Annals of Otology, Rhinology & Laryngology 119(8):514-525.

Management of Complex Cases of Petrous Bone Cholesteatoma

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Objectives: In a retrospective analysis of a quaternary referral neuro-otologic private practice, we identify complex cases of petrous bone cholesteatoma (ie, cases with encasement of vital structures such as the internal carotid artery, jugular bulb, and sigmoid sinus, with further extension to the clivus, sphenoid sinus, or rhinopharynx), review surgical approaches and techniques of management of vital structures, and propose the ideal surgical management.

Methods: We performed a retrospective case study of 130 cases of petrous bone cholesteatoma submitted to surgery between 1979 and 2009 to identify the complex cases and their classification, approach used, outcomes, and recurrences.

Results: Of 130 cases, 13 were complex. Facial palsy was the presenting feature in 11 cases, 7 of which presented with grade VI palsy. A long duration of facial palsy (more than 3 years) was seen in 5 cases. Clival involvement was seen in 6 cases; 1 case extended to the sphenoid sinus, and 1 to the rhinopharynx. The internal carotid artery was encased in 11 cases in the vertical and the horizontal parts. The jugular bulb was involved in 7 cases. Modified transcochlear approaches or infratemporal fossa approaches were used in all cases. There were no recurrences.

Conclusions: Classification is fundamental to choosing the right surgical approach. Transotic and modified transcochlear approaches hold the key to treating complex cases. Infratemporal fossa approach type B has to be used for extension into the clivus, sphenoid sinus, or rhinopharynx. Internal carotid artery, jugular bulb, and sigmoid sinus involvement should be identified before operation.

Key Words: dura, facial nerve, internal carotid artery, jugular bulb, lower cranial nerve, petrous bone cholesteatoma.

INTRODUCTION

One form of epidermoid involving the petrous portion of the temporal bone is called a petrous bone cholesteatoma (PBC). Slow-growing and locally destructive lesions, PBCs often remain asymptomatic until they involve the facial nerve or the labyrinth. Cases with encasement of one or more vital structures (internal carotid artery [ICA], jugular bulb, or sigmoid sinus) with or without further extension to the clivus, sphenoid sinus, or rhinopharynx are considered complex. According to their cause, they are divided into congenital and acquired forms, but as far as the management is considered, there is no difference.

PBC is difficult to manage. When the disease extends beyond the classic situation (Sanna classification), it becomes extremely difficult and requires a meticulously planned surgical strategy. It requires an extensive lateral skull base approach with complete knowledge of all of the lateral skull base approaches in order to remove disease from the vital structures (ICA, facial nerve, dura, jugular bulb, and sigmoid sinus) and to reach inaccessible areas (occipital condyle, ICA, sphenoid sinus, rhinopharynx). The application of various complex skull base procedures has a consequent risk of injury to vital neurovascular structures.

The Sanna classification[1] for PBC includes 5 classes: supralabyrinthine, infralabyrinthine, massive, infralabyrinthine-apical, and apical. Each class describes both the location and the extent of the lesion (Table 1). The extension of PBCs from the petrous bone into the clivus, sphenoid sinus, or rhinopharynx is rare, but can be extremely difficult to treat when present. These extensions require combination of an infratemporal fossa approach type B (IFTB) with a transotic (TO) approach or modified transcochlear approach type A (MTCA). The combination of infratemporal fossa approach type A (IFTA) and MTCA is called modified transcochlear approach type B (MTCB).[2] Therefore, a subclassification has been added to the existing classification.[3] Accordingly, extensions are designated "C" (clivus), "S" (sphenoid sinus), and "R" (rhinopharynx; Fig 1). This subclassification would help in planning the surgical approach.

High-resolution computed tomography (CT) of the temporal bone and cerebral magnetic resonance imaging (MRI) with gadolinium enhancement are the imaging techniques of choice for these lesions. They are important not only in diagnosis, but also in planning the surgical approach. Over time, cavity

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TABLE 1. SANNA'S CLASSIFICATION FOR PETROUS BONE CHOLESTEATOMA

Class	*Location*	*Extension*
Class I: supralabyrinthine	Geniculate ganglion of facial nerve	Anterior: horizontal part of ICA Posterior: posterior bony labyrinth Medial: IAC, petrous apex Inferior: basal turn of cochlea
Class II: infralabyrinthine	Hypotympanic and infralabyrinthine cells	Anterior: ICA vertical part, petrous apex, clivus Posterior: dura of posterior cranial fossa and sigmoid sinus Medial: IAC, lower clivus, occipital condyle Inferior: jugular bulb, lower cranial nerves
Class III: infralabyrinthine-apical	Infralabyrinthine compartment, ICA reaching to petrous apex	Anterior: ICA vertical and/or horizontal parts Posterior: posterior fossa through retrofacial air cells Medial: petrous apex, clivus, sphenoid sinus, rhinopharynx Inferior: jugular bulb, lower cranial nerves
Class IV: massive	Entire otic capsule	Anterior: ICA vertical and/or horizontal parts Posterior: posterior fossa dura and IAC Medial: petrous apex, superior and mid-clivus, sphenoid sinus Inferior: infralabyrinthine compartment
Class V: apical	Petrous apex	Anterior: Meckel's cave area and may involve nerve V Posterior: IAC and posterior cranial fossa Medial: superior or midclivus, sphenoid sinus Inferior: infralabyrinthine compartment

ICA — internal carotid artery; IAC — internal auditory canal.

obliteration after complete disease eradication has superseded the age-old open techniques because of the advantages of protection of exposed vital structures such as the ICA and the dura from infection and prevention of large draining cavities. Although obliteration of the cavity is an excellent technique, it carries the disadvantage of a lack of direct visualization of recurrence, and therefore necessitates following these patients with MRI with gadolinium enhancement.

MATERIALS AND METHODS

A retrospective case study was performed on 130 patients who received a diagnosis of and had treatment for PBC out of 4,500 cases of middle ear cholesteatoma from February 28, 1979, to May 20, 2009, at the Gruppo Otologico, Piacenza, Italy. Cases with involvement of one or more of the vital structures (ICA, jugular bulb, or sigmoid sinus) with further extension of the PBC to the clivus, sphenoid sinus, or rhinopharynx were considered complex.

All of these patients underwent a thorough otoneurologic evaluation followed by a pure tone audiometric evaluation. High-resolution CT of the temporal bone (coronal and axial) with bone window images with a width of 1 to 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 CT. Cerebral MRI (T1- or T2-weighted with gadolinium enhancement) was performed in cases suspected of extratemporal spread on CT until 1999, after which MRI was performed in all cases of PBC.

All of the lesions were classified by the Sanna classification[1] according to topographic location and extent on CT. Depending on the class of the PBC, management was planned. The surgical approach adopted was radical removal of the disease followed by obliteration of the cavity with temporalis muscle or abdominal fat with cul de sac closure of the external auditory canal (EAC).

Facial nerve function was graded before operation, immediately after operation, and 1 year after operation, according to the House-Brackmann (HB) grading system for facial nerves. The follow-up of these patients ranged from 3 months to 11 years (average, 4 years). Subsequently, the patients underwent an annual radiologic follow-up (CT and MRI) for detection of any recurrence for at least 5 years.

RESULTS

Of 130 cases of PBC, there were 13 complex cases (10%; 4 right-sided and 9 left-sided). Ten patients were male and 3 female (31 to 81 years of age; mean, 56 years).

Hearing loss was the commonest symptom; 7 patients had profound sensorineural hearing loss on presentation. Eleven patients presented with facial palsy (complete or grade VI in 7). The duration of facial paralysis in 4 patients was more than 3 years. Ear discharge, vertigo, tinnitus, and headache were the other presenting complaints.

There were 9 cases of acquired PBC, 1 iatrogenic PBC, and 3 congenital PBCs. Six cases were mas-

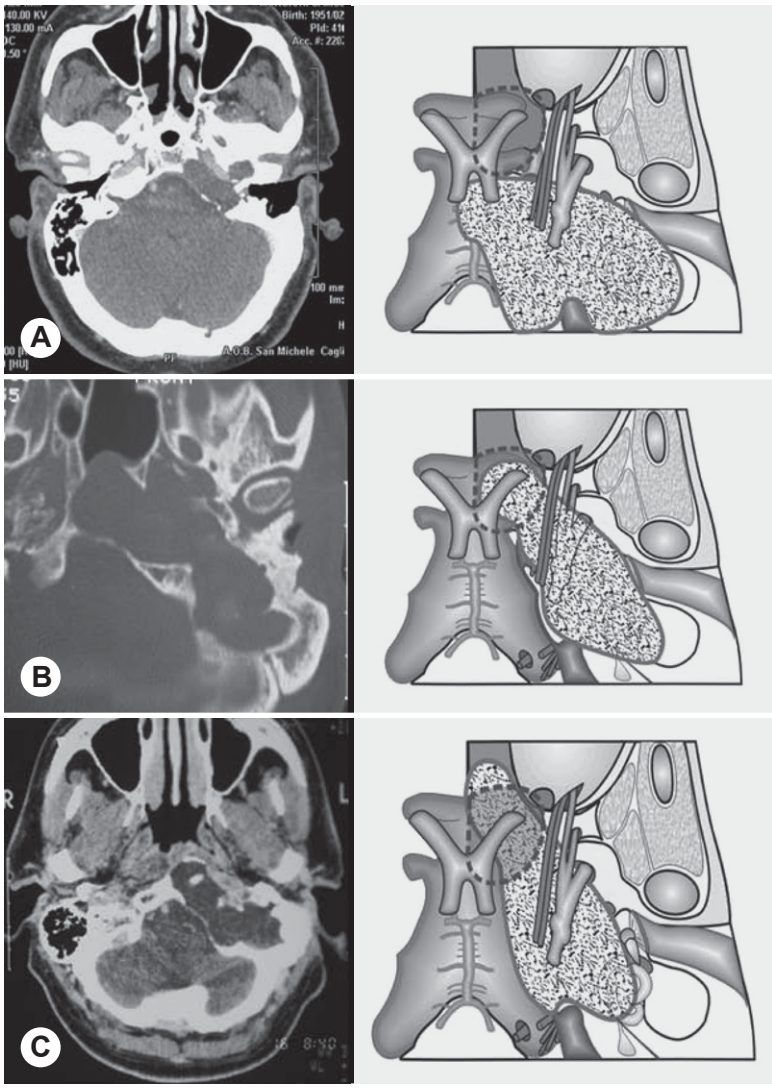

Fig 1. Extension of petrous bone cholesteatoma (PBC) to **A)** clivus, **B)** sphenoid sinus, and **C)** rhinopharynx. Computed tomography (CT) scan views (axial) are shown on left, and corresponding diagrams are shown on right.

sive, 3 were infralabyrinthine-apical, 2 were infralabyrinthine, 1 was supralabyrinthine, and 1 was apical. Clival involvement was seen in 6 cases. Involvements of the sphenoid sinus and rhinopharynx were seen in 1 case each. Involvement of the occipital condyle was seen in 2 cases, which also involved the soft tissues of the neck (Table 2).

MTCA was used in 8 cases. MTCB was performed in 3 cases. IFTA plus IFTB was performed in 1 case with sphenoid sinus involvement, and 1 patient with infralabyrinthine PBC underwent a retrolabyrinthine (RL) plus transmastoid (TM) approach (Table 2).

The facial nerve was involved in all but 1 case. Five patients with facial palsy of more than 3 years' duration did not undergo any treatment for facial nerve lesions. Sural nerve grafting was performed in 4 cases. Facial-hypoglossal anastomosis was done in 1 case primarily. One patient with apical PBC in which the facial nerve was involved in the internal auditory canal (IAC) underwent excision and sural nerve grafting, and had grade VI facial nerve function at 1 year after operation. Facial-hypoglossal nerve anastomosis was performed, after which the facial nerve function improved to grade IV. Facial nerve rerouting was done in 1 case, and decompression was done in 1 case (Table 3).

The ICA was involved in 11 of the 13 cases in the vertical and horizontal parts. The jugular bulb and sigmoid sinus were involved in 7 cases. The lower cranial nerves were involved in 4 cases (Table 2).

The complications included an intraoperative cerebrospinal fluid (CSF) leak in 2 cases. The patient with extension of an infralabyrinthine-apical PBC to the sphenoid sinus had a CSF leak with infection in the postoperative period. The other patient presented with hypoglossal nerve palsy, which persisted in the postoperative period.

There were no deaths and no recurrences. Of the 13 patients, 11 had at least 1 year of follow-up. One

TABLE 2. COMPLEX CASES OF PETROUS BONE CHOLESTEATOMA

Case No.	Class	Structures Involved				Extension	Approach
		FN	ICA	JB-SS	LCN		
1	IL	Y	N	Y	Y	Clivus, occipital condyle, and neck	RL+TM
2	Apical	Y	Y	N	N	Clivus	MTCA
3	IL-apical	Y	Y	Y	Y	Clivus and sphenoid sinus	IFT-A/B*
4	Massive	Y	Y	N	N		MTCA
5	Massive	Y	Y	N	N		MTCA
6	IL	Y	Y	Y	Y	Occipital condyle and neck	MTCA
7	SL	Y	Y	N	N		MTCA
8	Massive	Y	Y	N	N	Clivus	MTCB
9	Massive	Y	Y	Y	N		MTCA
10	IL-apical	Y	Y	N	N	Clivus	MTCB
11	Massive	Y	Y	Y	N		MTCA
12	IL-apical	Y	N	Y	N		MTCA
13	Massive	Y	Y	Y	Y	Clivus and rhinopharynx	MTCB

FN — facial nerve; ICA — internal carotid artery (horizontal and vertical); JB-SS — jugular bulb and sigmoid sinus; LCN — lower cranial nerves; IL — infralabyrinthine; RL+TM — retrolabyrinthine and transmastoid; MTCA — modified transcochlear type A; IFT-A/B — infratemporal fossa approach types A and B; SL — supralabyrinthine; MTCB — modified transcochlear type B.

*Patient had complications of cerebrospinal fluid leak and infection.

of the other 2 patients was operated on in May 2009, and the other, with extension to the rhinopharynx, developed carcinoma of the colon 6 months after the surgery and hence was lost to follow-up.

CASE REPORTS

Case 1 (Infralabyrinthine, With Clival Extension). A 59-year-old woman presented with complaints of left-sided decreased hearing, tinnitus, and vertigo that had lasted for 3 months. She had no history of ear discharge. The facial nerve was normal. On otoscopic examination, the tympanic membrane was intact, but there was an erosion of the posterior canal wall in the inferior aspect. A pure tone audiogram showed hearing within normal limits.

High-resolution axial and coronal CT revealed a large osteolytic lesion underlying the left mastoid cortex, eroding the bone over the sigmoid sinus and posterior fossa; it extended via the infralabyrinthine cells up to the lower clivus and to the occipital condyle. The middle ear, cochlea, vestibule, and semicircular canals were normal (Fig 2).

Cerebral MRI was suggestive of a hypointense lesion on T1-weighted images and a hyperintense lesion on T2-weighted images on the left side, occupying the infralabyrinthine region from the mastoid cortex, extending medially up to the occipital con-

TABLE 3. FACIAL NERVE INVOLVEMENT

Case No.	Duration of Palsy (mo)	Preop FN	Site of Involvement	Treatment	Postop FN
1		I	Uncovered in mastoid portion	Decompression	I
2		I	IAC	Sural nerve grafting and later facial hypoglossal anastomosis	IV
3		I	Mastoid portion	Anterior rerouting	II
4	15	III	IAC, labyrinthine portion, GG, tympanic portion, second genu	Sural nerve grafting	VI
5	8	IV	IAC, labyrinthine portion, GG, tympanic portion, second genu	End-to-end anastomosis	III
6	12	IV	Labyrinthine portion, GG, tympanic portion	No treatment	IV
7	4	VI	IAC, labyrinthine portion, GG	Sural nerve grafting	III
8	60	VI	IAC, labyrinthine portion, GG, tympanic portion, second genu, mastoid portion	No treatment	VI
9	5	VI	Labyrinthine portion, GG, tympanic portion	Sural nerve grafting	III
10	216	VI	Tympanic portion	No treatment	VI
11	6	VI	IAC, labyrinthine portion, GG, tympanic portion, second genu, mastoid portion	Nerves VII-XII anastomosis	VI
12	36	VI	IAC, labyrinthine portion, GG, tympanic portion	No treatment	VI
13	84	VI	IAC, labyrinthine portion, GG, tympanic portion	No treatment	VI

FN — facial nerve function (House-Brackmann grading); GG — geniculate ganglion.

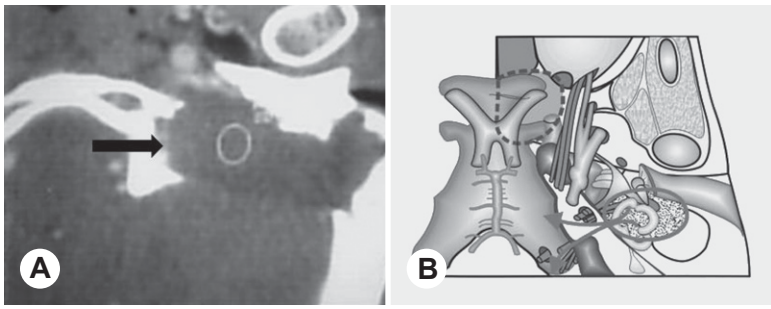

Fig 2. (Case 1) **A)** Axial high-resolution CT of temporal bone shows extent of PBC. Note intact labyrinthine bone. Arrow — erosion of occipital condyle. **B)** Diagram of infralabyrinthine PBC as viewed from top. Arrows — possible extensions of infralabyrinthine PBC (see Table 1).

dyle, and extending inferiorly into the neck (Fig 3). Magnetic resonance angiography was done to check the contralateral venous cerebral circulation, which was patent. The lesion was classified as infralabyrinthine, with extension to the lower clivus and the occipital condyle.

An RL plus TM approach was chosen, as the preoperative hearing was normal. Radical clearance of the disease was the aim, even if the hearing had to be compromised. During operation, the mastoid cortex was seen to be intact; the PBC had eroded the fallopian canal, leaving the facial nerve exposed from the second genu up to the stylomastoid foramen with preserved anatomic integrity of the nerve; therefore, the facial nerve was decompressed. The bone over the sigmoid sinus and the posterior fossa dura was eroded, and the cholesteatoma matrix was overlying it. The disease extended medially through the infralabyrinthine region, up to the lower clivus and the occipital condyle. The cochlea, the vestibule, and all of the semicircular canals were intact. Inferiorly, the cholesteatoma was overlying the digastric muscle. The matrix was present around the lower cranial nerves, but could be dissected off the nerves without injuring them. Complete clearance of the PBC was achieved. The cavity was obliterated with autologous abdominal fat, and blind sac closure of the EAC was performed.

The facial nerve in the postoperative period was grade I, with no deficit of the lower cranial nerves. The postoperative pure tone bone conduction results showed a high-frequency hearing loss. On postoperative neuroradiological follow-up, the patient was free of disease after 5 years and the facial nerve was grade I (Fig 4).

In this case, infralabyrinthine PBC with lower extension to the occipital condyle and the soft tissue of the neck posed difficulty in preservation of the jugular bulb and lower cranial nerves. It was an exceptional case in which the pneumatization and extent of the disease favored preservation of hearing (bone conduction), but even in such cases, hearing preservation should never be given preference over radical disease clearance.

Case 2 (Infralabyrinthine and Apical, With Clivus and Sphenoid Sinus Involvement). A 46-year-old man presented with a history of right ear discharge and complete hearing loss. The facial nerve was normal; there was no history of vertigo or tinnitus. There was a history of closed tympanoplasty performed 10 years earlier. On inspection, the patient had stenosis of the EAC with erosion of the posterior canal wall and discharge. Pure tone audiometry was suggestive of right-sided total deafness.

High-resolution CT and MRI were suggestive of a large PBC involving the right middle ear and mas-

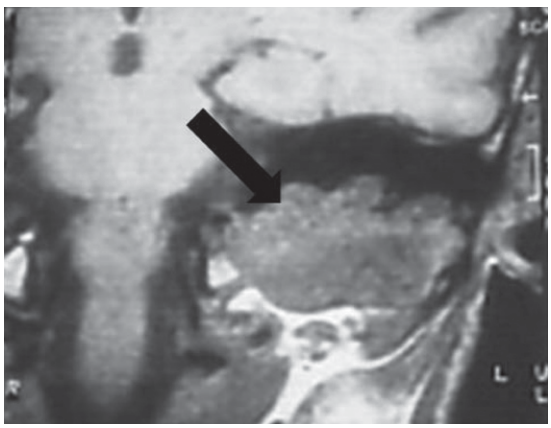

Fig 3. (Case 1) Coronal T1-weighted MRI. Note medial-to-lateral extent of lesion (arrow).

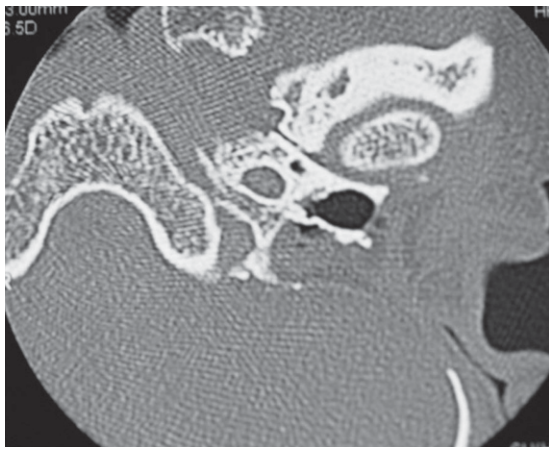

Fig 4. (Case 1) Postoperative axial CT shows no evidence of disease. Fat is seen obliterating cavity.

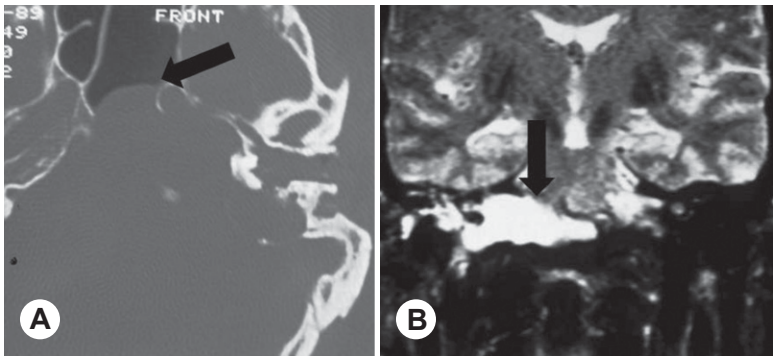

Fig 5. (Case 2) **A)** Axial CT scan shows PBC involving infralabyrinthine region, extending to petrous apex, clivus, and sphenoid sinus. Arrow — extension of PBC into sphenoid sinus. Note lesion medial to horizontal part of internal carotid artery (ICA). **B)** T2-weighted MRI with contrast enhancement shows extent of lesion (arrow).

toid, invading the petrous apex anteroinferior to the cochlea, infiltrating the clivus, and reaching the sphenoid sinus. The PBC was seen around the vertical and horizontal parts of the ICA (Fig 5). According to the radiologic findings, the lesion was classified as an infralabyrinthine apical type with extension to the sphenoid sinus.

Because of the anterior extension of the disease, IFTA plus IFTB was chosen. The facial nerve in the vertical part and horizontal part was dissected and was rerouted anteriorly. The PBC was found medial to the vertical part of the ICA, extending onto the mid-clivus and sphenoid sinus. Inferiorly, it extended toward the region of the jugular bulb. The internal jugular vein was ligated in the neck, and the sigmoid sinus was packed. The external walls of the jugular bulb and sigmoid sinus were removed. Bleeding from the inferior petrosal sinus was managed with Surgicel packing. The PBC was present medial to the jugular bulb, around the lower cranial nerves. The cholesteatoma was removed completely, and the cavity was obliterated with a vascularized temporalis muscle flap, as it was infected and fat obliteration carried a risk of further infection.

After operation, the patient developed a cavity infection and a CSF leak that were treated with medical therapy, compressive dressing, and lumbar drainage. The facial nerve function was grade II (HB) at 1 year on radiologic follow-up, and there was no evidence of any residual cholesteatoma at 15 years of follow-up (Fig 6).

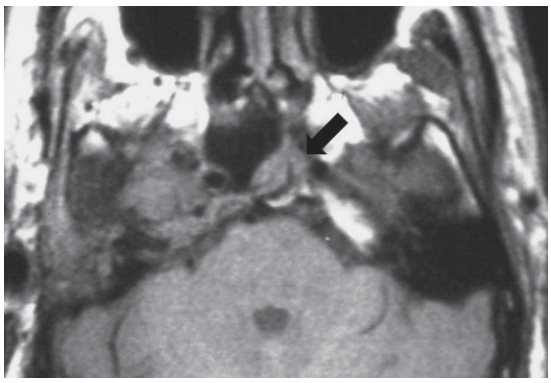

Fig 6. (Case 2) Postoperative MRI shows no evidence of residual disease. Arrow — temporalis muscle obliterating opening into sphenoid sinus.

In cases with massive anterior extension, IFTB is an excellent approach that provides very good access and control over the ICA in the vertical and horizontal portions along with access to the sphenoid sinus. In this case, IFTB was preferred over MTCB, as the preoperative facial nerve function was grade I.

Case 3 (Apical, With Clival Extension). A 43-year-old man presented with a history of left-sided progressive hearing loss and vertigo that had persisted for 1 year. There was a history of facial palsy that recovered in 10 to 15 days. On examination, the facial nerve was HB grade I. Pure tone audiometry suggested a left-sided high-frequency hearing loss.

High-resolution CT (Fig 7) and MRI revealed a well-defined, smooth osteolytic lesion at the left petrous apex, invading the IAC. The lesion was eroding the mid-clival region and was situated medial to the horizontal part of the ICA. It was hypointense on T1-weighted and hyperintense on T2-weighted images. The lesion was classified as an apical PBC of congenital origin with clival extension, as the middle ear and mastoid were normal.

MTCA was chosen to remove the disease. On exploration, a PBC was present medial to the vertical and the horizontal parts of the carotid artery, and the matrix of the cholesteatoma was adherent to the adventitia of the artery. The PBC was occupying the region of the petrous apex and the mid-clivus and

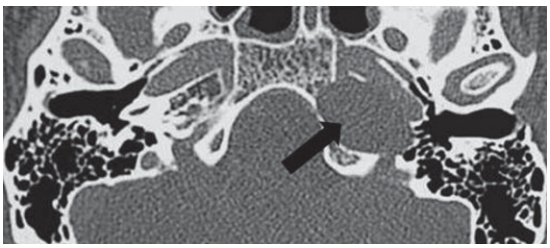

Fig 7. (Case 3) CT shows PBC occupying petrous apex, lying medial to horizontal part of ICA (arrow).

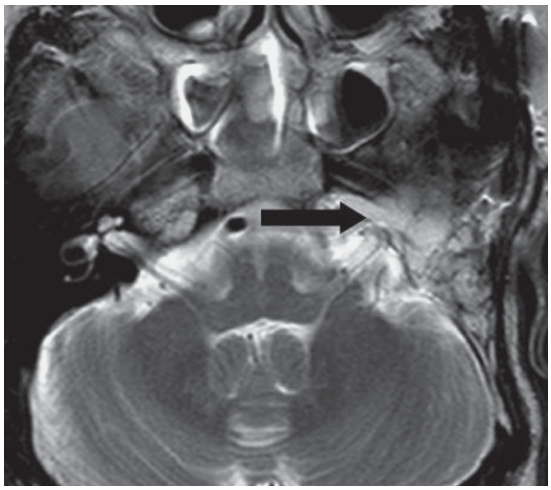

Fig 8. (Case 3) Postoperative T2-weighted MRI with contrast enhancement shows no evidence of disease. Fat is seen obliterating cavity (arrow).

was adherent to the acousticofacial bundle in the IAC. The matrix adherent to the carotid artery was dissected off the artery. The facial nerve was cut in order to gain access, the IAC was opened, and the disease adherent to the acousticofacial bundle was removed. The PBC was completely removed, and sural nerve grafting was performed. The eustachian tube was closed, and the cavity was obliterated with temporalis muscle and abdominal fat.

After 1 year, the facial nerve showed no signs of recovery, and remained grade VI. Hence, a faciohypoglossal anastomosis was performed. The facial nerve function 11 months after the hypoglossal-facial anastomosis was grade IV. There was no radiologic evidence of residual cholesteatoma 2 years after surgery (Fig 8).

An apical cholesteatoma is a rare pathologic entity that is congenital in origin. The diagnosis is usually delayed because of a lack of symptoms. Hearing preservation should not be given importance over clearance of the disease. Bone conduction can be preserved with use of IFTB.

Case 4 (Massive, With Clival Extension). A 58-year-old man presented with a history of facial palsy on the left side that had lasted for 6 years, along with gradually progressive hearing loss, tinnitus, and vertigo. He had developed a left gaze diplopia over the past few months. He underwent facial reanimation surgery 2 years earlier at another center, without any recovery. There was a history of open cavity mastoidectomy performed 20 years earlier.

On examination, the patient had a radical cavity; the facial nerve was grade VI (HB) along with left cranial nerve VI palsy. Pure tone audiometry indicated a profound sensorineural hearing loss on the left side.

High-resolution CT of the temporal bone with coronal and axial images revealed a large osteolytic lesion involving the cochlea, vestibule, and IAC, with erosion of the middle and posterior fossa dural plate, extending up to the middle and lower clivus. The lesion was seen medial to the horizontal portion of the ICA (Fig 9A).

Cerebral MRI with gadolinium enhancement revealed a hypointense lesion on T1-weighted images, occupying the petrous bone and the infratemporal fossa and extending superiorly to the cavernous part of the ICA. Posteriorly, the lesion extended into the cerebellopontine angle, lying adjacent to the brain stem. The dura of the middle fossa could be seen to be involved by the lesion. The lesion was classified as a massive PBC with clival extension.

MTCB was chosen, as the preoperative facial nerve function was grade VI. During operation, there was a massive cholesteatoma sac in the otic capsule, extending to the IAC, the petrous apex, and the clivus, which were removed. The facial nerve was interrupted from the IAC to the second genu. A large area of the middle and posterior fossa dura was uncovered. The adherent matrix was dissected, and suspected dural portions were bipolarized. The lesion was present medial to the vertical part, the genu, and the horizontal part of the ICA, extending to the cavernous part. The carotid artery was mobilized in its entire petrous part up to the anterior foramen lacerum, and the cholesteatoma medial to it was dissected. After clearance of the disease, the cavity was inspected with a 30° rigid endoscope to look for any residual disease. Facial nerve reconstruction was not possible because of long-standing facial palsy. An intraoperative dural tear with a CSF leak was repaired by inserting a free muscle plug

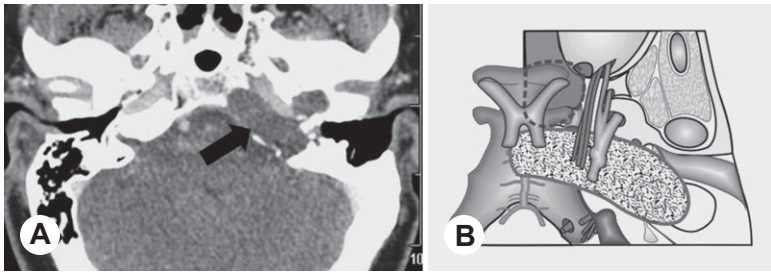

Fig 9. (Case 4) **A)** High-resolution CT axial cut demonstrates extent of massive PBC extending to clivus. Note previous open cavity mastoidectomy that was free of disease. **B)** Diagram of extent of disease. Note relation of trigeminal nerve and sixth nerve at petrous apex.

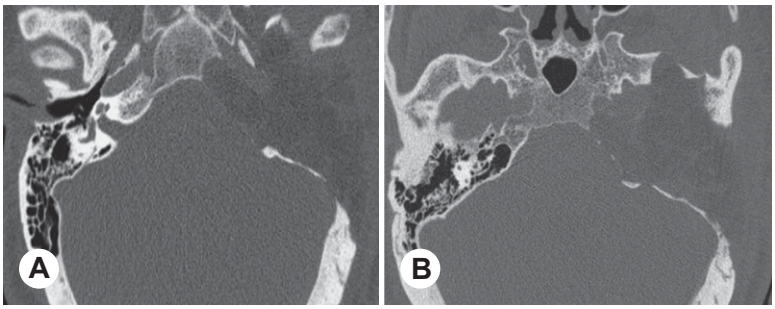

Fig 10. (Case 4) Postoperative high-resolution CT shows no evidence of residual PBC. **A)** Axial cut at level of horizontal portion of ICA shows fat medial to artery. **B)** Axial cut at level of cavernous portion of ICA shows no evidence of disease.

into the defect. The cavity was obliterated with autologous abdominal fat, and blind sac closure of the EAC was performed.

After operation, the patient had grade VI facial nerve paralysis. The nerve VI palsy had almost completely recovered at 1 month after operation. The neuroradiologic follow-up performed 3 months after the surgery did not reveal any residual lesion (Fig 10). This patient's operation was 5 months earlier, so long-term follow-up is not available at present.

A delay in diagnosis in this case was the main reason for the spread of the cholesteatoma. It is vital that all patients with radical cavities presenting with discharge, facial palsy, vertigo, and progressive hearing loss should be evaluated radiologically to rule out PBC.

Case 5 (Massive, With Clivus and Rhinopharynx). A 74-year-old man presented with a history of left-sided gradually progressive facial paralysis that had lasted for 7 years with progressive sensorineural hearing loss, tinnitus, and vertigo. The patient had had loss of consciousness 3 times in the 6 months before presentation. He had had otitis media during adolescence; it subsided after medical therapy.

Otoscopic examination revealed a dry attic perforation with a PBC visible behind an intact pars tensa in the anterior hypotympanum. The facial nerve was grade VI. There was a nerve XII palsy on the left, whereas nerves IX, X, and XI were normal. The nasal endoscopic examination suggested a bulge inferomedial to the left eustachian tube opening (Fig 11). Pure tone audiometry showed profound deafness on the left side.

High-resolution CT revealed a large osteolytic lesion occupying the left middle ear, mastoid, otic capsule, and petrous apex. The lesion extended toward the occipital condyle and the lower clivus posteriorly; anteriorly, it extended inferior to the sphenoid sinus, into the rhinopharynx. On cerebral MRI, it was hypointense on T1-weighting and hyperintense on T2-weighting. It extended anteriorly into the rhinopharynx and posteriorly to the occipital condyle (Fig 12).

MTCB was performed. During the operation, the facial nerve was disrupted. The cochlea was eroded by the disease. The ICA was engulfed by the lesion in the vertical and horizontal portions. The matrix was dissected off the ICA, and the lesion, which was found to reach the rhinopharynx, was removed from the retropharyngeal space. The jugular bulb and the lower cranial nerves, completely engulfed by the matrix, were cleaned. The suspected portions of the dura were bipolarized. There was an intraoperative CSF leak. A muscle plug was inserted into the dural defect and stabilized with fibrin glue. The cavity was obliterated with abdominal fat, and blind sac closure of the EAC was performed.

One month after the surgery, the facial nerve was grade VI, hypoglossal nerve palsy persisted, and CT showed no evidence of residual cholesteatoma. (We were unable to publish the image, as it was of poor quality.) The patient developed carcinoma of the colon 6 months after the surgery for PBC and was lost to follow-up.

Because of the long duration of the facial nerve paralysis, facial nerve reconstruction was not per-

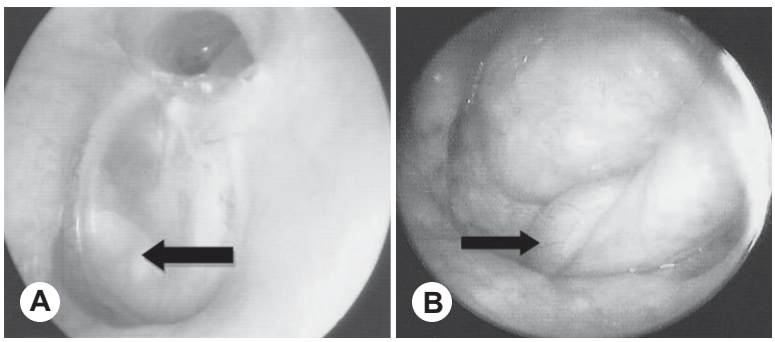

Fig 11. (Case 5) **A)** Dry attic perforation. Arrow — PBC mass behind intact pars tensa. **B)** Nasal endoscopic examination shows bulge (arrow) inferomedial to left eustachian tube opening.

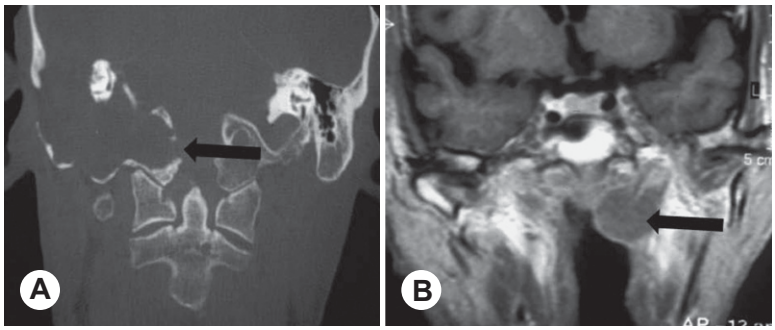

Fig 12. (Case 5) **A)** Coronal CT shows lesion extending to occipital condyle. Arrow — erosion of occipital condyle. **B)** Coronal T1-weighted MRI shows lesion extending to rhinopharynx. Arrow — lesion bulging in rhinopharynx.

formed in this case. It shows that MTCB can be used to remove a lesion extending to the rhinopharynx anteriorly and to the occipital condyle medially.

DISCUSSION

Cases of PBC with involvement of one or more vital structures (ICA, jugular bulb, or sigmoid sinus) with or without further extension of the PBC to the clivus, sphenoid sinus, and rhinopharynx are considered complex cases, not only because of the extent of surgical resection required to achieve complete removal, but also because of the morbidity associated with the involvement of vital structures and the risk of iatrogenic injury to these vital structures. Extensions to the sphenoid sinus, clivus, and rhinopharynx are tricky because of the special approaches required for their removal.

A delay in diagnosis is the main factor identified in the spread of these PBCs beyond the confines of the temporal bone. We have observed two main factors in this delay. First, radical mastoidectomy cases presenting with discharge are often labeled as discharging cavities and go undiagnosed until they present with facial palsy, vertigo, or sensorineural hearing loss. Second, cases of congenital PBC presenting with sudden-onset facial palsy are labeled as Bell's palsy because of the paucity of otoscopic findings. Bell's palsy is a diagnosis of exclusion and should be made after excluding other differential diagnoses. One of our patients (case 4) presented with facial palsy and a history of radical mastoidectomy performed 20 years earlier. He underwent plastic surgery for facial reanimation without its being noticed that he had a massive PBC. He remained without a diagnosis for 5 years before he presented to our clinic.

High-resolution CT with a bone window of 1- to 2-mm thickness (coronal and axial images) is the basic investigation for diagnosing PBC and planning management. Cerebral MRI (coronal and axial) with T1- and T2-weighted images with gadolinium enhancement should be performed in all cases of suspected or diagnosed PBC. It is imperative to interpret these images before embarking upon surgery, to know the extent of disease and plan the right surgical approach. The most important factors to be taken into consideration while treating complex PBC cases, in order of precedence, are 1) complete eradication of the disease; 2) preservation of facial nerve function, when present; 3) prevention of CSF leak and meningitis; and 4) obliteration of the cavity.

The choice of surgical approach is the most important step in surgical management of complex cases. It is based on the classification, which permits adoption of the correct approach, and on the preoperative facial nerve function. The two main approaches for these complex cases are the TO approach and MTCA.

Involvement of the horizontal part of the carotid artery and anterior extensions of the PBC (clivus, sphenoid sinus, rhinopharynx) should be managed by inclusion of IFTB in the TO approach or MTCA.

Sanna et al[2] modified the original transcochlear approach of House and Hitselberger[4] into types A, B, C, and D for various skull base lesions. MTCA is the basic approach, whereas MTCB incorporates the IFTB of Fisch[5] into the MTCA. Posterior rerouting of the facial nerve is associated with the morbidity of facial paresis because of disruption of the deep petrosal artery,[6] whereas the TO approach described by Fisch[5] maintains the facial nerve in its anatomic position, preserving its blood supply. These approaches should be chosen according to the preoperative facial nerve status. When the preoperative facial nerve function is grade I, we use a TO approach, whereas a modified transcochlear approach is chosen when a patient presents with facial paresis.

When a PBC extends to the sphenoid sinus or the rhinopharynx, there is a theoretical possibility of combining an endoscopic rhinological approach with a lateral skull base approach, but there are higher risks of CSF rhinorrhea and meningitis, and hence, we do not advocate this combination.

Once removal of the disease has been achieved,

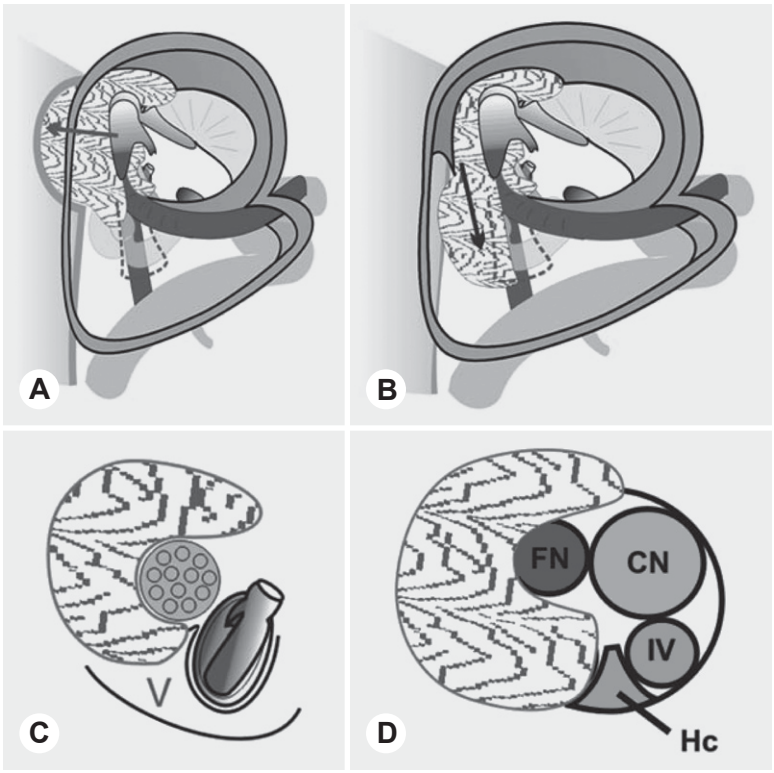

Fig 13. Diagrams of site and degree of involvement of facial nerve. **A)** Supralabyrinthine PBC (arrow) involving facial nerve in second genu, tympanic portion, first genu, and labyrinthine portion. **B)** Similar lesion (arrow) extending into IAC. **C)** Cut section of facial nerve at level of second genu shows PBC engulfing two thirds of circumference of nerve. V — vestibule. **D)** Cross section across IAC shows PBC engulfing two thirds of circumference of facial nerve (FN). CN — cochlear nerve; IV — inferior vestibular nerve; Hc — horizontal crest.

it is useful to carry out an endoscopic examination of the cavity with a 30° rigid endoscope to visualize the hidden areas that might not be accessible with the microscope. Sometimes, epithelium missed by conventional techniques can be found on endoscopic examination. In our experience, this is seldom found, if the correct approach is chosen.

Management of Facial Nerve. In any petrous bone lesions, the facial nerve has an intricate proximity to the lesion, and therefore, it is involved commonly, and often early, in these cases. Management depends on 1) the preoperative status; 2) the degree of facial nerve involvement; and 3) the site and extent of the lesion.

The preoperative status of the facial nerve is one of the most important aspects in planning the approach (ie, TO approach or MTCA). Normal facial nerve function does not rule out the possibility of involvement of the nerve. When the preoperative function is good (grades I and II), the surgeon can be optimistic of achieving a good outcome. In cases with preoperative facial nerve paralysis, the duration since the onset holds the key to the outcome.[7] A duration of less than 12 months would indicate good recovery.[8]

A second important determinant is the degree of involvement, which has to be assessed during operation. The vertical part of the facial nerve has a thicker covering (epineurium and perineurium) that is protective to the nerve as compared to the geniculate ganglion and the horizontal part. The facial nerve is more vulnerable to the disease process around the geniculate ganglion. Axon et al[6] observed a 100% incidence of facial paresis when the nerve was involved at the geniculate ganglion.

Intraoperative removal depends upon the degree of circumferential involvement of the nerve. An uncovered nerve has better results than a nerve enveloped or engulfed by the cholesteatoma.

The site of involvement (Fig 13) is important. The labyrinthine region is thin and has a poor blood supply and is therefore prone to earlier fibrous degeneration than are other sites. In addition, the labyrinthine portion of the facial nerve is delicate. In procedures in which the nerve is replaced by fibrous tissue, excision with a tension-free end-to-end anastomosis or grafting would be desired. The extent of the lesion is important when a larger segment is involved; tension-free end-to-end anastomosis cannot be performed, and hence, grafting would have to be performed after resection. Complete interruption of the nerve by the disease is a frequent finding, especially in massive PBC, which would require nerve grafting. From the treatment point of view, when the nerve is involved in the IAC or proximally, grafting is difficult. In patients with a long duration of facial palsy (more than 12 months), faciohypoglossal anastomosis is indicated.[7,8] When the lesion is medial to a facial nerve, anterior and/or posterior rerouting of

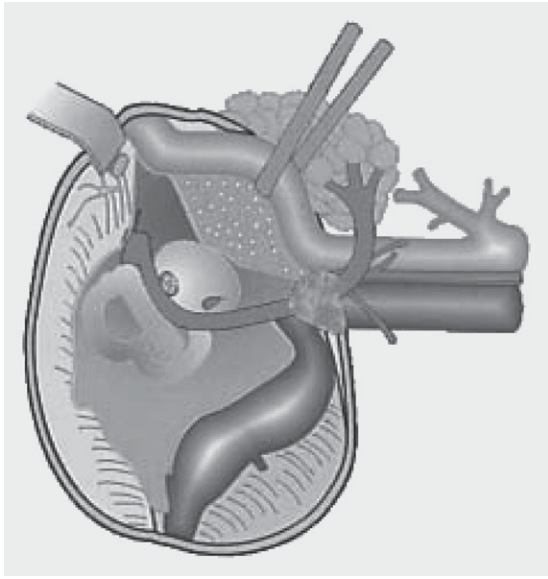

Fig 14. Diagram of mobilization of ICA and exposure achieved by infratemporal fossa approach type B.

the nerve can be performed. For infralabyrinthine lesions around the jugular foramen, a short rerouting as popularized by Glasscock or a long anterior rerouting as described by Hawthorne and Fisch[5] can be used, depending upon the disorder.[9]

Management of Dura. The matrix is often densely adherent to the dura of the middle and posterior fossae, and it might be a challenging task to achieve a radical removal. This is one of the major reasons for recurrences. Use of bipolar coagulation to devitalize the epithelium is the easiest and safest way to ensure complete clearance. Bipolarizing large areas of dura does not lead to dural necrosis if it is carefully performed.[6,8] There is a risk of opening the dura while removing the adherent matrix and causing an intraoperative CSF leak. These openings are usually small and can be managed by use of muscle plugs and obliteration of the cavity.

Management of ICA. Involvement of the ICA could be in its vertical and/or horizontal parts. Selection of the surgical approach is the most crucial aspect of management when involvement is suspected. A TO or modified transcochlear approach would enable control over the vertical portion and part of the horizontal portion. When the lesion involves the horizontal portion, IFTB and MTCB are the approaches of choice. Involvement of the carotid artery is more common in massive and infralabyrinthine-apical PBCs with anterior extension. Axon et al[6] observed that carotid artery involvement is more common in recurrent lesions. A PBC is a nonvascular lesion, which is less aggressive in terms of involvement of the artery. Moreover, the ICA has a thick adventitia that resists dissection of the matrix. Mobilization of the ICA is mandatory if the disease involves the artery for 360° (Fig 14). Dissection of the matrix of the vessel does not pose a difficulty,[6,10] but it should be performed with caution.

Management of Sigmoid Sinus and Jugular Bulb. The sigmoid sinus and the jugular bulb are difficult regions from which to remove the matrix because of their thin, fragile walls, and the lower cranial nerves are at risk of injury. In patients above the age of 65 years, one has to be very careful, as lower cranial nerve palsy in these patients could have grave consequences due to poor compensation. Therefore, in elderly patients, we prefer to leave matrix over the jugular bulb to avoid the risk of lower cranial nerve palsy. In these cases, follow-up is more rigorous. In cases in which clearance has to be achieved, it is imperative to control the internal jugular vein in the neck before dissection of the matrix. Ligation of the internal jugular vein in the neck and sigmoid sinus packing (extraluminal and intraluminal) enable removal of the lateral wall of the dome of the jugular bulb to clear the matrix. In these cases, it is always advisable before operation to ensure the contralateral cerebral venous patency with magnetic resonance angiography.

Management of CSF Leaks. CSF leaks resulting from dural tears do not need special repair, but can be swiftly resolved by inserting free muscle plugs into the subarachnoid space through the defect if the technique of cavity obliteration with abdominal fat and the cul de sac closure of the EAC is performed.

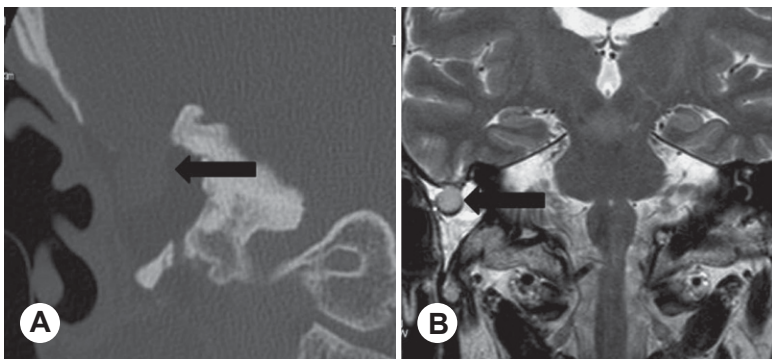

Fig 15. Comparison between **A)** coronal CT scan of temporal bone and **B)** T2-weighted MRI in demonstrating recurrence (arrows). MRI better detects recurrence.

Obliteration of these cavities with autologous abdominal fat is important, as this would protect the exposed area of dura from infection and prevent the problem of a postoperative draining cavity.[10] It also reduces the incidence of postoperative CSF leaks.

The major disadvantage of obliteration of the cavity is that a recurrence may not be detected, and it is mandatory to follow these patients radiologically. Residual lesions are better defined on MRI (Fig 15). We perform high-resolution CT and cerebral MRI (T1- and T2-weighted images with fat suppression) with gadolinium enhancement in suspected cases every year for at least 5 years.

CONCLUSIONS

1. The choice of surgical approach is fundamental, especially in dealing with complex PBC cases, and it must be based on a proper classification depending upon the extent of the disease.

2. Transotic and modified transcochlear approaches hold the key to complete clearance.

3. For extensions into the clivus, sphenoid sinus, or rhinopharynx, an infratemporal fossa approach should be combined with a transtemporal approach.

4. Carotid artery and jugular bulb involvement should be known before operation, and the lesions should be approached accordingly to ensure complete control over these vessels.

5. Obliteration is the solution to the problems related to a large cavity.

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