Chapter 10

Petrous Bone Cholesteatoma

10

10 Petrous Bone Cholesteatoma

Abstract

Petrous bone cholesteatomas (PBCs) are slow-growing expansile epidermoid lesions arising in the petrous portion of the temporal bone. In such cases, the first symptoms are facial nerve paralysis, vertigo, and deaf ear due to invasion of the facial nerve and labyrinth. Otoscopy may be irrelevant or only demonstrates pars flaccida perforation or an open mastoid cavity with evidence of suppurative discharge. Radiological examinations (computed tomography and magnetic resonance imaging scans) are fundamental to evaluate the extension of the lesion and to determine the surgical management. The classification proposed by Sanna divides PBCs into five groups based on the relationship of the disease to the labyrinthine block (supralabyrinthine, infralabyrinthine, infralabyrinthine-apical, massive, apical types). This radiological classification allows standardization in reporting and a clear planning of the surgical approach, which has to be suitable for a safer management of the facial nerve, dura, internal carotid artery, sigmoid sinus, and jugular bulb.

Keywords: petrous bone cholesteatoma, facial nerve, Sanna classification, transotic approach, transcochlear approach

Petrous bone cholesteatomas (PBCs) are slow-growing expansile epidermoid lesions arising in the petrous portion of the temporal bone with an incidence of 4 to 9% of all petrous pyramid lesions. These could be congenital, acquired, or iatrogenic. Congenital PBCs are most plausibly explained by the persistence of fetal epidermoid formation in the petrous bone or the middle ear from which it expands to the petrous bone. In such cases, the first symptoms are facial nerve paralysis, vertigo, and deaf ear due to invasion of the facial nerve and labyrinth. The acquired variety is due to the migration of squamous epithelium into the petrous bone secondary to a perforation in the tympanic membrane. The iatrogenic variety is due to the implantation of cholesteatoma after an otologic surgery. Fetid otorrhea, progressive facial palsy, vertigo, and hearing loss of any type (conductive, sensorineural, mixed) are usually encountered in these patients.

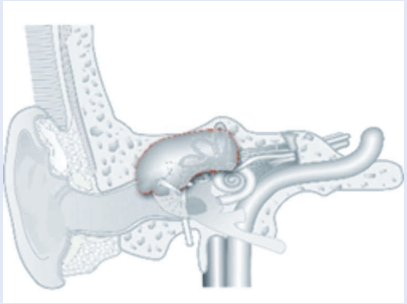
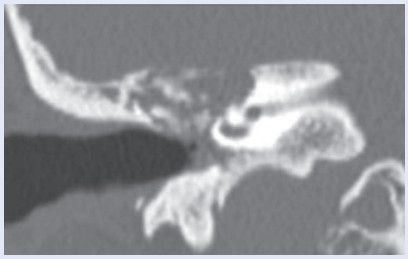
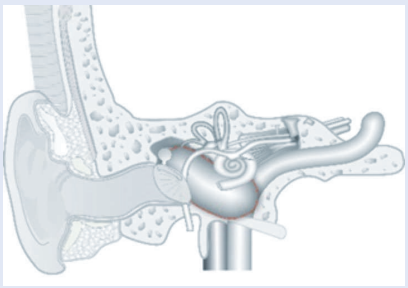
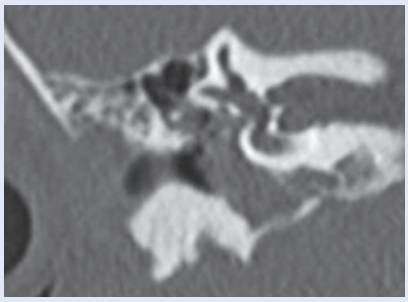
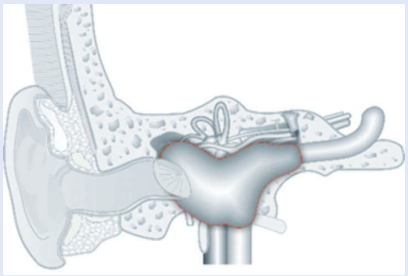
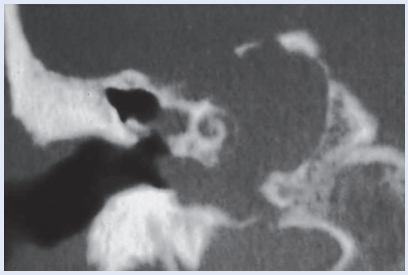
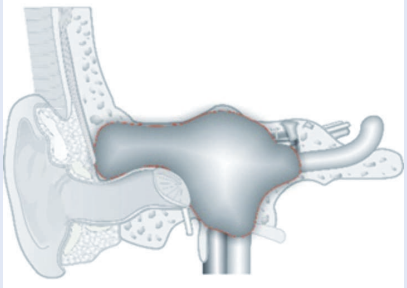
Otoscopy may be irrelevant or only demonstrates pars flaccida perforation or an open mastoid cavity with evidence of suppurative discharge. A computed tomography (CT) scan and magnetic resonance imaging (MRI) are fundamental to evaluate the extension of the lesion and to determine the surgical management.

The rarity of these lesions, slow and silent growth pattern, their complex location in the skull base, proximity to vital neurovascular structures (facial nerve, internal carotid artery, sigmoid sinus, jugular bulb, lower cranial nerves, dura), and tendency to recur make PBCs very challenging to diagnose and treat. PBCs have shown to be locally aggressive by involving the petrous bone and the areas surrounding it, such as the clivus, nasopharynx, sphenoid sinus, and the infratemporal fossa, and even extending intradurally. Also, the close proximity of the disease to the labyrinth and the facial nerve puts to risk both hearing and facial nerve function, which is reflected in the high incidence of facial nerve palsy (34.6–100%) seen in the important series reported in literature.

Surgery remains the mainstay of treatment of PBCs. The choice of surgical approach has evolved from radical petromastoid exenteration with marsupialization of the cavity to closed and obliterative techniques following complete eradication Advancements in neuroradiology and microscopic lateral skull base surgery have made it possible today to completely extirpate these lesions safely with minimal recurrences and perioperative morbidity. The primary objective in surgical approaches for PBCs is to ensure total disease eradication along with complete control and safety of the surrounding important neurovascular structures. The development of the transotic and transcochlear approaches, combined with various other skull base approaches, has helped achieve both these objectives and is considered the mainstay of surgery for PBCs.

The classification proposed by Sanna divides PBCs into five groups based on the relationship of the disease to the labyrinthine block. This radiological classification allows standardization in reporting and a clear planning of the surgical approach (▶ Table 10.1, ▶ Table 10.2). Also see ▶ Fig. 10.1, ▶ Fig. 10.2, ▶ Fig. 10.3, ▶ Fig. 10.4, ▶ Fig. 10.5, ▶ Fig. 10.6, ▶ Fig. 10.7, ▶ Fig. 10.8, ▶ Fig. 10.9, ▶ Fig. 10.10, ▶ Fig. 10.11, ▶ Fig. 10.12, ▶ Fig. 10.13, ▶ Fig. 10.14, ▶ Fig. 10.15, ▶ Fig. 10.16, ▶ Fig. 10.17, ▶ Fig. 10.18, ▶ Fig. 10.19, ▶ Fig. 10.20, ▶ Fig. 10.21, ▶ Fig. 10.22, ▶ Fig. 10.23, ▶ Fig. 10.24, ▶ Fig. 10.25, ▶ Fig. 10.26, ▶ Fig. 10.27, ▶ Fig. 10.28, ▶ Fig. 10.29, ▶ Fig. 10.30, ▶ Fig. 10.31, ▶ Fig. 10.32, ▶ Fig. 10.33, ▶ Fig. 10.34, ▶ Fig. 10.35, ▶ Fig. 10.36, ▶ Fig. 10.37, ▶ Fig. 10.38, ▶ Fig. 10.39, ▶ Fig. 10.40, ▶ Fig. 10.41, ▶ Fig. 10.42, ▶ Fig. 10.43, ▶ Fig. 10.44.

Table 10.1 Updated Sanna classification of petrous bone cholesteatomas

Updated Sanna Classification of Petrous Bone Cholesteatomas (2016)		
Class	**Cholesteatoma location**	**Relations, extension, and features**
Class I: Supralabyrinthine 	Centered on the geniculate ganglion area of the FN and the anterior epitympanum 	Superior: Tegmen or dura Inferior: Semicircular canals, apical turns of the cochlea Medial: Limited extension beyond the otic capsule into the petrous apex Lateral: Antrum, epitympanum and further into middle ear Anterior: Horizontal part of the pICA Posterior: Posterior bony labyrinth Features: Usually associated with fistula of the semicircular canals, erosion of tegmen, involvement of the FN
Class II: Infralabyrinthine 	Centered on the infracochlear, infralabyrinthine, and hypotympanic cells 	Superior: Basal turn of the cochlea, vestibule Inferior: Jugular bulb, lower cranial nerves, occipital condyle Medial: Limited extension beyond the otic capsule into the petrous apex Lateral: Hypotympanum and further into middle ear, retrofacial cells Anterior: Vertical and horizontal part of pICA Posterior: Posterior semicircular canal, IAC Features: Fistula of the semicircular canals, erosion of the cochlea, jugular bulb, carotid canal, involvement of the lower cranial nerves
Class III: Infralabyrinthine-Apical 	Involves infralabyrinthine cell tracts extending medially into the petrous apex 	Superior: Basal turn of the cochlea, vestibule Inferior: Jugular bulb, lower cranial nerves, occipital condyle Medial: Extension into the petrous apex, lower clivus, along the greater wing of sphenoid into the foramen spinosum, foramen ovale; may extend up to sphenoid sinus Lateral: Hypotympanum and further into middle ear, retrofacial cells Anterior: Vertical and horizontal part of pICA Posterior: IAC, dura of the posterior cranial fossa (posterolaterally) Features: Fistula of the semicircular canals, erosion of the cochlea, jugular bulb, involvement of the lower cranial nerves, extensive destruction of the carotid canal, involvement of the internal auditory canal
Class IV: Massive 	Centered on the otic capsule 	Superior: Dura of the middle fossa, may extend intradurally Inferior: Hypotympanic cells, infralabyrinthine cells, jugular bulb, lower cranial nerves Medial: Extension into the petrous apex, along the greater wing of sphenoid into the foramen spinosum, foramen ovale; may extend up to sphenoid sinus Lateral: Middle ear, antrum, retrofacial cells Anterior: Vertical and horizontal part of pICA Posterior: IAC, dura of the posterior cranial fossa, may extend intradurally Features: various degrees of destruction of the otic capsule, involvement of FN

▶

Table 10.1 continued

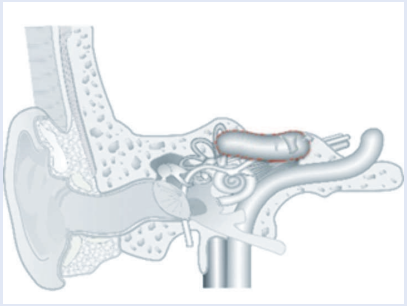
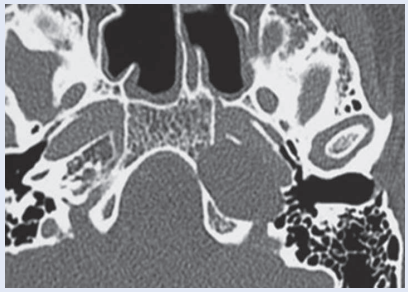
Updated Sanna Classification of Petrous Bone Cholesteatomas (2016)		
Class	**Cholesteatoma location**	**Relations, extension, and features**
Class V: Apical 	Centered on the petrous apex 	Superior: Dura of the middle fossa, Meckel's cave, may extend intradurally Inferior: Hypotympanic cells, infralabyrinthine cells, jugular bulb, lower cranial nerves, infratemporal fossa Medial: Extension into the spheno-petro-clival junction, midclivus, along the greater wing of sphenoid into the foramen spinosum, foramen ovale; may extend up to sphenoid sinus Lateral: Otic capsule Anterior: Horizontal part of pICA and foramen lacerum Posterior: IAC, dura of the posterior cranial fossa, may extend intradurally Features: Otic capsule may be eroded medially, erosion of horizontal petrous carotid, clivus and intradural extensions into middle fossa or posterior fossa; extensions also possible into sphenoid, nasopharynx, or infratemporal fossa

Table 10.2 Updated Sanna subclassification of petrous bone cholesteatomas

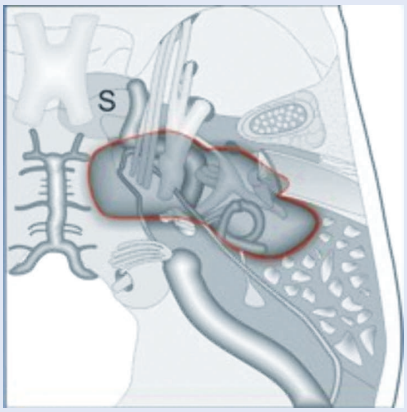
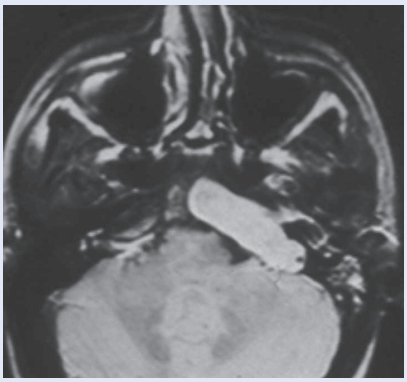
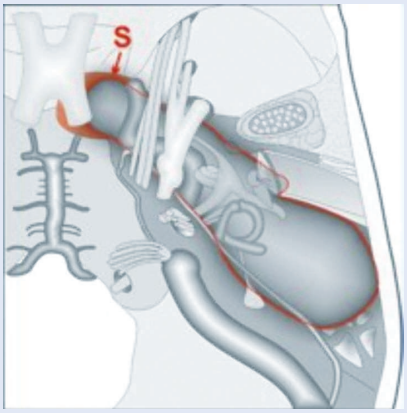
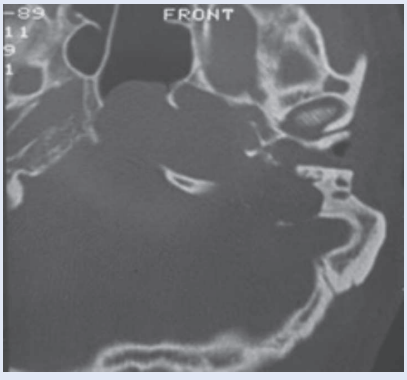
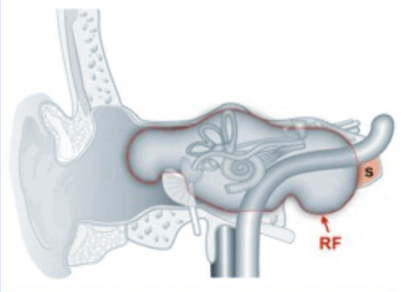
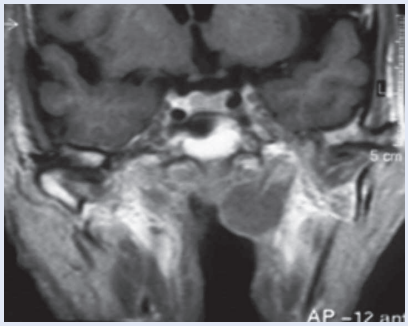
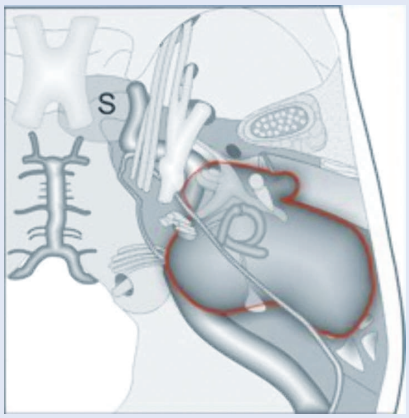
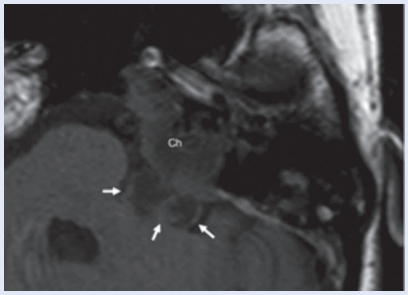
Updated Subclassification of Petrous Bone Cholesteatomas (2016)	
Subclasses	**Relations and features**
Clivus (C) 	Superior and mid clival extensions are seen from massive, infralabyrinthine-apical and apical PBC whereas the lower clival involvement is a feature of infralabyrinthine-apical PBC 
Sphenoid sinus (S) 	Sphenoid sinus involvement is seen from anteromedial extensions of massive, infralabyrinthine-apical and apical PBC; it is a rare extension 
Nasopharynx (N)	It is the rarest extension of the PBC; it is an extension of infralabyrinthine-apical or massive PBC,

Table 10.2 continued

Updated Subclassification of Petrous Bone Cholesteatomas (2016)	
Subclasses	**Relations and features**
	which may extend through the clivus beneath the sphenoid sinus into the nasopharynx 
Intradural (I) 	Intradural extensions may arise from the massive, infralabyrinthine-apical and apical PBCs usually into the posterior cranial fossa and rarely into the middle cranial fossa 

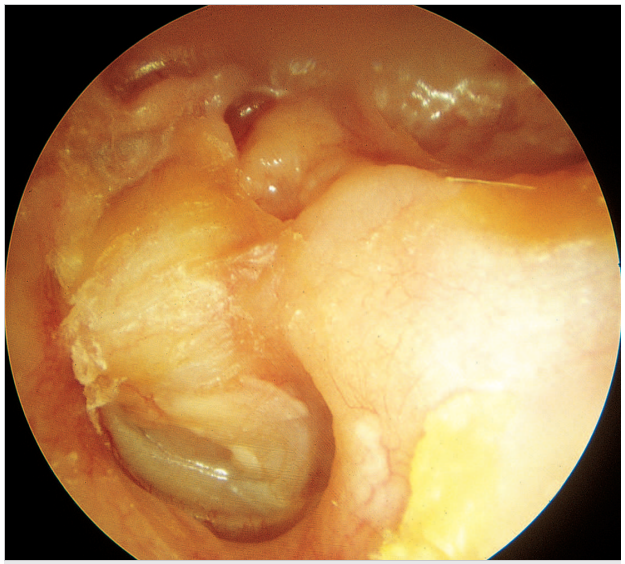


Fig. 10.1 Left acquired or iatrogenic supralabyrinthine petrous bone cholesteatoma in a radical cavity. A whitish retrotympanic mass is seen at the level of the second portion of the facial nerve. The patient presented with progressive facial nerve paralysis and total hearing loss. A correct diagnosis depends not only on otoscopy but also on the symptomatology (facial paralysis, anacusis) and a high-resolution CT scan.

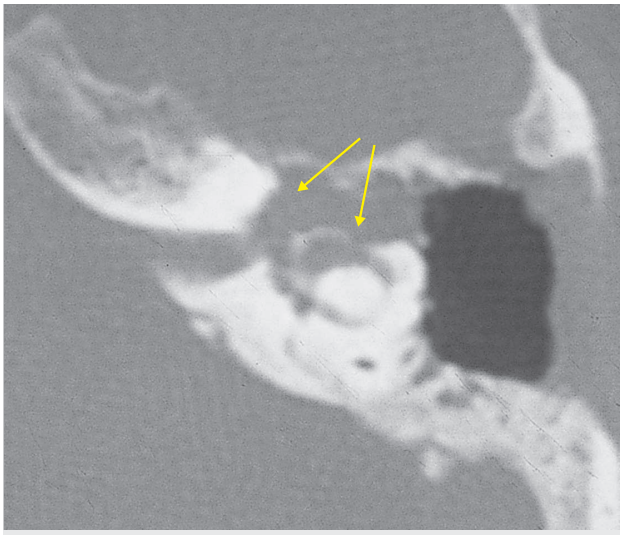


Fig. 10.2 CT scan of the case presented in ▶ Fig. 10.1, axial section. Involvement of the lateral semicircular canal and the vestibule (*arrows*) is well visualized. The cholesteatoma invades the cochlea anteriorly, while medially it reaches the fundus of the internal auditory canal. The posterior semicircular canal is not invaded.

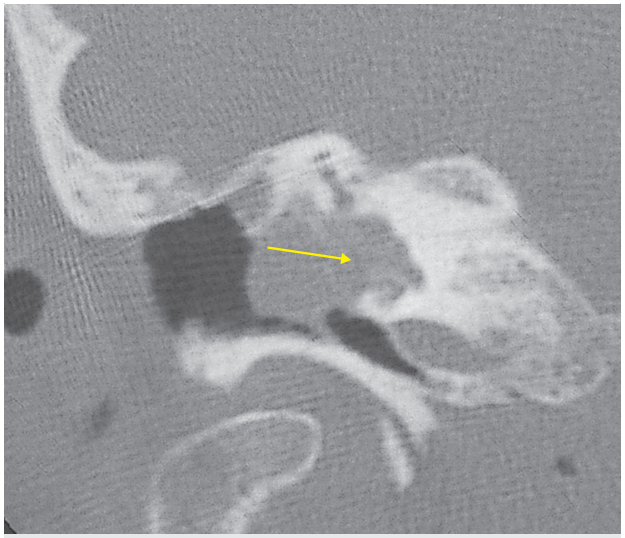


Fig. 10.3 CT scan of the case presented in ▶ Fig. 10.1, coronal section. The medial extension of the cholesteatoma can be appreciated (*arrow*).

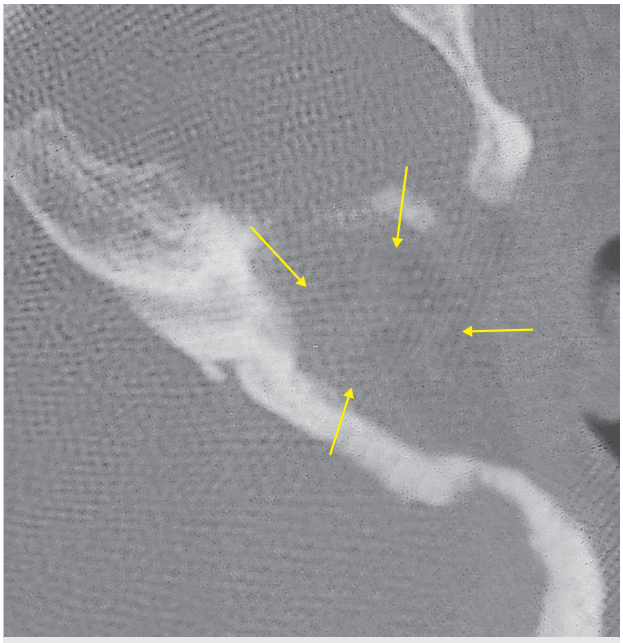


Fig. 10.4 Postoperative CT scan. A transcochlear approach was performed and the operative cavity was obliterated with abdominal fat (*arrows*).

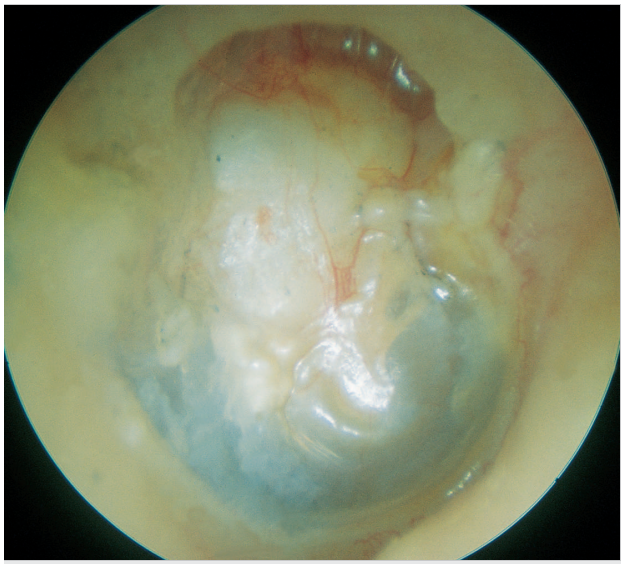


Fig. 10.5 Right acquired supralabyrinthine petrous bone cholesteatoma. A whitish mass is present in the mastoid cavity of an open tympanoplasty. The mass occupies the whole epitympanum and extends interiorly behind the tympanic membrane. The patient presented with ipsilateral facial paralysis and conductive hearing loss.

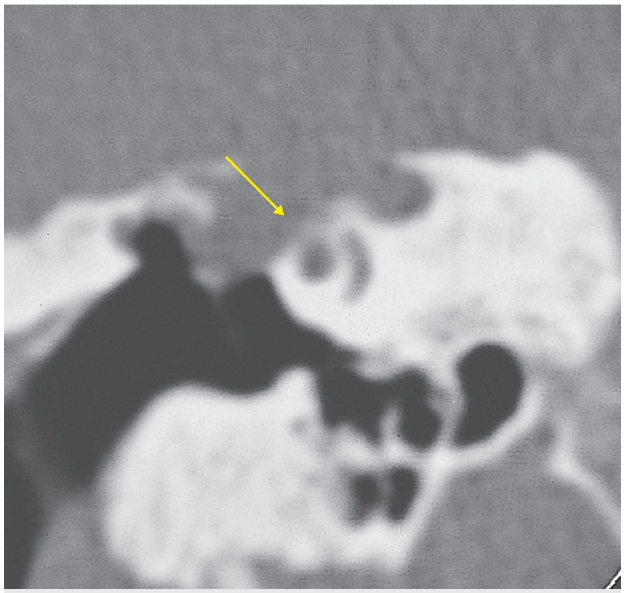


Fig. 10.6 CT scan of the case presented in ▶ Fig. 10.5. The cholesteatoma invades the cochlea (*arrow*). Total removal of the pathology was accomplished using a transcochlear approach with obliteration of the operative defect using abdominal fat. The external auditory canal was closed as cul de sac. The facial nerve was infiltrated at the level of the geniculate ganglion and was repaired using a sural nerve graft.

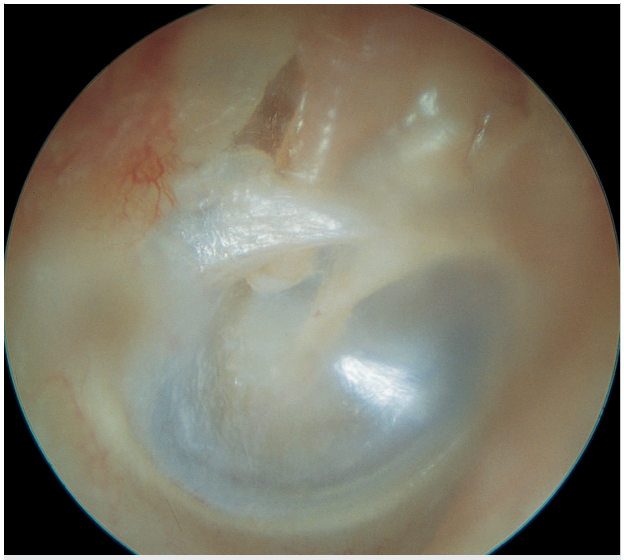


Fig. 10.7 Another example of right acquired supralabyrinthine petrous bone cholesteatoma. The patient presented with right facial nerve paralysis. Otoscopy reveals a right epitympanic erosion.

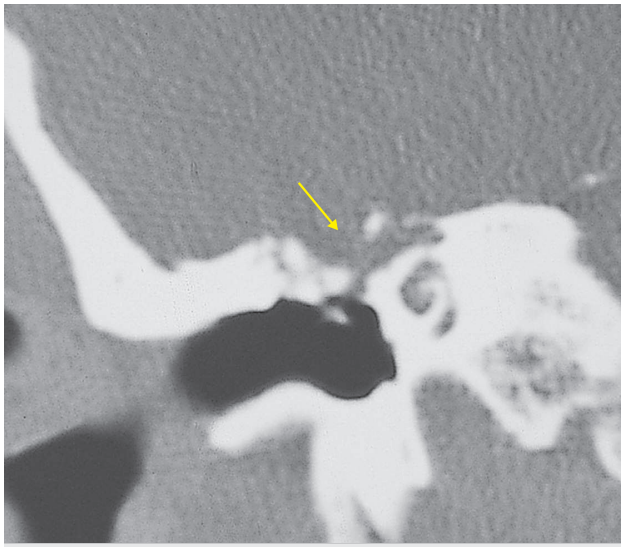


Fig. 10.8 CT scan of the case presented in ▶ Fig. 10.7, coronal view. Typical location and erosion of acquired small supralabyrinthine petrous bone cholesteatoma (*arrow*).

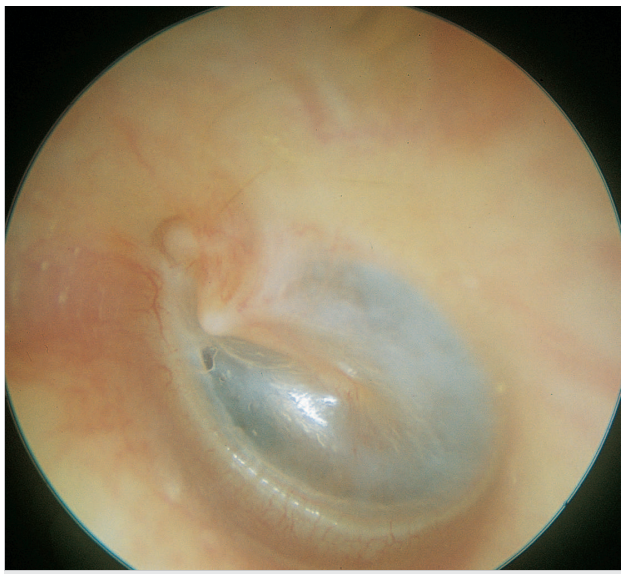


Fig. 10.9 Left congenital supralabyrinthine petrous bone cholesteatoma with extension toward the apex.Otoscopy is negative. The patient complained of progressive facial nerve paralysis of 5 years' duration as well as conductive hearing loss (▶ Fig. 10.10, ▶ Fig. 10.11).

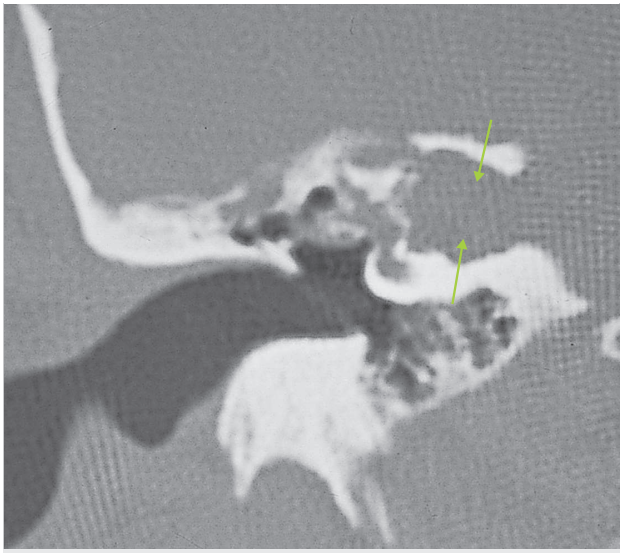


Fig. 10.10 CT scan of the case presented in ▶ Fig. 10.9. Coronal view showing extension of the cholesteatoma into the internal auditory canal (*arrows*).

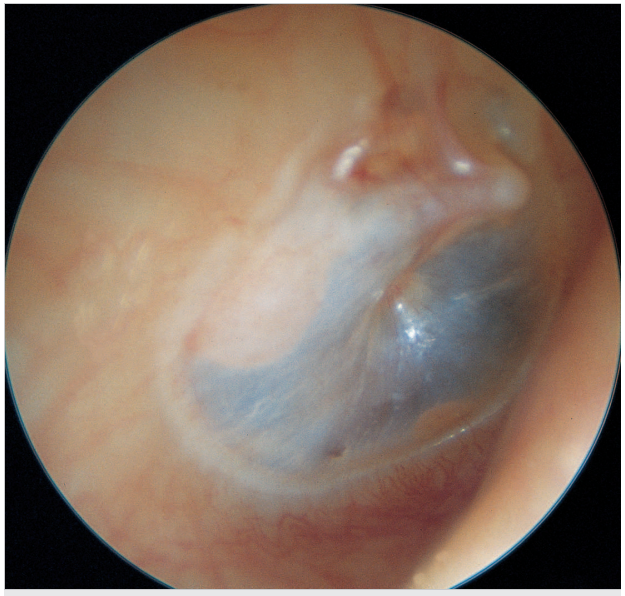


Fig. 10.11 Right congenital infralabyrinthine apical petrous bone cholesteatoma in a 30-year-old female patient. In the posterosuperior quadrant, a white retrotympanic view is observed. The patient had complained of right anacusis since childhood and instability of 1-year duration. The facial nerve was normal.

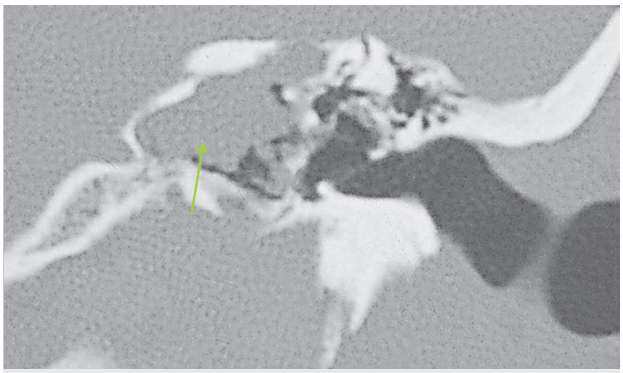


Fig. 10.12 CT scan of the case presented in ▶ Fig. 10.11. Coronal view demonstrating the involvement of the infralabyrinthine apical compartment by the cholesteatoma (*arrow*).

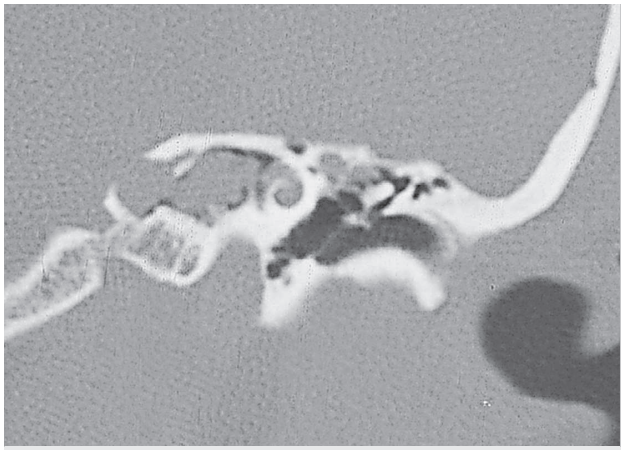


Fig. 10.13 CT scan of the case presented in ▶ Fig. 10.11. A more anterior coronal view at the level of the cochlea.

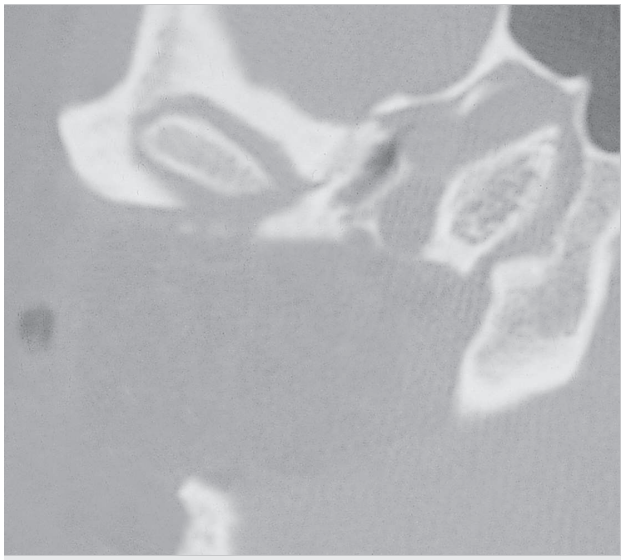


Fig. 10.14 Postoperative CT scan showing total removal of the cholesteatoma through the transcochlear approach and obliteration of the operative cavity using abdominal fat.

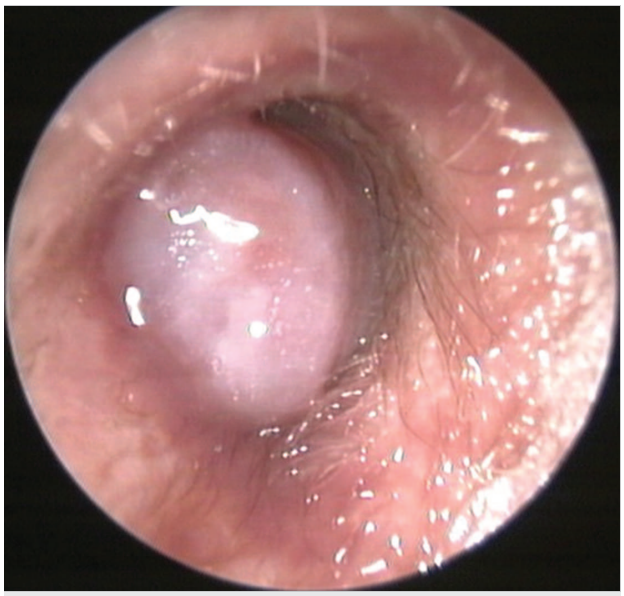


Fig. 10.15 Polyp in the external auditory canal in a patient who had undergone a myringoplasty. The patient presented with otorrhea and sensorineural hearing loss.

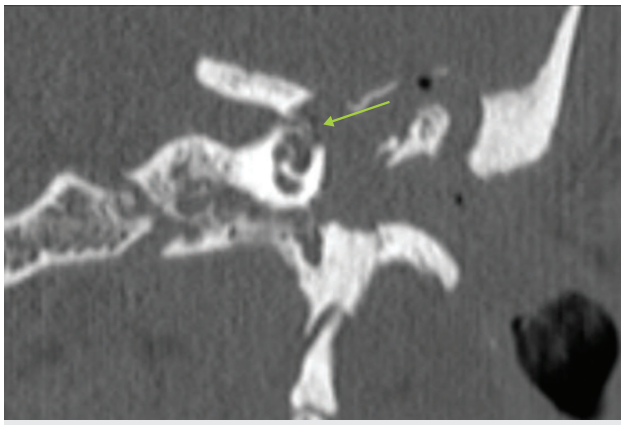


Fig. 10.16 CT scan of the case on ▶ Fig. 10.15, coronal view. A cholesteatoma involving the basal turn of the cochlea is visible (*arrow*).

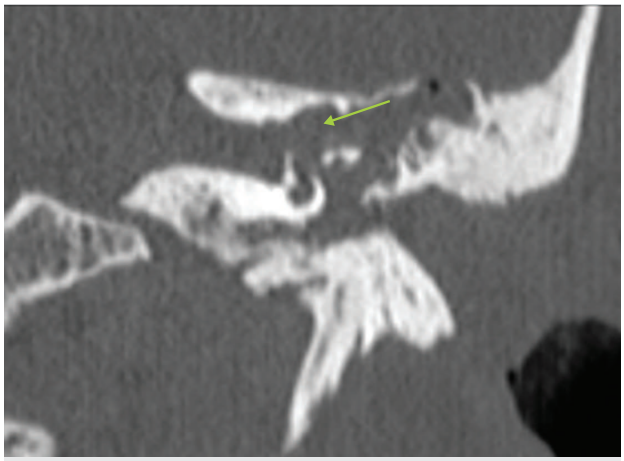


Fig. 10.17 CT scan of the same case, coronal view. The PBC (massive type) involves the labyrinth (*arrow*).

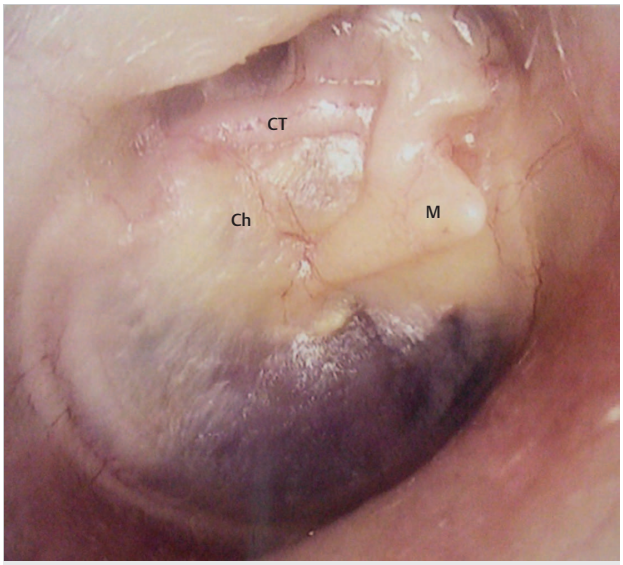


Fig. 10.18 Right ear. This 60-year-old male patient presented with anacusis and facial nerve palsy. The otoscopy showed a dry clean retraction in the attic. Presence of whitish matter behind the tympanic membrane was the only clue for the presence of cholesteatoma. The CT scan (see ▶ Fig. 10.19, ▶ Fig. 10.20) revealed the presence of a supralabyrinthine PBC extended to the petrous apex. A modified transcochlear type A approach was performed. Ch, cholesteatoma; CT, chorda tympanic nerve; M, malleus.

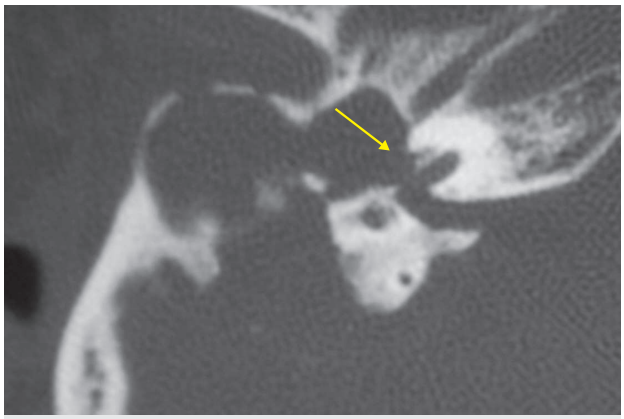


Fig. 10.19 CT scan of the same case, axial view. The cholesteatoma causes erosion of the cochlea and involvement of the facial nerve (*arrow*).

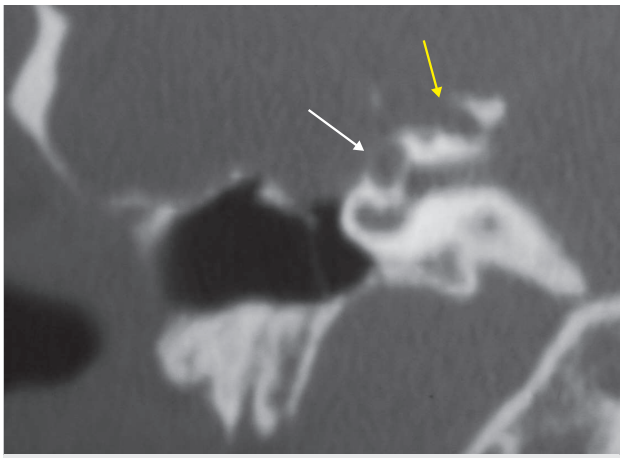


Fig. 10.20 CT scan of the same case, coronal view, shows severe erosion of the otic capsule (*white arrow*) and extensive extension of the cholesteatoma medially to the labyrinth, toward the internal auditory canal (*yellow arrow*).

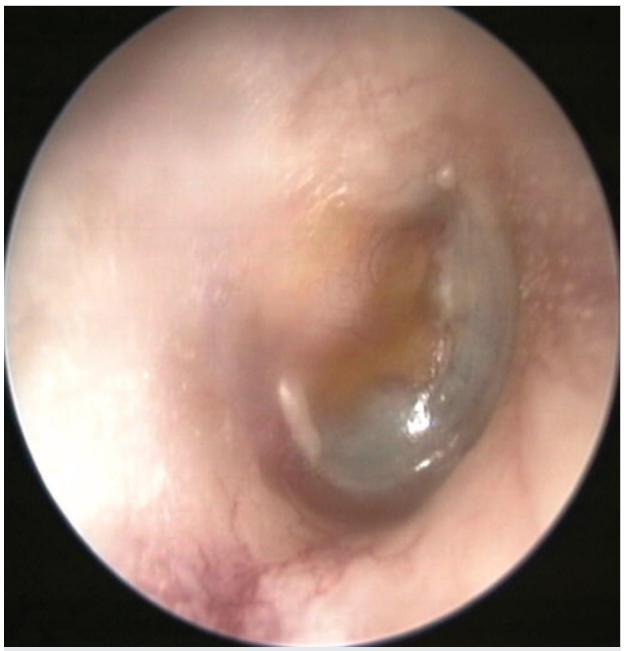


Fig. 10.21 Right ear. This 24-year-old man presented to our institution after two tympanoplasties for cholesteatoma. He complained of persistent otorrhea, severe hearing loss (mixed type), and vertigo. CT scan revealed the presence of a supralabyrinthine PBC with involvement of the semicircular canals. A transotic approach with preservation of the cochlea was performed. Facial nerve was grade I House–Brackmann scale after surgery.

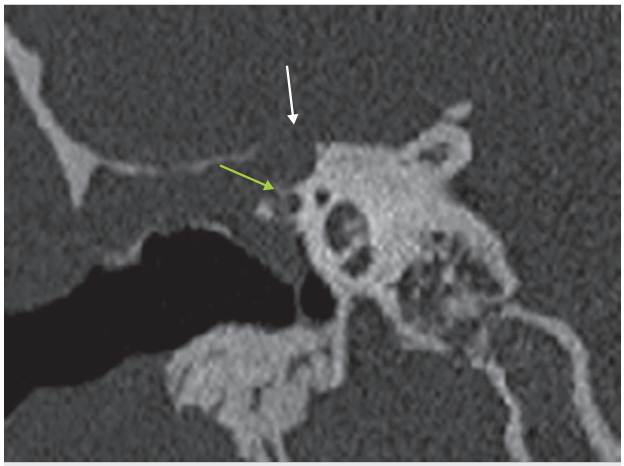


Fig. 10.22 CT scan, coronal view. The cholesteatoma involves the supralabyrinthine compartment, causing erosion of the middle fossa dura plate (*white arrow*). The cochlea is not invaded by the disease, while the second portion of the facial nerve is in close contact with the pathology (*green arrow*). However, facial nerve function is normal.

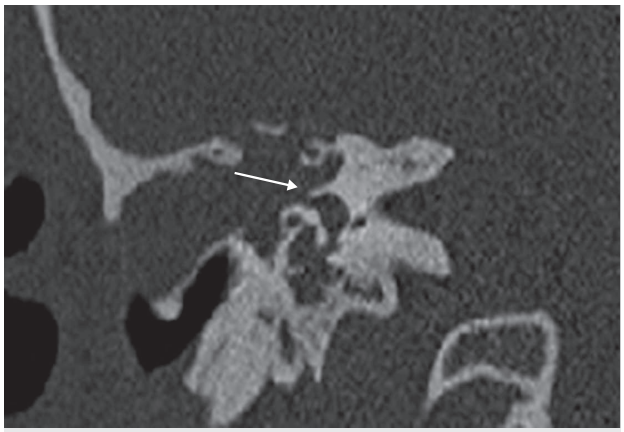


Fig. 10.23 CT scan, coronal view. The cholesteatoma involves the lateral semicircular canal (*arrow*).

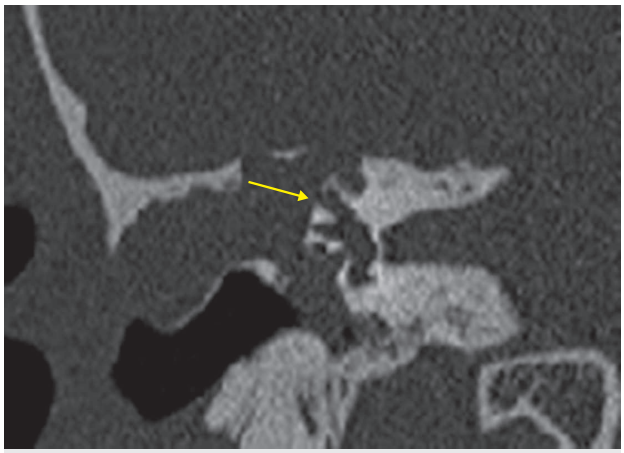


Fig. 10.24 CT scan, coronal view. The cholesteatoma involves the superior semicircular canal (*arrow*).

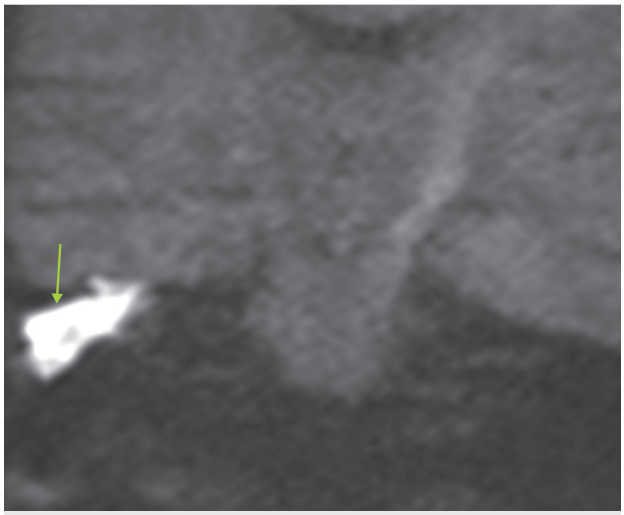


Fig. 10.25 MRI scan, diffusion-weighted image (DWI), coronal view. The cholesteatoma is typically hyperintense in this sequence (*arrow*). The DWI technique is particularly important in the follow-up, because it allows the detection even of small (> 2 mm) residual/recurrent disease.

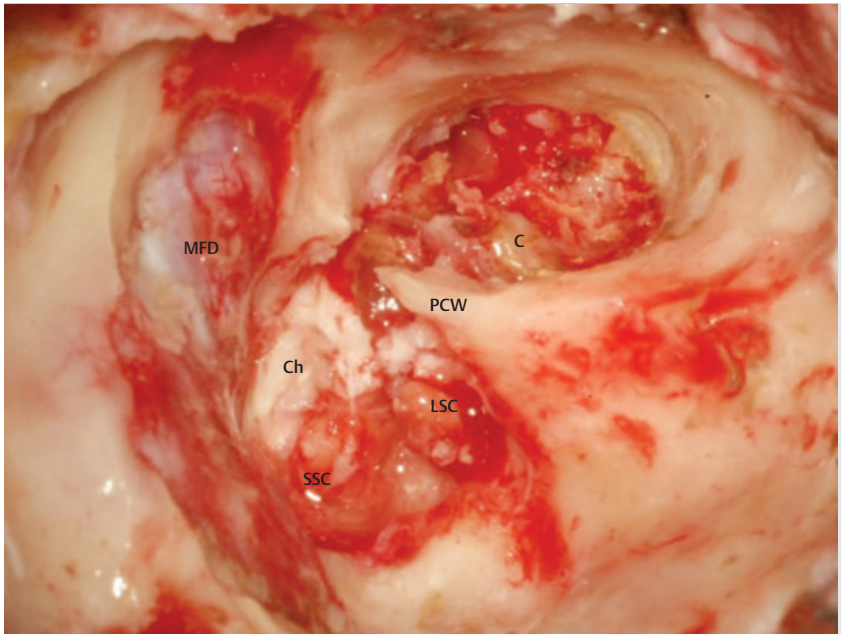


Fig. 10.26 Intraoperative picture of the same case. Blind sac closure of the external auditory canal, removal of the tympanic membrane and the ossicles, and mastoidectomy have been performed. The cholesteatoma (Ch) involves both the lateral (LSC) and the superior semicircular canal (SSC). C, cochlea; MFD, middle fossa dura; PCW, posterior canal wall.

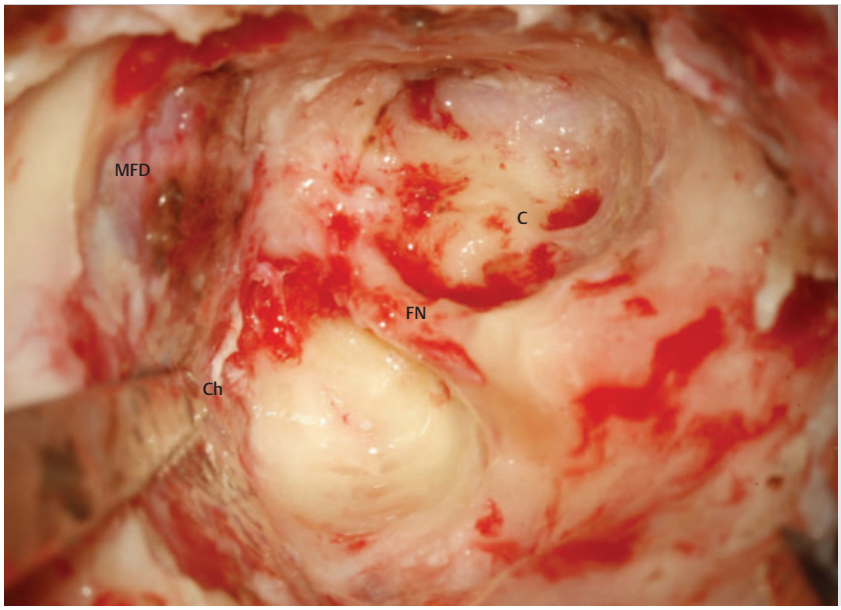


Fig. 10.27 Surgical view after labyrinthectomy. There is still some cholesteatoma matrix on the middle fossa dura, which is gently elevated with a retractor. The third portion of the facial nerve is completely skeletonized. After having removed all the cholesteatoma, the cavity is obliterated with abdominal fat. C, cochlea; Ch, cholesteatoma; FN, facial nerve; MFD, middle fossa dura.

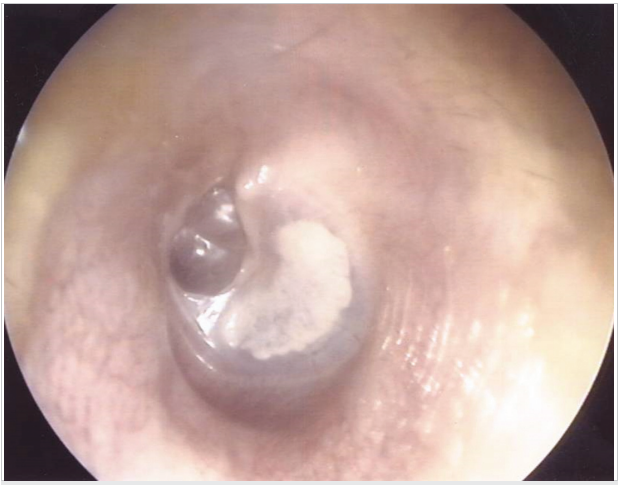


Fig. 10.28 Left ear. Otoscopy showed myringosclerosis of the posterior quadrants and a retraction pocket on the anterior quadrant. The patient complained of left progressive hearing loss evolved in anacusis and facial nerve paralysis for the last 2 years. A CT scan revealed the presence of a PBC on both sides: massive type on the left side (see ▶ Fig. 10.29, ▶ Fig. 10.30) and supralabyrinthine type on the right side (▶ Fig. 10.32). Surgery on this side was performed through a transcochlear approach.

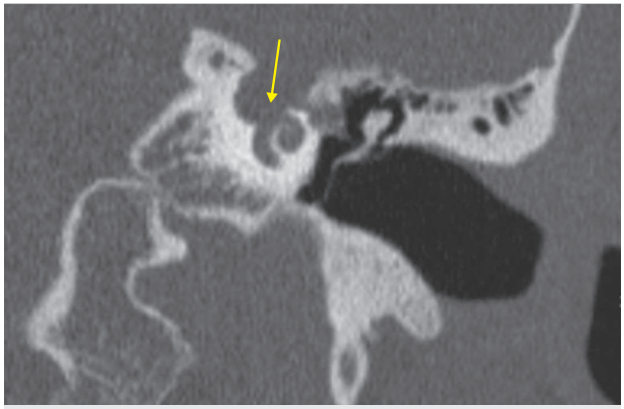


Fig. 10.29 CT scan of the same case, coronal view. The cholesteatoma has eroded the basal turn of the cochlea (*arrow*).

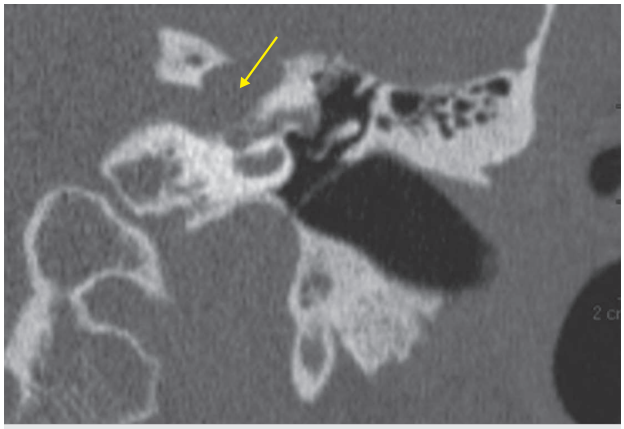


Fig. 10.30 CT scan of the same case, coronal view. The cholesteatoma has engulfed the labyrinth, going medial toward the roof of the internal auditory canal (*arrow*).

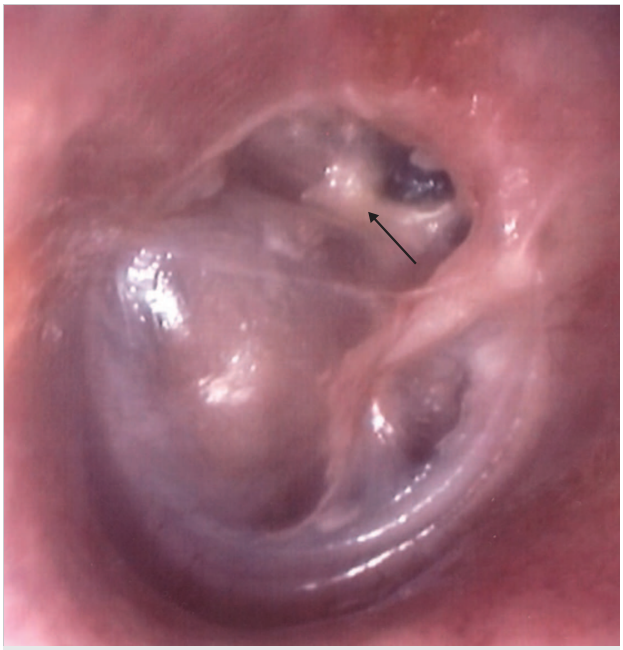


Fig. 10.31 Right ear, same case. The posterior half of the tympanic membrane is scarred, and touched to the promontory. The posterosuperior quadrant goes deep toward the attic, touching the superstructure of the stapes (*arrow*). Hearing function is normal, even though the cholesteatoma has determined a fistula of the lateral semicircular canal (see ▶ Fig. 10.32). A transmastoid + middle fossa approach was chosen with the purpose of preserving right hearing function.

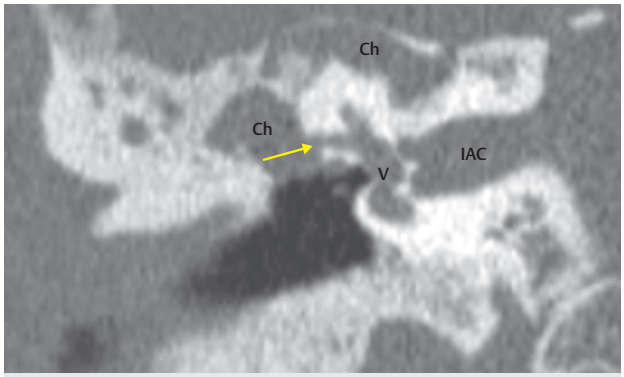


Fig. 10.32 CT scan of the same case, coronal view. A fistula in the lateral semicircular canal is clearly seen (*arrow*). The cholesteatoma infiltrates medially along the middle fossa plate to reach the petrous apex. Ch, cholesteatoma; IAC, internal auditory canal; V, vestibule.

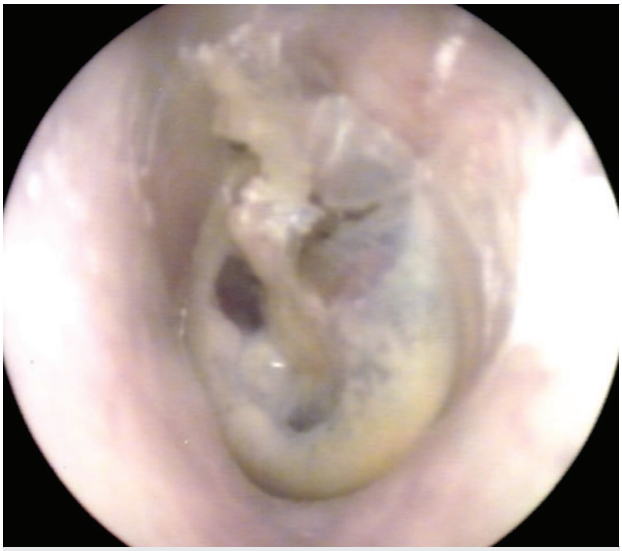


Fig. 10.33 Left ear. Otoscopy shows diffuse myringosclerosis of the tympanic membrane with an anterosuperior retraction pocket. This patient underwent two myringoplasties elsewhere. The patient complained of left anacusis and facial nerve palsy (both lasting for 5 years). Lower cranial nerves paralysis has developed since 2 months before our consultation. CT and MRI scans revealed the presence of a massive PBC that extended intracranially and toward the clivus and sphenoid sinus. A single-stage modified transcochlear approach was performed for the removal of the disease.

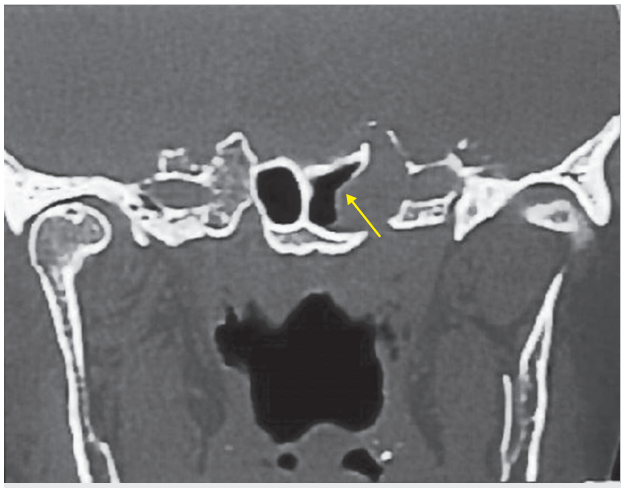


Fig. 10.34 CT scan of the same case, coronal view. The cholesteatoma invades the sphenoid sinus (*arrow*).

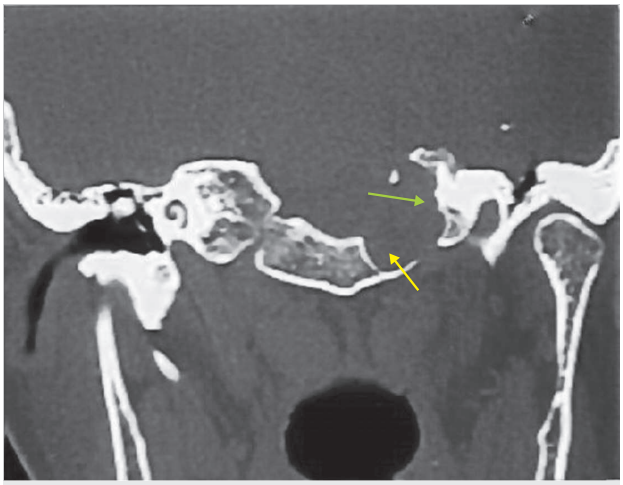


Fig. 10.35 CT scan of the same case, coronal view. The cholesteatoma erodes the clivus (*yellow arrow*). The cochlea is not recognizable due to massive engulfment by the disease (*green arrow*).

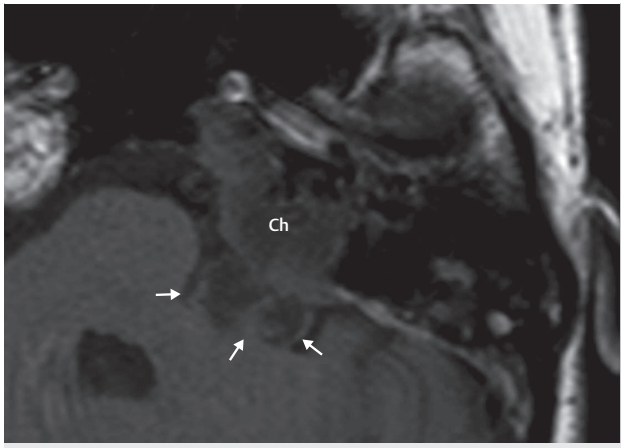


Fig. 10.36 MRI of the same case (T1W), axial view. The cholesteatoma causes compression of the brain stem protruding from the petrous apex (*arrows*). Ch, cholesteatoma.

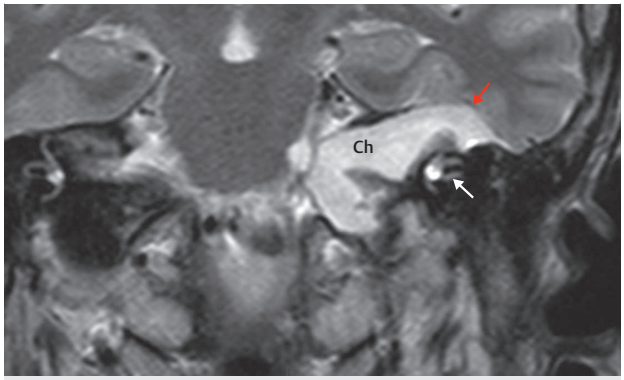


Fig. 10.37 MRI of the same case (T2W), coronal view, which shows a large petrous apex lesion elevating the middle fossa dura to compress the temporal lobe (*red arrow*). The vestibule and the semicircular canal can be identified (*white arrow*). Ch, cholesteatoma.

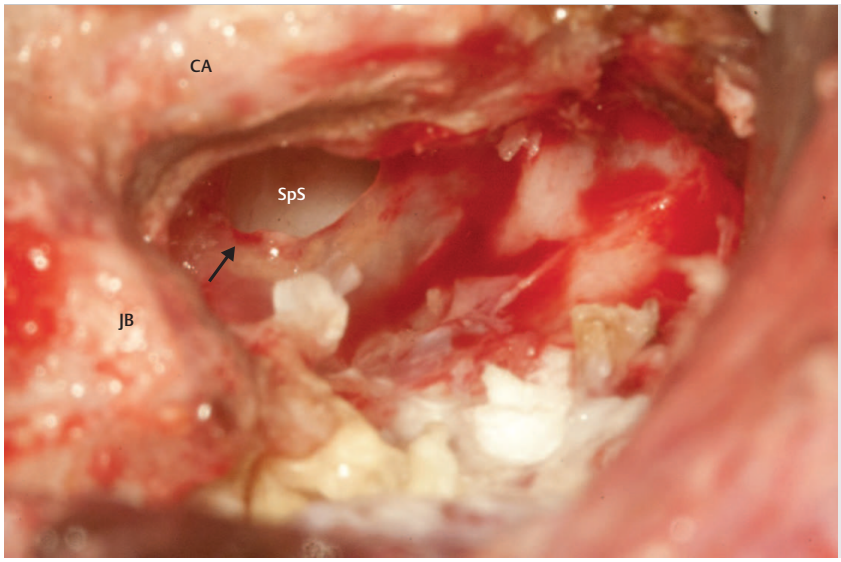


Fig. 10.38 Intraoperative picture. The cholesteatoma has been nearly completely removed from the area of the sphenoid sinus. The opening of sphenoid sinus is seen in the petroclival area medially to the carotid artery (*arrow*). CA, carotid artery; JB, jugular bulb; SpS, sphenoid sinus.

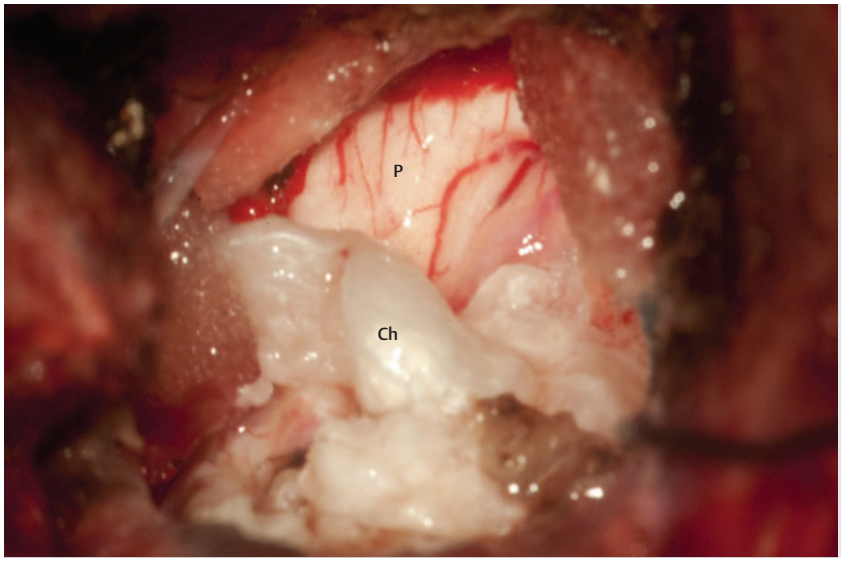


Fig. 10.39 Intraoperative picture. The intradural portion of the cholesteatoma attached to the pons is shown. Ch, cholesteatoma; P, pons.

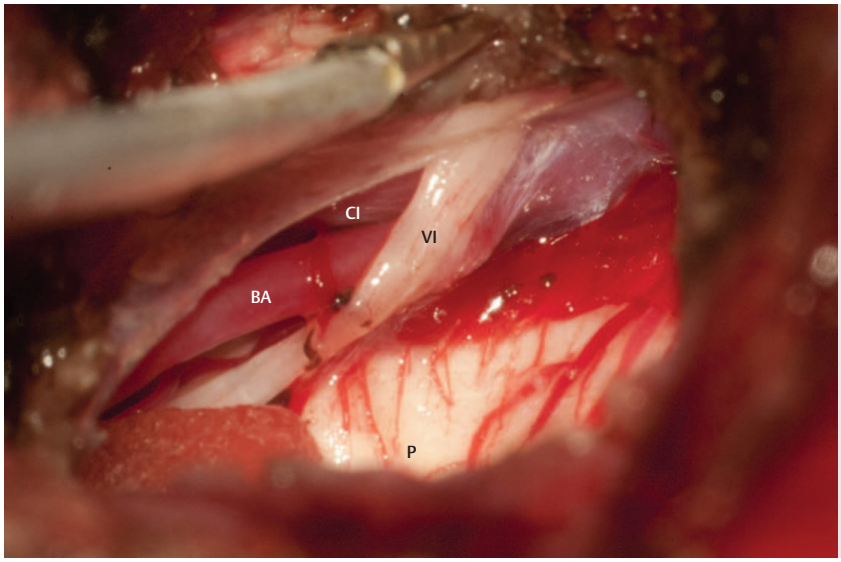


Fig. 10.40 Intraoperative picture. The majority of cholesteatoma is removed from the cerebellopontine angle. BA, basilar artery; Cl, clivus; P, pons; VI, abducens nerve.

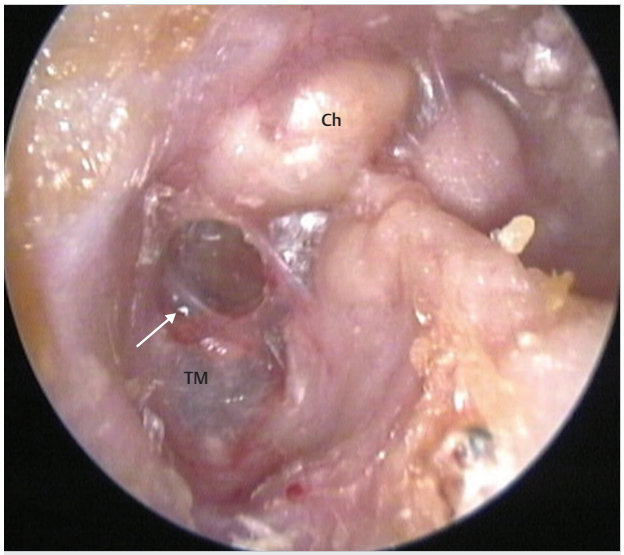


Fig. 10.41 A case of residual cholesteatoma in an open cavity is shown. The patient underwent cholesteatoma surgery with canal wall down technique elsewhere. Residual cholesteatoma (Ch) is seen at the tegmen of the cavity, just superiorly to the tympanic membrane. A deep dimple (*arrow*) is seen in the superior part of the tympanic membrane (TM), and the facial ridge remains very high.

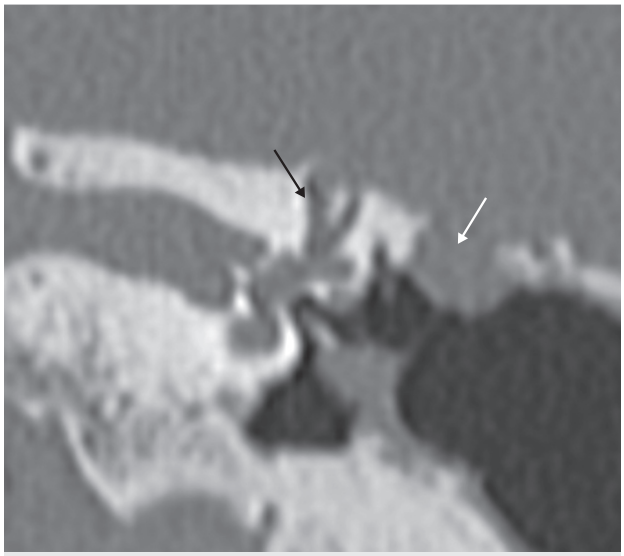


Fig. 10.42 CT scan of the same case, coronal view, shows the presence of soft tissue eroding the tegmen of the antrum (*white arrow*). The cholesteatoma infiltrates medially to the superior semicircular canal eroding the ampulla (*black arrow*).

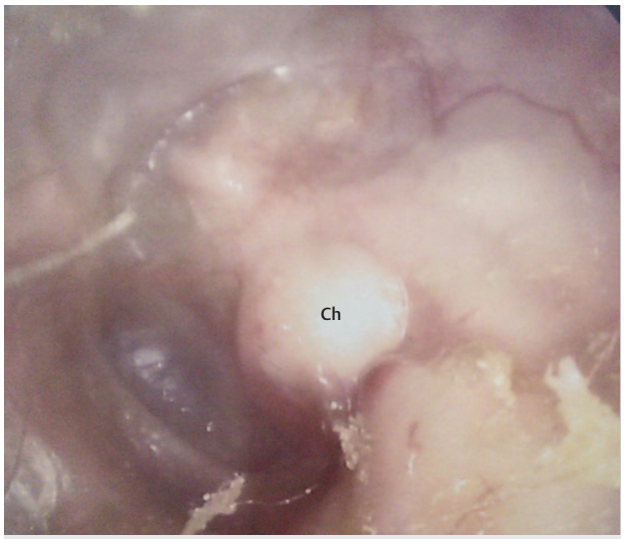


Fig. 10.43 A case similar to that of ▶ Fig. 10.41. Residual cholesteatoma in a canal wall down tympanoplasty. Otoscopy shows a whitish mass in the canal wall cavity located beneath the posterosuperior quadrant of the tympanic membrane. Ch, cholesteatoma.

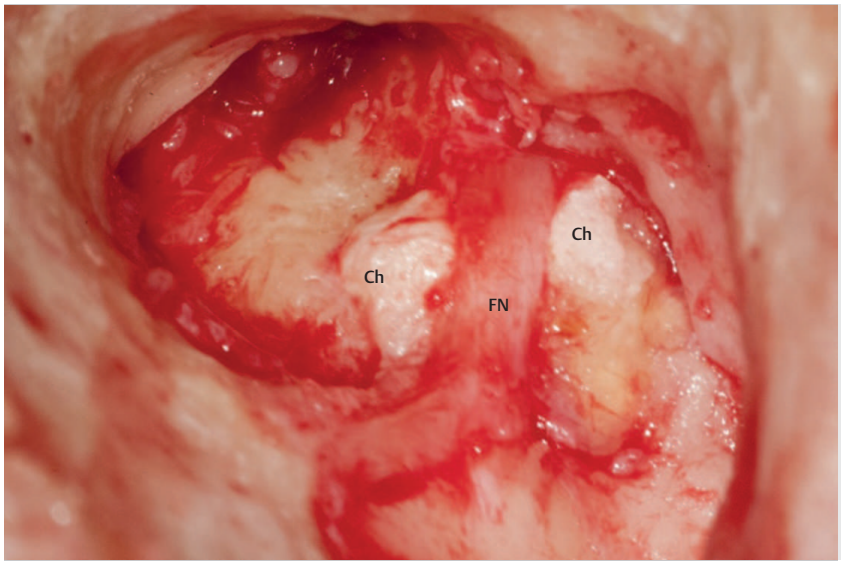


Fig. 10.44 Intraoperative picture. The cholesteatoma is medial to the facial nerve and involves both the cochlea and the labyrinth. A transotic approach was performed, thus leaving the facial nerve in situ. Ch, cholesteatoma; FN, facial nerve.

10.1 Surgical Management

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 aims at preoperatively diagnosing the extension of PBC beyond the temporal bone (clivus, sphenoid sinus rhinopharynx, intradural space). 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 disease and preoperative facial nerve function. The approach is chosen depending on the type of PBC and its extent, which should be determined according to the CT scan and MRI findings.

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 cerebrospinal fluid (CSF) leak and meningitis, (4) cavity obliteration, and (5) hearing preservation whenever feasible.

In supralabyrinthine PBC, 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. In infralabyrinthine PBC, bone conduction can be preserved with subtotal petrosectomy and blind sac closure of the external auditory canal with cavity obliteration. Hearing preservation is usually not possible in infralabyrinthine-apical and massive PBC; 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, rhinopharynx, and intradural space depending on the type used. The posterior rerouting of the facial nerve carries the disadvantage of postoperative facial paresis. Therefore, we prefer a transotic approach when facial nerve function is normal. Whenever the cholesteatoma involves the apical portion of the temporal bone or when it extends further to the clivus, sphenoid sinus, or rhinopharynx, an infratemporal fossa approach type B or modified transcochlear approach type A is incorporated into the transotic approach depending on the preoperative status of the facial nerve.

10.1.1 The Transotic and Modified Transcochlear Approaches

The original transcochlear approach described by House and Hitselberger (1976) includes identification of the internal auditory canal, posterior rerouting of the facial nerve, and removal of the cochlea and petrous apex with preservation of the middle ear and external auditory canal.

Fisch (1978) described the transotic approach in which he removed the external auditory canal and middle ear but keeps the facial nerve in situ.

Our modified transcochlear approach, on the other hand, combines the removal of the external auditory canal and middle ear with the posterior rerouting of the facial nerve, thus removing the major impediment to anterior extension of the approach. This allows better control of the vertical and horizontal intrapetrous internal carotid artery and facilitates the total removal of the petrous apex. The extensive anterior bone removal provides an excellent control of the ventral surface of the brainstem without cerebellar and brainstem retraction. We have further classified this approach into four types according to the extension. Type A represents the basic approach; types B, C, and D denote the anterior, superior, and inferior extensions, respectively.

The transotic and modified transcochlear approaches have been shown in ▶ Fig. 10.45 and ▶ Fig. 10.46.

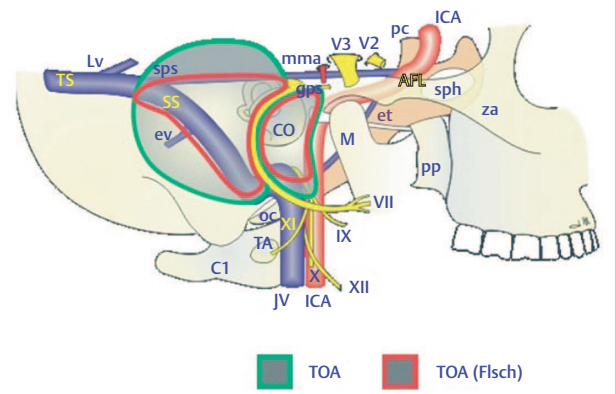


Fig. 10.45 A schematic drawing showing the outlines of the transotic approach. C1, first cervical vertebra; CO, cochlea; ET, Eustachian tube; EV, emissary vein; FN, facial nerve; IAC, internal auditory canal; ICA, internal carotid artery; IX, glossopharyngeal nerve; JV, jugular vein; Lv, Labbé's vein; M, mandible; MMA, middle meningeal artery; OC, occipital condyle; pc, posterior clinoid; pp, pterygoid process; SS, sigmoid sinus; SPH, sphenoid; TA, transverse process of the atlas; TS, transverse sinus; V2, second branch of the trigeminal nerve; V3, third branch of the trigeminal nerve; VII, facial nerve; XII: Hypoglossal nerve, XI, accessory nerve; ZA, zygomatic process.

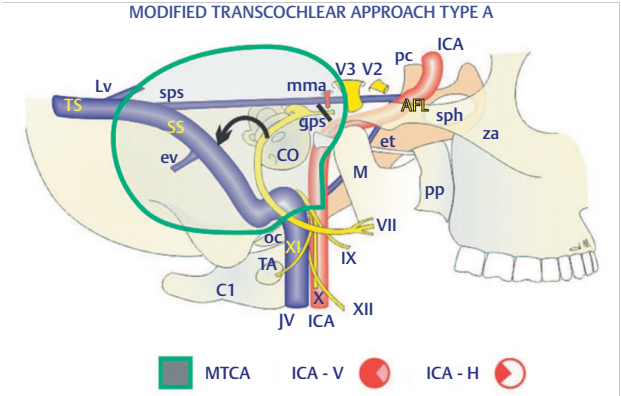


Fig. 10.46 A schematic drawing showing the outlines of the modified transcochlear approach type A. About three-fourths of the vertical internal carotid artery (ICA-V) are under control in contrast to only one-fourth of the horizontal internal carotid artery (ICA-H) using this approach.

Indications of the Transotic Approach

- Some cases of cerebellopontine angle tumors with anterior extension and preoperative normal facial nerve function (i.e., epidermoids).
- PBCs.
- Some cases of petrous bone tumors with preoperative normal facial nerve function.

Indications of the Modified Transcochlear Approach (Type A—Basic Type)

- *Extradural lesions*: extensive petrous bone apex lesions with preoperative facial nerve and inner ear compromise, that is:
  - PBC of the massive infralabyrinthine apical and supralabyrinthine types.
  - Recurrent acoustic neurinoma with petrous bone invasion.
  - Extensive facial nerve tumors.
  - Cholesterol granuloma (only with preoperative facial and hearing impairment).
- Intradural lesions:
  - Large clival and petroclival lesions lying ventral to the brainstem, that is, petroclival meningiomas.
  - Previously irradiated petroclival meningiomas.
  - Residual or recurrent nonacoustic lesions of the posterior fossa with anterior extension into the prepontine cistern, particularly those with encasement of the vertebrobasilar artery or perforating arteries, or both, that is, huge posterior fossa epidermoids
  - Recurrent acoustic neurinomas with facial nerve paralysis.
- *Transdural lesions* invading the petrous apex as en plaque meningiomas, or primary clival or temporal bone lesions with secondary posterior fossa extension as chordomas, chondrosarcomas, and extensive glomus jugulare tumors.
- Some cases of epidermoids.

Surgical Technique

A wide **C**-shaped postauricular incision is made. The incision starts 3 cm above the auricle, curves posteriorly to approximately 4 to 5 cm posterior to the auricular sulcus, and ends inferiorly at the level of the mastoid tip. The skin and subcutaneous tissues are elevated and the muscoloperiosteal layer is incised in a **T**-shaped pattern. A small Palva flap based anteriorly is outlined.

The external auditory canal is transected and closed as a cul de sac. The anterior skin flap is retracted using skin hooks, whereas the muscoloperiosteal layer is raised and is kept retracted using 1/0 silk sutures.

An extended mastoidectomy is performed, with removal of bone 2 to 3 cm posterior to the sigmoid sinus and over the middle fossa dura. The external auditory canal (posterior and superior walls) is drilled and the facial nerve is skeletonized. The inferior tympanic bone is also drilled.

The labyrinthectomy is performed and the internal auditory canal is identified. Bone superior and inferior to the internal auditory canal is drilled, creating two deep troughs around the canal.

The retrofacial air cells are drilled. The anterior wall of the external auditory canal and the tympanic bone are thinned out. The internal carotid artery is identified in front of the cochlea.

The cochlea is drilled. The internal carotid artery is better identified. All the bone inferior and superior to the internal auditory canal is completely removed.

In case of a modified transcochlear approach, the facial nerve is freed from the third to the fist portion and posteriorly rerouted together with all the contents of the internal auditory canal.

For intradural cases, the dura is next opened and the pathology is dealt with.

The surgical technique has been shown in ▶ Fig. 10.47, ▶ Fig. 10.48, ▶ Fig. 10.49, ▶ Fig. 10.50, ▶ Fig. 10.51, ▶ Fig. 10.52, ▶ Fig. 10.53, ▶ Fig. 10.54, ▶ Fig. 10.55, ▶ Fig. 10.56, ▶ Fig. 10.57, ▶ Fig. 10.58, ▶ Fig. 10.59, ▶ Fig. 10.60, ▶ Fig. 10.61, ▶ Fig. 10.62, ▶ Fig. 10.63, ▶ Fig. 10.64, ▶ Fig. 10.65, ▶ Fig. 10.66, ▶ Fig. 10.67, ▶ Fig. 10.68, ▶ Fig. 10.69, ▶ Fig. 10.70.

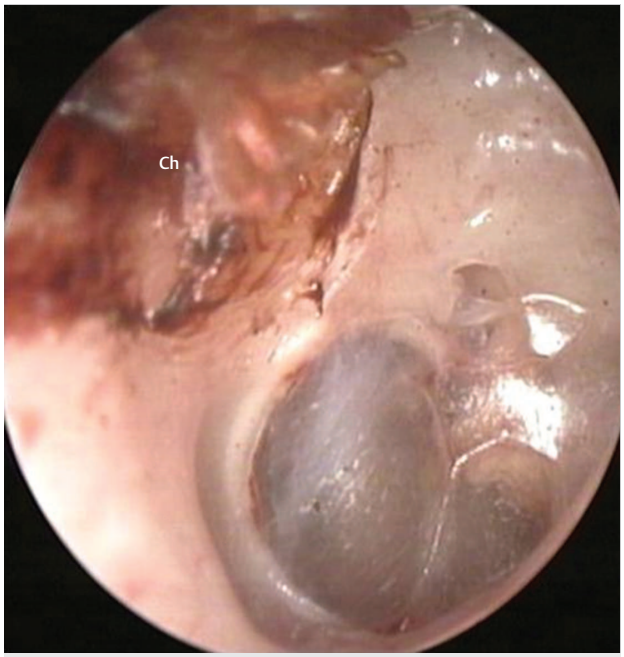


Fig. 10.47 Otoscopy of a case of massive cholesteatoma (Ch) is shown. The patient underwent canal wall down tympanoplasty elsewhere, and the mastoid cavity is filled with debris. No cholesteatoma is seen in the tympanic cavity. Audiometry showed moderate to severe mixed hearing loss. Facial nerve function was normal (grade I House–Brackmann scale), so a transotic approach was performed.

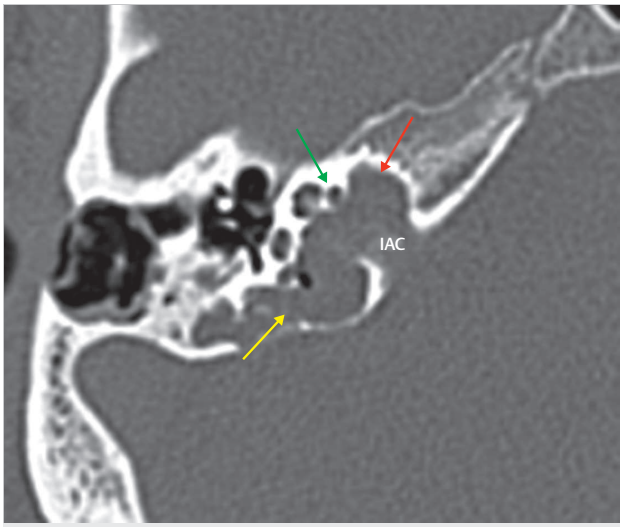


Fig. 10.48 CT scan of the same case, axial view. The cholesteatoma engulfs the labyrinth (*yellow arrow*) and the basal turn of the cochlea (*green arrow*), reaching the petrous apex (*red arrow*) and the internal auditory canal (IAC).

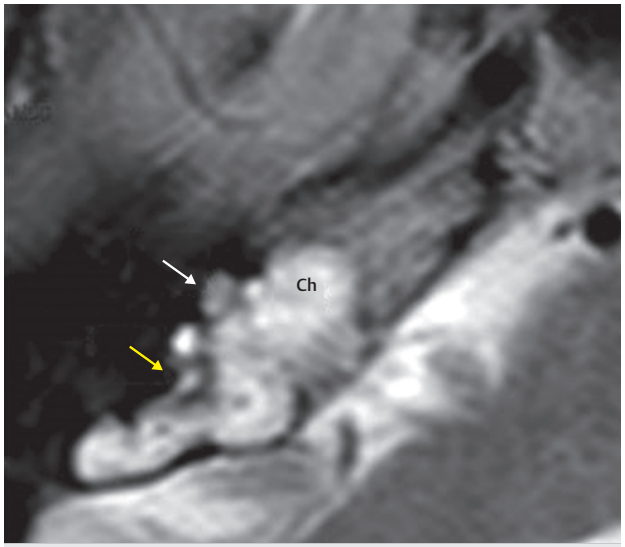


Fig. 10.49 MRI (T2W), axial view. The cholesteatoma (Ch) has advanced far medially, eroding the posterior labyrinth to reach the petrous apex. The apical turn of the cochlea (*white arrow*) and the posterior semicircular canal (*yellow arrows*) seem spared.

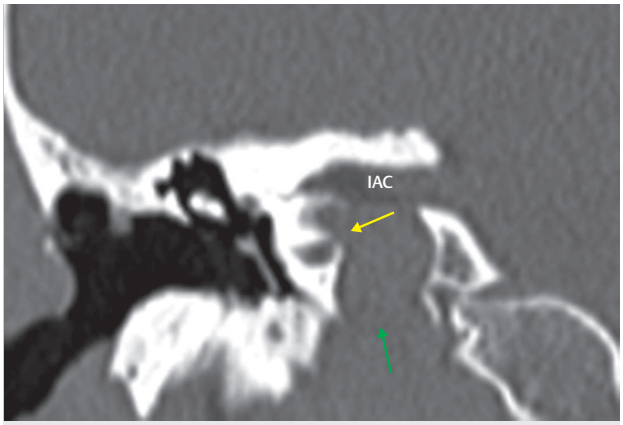


Fig. 10.50 CT scan of the same case, coronal view. The cholesteatoma invades the labyrinth (*yellow arrow*), the jugular bulb (*green arrow*), and the roof of the internal auditory canal (IAC).

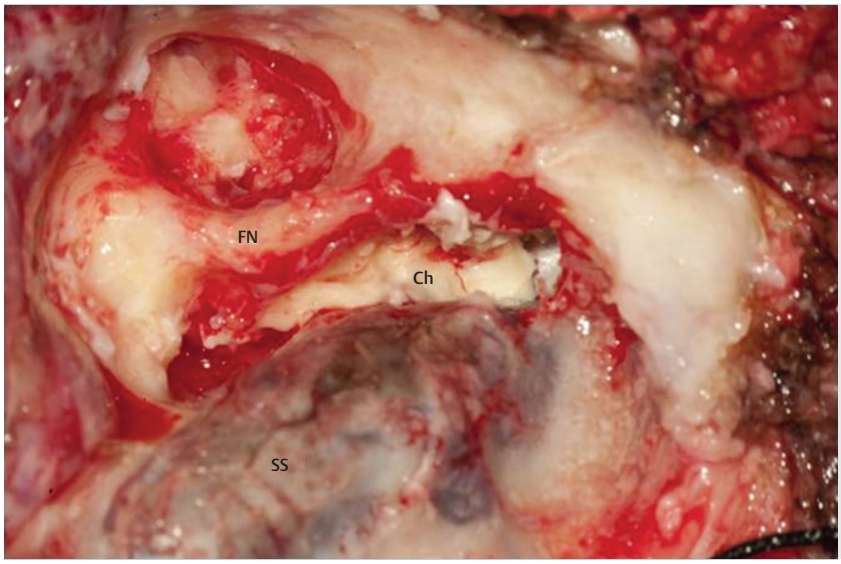


Fig. 10.51 Intraoperative image. Blind sac closure of the external auditory canal, removal of the tympanic membrane and the ossicles, and extended mastoidectomy have been performed. Further removal of the bone exposed huge cholesteatoma located medially to the labyrinth. The sigmoid sinus is exposed to enhance access to the digastric ridge. Ch, cholesteatoma; FN, facial nerve; SS, sigmoid sinus.

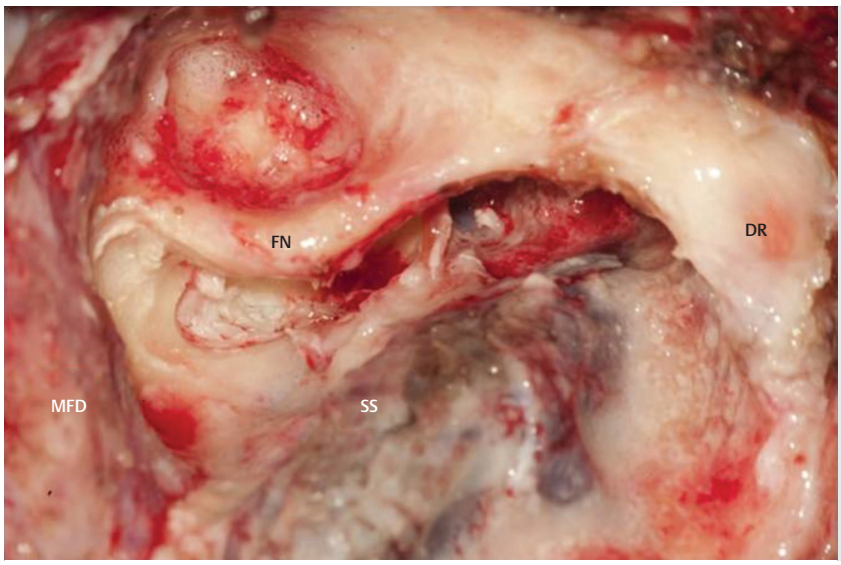


Fig. 10.52 Intraoperative image. The drilling is advanced superiorly by removing lateral and posterior semicircular canals. The cholesteatoma goes medial to the labyrinth toward the internal auditory canal. Facial nerve is well skeletonized. DR, digastric ridge; FN, facial nerve; MFD, middle fossa dura; SS, sigmoid sinus.

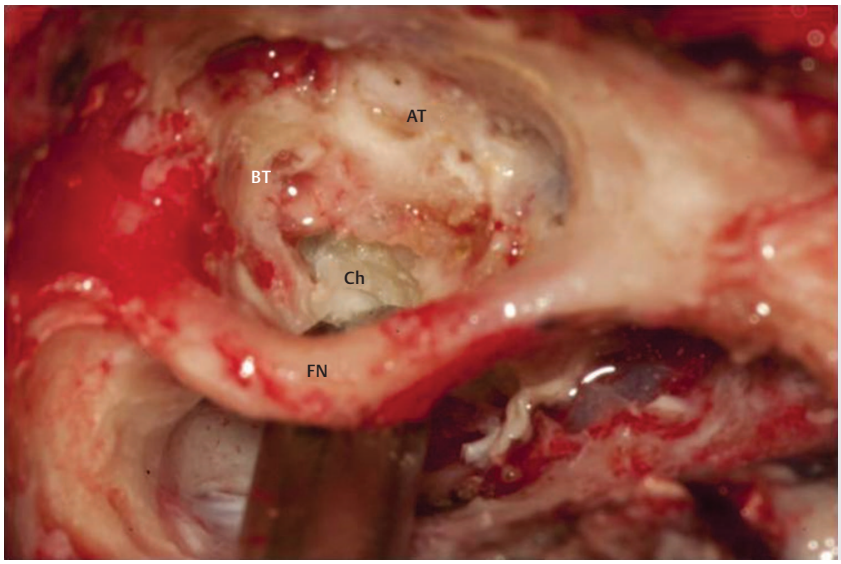


Fig. 10.53 Intraoperative image. The cholesteatoma has been removed from the labyrinth and the subfacial space. The cholesteatoma invading the modiolus is seen. AT, apical turn of cochlea; BT, basal turn of cochlea (upper pole); Ch, cholesteatoma; FN, facial nerve.

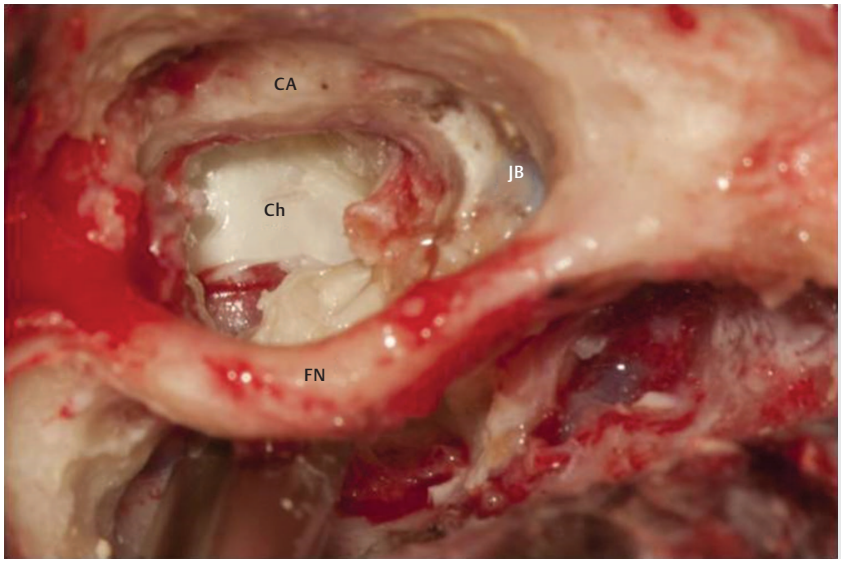


Fig. 10.54 Intraoperative image. Cochlea has been completely removed. Debris of the cholesteatoma filling the petrous apex is partially evacuated. The position of the jugular bulb located comparatively inferiorly permits access to the petrous apex from the area under the facial nerve. CA, internal carotid artery; Ch, cholesteatoma; FN, facial nerve; JB, jugular bulb.

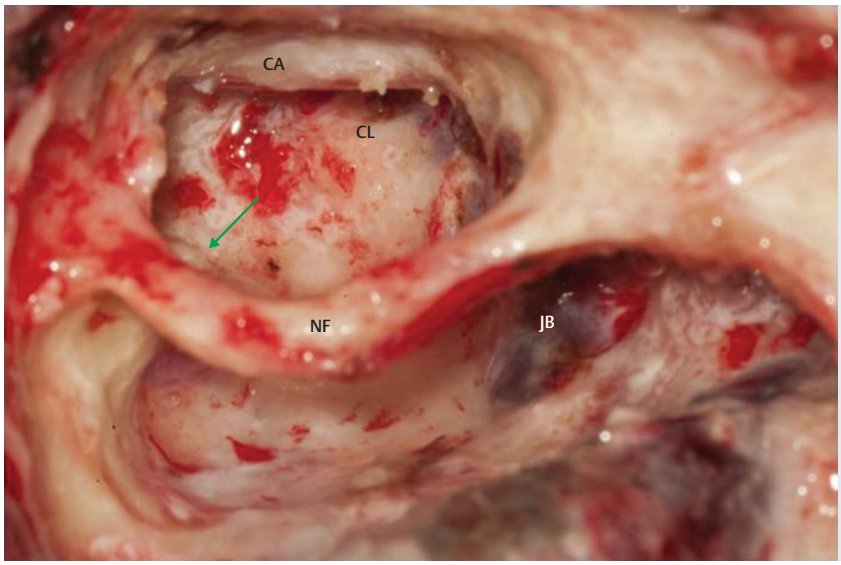


Fig. 10.55 Cleaning of the area anterior to the internal auditory canal (*arrow*) has been accomplished. The clivus is exposed. The cavity is free from the cholesteatoma. CA, internal carotid artery; FN, facial nerve; CL, clivus; JB, jugular bulb.

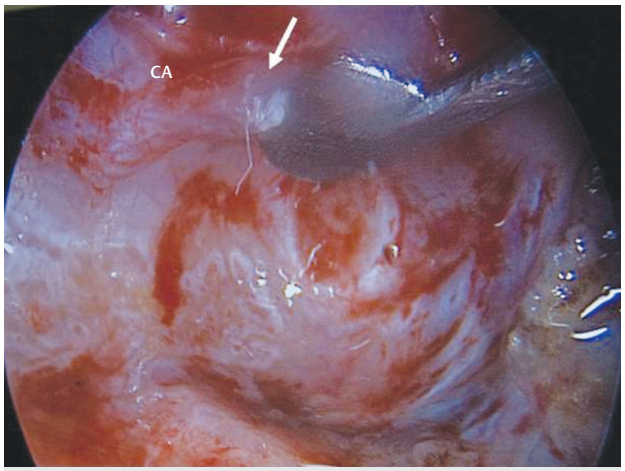


Fig. 10.56 With a 30-degree endoscope, the area under the internal carotid artery (CA) could be explored to check the presence of any residual pathology. In this case, a small piece of skin is detached from the bony wall of the artery.

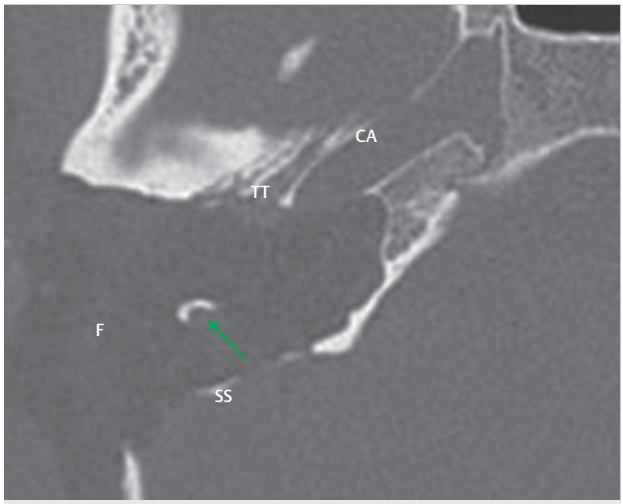


Fig. 10.57 Postoperative CT shows extensive removal of bone from the petrous apex. The mastoid segment of the facial nerve is left in the fallopian canal (*arrow*). The posterior fossa dura is partially exposed, and the carotid artery is uncovered posterolaterally. CA, carotid artery; F, abdominal fat; SS, sigmoid sinus; TT, tensor tympani.

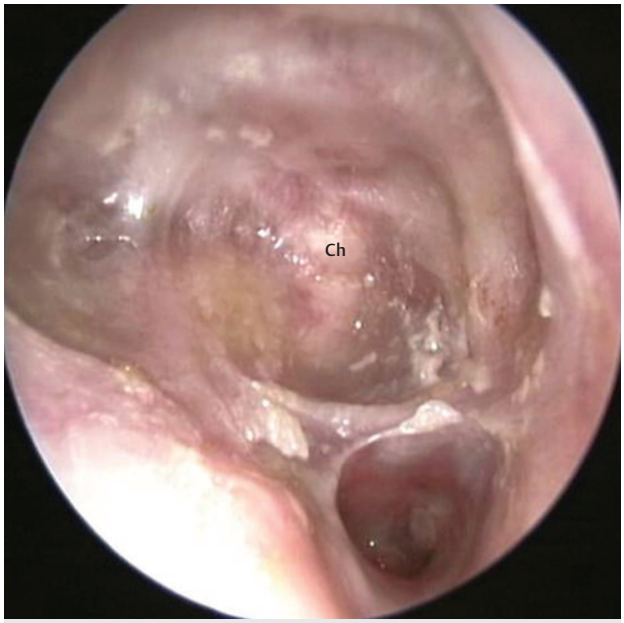


Fig. 10.58 Otoscopy of a case of massive PBC. The patient underwent six tympanoplasties elsewhere. A huge cavity is seen with the presence of a whitish mass strongly suggestive of a recurrent cholesteatoma (Ch). The patient complained of recurrent otorrhea and hearing loss (mixed type). Facial nerve function was normal.

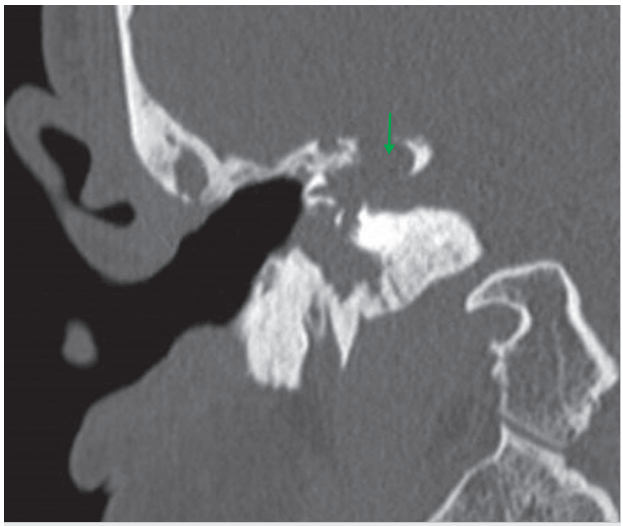


Fig. 10.59 CT scan of the same case, coronal view. Massive PBC with invasion of the labyrinth and the roof of the internal auditory canal (*arrow*).

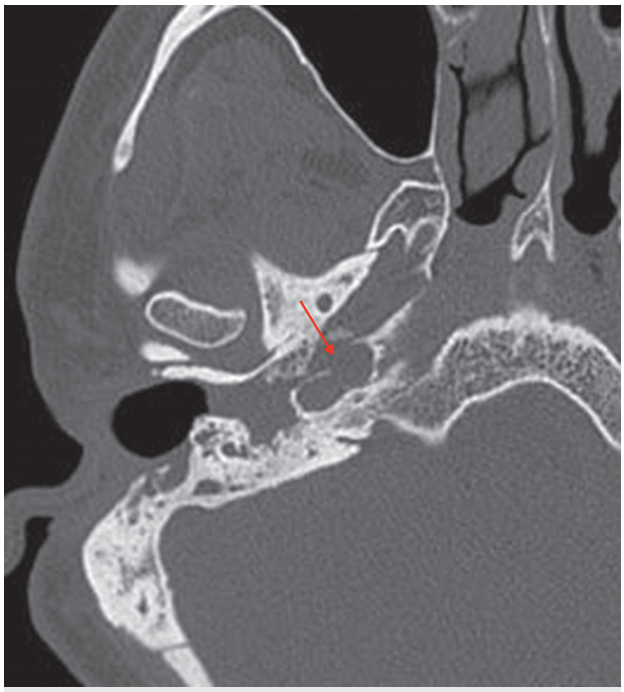


Fig. 10.60 CT scan of the same case, axial view. The cholesteatoma extends anteromedially to the vertical portion of the internal carotid artery. Considering this anterior extension, an infratemporal fossa approach type B was incorporated into the transotic approach for the removal of the disease.

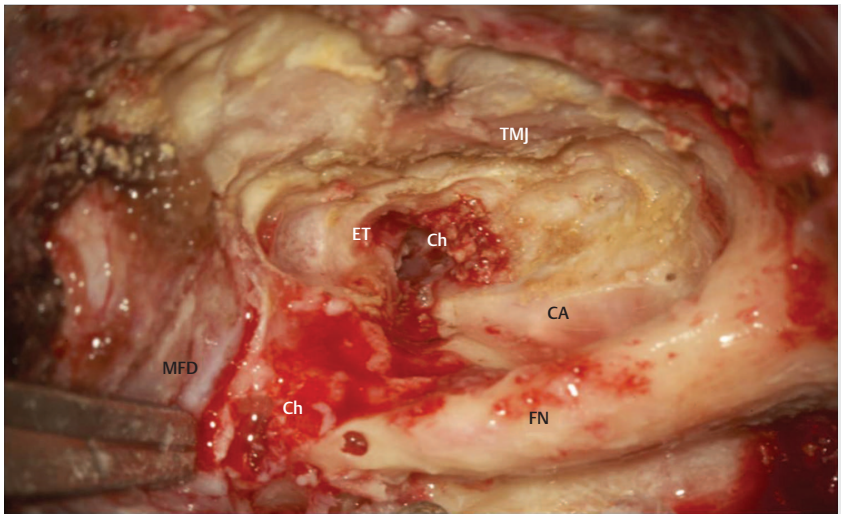


Fig. 10.61 Intraoperative picture. The cholesteatoma has been removed from the tympanic cavity and the mastoid. The labyrinth and the cochlea, involved by the cholesteatoma, have been removed too. The cholesteatoma infiltrates the Eustachian tube and the area anterior to the carotid artery. CA, carotid artery; Ch, cholesteatoma; ET, Eustachian tube; FN, facial nerve; MFD, middle fossa dura; TMJ, temporomandibular joint.

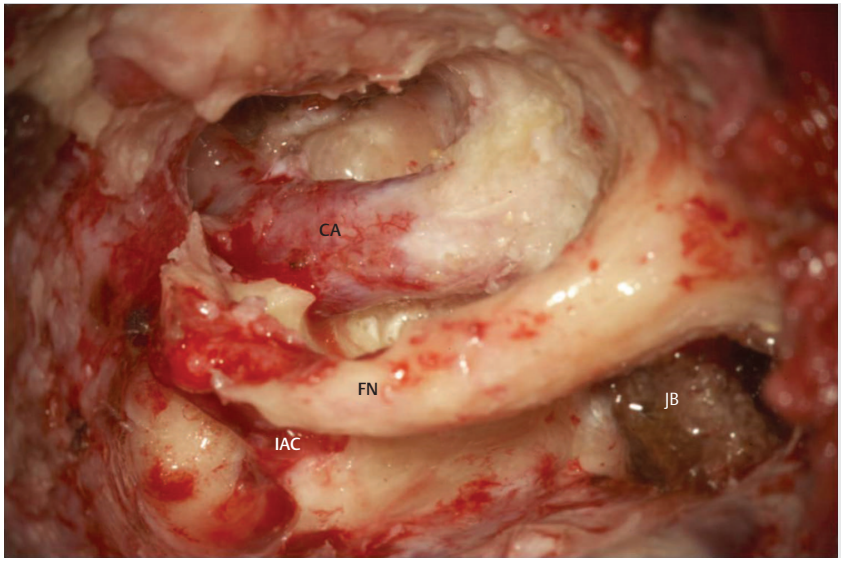


Fig. 10.62 Intraoperative picture. View after complete removal of the cholesteatoma. The carotid artery is under control until its horizontal part (*arrow*). CA, carotid artery; FN, facial nerve; IAC, internal auditory canal; JB, jugular bulb.

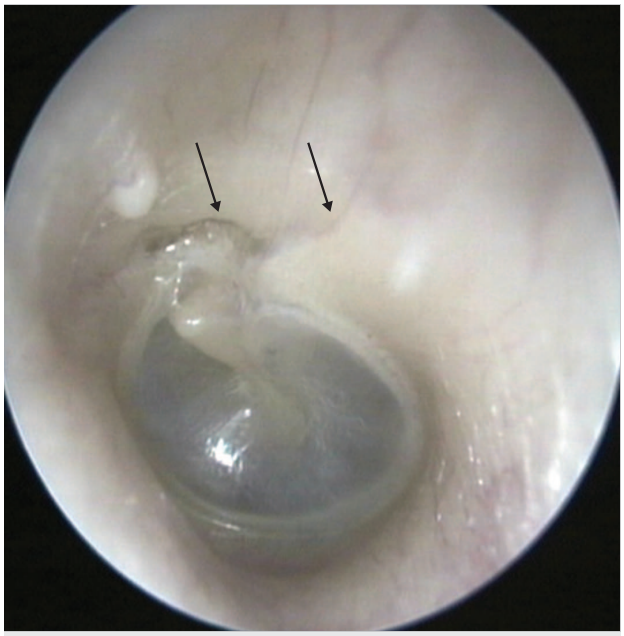


Fig. 10.63 Case of a supralabyrinthine PBC. This 26-year-old female patient was evaluated for left progressive facial palsy for the last 1 year (near total, grade V House–Brackmann scale) and moderate to severe mixed hearing loss. Ten years before, she had head trauma with temporal bone fracture (*arrows*) and temporary left facial nerve palsy. The patient underwent PBC removal through a modified transcochlear approach type A with facial nerve reconstruction.

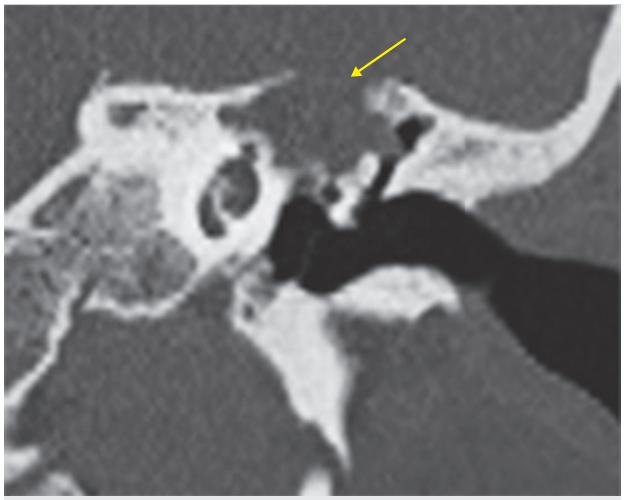


Fig. 10.64 CT scan of the same case, coronal view, shows supralabyrinthine PBC (*arrow*).

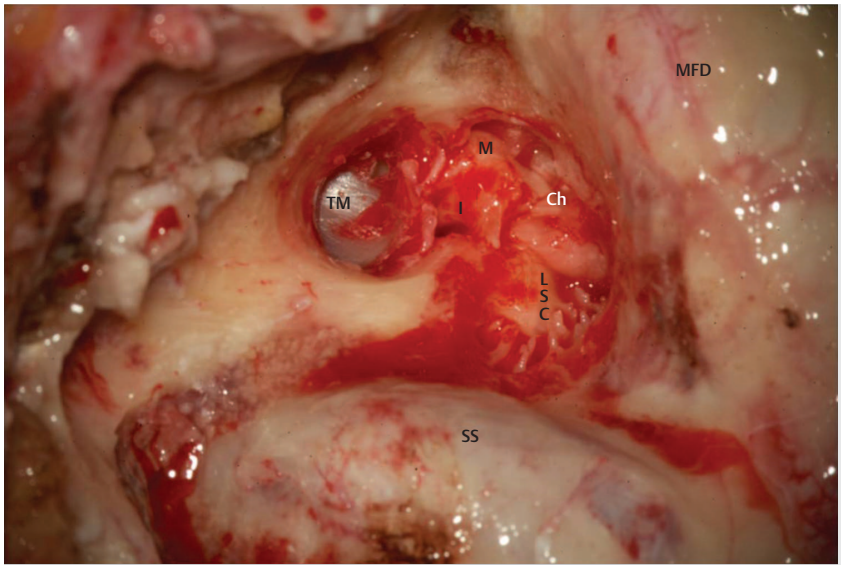


Fig. 10.65 Intraoperative picture. Blind sac closure of the external auditory canal and extended mastoidectomy have been performed. Cholesteatoma is seen in the supralabyrinthine area, infiltrating the middle fossa dura. Ch, cholesteatoma; I, incus; LSC, lateral semicircular canal; M, malleus; MFD, middle fossa dura; SS, sigmoid sinus; TM, tympanic membrane.

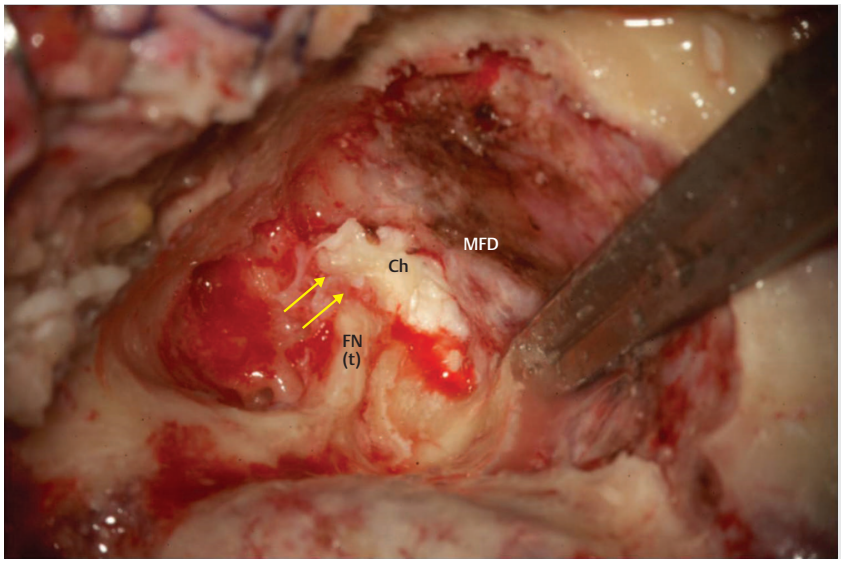


Fig. 10.66 Intraoperative picture. The cholesteatoma involves the supralabyrinthine area toward the internal auditory canal. The second portion of the facial nerve is engulfed by the pathology, leading to interruption of the nerve itself (*arrow*). The labyrinth has been previously drilled. Ch, cholesteatoma; FN (t), tympanic portion of the facial nerve; MFD, middle fossa dura.

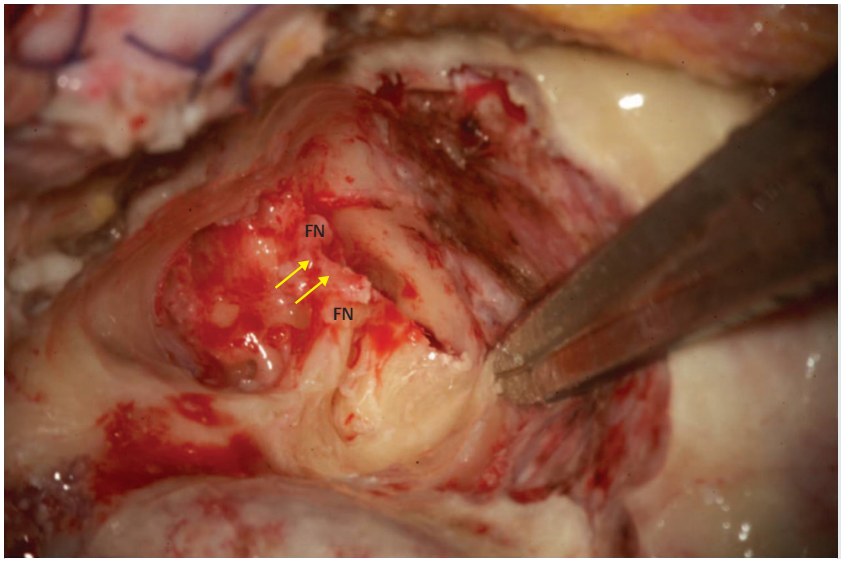


Fig. 10.67 Intraoperative view after removal of the cholesteatoma. The facial nerve (FN) is interrupted on its second portion (*arrow*). The proximal stump of the facial nerve is replaced by fibrous tissue, so a healthy portion of the nerve should be found for the anastomosis with the distal stump.

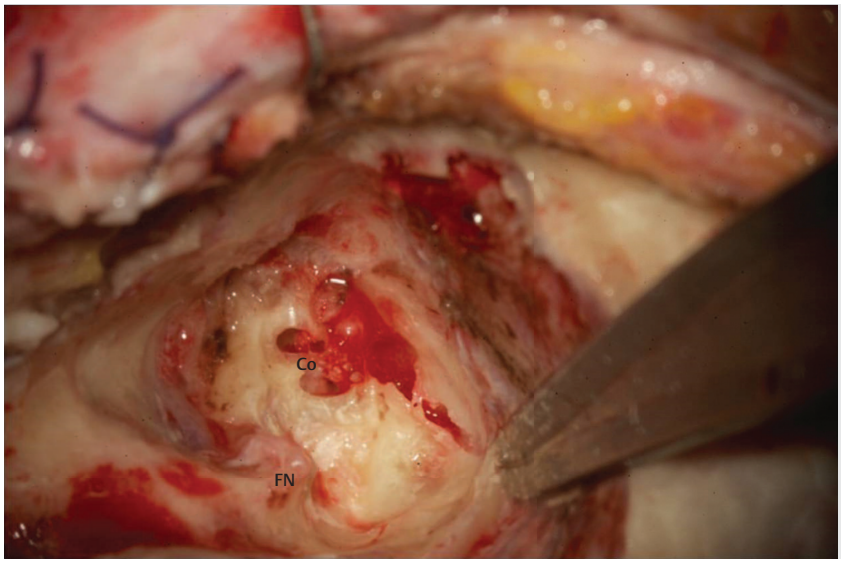


Fig. 10.68 Intraoperative picture. The third and second healthy portions of the facial nerve (FN) are detached from the fallopian canal and rerouted posteriorly. The cochlea (Co) is drilled to reach the fundus of the internal auditory canal and the first portion of the facial nerve.

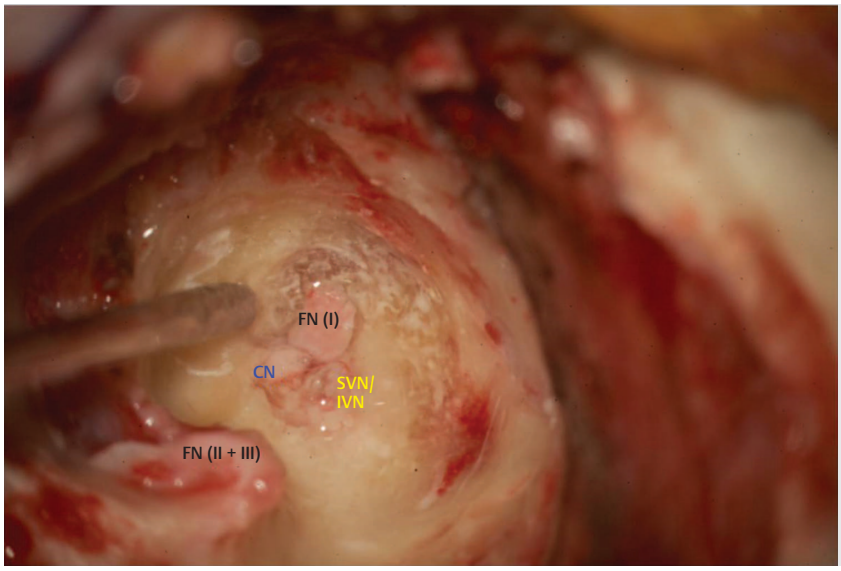


Fig. 10.69 Intraoperative picture after removal of the cochlea and opening of the internal auditory canal. From this view, the first portion of the facial nerve (FN I) is just superior to the cochlear nerve (CN) and anterior to the vestibular nerves (SVN/IVN). The facial nerve, in its second and third portion (FN II + III), has been posteriorly rerouted.

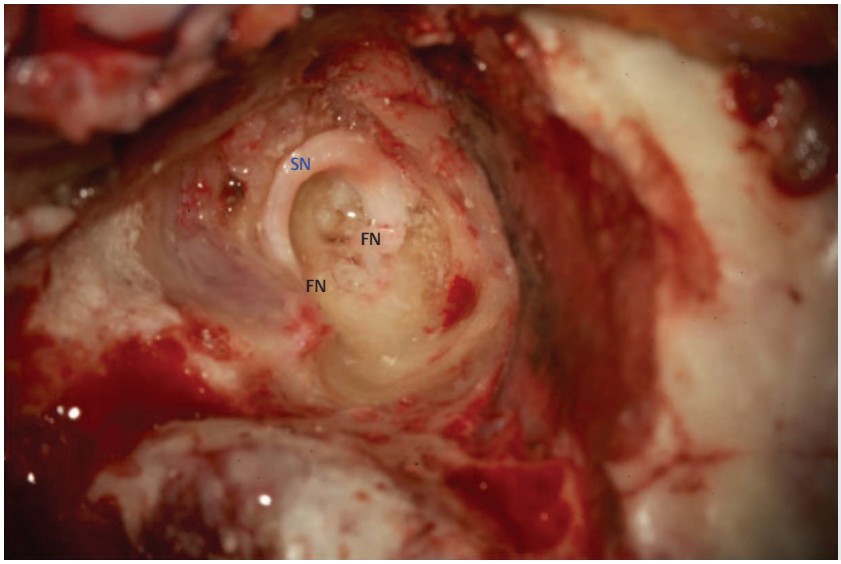


Fig. 10.70 Intraoperative picture. A sural nerve graft (SN) is used to restore continuity of the facial nerve. The two ends are fixed with temporalis fascia and fibrin glue. The cavity is further obliterated with abdominal fat.

10.1.2 Problems in Surgery

Hearing preservation. Hearing preservation would be important in exceptional cases, that is, 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. If the cochlea is not destroyed beyond what is essential for the clearance of the disease, a cochlear implant should be used in ears with an accessible cochlear lumen even after labyrinthectomy. Another option is a BAHA if bone conduction is preserved in the ipsilateral ear.

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

- *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 or facial-trigeminal (masseteric nerve) anastomosis*: In patients with long duration of facial palsy (>12 months).

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 the rhinopharynx, it is important to perform a complete control of the internal carotid artery to mobilize the artery if necessary. PBC are less aggressive in terms of arterial involvement and are easier to dissect as compared with other tumors (i.e., tympanojugular paragangliomas). 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. Dealing with the jugular bulb in cases where it is involved is a well-thought-out strategy. Preoperative imaging must be carefully analyzed for two aspects: the relationship of the lesion with the jugular bulb and the patency of the contralateral venous drainage system by MR venography. In the presence of a hypoplasia of the contralateral venous system, sacrifice of the bulb means occlusion of the main venous drainage of the brain, with the consequent risk of benign intracranial hypertension or venous infarction of the temporal lobe. In such cases, damage to the jugular bulb or the sigmoid sinus must be avoided at all cost. 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 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. 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 (T1 W and T2 W images with fat suppression, diffusion-weighted images, non–echo planar imaging [EPI]) with gadolinium enhancement every year for at least 10 years.

Summary

When a patient presents with hearing loss (sensorineural or mixed) and/or facial nerve paralysis with or without a retrotympanic mass, the probability of a PBC should be considered. In such cases, it is necessary to perform a high-resolution CT scan of the temporal bone. The ideal treatment for PBC is radical surgical removal, although destruction of the labyrinth and rerouting of the facial nerve may be required. The status of the contralateral ear must also be considered. The transotic and modified transcochlear approaches are the most appropriate for the removal of PBC. These approaches offer direct lateral access to the petrous bone and allow the removal of all types of PBC with their possible extension into the clivus, sphenoid sinus, or intradural space. In addition, they have the advantage of minimizing the occurrence of CSF leak and allow control of the different vital structures, including the internal carotid artery. Closure of the external auditory canal as a cul de sac and obliteration of the operative cavity with abdominal fat avoid the risk of infection and the need for frequent toilet of a very deep cavity. The advances in hearing rehabilitation has given further options in cases with preserved cochlea and/or preserved bone conduction. The middle cranial fossa approach can be used in few cases with small supralabyrinthine PBC and noncompromised inner ear function.