.
Surgical Approach Used in Relation to the Class Type.

Surgical Approach	MC	SLC	ILC	ILA
MTCA	18	8	2	—
ETLA	12	15	—	1
Middle Fossa	2	1	—	—
Transotic	4	8	—	1
Subtotal petrosectomy	3	7	—	—
Subtotal petrosectomy + obliteration	—	—	3	—
Radical pertomastoidectomy	3	2	—	—
Retrolabyrinthine	—	—	2	—
Infratemporal fossa (B)	—	—	—	1
Total	42	41	7	3

MC = massive cholesteatoma; SLC = supralabyrinthine cholesteatoma; ILC = infralabyrinthine cholesteatoma; ILA = infralabyrinthine apical; MTCA = modified transcochlear approach; ETLA = enlarged translabyrinthine approach.

CONCLUSION

Petrous bone cholesteatoma presents difficulties in its diagnosis and treatment. Adequate history-taking and high clinical suspicious with the advancement of imaging techniques made its diagnosis more feasible. Improvement of the lateral skull base approaches rendered safe, adequate, and complete removal of the tumor.

Radical removal of the disease is always our aim in cholesteatoma surgery and takes priority than hearing preservation. So, we do not hesitate to remove the otic capsule when it is mandatory to achieve our goal. The middle fossa is indicated in an attempt to preserve the hearing in small supralabyrinthine lesions without any radiologic evidence of cochlear or labyrinthine fistula. The modified transcochlear approach is our technique of choice for massive cholesteatoma. Radical petromastoid exenteration with fat obliteration of the cavity is performed in small infralabyrinthine cholesteatoma.

TABLE VIII.
Preoperative versus Postoperative Hearing.

Type of Hearing	Preoperative Hearing	Postoperative Hearing
Normal	1	0
Conductive hearing loss	27	4
Mixed	23	6
Sensorineural hearing loss	8	0
Anacusis	34	83

TABLE IX.
Postoperative Complications.

Postoperative Complications	No. of Cases	Percent
Facial palsy	6 of 42	14
Anacusis	49 of 59	83
Cavity infection	1 of 93	1
Meningitis	1 of 93	1
Abdominal hematoma	1 of 93	1

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