

Chapter 44

Clinical and Surgical Pathology of the Petrous Apex

Sampath Chandra Prasad, Fernando Mancini, Mario Sanna

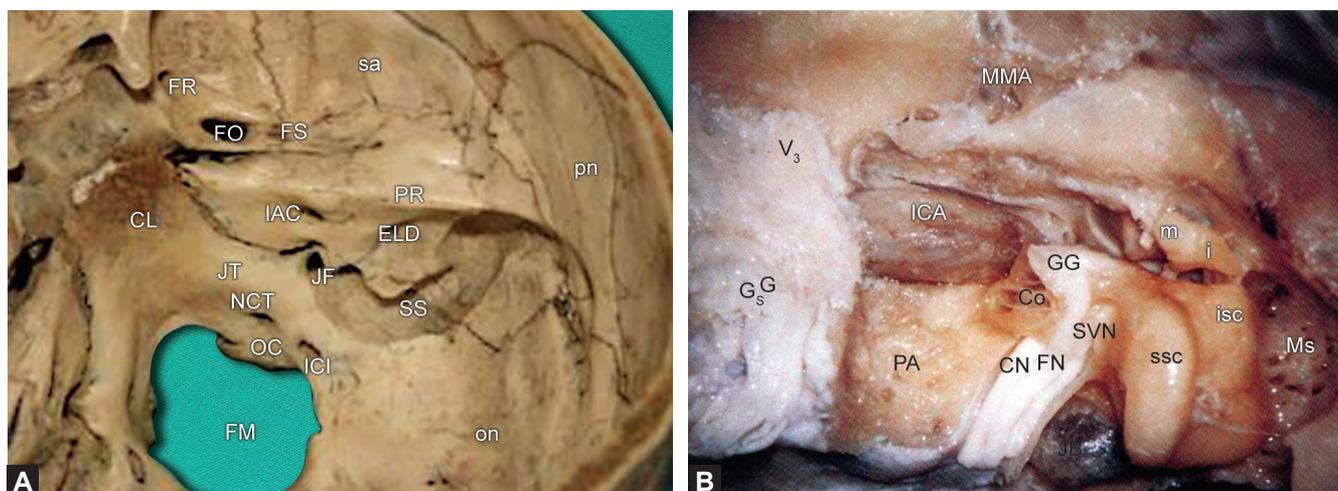
INTRODUCTION

The petrous apex (PA) is a pyramid shaped part of the temporal bone that is surrounded by the brain, complex neurovascular structures, and the organ of hearing. Multiple pathologic processes may affect the PA that ranges from infection like petrous apicitis to complex tumors like chondrosarcomas. Diseases affecting the PA not only tend to have serious morbidities due to the affliction of important structures in the vicinity, but they are also difficult to approach surgically due to the deep location in the skull base. The presence of the internal carotid artery (ICA) and the facial nerve in the close vicinity adds to the surgical challenge. The treating surgeon must have a thorough understanding of the anatomy, clinical pathology, and skull base approaches to effectively deal with such lesions. In the last decade, rapid advances have been made in lateral and endoscopic skull base surgery in terms of accurate mapping of the anatomy of the PA and the surrounding areas and formulation of appropriate surgical approaches. Facial nerve monitoring and interventional neuroradiology have greatly improved the safety of surgical procedures. Management of the diseases of the PA also gets complicated due to the delayed and variable presentation of symptoms. Due to the variability of symptoms, it is difficult to diagnose them based on clinical findings alone and hence they remain undetected for many months or years before a diagnosis is made. With the advent of modern skull base surgery, advances in neuroimaging, and neuroanesthesia, it is today possible to precisely locate and extirpate almost all types of tumors.¹

ANATOMY OF THE PA

The PA is a part of the central skull base and is defined as the portion of the petrous temporal bone anteromedial

to the bony labyrinth and lateral to the petro-occipital fissure.² The anteromedial margin of the PA forms the posterior limit of the middle cranial fossa (MCF) (Figs. 44.1A and B). The PA is divided into a larger anterior triangular (principally consisting of bone marrow or air cells) and a smaller posterior quadrangular (derived from the dense bone of the otic capsule) compartments (Fig. 44.2). At the junction of the PA with the sphenoid and the occipital bone, it is separated from the clivus by an ovoid horizontal gap, the foramen lacerum, which contains a bridge of dense fibrous tissue and cartilage (Figs. 44.3A and B). Above the foramen lacerum, the petrous ICA exits the medial opening of the carotid canal to continue as the cavernous portion. At the superomedial surface of the PA, there is an important landmark, a shallow depression on which lies the trigeminal ganglion within the Meckel's cave (Fig. 44.4). It is due to this close relationship of the nerve to the PA that trigeminal symptoms are often associated with lesions in this area. The Dorello's canal carrying the sixth cranial nerve is an anatomic channel from the dural margin along the petroclival junction to the posterior cavernous sinus (Figs. 44.3A and B). Lesions of the PA or petroclival junction are notorious for invading or compressing the Dorello's canal and causing the sixth cranial nerve palsy and diplopia. The inferior extracranial surface of the PA is intimately related to the nasopharynx. The posteromedial margin of the PA meets the clivus that is formed by the basiocciput and sphenoid. Invasive neoplasms of the nasopharynx, thus, can traverse the sinus of Morgagni and readily gain access to the bony skull base including the PA.³ In approximately one-third of cases, the PA is aerated by tracts (peritubal, posteromedial, and subarcuate) extending superior and inferior to the cochlea in order to communicate with the middle ear cleft.²



Figs. 44.1A and B: (A) The PA and its relations. (B) Superior view of the PA in a dissected specimen.

(PA: Petrous apex; FO: Foramen ovale; FR: Foramen rotundum; FS: Foramen spinosum; IAC: Internal auditory canal; SS: sigmoid sinus; JF: Jugular foramen; FM: Foramen magnum; OC: Occipital condyle; OB: Occipital bone; PB: Parietal bone; SB: Sphenoid bone; CL: Clivus; JT: Jugular tubercle; PA: Petrous apex; G_sG: Gasserian ganglion; V₃: third branch of the trigeminal nerve; ICA: Internal carotid artery; MMA: Middle meningeal artery; Co: Cochlea; GG: Geniculate ganglion; CN: Cochlear nerve; FN: Facial nerve; SVN: Superior vestibular nerve; SSC: Superior semicircular canal; LSC: Lateral semicircular canal; M: Malleus; I: Incus; Ms: Mastoid; ON: Occipital bone; ICI: Posterior Condylar Foramen; SA: Squamous bone; PN: Parietal bone; PR: Petrous Ridge; ELD: Endolymphatic Duct; NCT: Anterior Conylar Foramen).

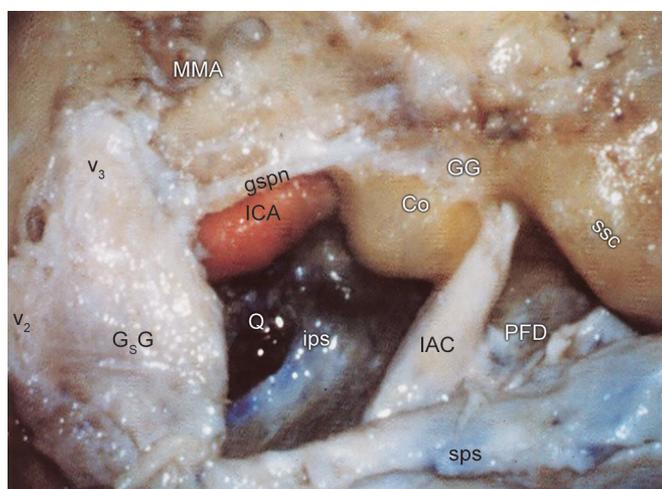


Fig. 44.2: The petrous apex is divided into an anterior triangular and a posterior quadrangular (Q) compartments.

(G_sG: Gasserian Ganglion; V₂: Maxillary branch of Trigeminal Nerve; V₃: Mandibular branch of Trigeminal Nerve; MMA: Middle Meningeal Artery; GSPN: Greater Superficial Petrosal nerve; ICA: Internal Carotid Artery; IPS: Inferior Petrosal Sinus; SPS: Superior Petrosal Sinus; PFD: Posterior Fossa Dura; SSC: Superior Semicircular Canal; Co: Cochlea; IAC: Internal Auditory Canal.)

The ICA is the most prominent structure in the PA. The petrous ICA enters the temporal bone into the carotid canal and extends till the foramen lacerum. It has three sections: a vertical portion, the genu, and the horizontal

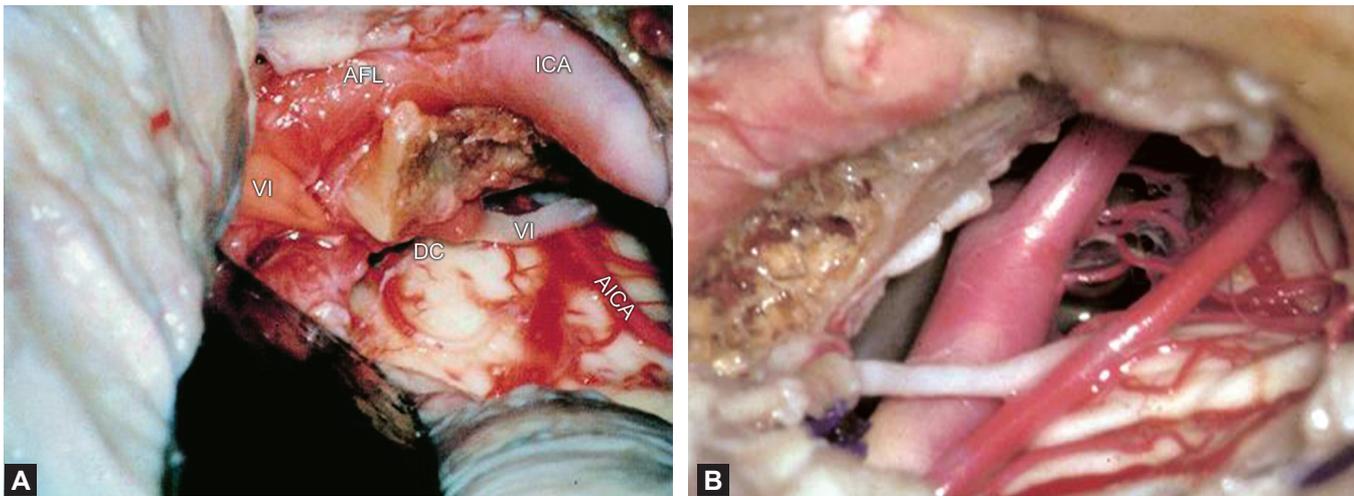
portion (Fig. 44.4). At its entrance, the vertical ICA lies anterior to the cochlea. The anteromedial part of the roof of the horizontal ICA is formed by a thin plate of the bone that separates the artery from the trigeminal ganglion (Figs. 44.5 and 44.6).

DISEASES AFFECTING THE PA

A variety of diseases affects the PA and they can arise from the bone, meninges, pneumatized air cells, middle ear, cranial nerves, endolymphatic sac, and the paraganglionic chemoreceptors of the jugular bulb at the skull base. Razek et al.⁴ proposed a classification of the lesions of the PA based on the type of lesions. We have modified it to include additional lesions that make the list more comprehensive (Table 44.1).

In the following sections, we will discuss some of the most important lesions and the relevant surgical approaches.

Petrous Apex Cholesterol Granuloma (PACG)—A PACG is a benign lesion that primarily develops as a result of a foreign body giant cells response to cholesterol deposits. This develops into a cavity with a fibrous lining and filled by a golden to brownish yellow fluid that contains lipids, cholesterol crystals surrounded by foreign body giant cells, fibrous tissue reaction, vascular proliferation, and chronic inflammation. Cholesterol granulomas are the most



Figs. 44.3A and B: (A) The course of the right petrous internal carotid artery (ICA) and the anterior foramen lacerum (AFL) can be identified in this dissected specimen. Also seen are the abducens nerve (VI) in the Dorello's canal (DC) and the anterior inferior cerebellar artery (AICA). (B) The petrous apex has been drilled out and the basilar artery is identified.

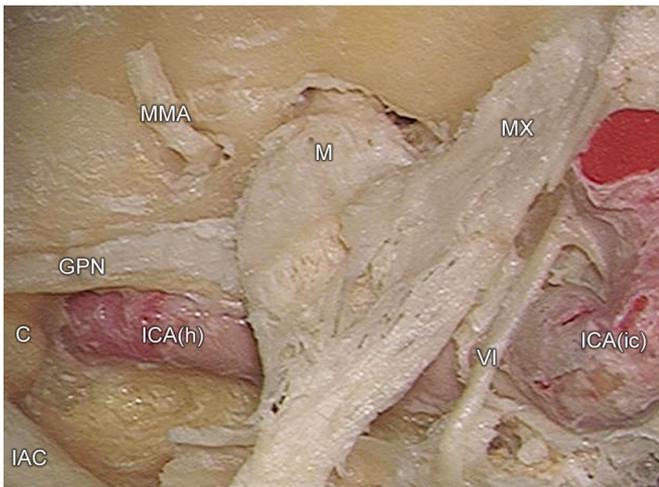


Fig. 44.4: The course of the horizontal segment of the left petrous internal carotid artery ICA(h) and its relation to Meckel's cave is seen. (MMA: Middle meningeal artery; M: Mandibular nerve; MX: Maxillary nerve; VI: Abducens nerve; ICA(ic): Intracranial internal carotid; C: Cochlea; IAC: Internal auditory canal; GPN: Greater Superficial Petrosal Nerve).

common of the lesions arising in the PA. Classically, they occur in patients with a pneumatized PA and a long-standing history of otitis media. But pneumatization of the PA itself has been reported to be in the range of 9–30% of temporal bones and this accounts for the rarity of the disease.

There are two theories to explain the origin of PACG: the older obstruction-vacuum theory and the exposed marrow theory proposed by Jackler et al.²² The pathogenesis in both the theories is illustrated in (Flowchart 44.1). The exposed marrow theory has been supported by evidence from

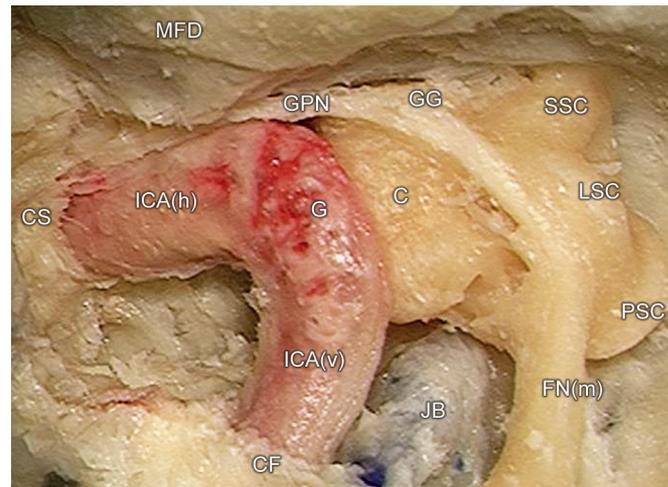


Fig. 44.5: All the bone around the intratemporal internal carotid artery (ICA) of a left temporal bone has been drilled out from the point of its entry at the carotid foramen (CF) to its exit at the cavernous sinus (CS). Note the close relation of the vertical portion ICA(v) and the genu (G) of the ICA to the cochlea (C) and that of the horizontal portion ICA(h) to the middle fossa dura (MFD).

(GPN: Greater petrosal nerve, C: Cochlea, GG: Geniculate ganglion, JB: Jugular bulb, SSC: Superior semicircular canal, LSC: Lateral semicircular canal, PSC: Posterior semicircular canal, FN(m): Mastoid segment of the facial nerve.)

recent studies.^{23,24} Clinical features of PACG may overlap with some of the other inner ear pathologies, and hence radiological diagnosis is often crucial in clinching the diagnosis. Many times the diagnosis may be incidental. In our series²⁵ the following symptoms were noted: hearing loss (60%), vertigo and tinnitus (27%), diplopia (40%), headache (27%), facial paresthesia (13%), trigeminal neu-

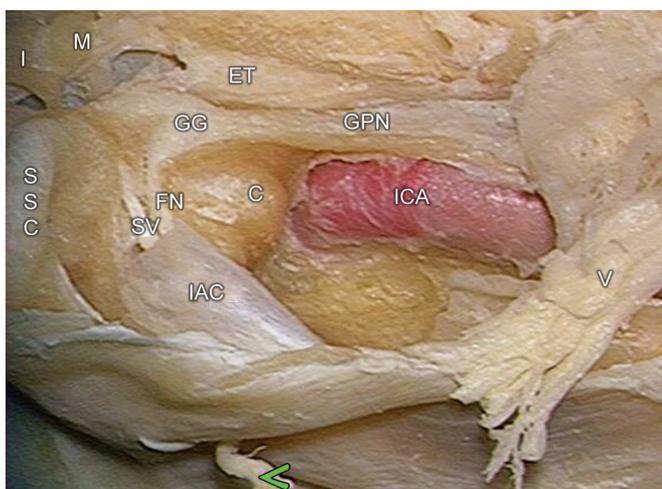


Fig. 44.6: The left petrous apical contents as seen from the middle cranial fossa. The bone overlying the internal auditory canal has been removed. The relationship of the ICA to the greater superficial petrosal and the trigeminal nerves can be seen. (IAC: Internal Auditory Canal; SV: Superior vestibular nerve, SSC: Superior semicircular canal, GPN: Greater petrosal nerve, ICA: Internal carotid artery, M: Malleus, I: Incus, V: Trigeminal nerve, GG: Geniculate Ganglion; FN: Facial Nerve; C: Cochlea; ET: Eustachian Tube).

ralgia (7%), and hemifacial spasm (7%). Other signs like seizures and signs of brainstem compression may appear in large lesions. Magnetic resonance imaging (MRI) is a reliable tool for the diagnosis of this lesion. Although most other PA lesions have low or intermediate signal intensity on T1-weighted images, PACG is usually hyperintense on both T1- and T2-weighted images (Figs. 44.7A and B). Computed tomography (CT) scan will show an isodense lesion, with evidence of bone erosion.

Management of PACG can be conservative or surgical (Flowchart 44.2). Small lesions that are asymptomatic or those that are discovered incidentally can be managed by a wait-and-scan policy. In our series of 30 patients, 13 were managed by wait-and-scan and in this group only one lesion showed growth during the follow-up. Larger lesions or those that induce symptoms must be operated upon.

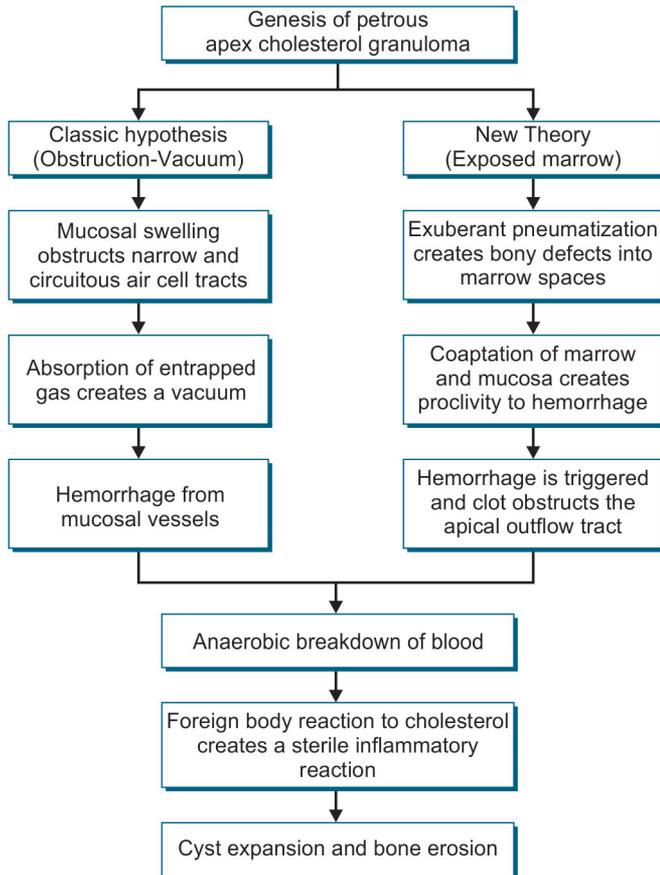
The main goal of surgery is drainage and ventilation of the cavity after removal of the lesion. There are several surgical approaches that have been proposed in literature including the MCF approach, suboccipital approach, infralabyrinthine (IL) approach, infracochlear (IC) approach, transotic (TO) approach, infratemporal fossa approach (ITFA) type B, and the transnasal-transsphenoidal approaches. The factors influencing the choice of surgical approach are preoperative hearing, location and extent of the lesion, relationship with neurovascular structures,

Table 44.1: Lesions of the petrous apex.

Type	Lesion
Developmental	Cholesteatoma
	Cephalocele
	Petrous bone anomalies ^{5,6}
Obstructive	Cholesterol granuloma
	Effusion
	Mucocele
Inflammatory	Petrous apicitis
	Osteomyelitis (bacterial, fungal, or tubercular)
	Inflammatory pseudotumor
	Wegener's granulomatosis
	Sarcoidosis ⁷
Tumors	Benign
	Meningioma
	Schwannoma
	Paraganglioma
	Chondroma
	Chondroblastoma
	Myxoma
	Osteoblastoma
	Osteoclastoma (Giant Cell Tumor) ⁸
	Facial nerve neuroma
	Lipoma
	Osteoma ⁹
	Dermoid cyst ¹⁰
	Malignant
	Chondrosarcoma
	Chordoma
	Endolymphatic sac tumor
	Squamous cell carcinoma
	Metastasis
	Plasmacytoma
	Lymphoma
	Leukemia ^{11,12}
Nasopharyngeal carcinoma	
Rhabdomyosarcoma	
Langerhans cell histiocytosis	
Plasma cell myeloma ¹³	
Malignant germ cell tumor ¹⁴	
Melanoma ¹⁵	
Vascular lesions	Petrous internal carotid artery aneurysm
	Intraosseous dural arteriovenous fistula
Osseous dysplasia	Fibrous dysplasia
	Paget disease
Pseudolesions	Asymmetric marrow
	Effusion
	Pseudofracture
	Infantile pseudolesion
Fractures	Transverse, longitudinal, or mixed
Syndromes	Maffucci's syndrome ^{16,17}
	Gorham–Stout syndrome ^{18,19}
	Ollier's disease ^{20,21}
Contiguous lesions from surrounding structures	Sinonasal tumors
	Parotid tumors
	Skull base tumors

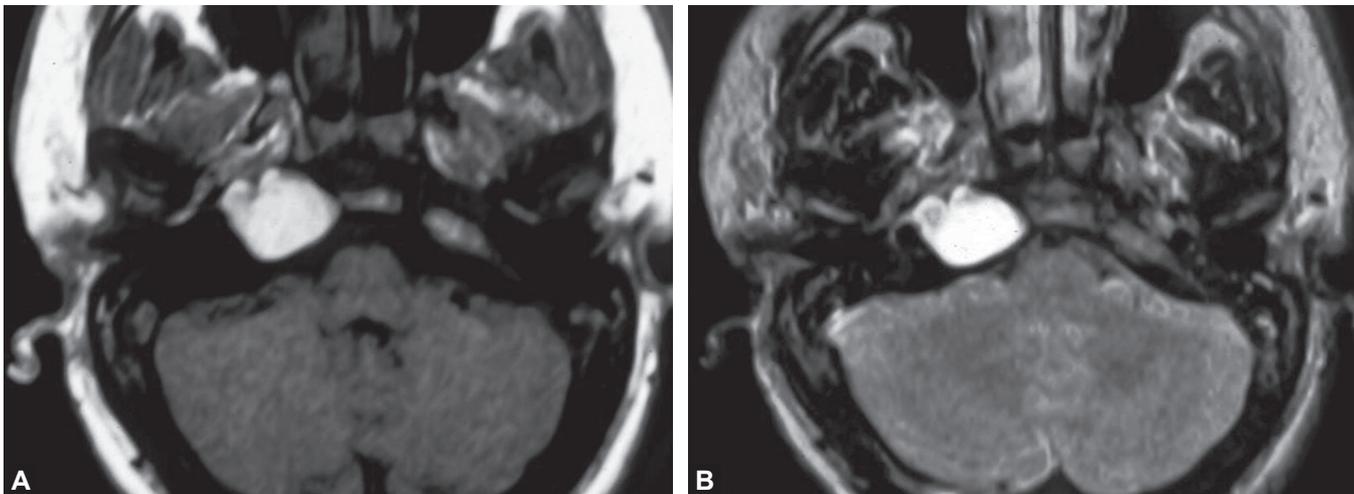
Note: Modification of Razek et al.'s⁴ classification of lesions of the petrous apex.

Flowchart 44.1: The two theories of formation of petrous apex cholesterol granuloma.

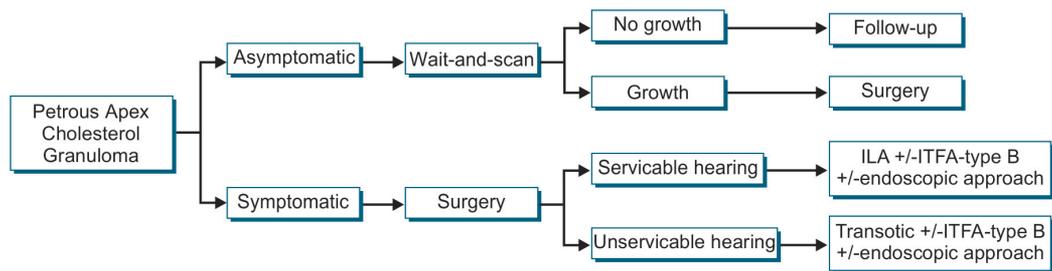


Source: From Jackler RK, Cho M. A new theory to explain the genesis of petrous apex cholesterol granuloma. *Otol Neurotol.* 2003; 24:96-106.

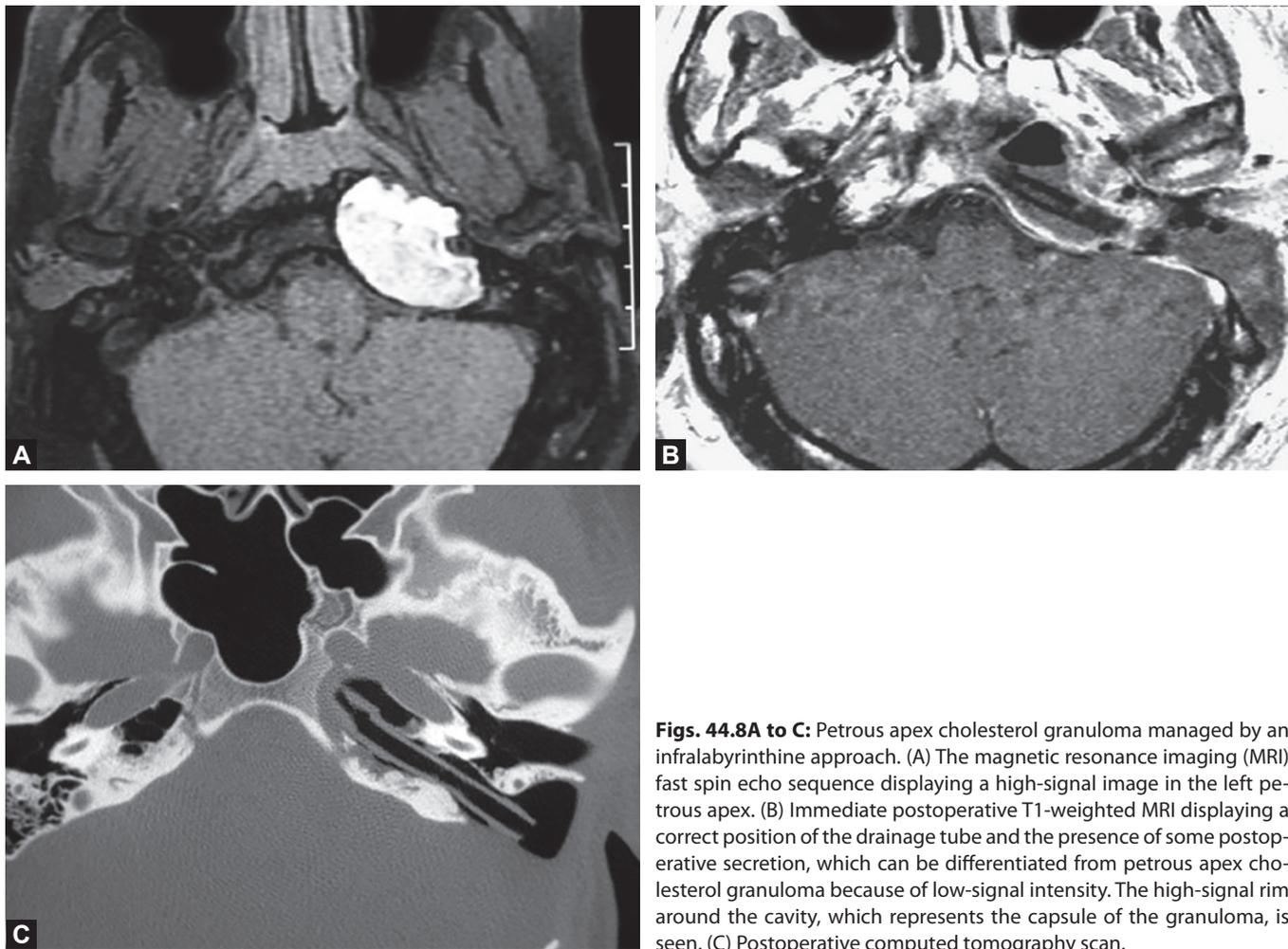
and anatomical variations (e.g. high jugular bulb). We do not favor MCF approaches due to the limited control of the ICA, difficulty in placing a drain through this approach (especially when aeration of the cavity becomes important in subtotal removals), and due to the risk of chemical meningitis by way of contamination of the subarachnoid spaces by the fluid content of the cyst. In case of preoperative serviceable hearing, IL and IC approaches are appropriate choices. The IL approach (Figs. 44.8 and 44.9) is the best option in serviceable hearing because it does not involve manipulation of the tympanic membrane (TM) and ossicular chain as the drainage tube is positioned in the mastoid cavity. The risk of injury to the ICA is minimal and revision surgery, when necessary, can be easily accomplished by a postauricular incision without manipulating the TM and the ossicular chain. Complications include sensorineural hearing loss and facial nerve injury. The IC approach provides adequate access to the PA, but has certain drawbacks. It requires maneuvers on the TM, enlargement of external auditory canal followed by reconstruction of the tympanic plate with bone patè. There is also a risk of ICA injury during exposure. The catheter is positioned close to the TM and revision surgery requires the elevation of the TM. The main handicap with both the IL and IC approaches arises in the presence of a high jugular bulb. To avoid injury to the jugular bulb, particularly when it is dominant, we prefer to approach the lesions using an ITFA type B (Figs. 44.10A to C). This approach presents excellent exposure, complete control of the intrapetrous ICA, and



Figs. 44.7A and B: The magnetic resonance imaging (MRI) of a patient with a right petrous apex cholesterol granuloma. A hyperintense lesion can be seen both in T1 (A) and T2 (B) weighted MRI.

Flowchart 44.2: Algorithm for surgical management of petrous apex cholesterol granuloma.

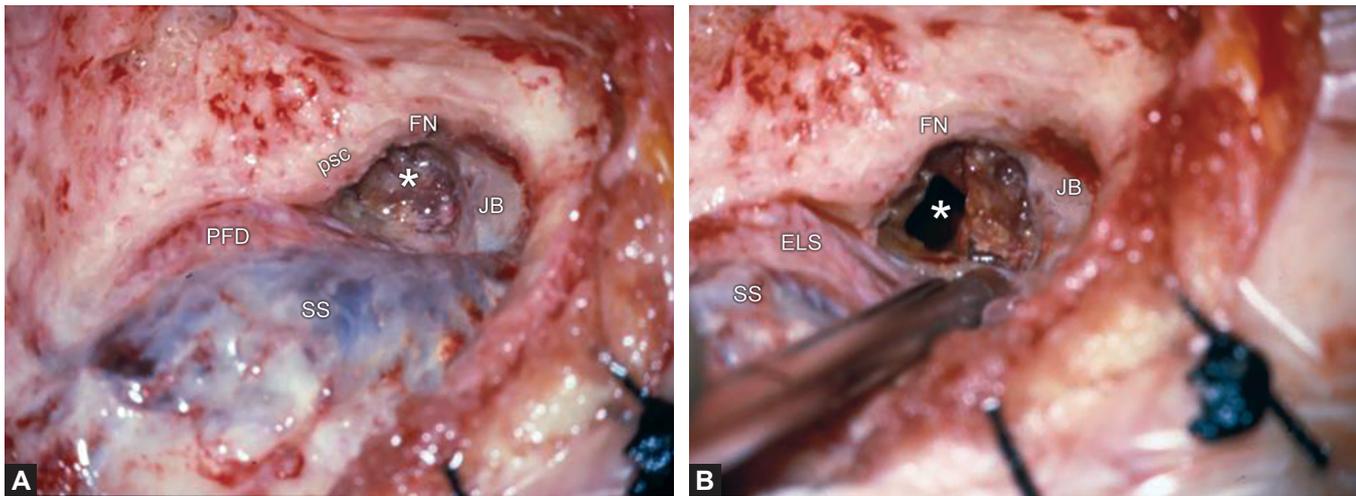
(ILA: Infralabyrinthine approach; ITFA: Infratemporal fossa approach)



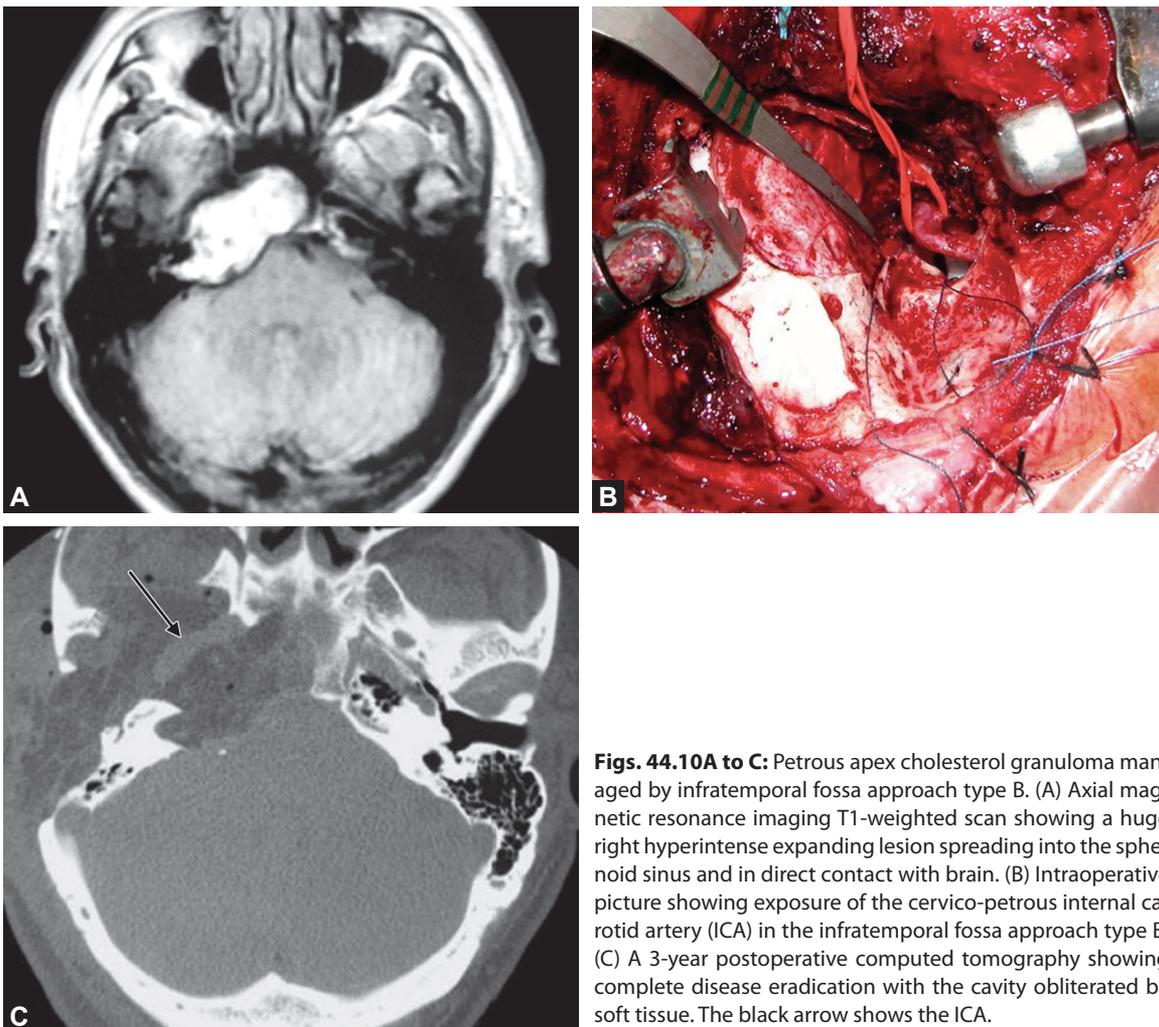
Figs. 44.8A to C: Petrous apex cholesterol granuloma managed by an infralabyrinthine approach. (A) The magnetic resonance imaging (MRI) fast spin echo sequence displaying a high-signal image in the left petrous apex. (B) Immediate postoperative T1-weighted MRI displaying a correct position of the drainage tube and the presence of some postoperative secretion, which can be differentiated from petrous apex cholesterol granuloma because of low-signal intensity. The high-signal rim around the cavity, which represents the capsule of the granuloma, is seen. (C) Postoperative computed tomography scan.

the possibility of complete cyst removal from the clivus and sphenoid sinus. The disadvantages of ITFA type B are blind sac closure of the external auditory canal with loss of conductive hearing and retraction of mandibular condyle with section of V3 branch of the trigeminal nerve, leading to difficulty in mastication.

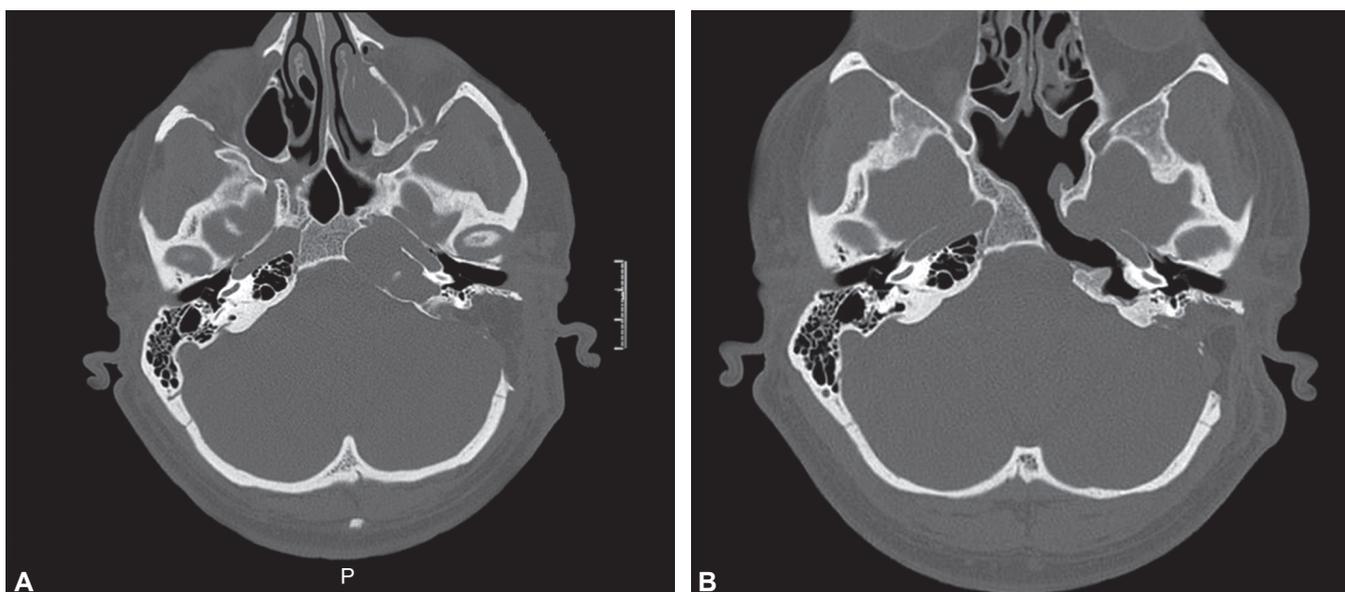
The translabyrinthine (TL) and TO approaches are employed for patients without serviceable hearing and huge lesions. These approaches allow a wide and direct exposure of the PA. We advocate combining the ITFA type B with the TL or TO approach for those lesions extending anteriorly to the ICA and eroding the clivus to achieve



Figs. 44.9A and B: (A) Infralabyrinthine approach showing isolation of the lesion between the facial nerve, jugular bulb, and the posterior semicircular canal and (B) after opening the cavity and drainage of contents.
(PFD: Posterior Fossa Dura; PSC: Posterior Semicircular Canal; FN: Facial Nerve; JB: Jugular Bulb; ELS: Endolymphatic Sac; *: Lesion).



Figs. 44.10A to C: Petrous apex cholesterol granuloma managed by infratemporal fossa approach type B. (A) Axial magnetic resonance imaging T1-weighted scan showing a huge right hyperintense expanding lesion spreading into the sphenoid sinus and in direct contact with brain. (B) Intraoperative picture showing exposure of the cervico-petrous internal carotid artery (ICA) in the infratemporal fossa approach type B. (C) A 3-year postoperative computed tomography showing complete disease eradication with the cavity obliterated by soft tissue. The black arrow shows the ICA.



Figs. 44.11A and B: In this case of petrous apex cholesterol granuloma, an endoscopic approach was performed after an infralabyrinthine approach failed to completely drain the lesion. (A) The computed tomography scan showing the lesion involving the petrous apex and reaching the sphenoid. Note that a transmastoid infralabyrinthine approach has been performed. (B) Postoperative picture showing drainage from both the lateral and endoscopic approaches.

complete removal of the cyst wall. If the PACG extends into sphenoid sinus or abuts its posterior wall, a transnasal endoscopic approach is indicated (Figs. 44.11A and B).

Petrous Bone Cholesteatoma (PBC)— The PBC is an epidermoid cyst of the petrous portion of the temporal bone. The PBC in the PA is less frequent than secondary cholesteatoma and the slow-growing nature of this lesion leads to a delayed diagnosis. Apart from this, congenital PBC presenting with sudden onset facial palsy is often misdiagnosed as Bell's palsy due to the normalcy on otoscopy. Sanna et al.^{26,27} proposed a classification for PBC that is widely used (Table 44.2). Of the categories in the classification, the infralabyrinthine-apical, massive, and apical PBC affect the PA. The PBCs can be either congenital or acquired. The term congenital cholesteatoma is used to describe lesions that are believed to arise from epithelial cell rests within the temporal bone. The acquired type is the result of a medial invasion of cholesteatoma from the tympanomastoid region. Supralabyrinthine PBCs were the most common type, with 92 (45.8%) cases followed by the massive PBCs with 72 (35.8%) cases (Table 44.3). Apical PBCs were rare with just four (2%) of cases in the series. Two of the main differentiating factors between congenital and acquired PBC are the following: while in acquired PBC, the cavity connects to the external auditory canal and hence is susceptible to secondary bacterial

invasion, and in the congenital form the lesion is essentially sterile. Second, the mastoid will be more sclerosed in the acquired form due to long-standing infection in comparison to congenital PBC where the pneumatization is normal. The most common clinical presentation of PBC is hearing loss and facial palsy and is common to both congenital and acquired cholesteatomas. In one of the largest series of PBC published ($n=246$), we found an incidence of 64% of hearing loss and 53% of facial palsy. The commonest sites for the involvement of the facial nerve in our series were the tympanic part (94%), geniculate ganglion (84%), and the labyrinthine portion (69%). The facial nerve was involved in multiple segments in most of the patients.

On CT scan, PBCs appear as a nonenhancing, expansile lesions with bone destruction. On MRI they show intermediate to low-signal intensity on T1-weighted images and high-signal intensity on T2-weighted images. There is no enhancement after contrast administration. Diffusion-weighted imaging is useful in diagnosis of cholesteatomas, as the lesions often show restricted diffusion, a feature that can be particularly beneficial in detection of recurrent cholesteatomas after surgical resection.⁴

The main factors to be taken into consideration while treating PBC are complete eradication of the disease, preservation of facial nerve function, prevention of cerebrospinal fluid leak, and hence meningitis, cavity obliteration, and

Table 44.2: Updated Sanna classification for Petrous Bone Cholesteatomas

<i>Updated Sanna Classification of Petrous Bone Cholesteatomas</i>		
<i>Class</i>	<i>Cholesteatoma location</i>	<i>Relations and Features</i>
Class I: Supralabyrinthine (Figs. 44. 1A and B)	Centered on the geniculate ganglion area of the FN and the anterior epitympanum	<p>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</p> <p>Features: Usually associated with fistula of the semicircular canals, erosion of tegmen, involvement of the FN</p>
Class II: Infralabyrinthine (Figs. 44. 2A and B)	Centered on the infracochlear, infralabyrinthine and hypotympanic cells	<p>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</p> <p>Features: Fistula of the semicircular canals, Erosion of the cochlea, jugular bulb, carotid canal, involvement of the lower cranial nerves</p>
Class III: Infralabyrinthine-Apical (Figs. 44. 3A and B)	Involves infralabyrinthine cell tracts extending medially into the petrous apex	<p>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)</p> <p>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</p>
Class IV: Massive (Figs. 44. 4A and B)	Centered on the otic capsule	<p>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</p> <p>Features: various degrees of destruction of the otic capsule, involvement of FN</p>

Contd...

Contd...

Class V: Apical (Figs. 44. 5A and B)	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 sphenopetroclival 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.
<i>Subclasses</i>	<i>Relations and Features</i>	
Clivus (C) (Figs. 44. 6A to D)	Superior and mid clival extensions are seen from massive, infralabyrinthine-apical and apical PBC whereas the lower clival involvement is a feature of infralabyrinthine-apical PBC	
Sphenoid Sinus (S) (Figs. 44.7A to D)	Sphenoid sinus involvement is seen from anteromedial extensions of massive, infralabyrinthine-apical and apical PBC; it is a rare extension	
Nasopharynx (N) (Figs. 44.8A to C)	It is the rarest extension of the PBC; it is an extension of infralabyrinthine-apical or massive PBC, which may extend through the clivus beneath the sphenoid sinus into the 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.	

(FN: Facial nerve; pICA: Petrous Internal Carotid Artery; IAC: Internal Auditory Canal; CO: Cochlea; M: Mandible; ICA: Internal Carotid Artery; PC: Paraclival Carotid; MMA: Middle Meningeal Artery; SS: Sigmoid Sinus; TS: Transverse Sinus; EV: Emissary Vein; Lv: Labbe's Vein; JV: Jugular Vein; TA: Transverse process of the atlas; C1: 1st cervical vertebra; OC: Occipital Condyle; PP: Pterygoid Plate; ET: Eustachian Tube; SPH: Sphenoid; ZA: Zygomatic process; V3: 3rd branch of the Trigeminal nerve; V2: 2nd branch of the trigeminal nerve; VII: FN, IX: Glossopharyngeal nerve; XII: Hypoglossal nerve; Ch: Cholesteatoma).

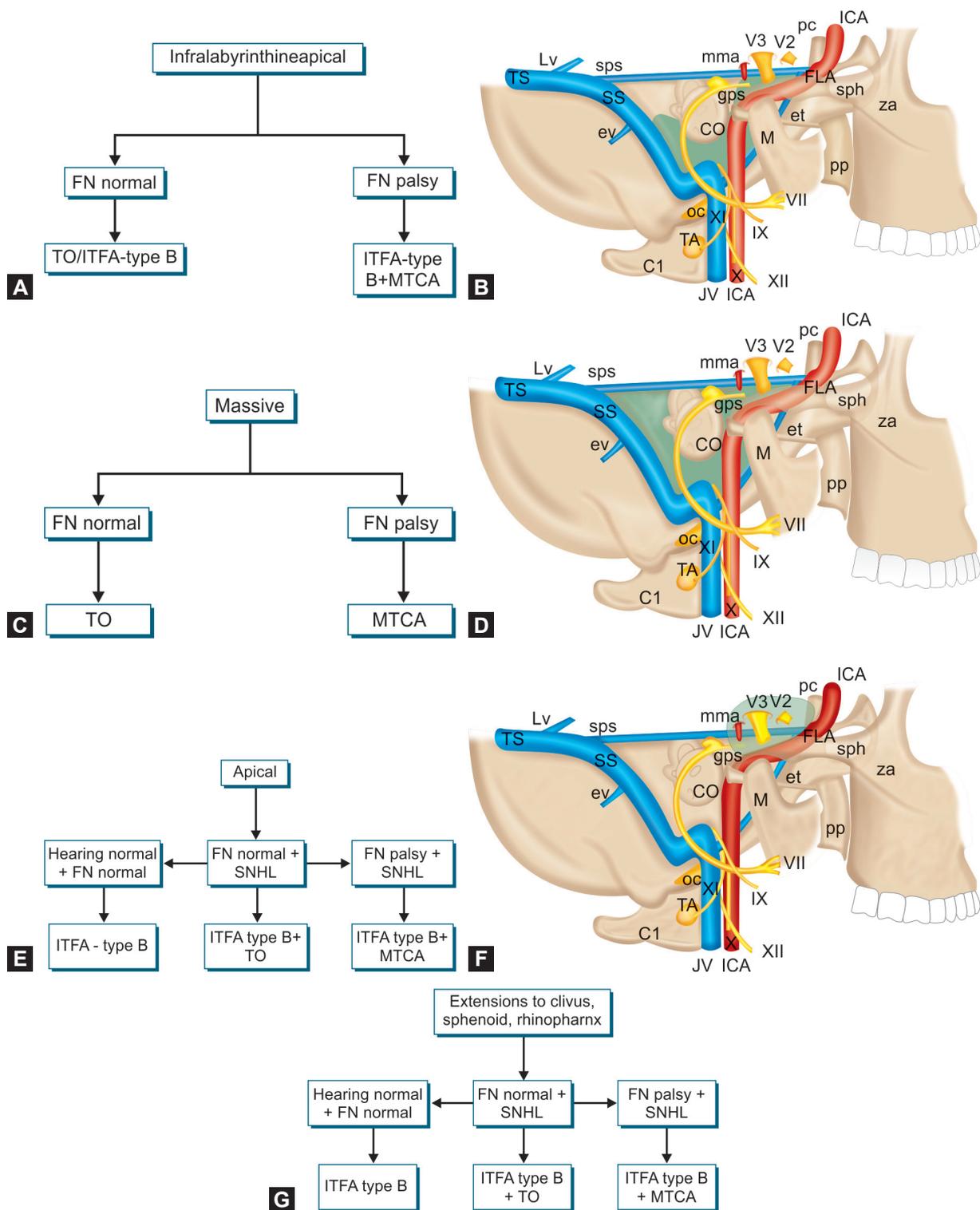
Table 44.3: Sanna classification of PBCs versus the approach used in this series

<i>Petrous Bone Cholesteatomas at the Gruppo Otologico</i>						
	<i>SL</i>	<i>IL</i>	<i>IL-A</i>	<i>M</i>	<i>A</i>	<i>Total</i>
Total	92 (45.8%)	18 (8.9%)	15 (7.5%)	72 (35.8%)	4 (2%)	201 (100%)

(SL: Supralabyrinthine; M: Massive; IL: Infralabyrinthine; IL-A: Infralabyrinthine-apical, A: Apical)

hearing preservation whenever feasible. The choice of surgical approach has evolved from radical petromastoid exenteration with marsupialization of the cavity to closed and oblitative techniques following complete eradication. The decision regarding the surgical approach depends on several factors, but the most significant of which are the

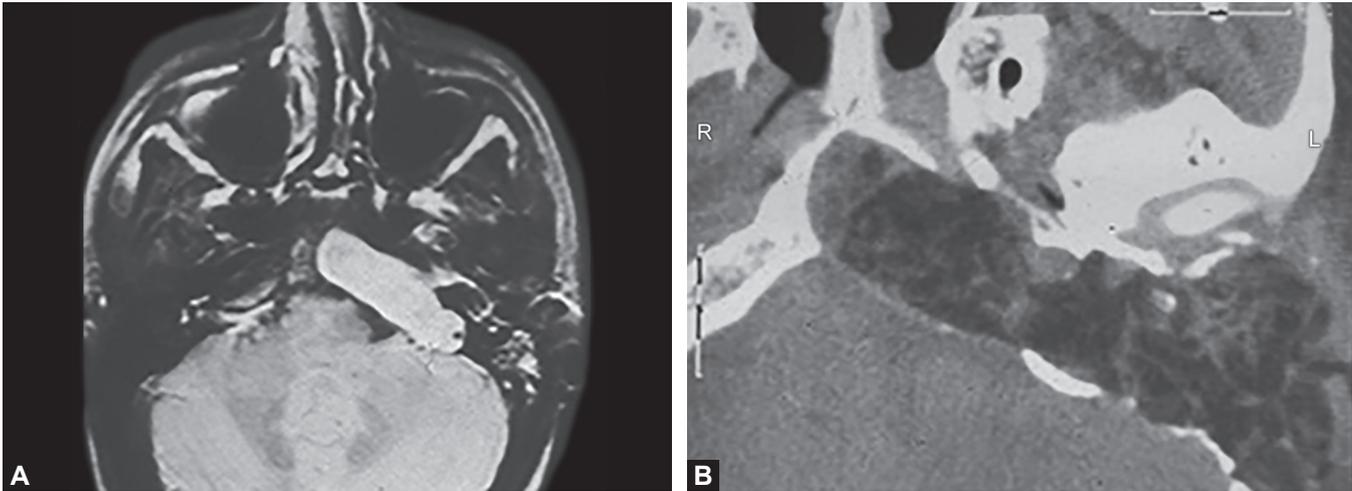
extent of the disease and preoperative facial nerve function. The approach is chosen depending on the type of PBC and its extent, which should be determined according to the CT scan and MRI findings. Algorithms for the management of infralabyrinthine-apical, massive, and apical PBCs presented in Figure 44.12. to 44.14 are representative



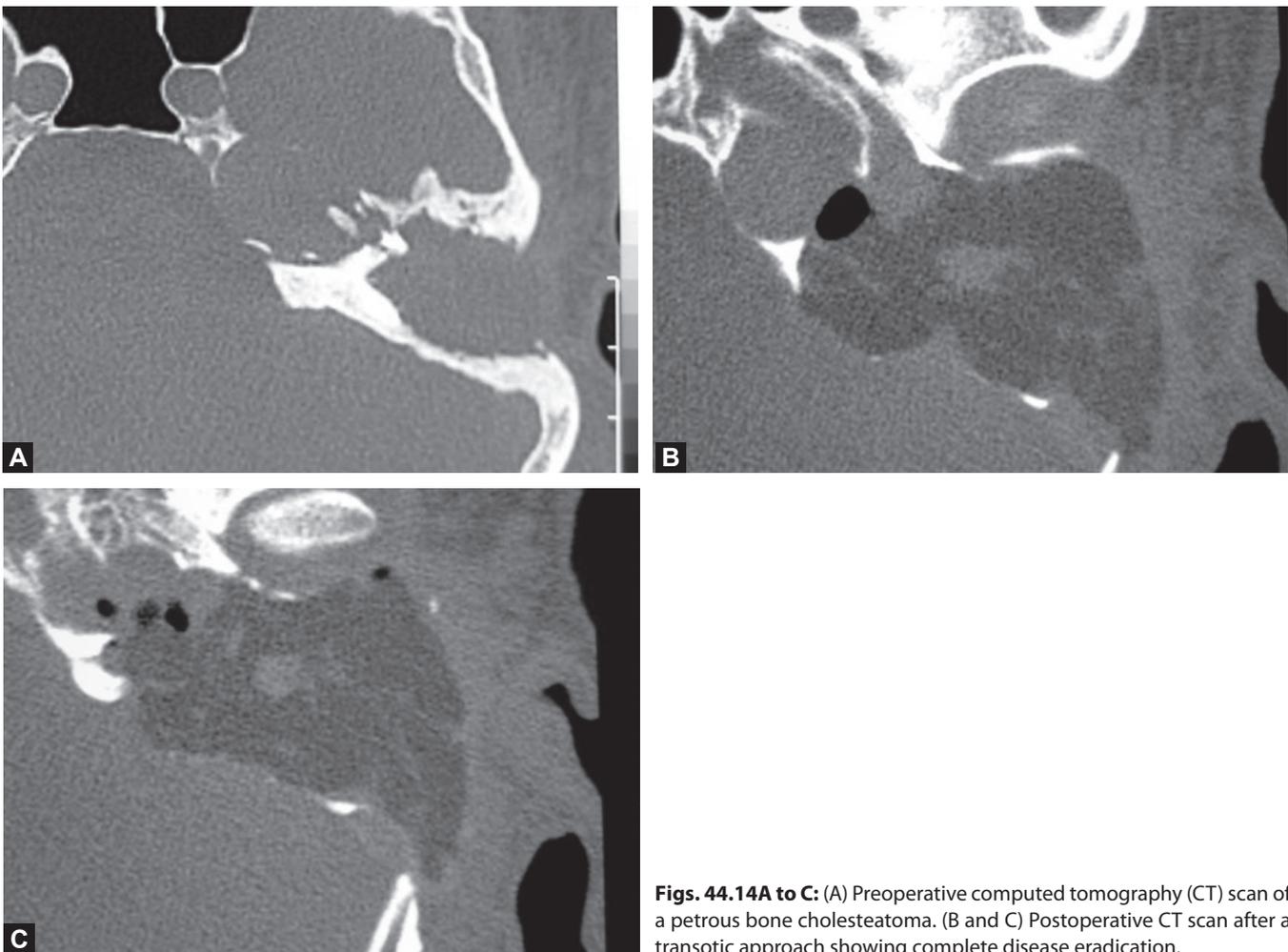
Figs. 44.12A to G: Algorithm for surgical management of the petrous bone cholesteatoma with illustrations.

pictures of our management of some of the difficult cases that we encountered.

Meningioma involving the PA—Meningiomas involving the PA and the adjacent clivus are some of the most



Figs. 44.13A and B: (A) Preoperative magnetic resonance imaging of a right petrous bone cholesteatoma, massive type with extensions into the clivus and posterior fossa. (B) Postoperative computed tomography showing the extent of bone removed by a modified transcochlear approach with complete clearance of the disease.

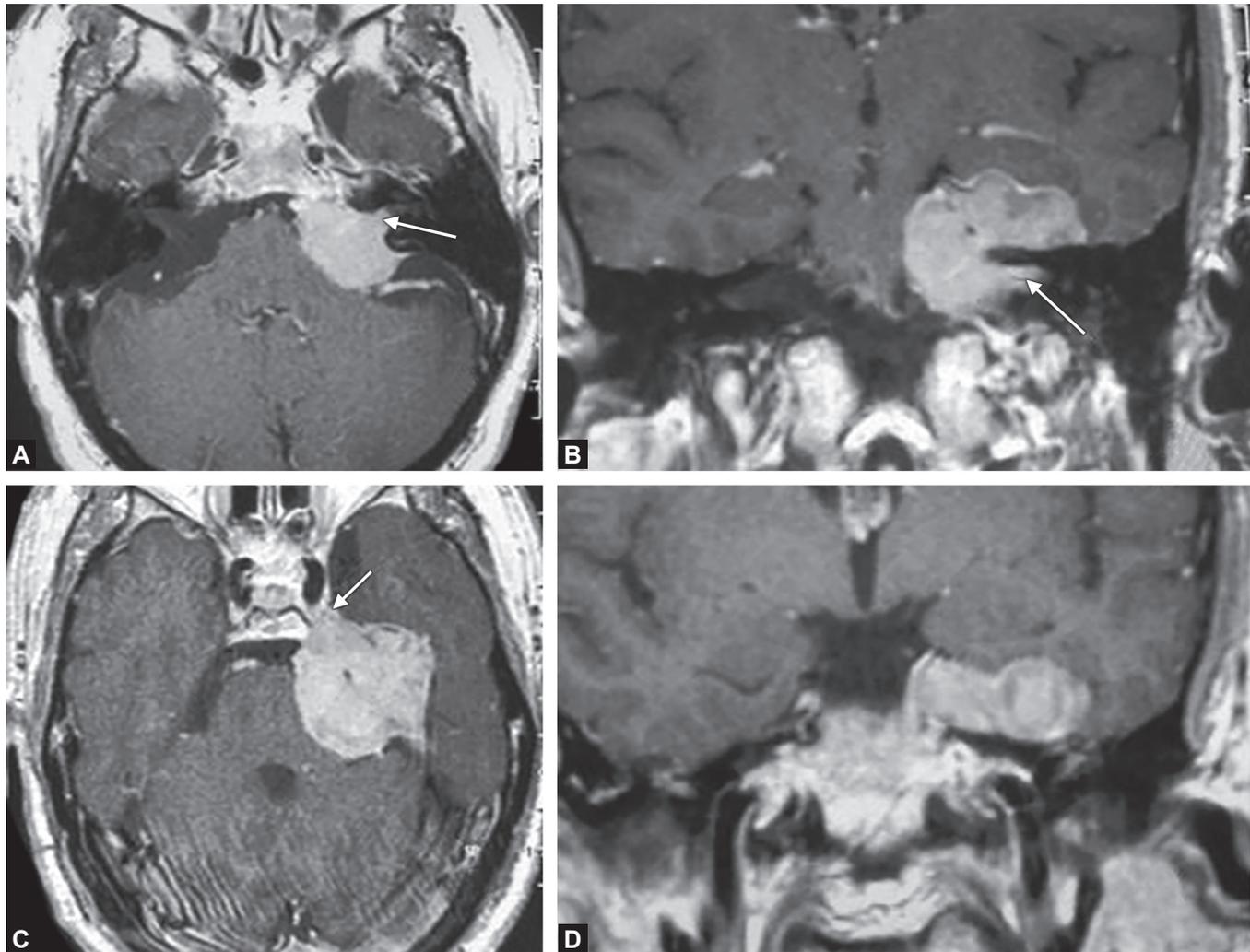


Figs. 44.14A to C: (A) Preoperative computed tomography (CT) scan of a petrous bone cholesteatoma. (B and C) Postoperative CT scan after a transotic approach showing complete disease eradication.

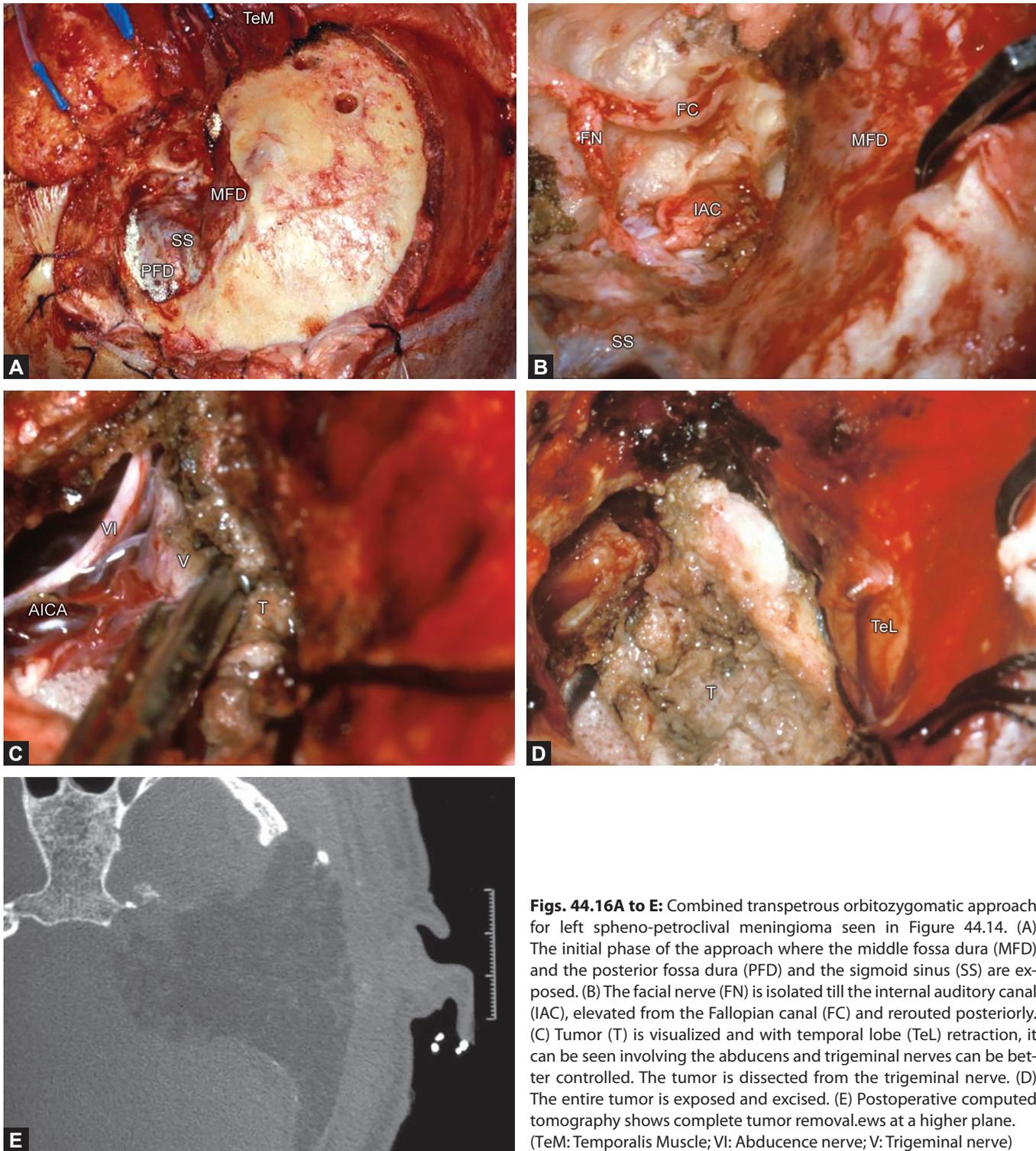
difficult lesions to manage in the area. The petroclival and posterior petrous face meningiomas involve the PA. Petroclival meningiomas arise from the clivus or petroclival junction medial to the trigeminal nerve. They may extend along the petrous pyramid, middle fossa, or into the cavernous sinus. Large tumors displace or encase the basilar artery or its branches. As with most other lesions of the PA, in our series, hearing loss (76.5%), tinnitus (44.4%), instability (51.8%), and vertigo (22.2%) were the most common presenting complaints. Other features include facial paresis, ataxic gait, trigeminal neuralgia, facial anesthesia, and headaches. On CT, meningiomas appear to be hyperattenuating to brain, iso- to hypointense on T1-weighted

images, and iso- to hyperintense on T2-weighted images. They also demonstrate postcontrast enhancement (Figs. 44.15A to D). Meningiomas may cause hyperostosis of the PA, a finding that is most evident in CT.⁴ Angiography of both vertebral and ICA is done to determine the major arterial branches and the blood supply to the tumor.

The adoption of a particular approach for petroclival meningiomas depends on the location of the tumor. Tumors of the upper and middle clival area tend to involve the PA. The upper clival tumors can be approached by the frontotemporal orbitozygomatic approach (Figs. 44.16A to E) or the middle fossa transpetrous approach as described by House et al. in 1986.²⁸ Tumors of the middle



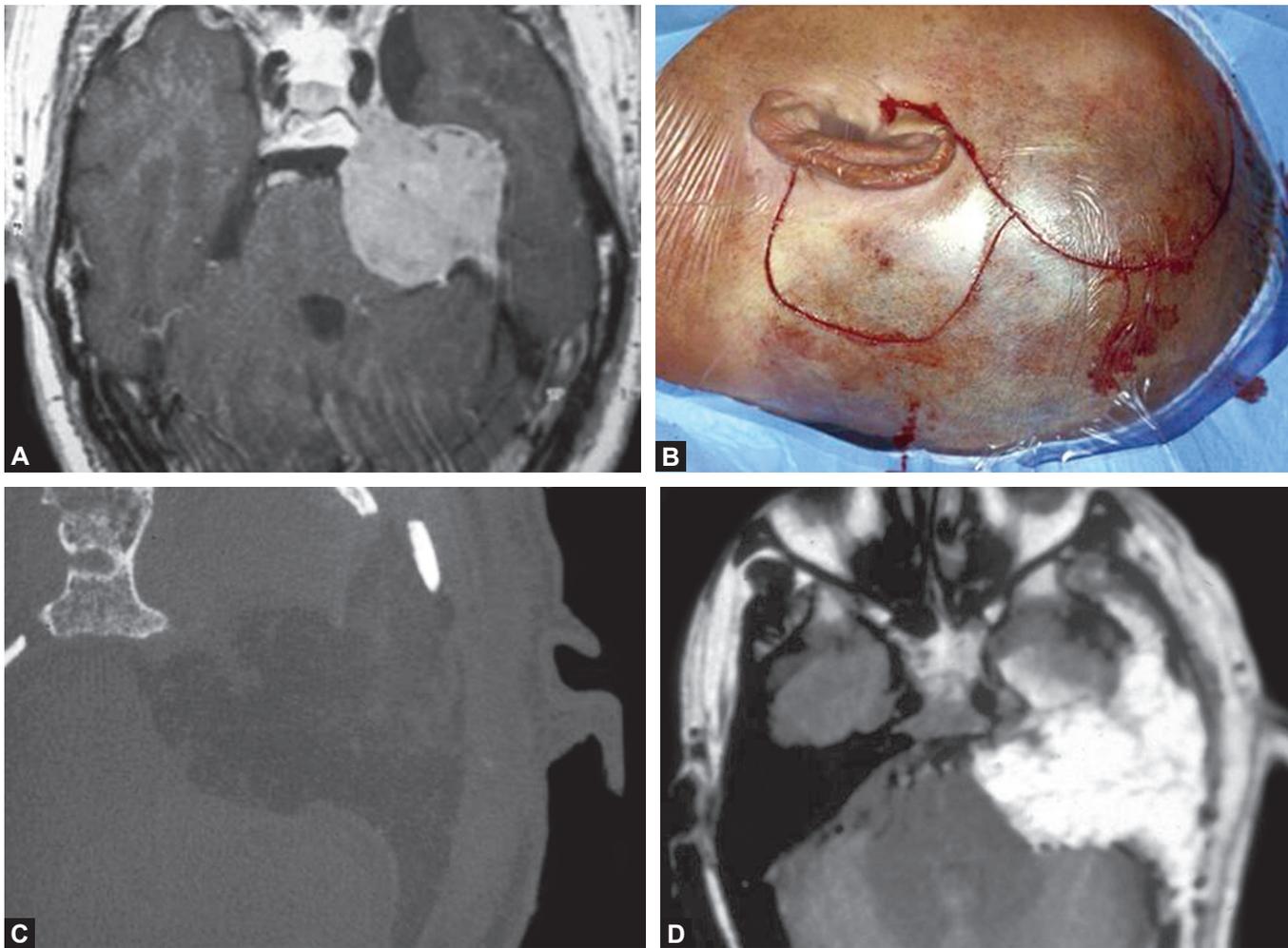
Figs. 44.15A to D: Left petroclival meningioma; (A) Preoperative T1-weighted magnetic resonance imaging with gadolinium enhancement, axial view, shows the tumor anterior to the internal auditory canal (arrow) and growing into the prepontine cistern. (B) Coronal view shows tumor involvement of both the middle and posterior cranial fossae and into the internal auditory canal (arrow). (C) Axial and (D) coronal view.



Figs. 44.16A to E: Combined transpetrous orbitozygomatic approach for left sphenopetroclival meningioma seen in Figure 44.14. (A) The initial phase of the approach where the middle fossa dura (MFD) and the posterior fossa dura (PFD) and the sigmoid sinus (SS) are exposed. (B) The facial nerve (FN) is isolated till the internal auditory canal (IAC), elevated from the Fallopian canal (FC) and rerouted posteriorly. (C) Tumor (T) is visualized and with temporal lobe (TeL) retraction, it can be seen involving the abducens and trigeminal nerves can be better controlled. The tumor is dissected from the trigeminal nerve. (D) The entire tumor is exposed and excised. (E) Postoperative computed tomography shows complete tumor removal. (TeM: Temporalis Muscle; VI: Abducence nerve; V: Trigeminal nerve)

clivus can be adequately approached by means of modified transcochlear (Figs. 44.17A to D), retrolabyrinthine, suboccipital, or the TL approaches, of which the modified

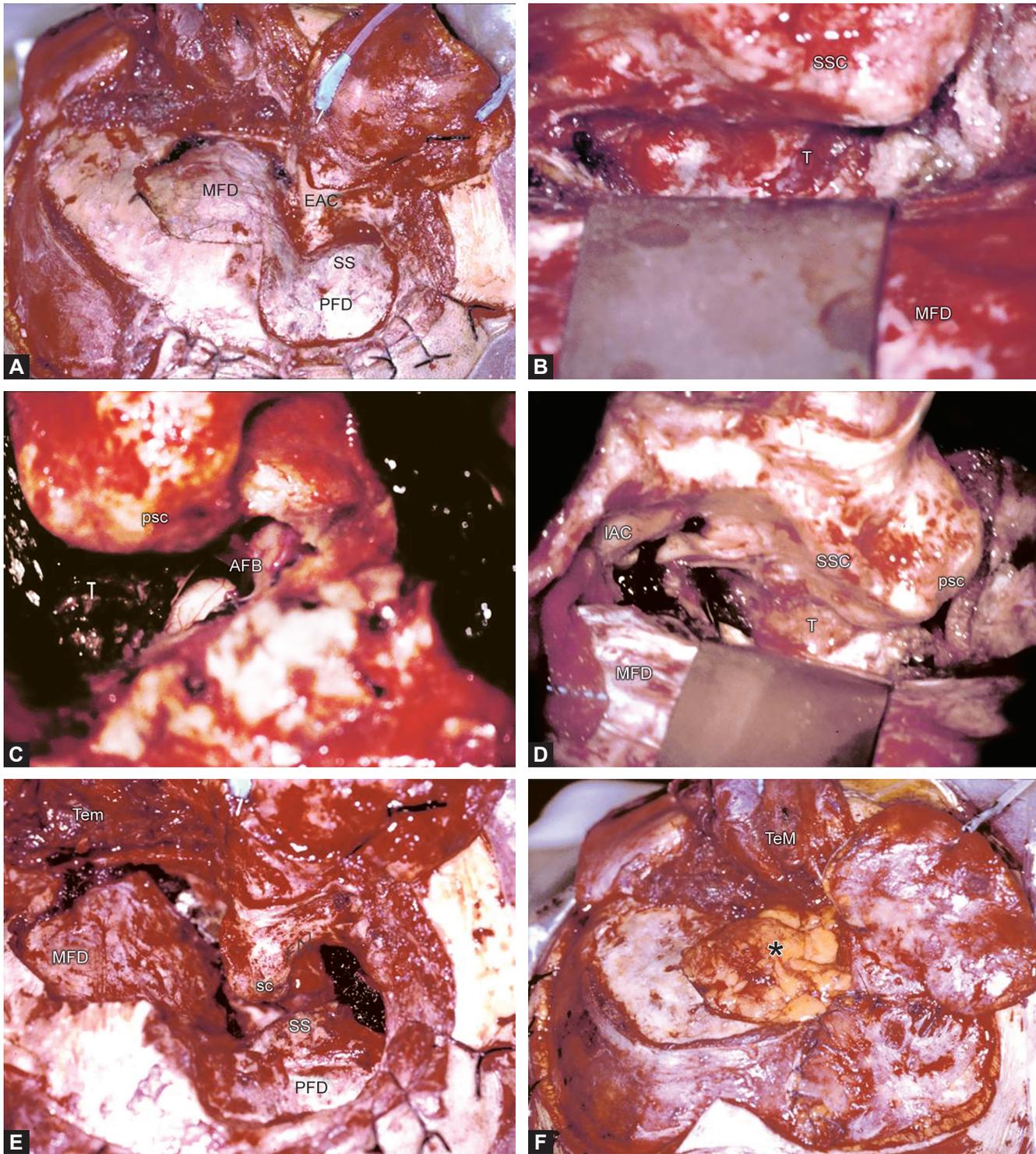
transcochlear approach type A is the safest. The modified transcochlear approach offers a wide and safe exposure, needs no cerebellar or brainstem retraction, permits rem-



Figs. 44.17C and D: Sphenopetroclival meningioma excised using a modified transcochlear-transtentorial approach. (A) In this T1-weighted image, the lesion is seen to involve the petrous apex, middle, and posterior fossa. (B) Skin incision. (C) The computed tomography scan shows complete tumor removal. (D) The magnetic resonance imaging shows the area of the tumor replaced with fat.

oval of the invaded dura or bone, and can be extended to afford excellent control of anterior, superior, or inferior tumor extensions. The disadvantages are sacrifice of hearing and facial nerve impairment. The hearing preservation surgeries, however, provide restricted access to the tumor and also involve retraction of the cerebellum. The posterior petrous face meningiomas can be divided into lesions lying anterior, posterior, or centered on the internal auditory canal. Lesions lying anterior to the internal auditory canal tend to involve the PA and they can be tackled by the following policy: small lesions are managed by a middle fossa transpetrous approach and larger lesions not reaching the midline can be managed by a retrolabyrinthine subtemporal transapical approach (Figs. 44.18A to F) with or without transtentorial extension.

Paraganglioma involving the PA—Tympanojugular paragangliomas (TJPs) are benign tumors involving the middle ear cleft and the skull base. The Fisch class C3 and C4 tumors are generally considered as large tumors and it is this subset of tumors that have the potential to involve the PA. As TJPs grow out of the jugular foramen, they tend to follow two paths; one extending either into the carotid canal, the PA, and the clivus (Fig. 44.19) and two; into the intradural space through the medial wall of the jugular bulb involving the lower cranial nerves in the process. As a rule, ITFA type A with transcondylar-transtubercular extension can be used for C2–C4 tumors. If the tumor involves the clivus or foramen magnum, additional procedures such as modified transcochlear approach or the extreme lateral transcondylar approaches are necessary. If TJPs involve



Figs. 44.18A to F: Retrolabyrinthine subtemporal transapical approach for a right posterior petrous face meningioma seen in Figures 44.10A to D. (A) Retrolabyrinthine approach with middle fossa craniotomy has been performed. The middle fossa dura (MFD), posterior fossa dura (PFD), and the sigmoid sinus (SS) are exposed. The external auditory canal (EAC) is identified. (B) The middle fossa dura is elevated. The tumor can be identified involving the petrous apex. (C) At a lower magnification, a panoramic view of the tumor in the posterior fossa is seen. The acousticofacial bundle (AFB) and the posterior semicircular canal (PSC) are identified. (D) The dura of the posterior fossa has been opened from the subtemporal aspect and the tumor (T) identified; IAC: Internal Auditory Canal; SSC: Superior Semicircular Canal; (E) View after total tumor removal. (F) The retrolabyrinthine aspect has been obliterated with abdominal fat and the U-shaped musculoperiosteal flap has been repositioned and sutured. Abdominal fat (*) is also placed to obliterate the subtemporal opening. The temporalis muscle (TeM) is then sutured back to close the defect.

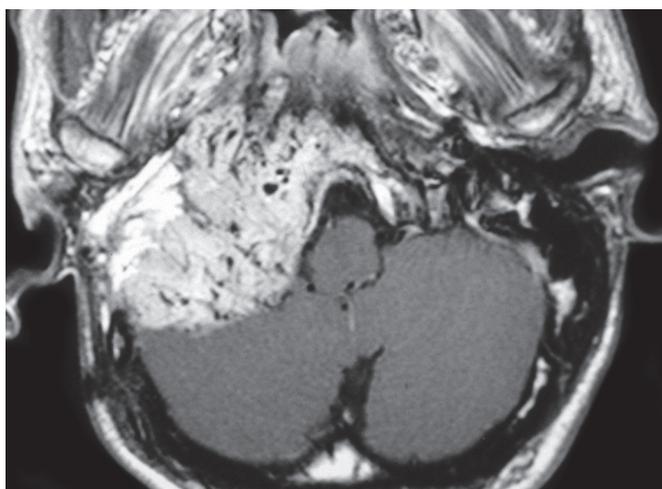


Fig. 44.19: Giant sized tumor compressing the cerebellum and invading clivus.

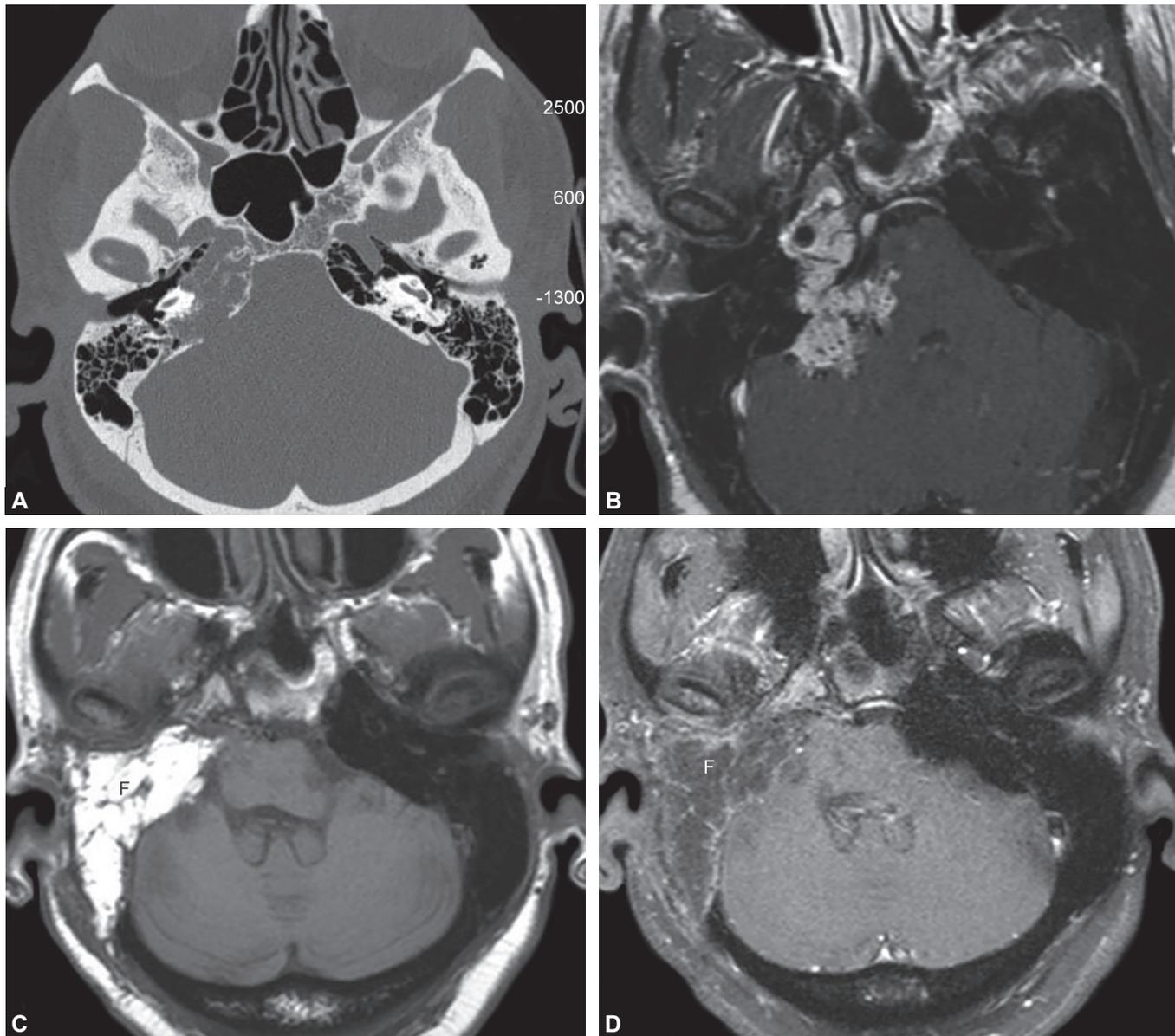
the cavernous sinus, it should be intentionally left intact to avoid compromising cranial nerves III, IV, and VI. For tumors extending to the foramen magnum and lower clivus, modified transcochlear approach type D or the extreme lateral approach may be used. Tympanojugular paragangliomas frequently involve the ICA due to their close anatomical proximity (Figs. 44.20A to D).^{29,30} When indicated, the tumor must be dissected from the arterial wall. This can be accomplished by subperiosteal and sub-adventitial dissection. When the tumor encapsulates the artery causing stenosis, manipulation without proper endovascular intervention may give rise to severe bleeding, incomplete removal, or cerebral vascular accident. The balloon occlusion is performed in the case where the ICA is infiltrated by the tumor and the collateral blood flow is sufficient. In cases with insufficient collateral blood flow, we have developed the technique of intra-arterial stenting of the ICA.³¹ It reinforces the ICA so that the surgeon can easily establish a cleavage plane on the external surface of the stent and more aggressively remove the tumor without the risk of blowout (Figs. 44.21 and 44.22). This new technique can allow reappraisal of selected cases previously suited only for subtotal resection.³²

Chordoma and chondrosarcoma—Chordomas are a rare and unusual group of malignant tumors that are believed to arise from the persistent rests of embryonal notochord. It is believed to have a male preponderance. The pathogenesis of chordomas also remains unclear. Chondrosarcomas of the skull base are rare slow growing locally aggressive malignant tumors that constitute 0.15% of all intracranial neoplasms. It has been hypothesized

that cranial base chondrosarcomas may originate from multipotential mesenchymal cells or from embryonal cartilage remnants of skull synchondroses.³³ The most common tumor sites of origin of both the entities nearly overlap and have been reported to be the petroclival, petro-occipital, spheno-occipital, and the sphenopetrosal synchondroses. Both chordomas and chondrosarcomas cause symptoms primarily related to cranial nerve involvement like dysfunction of extraocular movement with diplopia, hearing loss, dizziness and tinnitus, sensory disturbances of the face, and dysphagia. Pain and headache may be present due to erosion of bone or pressure due to the tumor. The CT most commonly reveals bone destruction and a soft tissue mass with tumoral calcification, the latter being encountered in 56% of tumors. On T1-weighted MRI, chondrosarcomas have a low-to-intermediate signal intensity and are isointense or hypointense to gray matter. On proton density and T2-weighted images, they have high-signal intensity and are hyperintense to gray matter. Contrast enhancement is typically heterogeneous (Figs. 44.23A and B). The radiologic distinction of chondrosarcoma and chordoma is usually not possible.

Surgery is considered the standard of care in the treatment of skull base chordomas and chondrosarcomas. Proton beam radiotherapy or radiosurgery is often used as adjuvant. The surgical intervention needs to be aggressive in these lesions. Small lesions with good preoperative hearing can be managed by the middle fossa transpetrous approach with extensive removal of the bone of the PA (Figs. 44.24A and B). Larger lesions with involvement of the vertical or the inferolateral aspects of the horizontal ICA necessitate a wider approach such as the ITFA type B (Figs. 44.25A to D) or its combination with a TO approach (Figs. 44.26A to D). If the preoperative hearing is good, then a retrolabyrinthine subtemporal transtentorial approach can be employed (Figs. 44.27 and 44.28). Extensive transdural cases will necessitate the addition of a modified transcochlear approach. In two of our series of 13 patients with chondrosarcomas, the petrous ICA showed angiographic signs of stenosis. One of the patients underwent permanent balloon occlusion of the ICA in the presence of adequate collateral flow through the circle of Willis. The second patient underwent stenting of the cervico-petrous ICA because of inadequate contralateral circulation in the circle of Willis (Figs. 44.29A to D).

Facial nerve neuroma—Facial nerve neuromas are uncommon benign neoplasms of Schwann cells. They



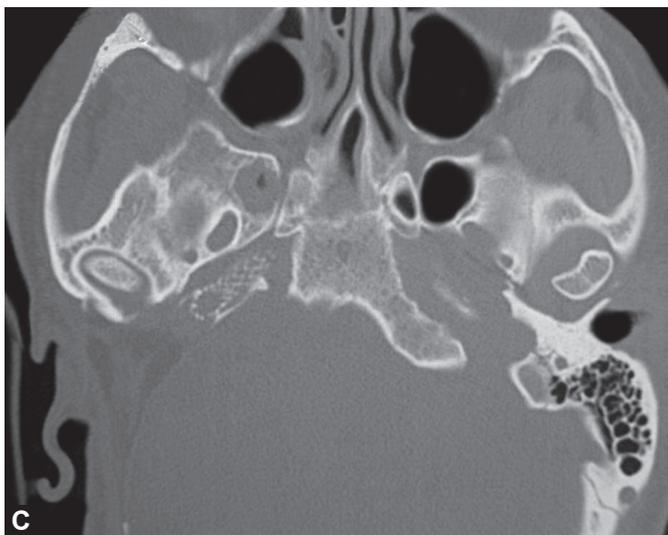
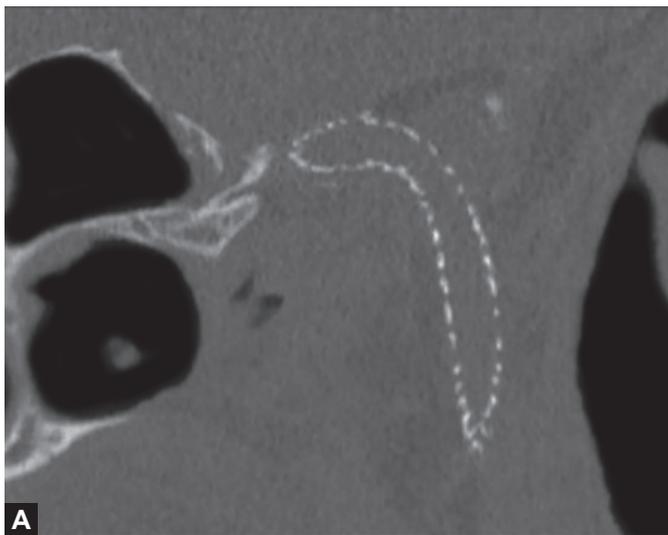
Figs. 44.20A and D: (A) The portion of the petrous apex medial to the internal carotid artery is involved by the tumor. Therefore, this tumor is classified as the Fisch class C3. (B) Note the vertical portion of the internal carotid artery completely encased by the tumor. (C) Postoperative T1 enhanced image, F: fat. (D) The magnetic resonance imaging (MRI), fat suppression axial view shows no residual tumor, (F: Fat).

compose only 0.8% of all intrapetrous mass lesions.³⁴ Although tumors involve all portions of the facial nerve, the geniculate ganglion is the commonest site and hence also most likely to involve the PA. Symptoms in facial nerve neuroma have an insidious onset that also depends on the location and extent of the lesion. Progressive or sudden facial palsy is a common symptom. In our series of 103 facial nerve tumors, only 76% of our patients presented

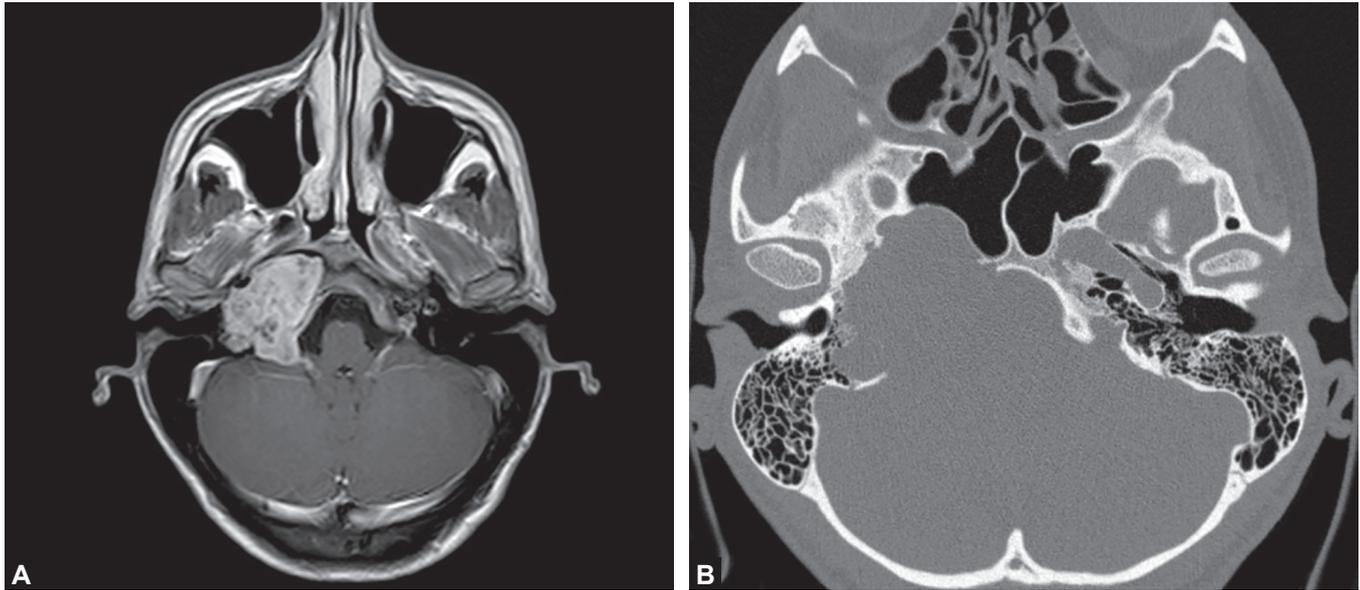
with the classic gradual facial palsy. Other presentations included sudden, recurrent, or hemifacial palsy. Facial nerve function was normal in 9.5% of the cases. In about 5% of patients with Bell's palsy a facial nerve neuroma is found to be the cause.³⁴ Due to the proximity of the tumor to the cochlea, a sensorineural or conductive hearing loss can occur, depending on whether the tumor originates proximal or distal to the geniculate ganglion. The CT scan



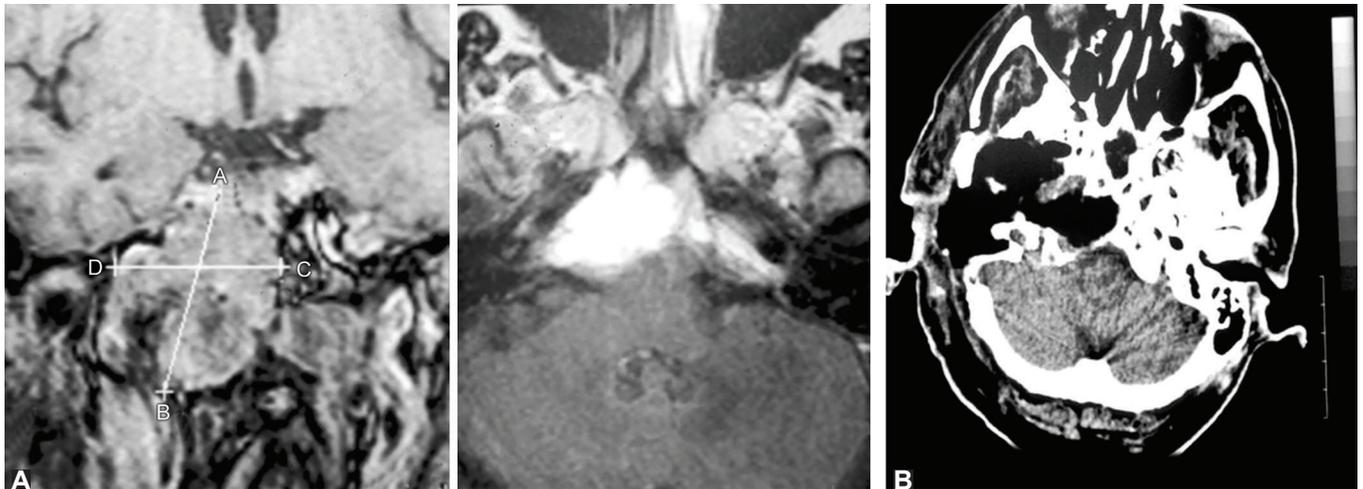
Fig. 44.21: Here the ICA has been anteriorly displaced. Note that the intra-arterial stent appears clearly through the thinned arterial wall. (ICA: Internal carotid artery, T: Tumor).



Figs. 44.22A to C: Postoperative computed tomography scans, coronal view (A) and axial views (B and C) showing the intra-arterial stent inserted into vertical and horizontal portions of the internal carotid artery.



Figs. 44.23A and B: (A) Axial T1-weighted magnetic resonance imaging with gadolinium enhancement showing right petroclival chondrosarcoma. Tumor shows heterogeneous enhancement and appears to be confined to the extradural space even though posterior cranial fossa dura is pushed medially. The internal carotid artery is encased by the tumor and anteriorly displaced. (B) Axial computed tomography scan with bone windows of the same tumor showing irregular bone erosion of the petrous apex.

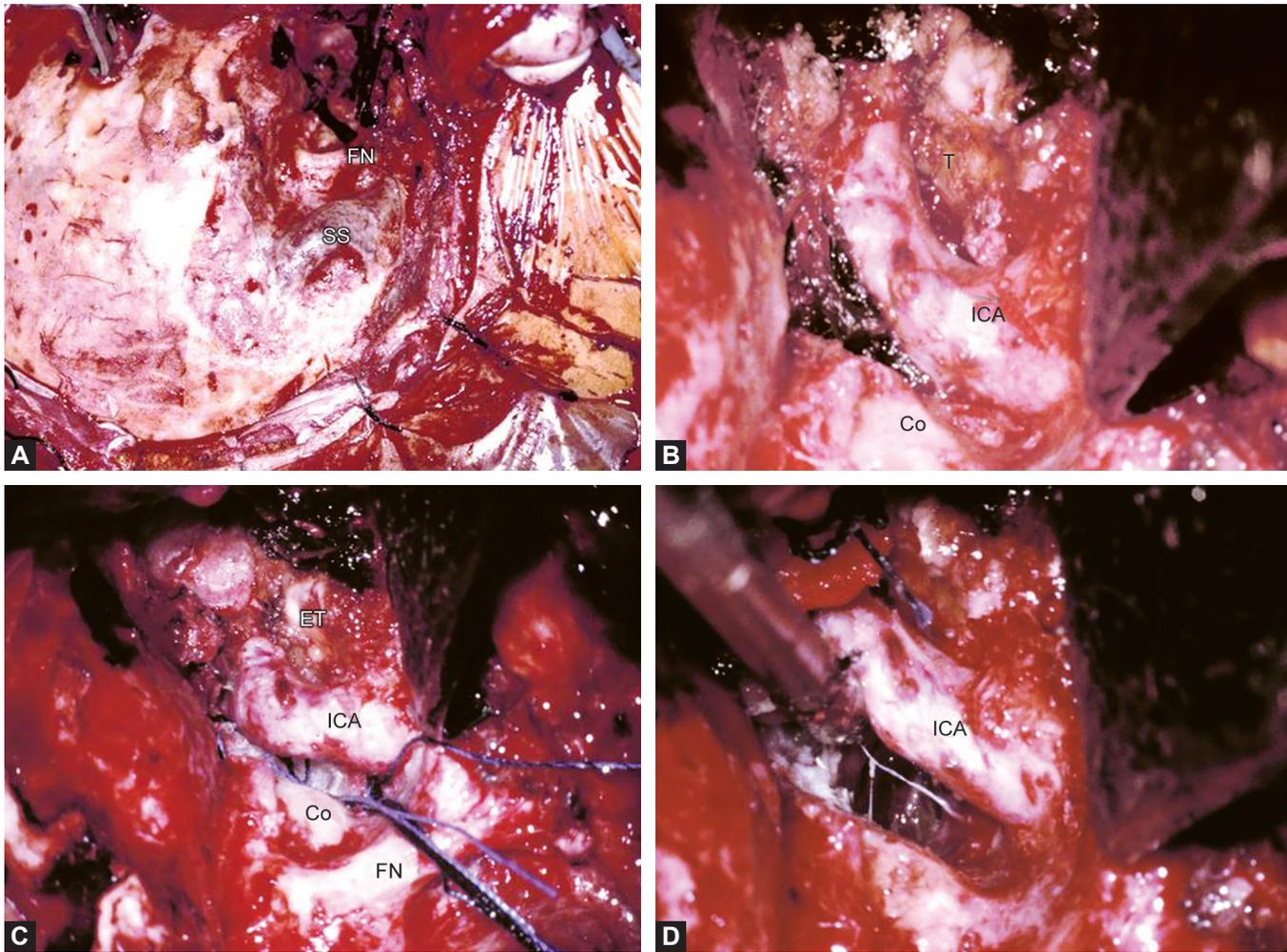


Figs. 44.24A to B: (A) The magnetic resonance imaging showing a right clival chordoma extending into the petrous apex and the clivus; (B) Postoperative computed tomography scan shows a complete tumor removal after a left middle fossa transpetrous approach.

shows enlargement of the facial canal, bone erosion, or an enhancing soft tissue mass and MRI shows hypo- or isointense on T1-weighted imaging, hyperintense, or isointense on T2-weighted imaging with marked contrast enhancement.

Therapeutic options for patients with facial nerve neuromas include surgical intervention, observation, and radiotherapy. Nowadays, surgical resection with facial

nerve repair is usually the standard management for patients with poor facial function [House-Brackmann (HB) grade III or worse]. In patients presenting with normal or near-normal facial nerve function, initial observation with periodic examination and imaging is usually recommended. However, on rare occasions surgeons can be faced with a situation in which the management deci-

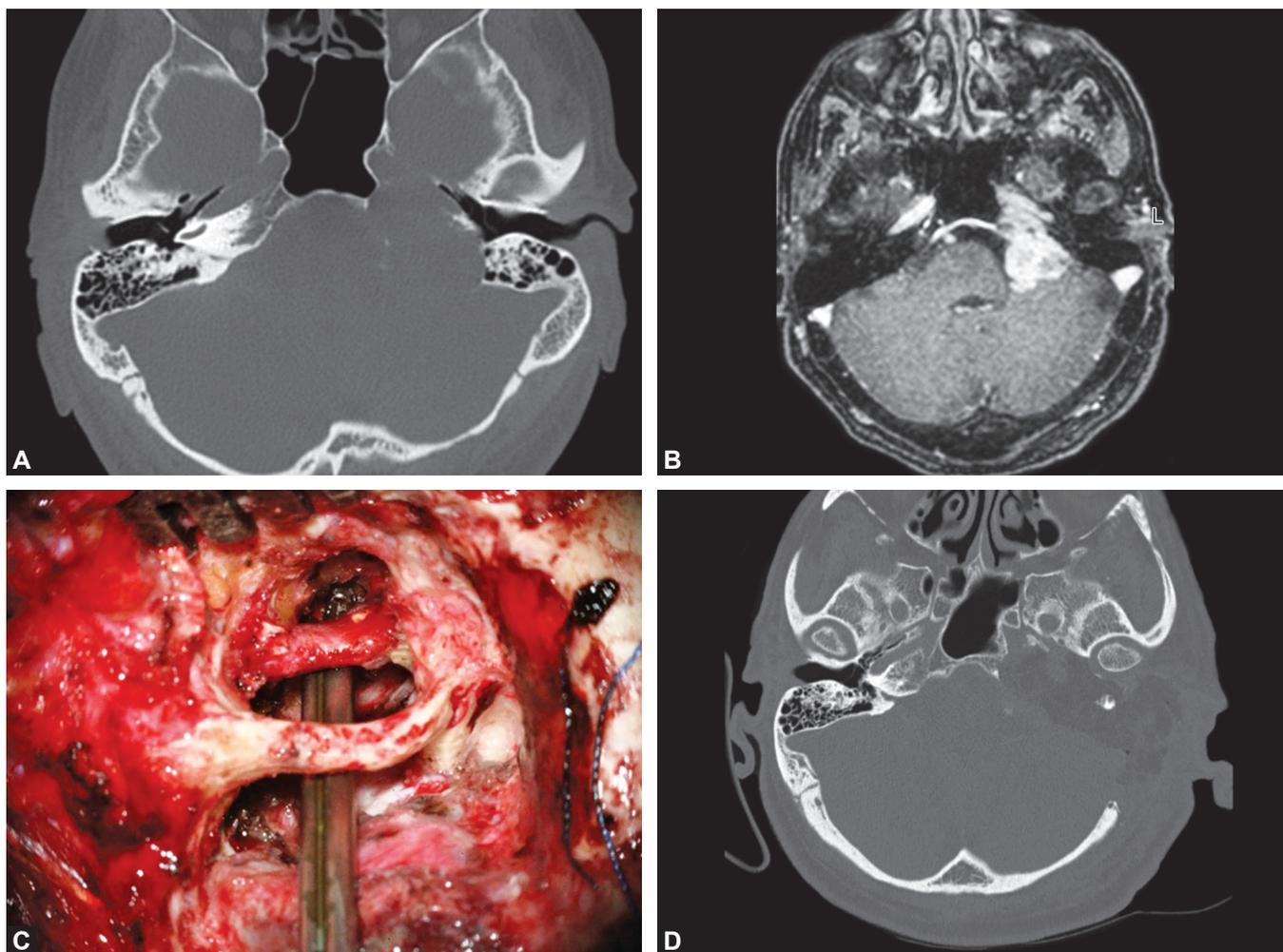


Figs. 44.25A to D: The infratemporal fossa approach type B performed to excise the chordoma in Figure 44.24. (A) The approach has been performed. (B) The internal carotid artery has been exposed anterior to the cochlea. Note the tumor occupying the petrous apex. (C) Neurosurgical cottonoids placed in the petrous apex for hemostasis. (D) The internal carotid artery is displaced laterally to ascertain total tumor removal. (T: Tumor; ICA: Internal Carotid Artery; Co: Cochlea; ET: Eustachian Tube; FN: Facial Nerve; SS: Sigmoid Sinus).

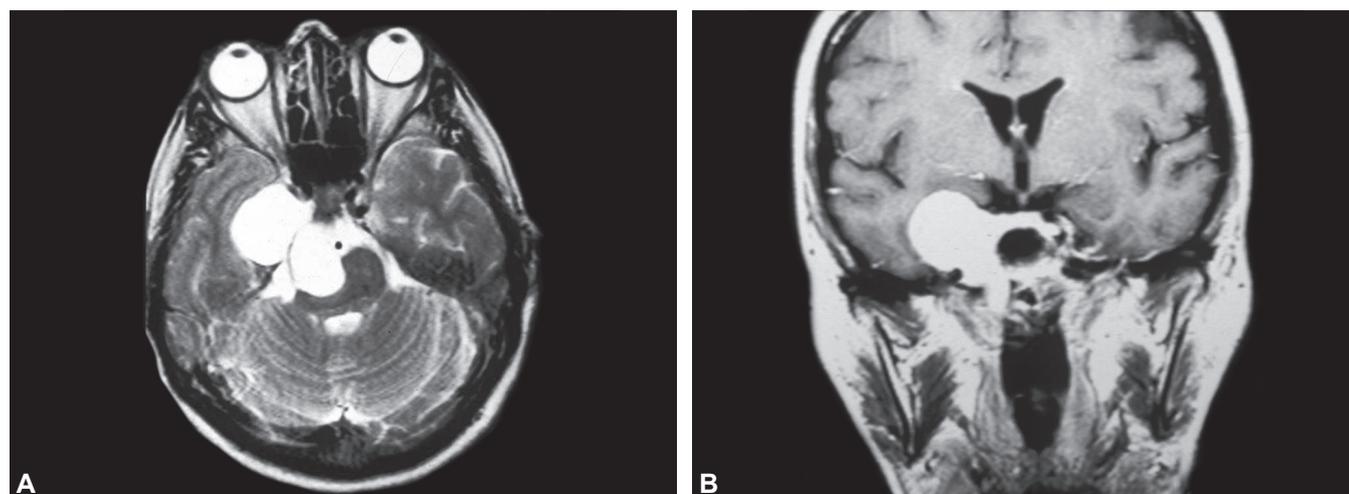
sion-making process is particularly challenging. In these complex cases treatment should be individualized. We recommend early surgical intervention regardless of the preoperative facial and hearing functions in the following cases: intratemporal facial nerve neuromas extending with a large tumor component into the parotid, multiple-segment tumors extending in both the cerebellopontine angle and the MCF, fast-growing tumors, and large tumors with temporal lobe compression. The surgical approaches of tumors involving the PA include MCF-transpetrous approach, combined MCF-transmastoid approach, transcochlear approach, or the TL approach. In most of the cases, it becomes necessary to reconstruct the cut sections of

the nerve after tumor removal. We prefer to use the sural nerve as a cable nerve graft to replace the lost segment of the nerve. A maximum recovery of the postoperative facial nerve function to HB grade III can be expected after nerve grafting.^{35,36}

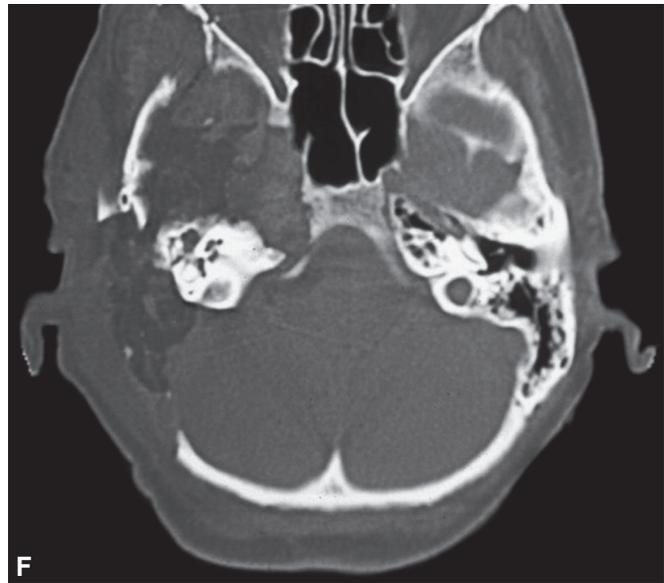
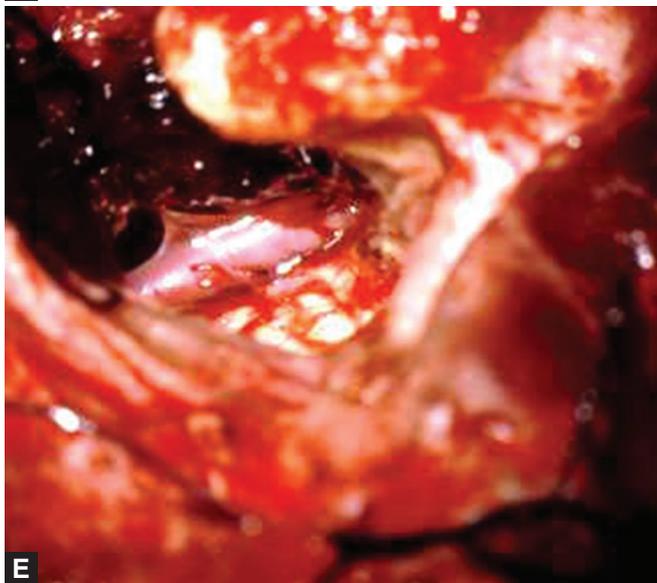
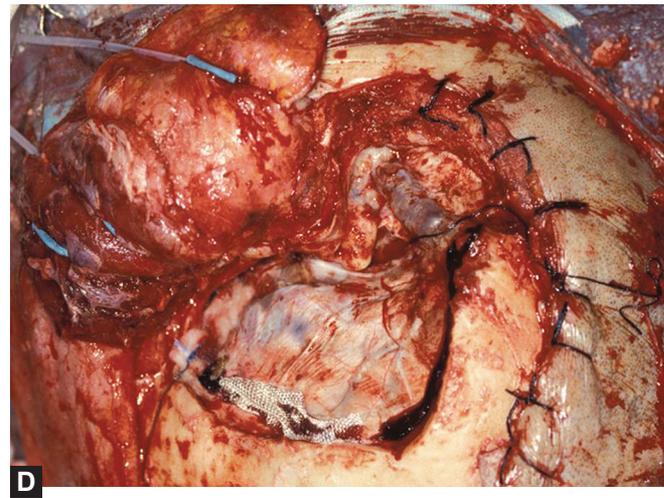
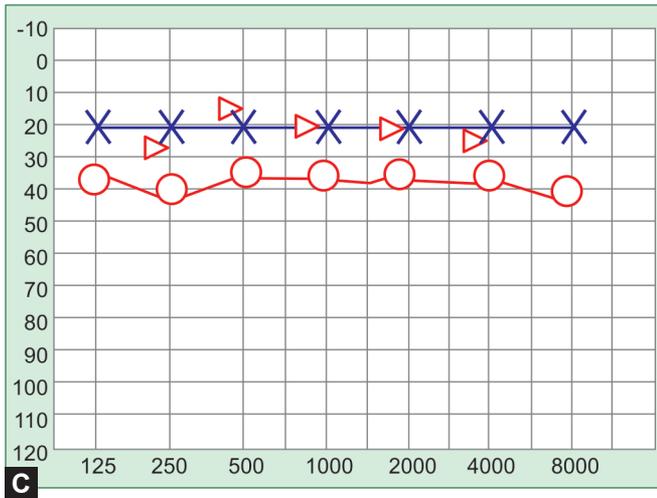
Trigeminal schwannoma—These are rare tumors of the Schwann cells. This is one of the important lesions involving the Meckel's cave area. Naturally, clinical presentation corresponds to a trigeminal nerve dysfunction: neuralgia, neurasthenia, or numbness. If the lesion is large, then mass effect symptoms may be present. Depending on their location they can be classified as: Type I - Predominantly middle fossa tumor (Fig. 44.30); the



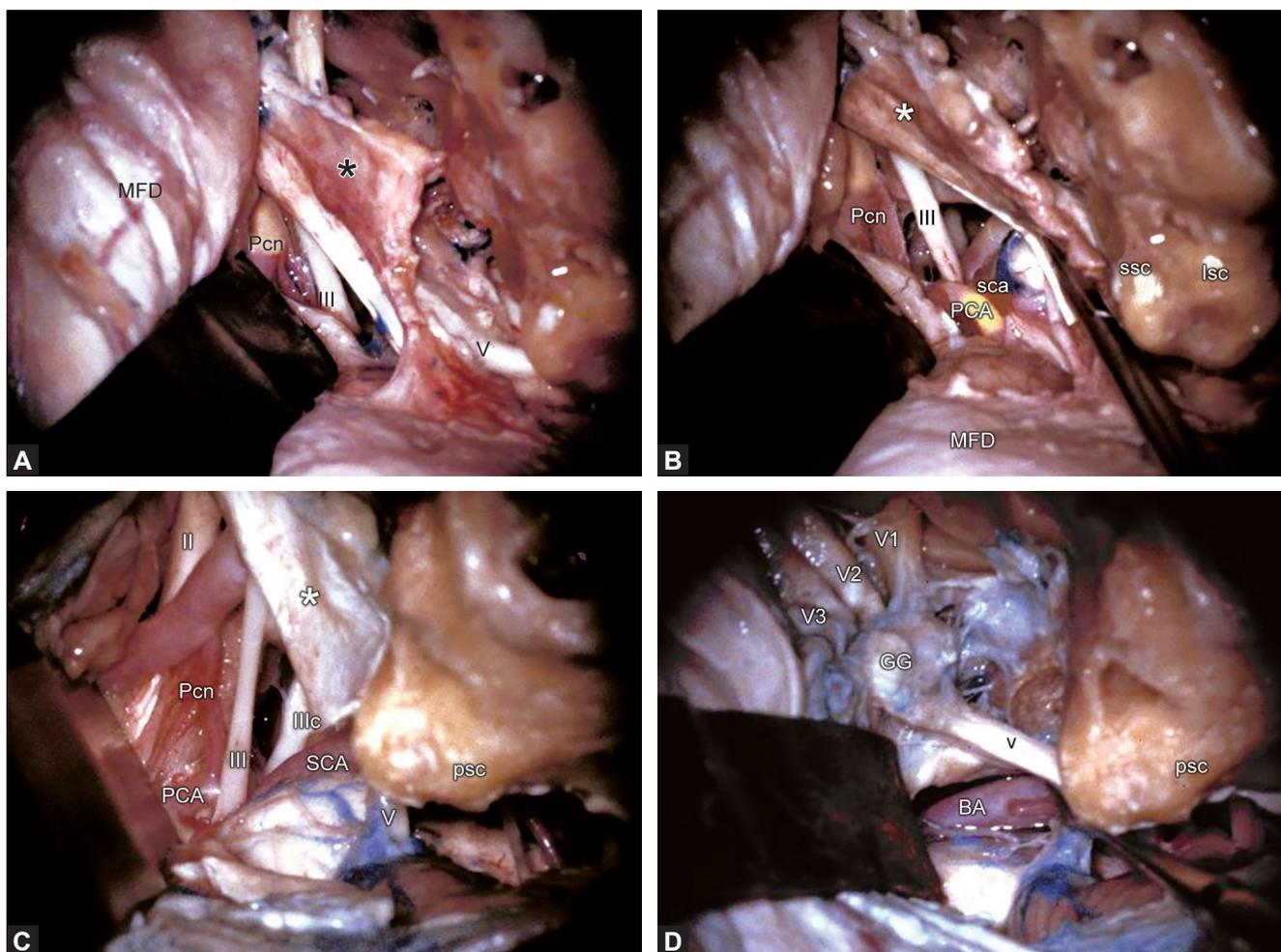
Figs. 44.26A and D: (A) Preoperative computed tomography (CT) and (B) T1-weighted magnetic resonance imaging with contrast of a chondrosarcoma involving the left petrous apex. (C) Using an infratemporal fossa approach type B with transotic approach the tumor was completely excised. (D) Postoperative CT scan showing complete tumor removal.



Figs. 44.27A to B:



Figs. 44.27A to G: (A) Axial and (B) coronal images of T2-weighted magnetic resonance imaging of a right petroclival chordoma. (C) Audiogram shows good hearing in the right ear. (D and E) A retrolabyrinthine subtemporal transapical approach was done with preservation of the middle ear and inner ear. The petrous apex has been drilled out and the basilar artery is visible. (F and G) Postoperative computed tomography scans showing complete tumor removal.

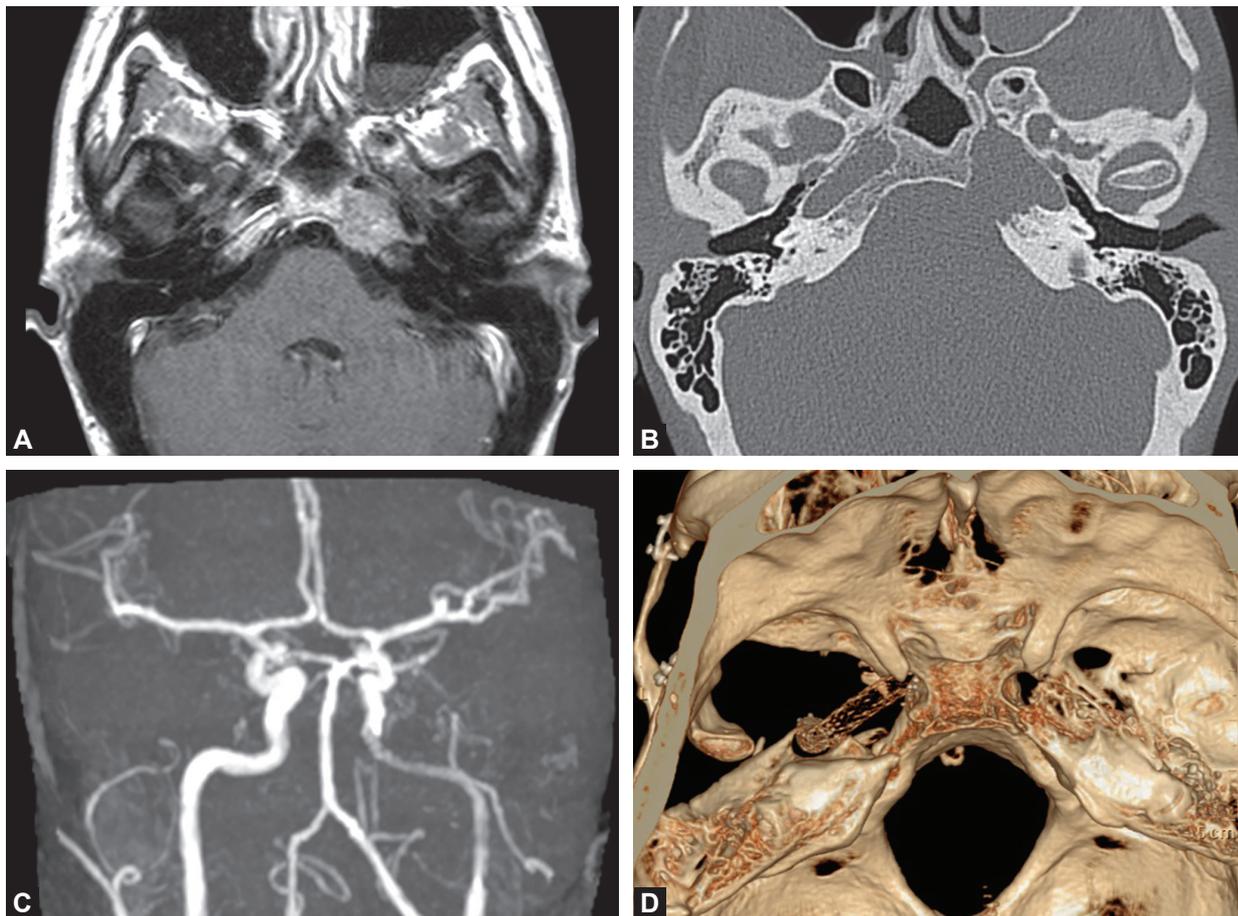


Figs. 44.28A to D: An anatomical dissection of retrolabyrinthine subtemporal transtentorial approach. (A) Following a retrolabyrinthine craniotomy, the middle fossa dura (MFD) is incised from the posterior to anterior, taking care not to injure the vein of Labbe. (B) The tentorium (*) is incised, taking care not to injure the trochlear nerve. (C) The tentorium is incised further till the tentorial notch is reached and cranial nerves II and III are exposed (IIIc is contralateral oculomotor nerve). The superior cerebellar artery (SCA), posterior cerebral artery (PCA), and the posterior communicating artery (Pcn) are seen. (D) The branches of the trigeminal nerve (V1, V2, and V3) and the Gasserian ganglion (GG) at the level of the lateral wall of the cavernous sinus. The basilar artery (BA) is also seen. (SSC: Superior semicircular canal, PSC: Posterior semicircular canal, and LSC: Lateral semicircular canal).

approach is frontotemporal orbitozygomatic (Figs. 44.31A to D). Type II - Predominantly posterior fossa tumor with limited middle fossa involvement (Fig. 44.32); modified transcochlear approach type C (Figs. 44.33A and B), retrolabyrinthine subtemporal transtentorial approach (for hearing preservation), or the middle fossa transpetrous approach. Type III - Significant middle and posterior fossa components (Fig. 44.34); combined frontotemporal orbitozygomatic and transpetrous approaches (Figs. 44.35A and B). Type IV - Extradural tumor involving the infratemporal fossa and surrounding structures (Fig. 44.36); the

approach used is an infratemporal transzygomatic approach (ITFA type D). A schematic drawing of the different approaches to the Meckel's cave is shown in (Fig. 44.37).¹ Another example of a massive dermoid in the Meckel's cave area excised by a TL approach with transapical, subtemporal orbitozygomatic is shown in (Figs. 44.38A to F).

Osteoclastoma (Giant Cell Tumor, GCT)—Giant Cell Tumors are a group of rare benign neoplasms that are most commonly found in the epiphysis of long bones. Of the total 1–2% of these lesions present in the head and neck with the skull base being a commonly reported site



Figs. 44.29A to D: (A) Axial T1-weighted magnetic resonance imaging (MRI) with gadolinium enhancement showing chondrosarcoma involving the left petrous apex. Note that the internal carotid artery (ICA) appears compressed. (B) Axial computed tomography (CT) of the same lesion showing erosion of the carotid canal. (C) The MRI with maximum intensity projections demonstrates narrowing of the cervical and petrous portions of the ICA. (D) A 3D shaded surface display reconstruction from the CT scan showing surgical defect that resulted from infratemporal fossa approach type B approach. Note the mesh of the stent inside the internal carotid artery.

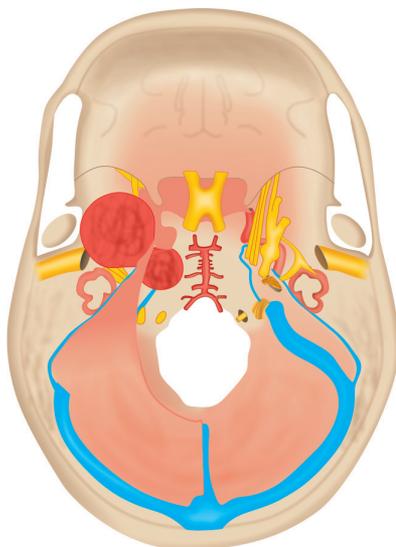
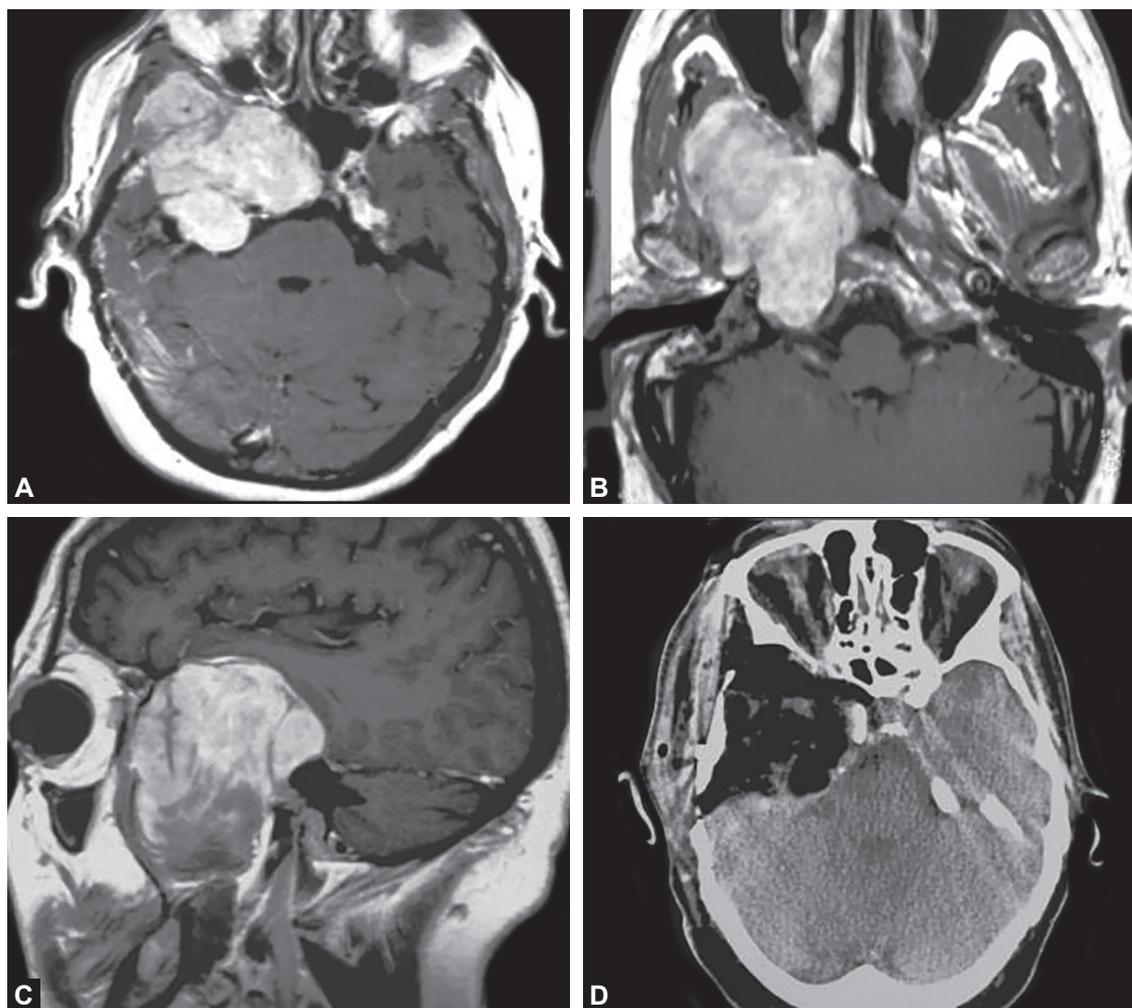


Fig. 44.30: Schematic diagram of a type I trigeminal schwannoma with a predominantly middle fossa component.



Figs. 44.31A to D: Massive right trigeminal schwannoma. (A) Axial magnetic resonance imaging showing the tumor extending into the sphenoid sinus and abutting the orbit. (B) Axial view at a lower cut shows extensive involvement of the infratemporal fossa. (C) Sagittal view shows the impingement of the middle fossa by the tumor. (D) Postoperative computed tomography scan after total removal via a combined infratemporal type C orbitozygomatic approach.

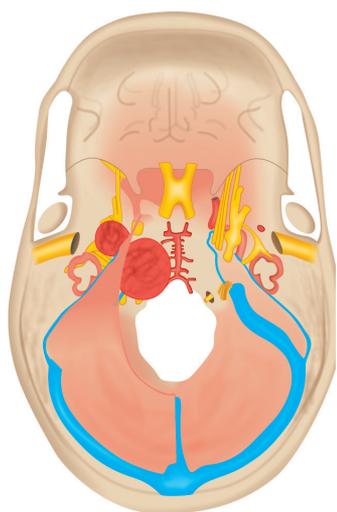
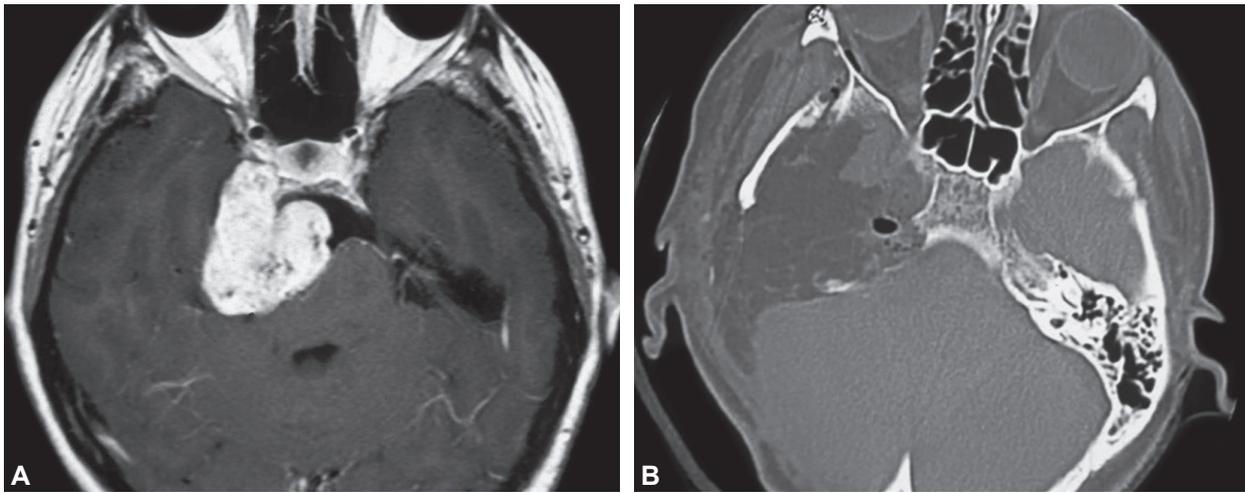


Fig. 44.32: Schematic diagram of a type II trigeminal schwannoma with a predominantly posterior fossa component.



Figs. 44.33A and B: (A) Preoperative and (B) postoperative pictures of a type II trigeminal schwannoma that was excised completely using a modified transcochlear approach.

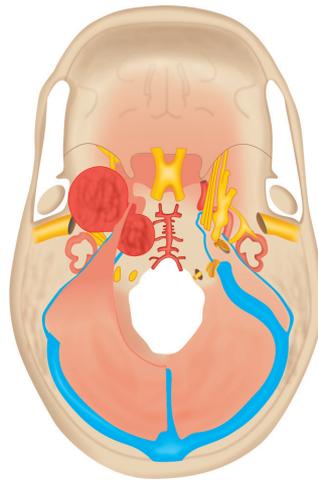
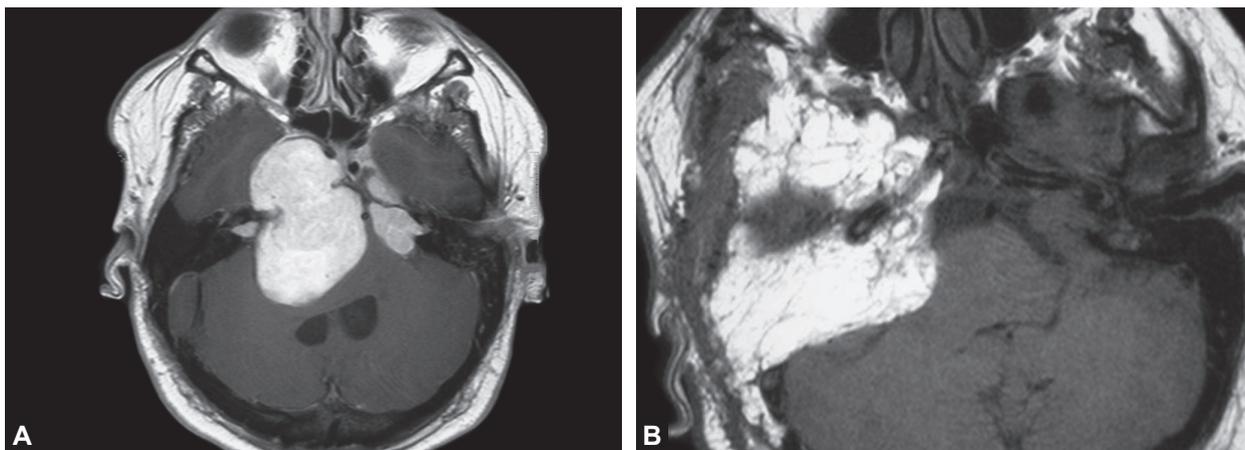


Fig. 44.34: Schematic diagram of a type III trigeminal schwannoma with significant middle fossa and posterior fossa components.



Figs. 44.35A and B: (A) Preoperative and (B) postoperative magnetic resonance imaging scans of a type III trigeminal schwannoma that was completely excised using a combined frontotemporal orbitozygomatic and transpetrous approaches.

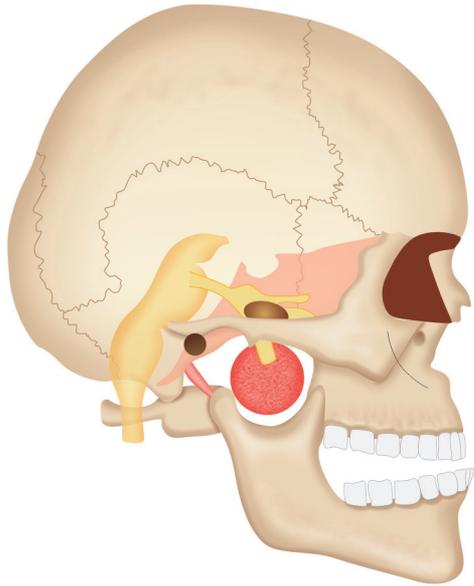


Fig. 44.36: Schematic diagram of a type IV trigeminal schwannoma where the tumor is extradural and infratemporal.

(temporal, sphenoid, and ethmoid bones). In the skull base the temporal bone is a common site of occurrence of GCTs. Although benign, these tumors have a locally destructive character that can be potentially dangerous in the presence of the intricate neurovasculature of the temporal bone and skull base.⁸ Hearing loss, tinnitus, and subcutaneous masses are the most commonly reported symptoms in GCTs of the skull base. The TM is often intact as the tumor is anteromedial to it. The radiological picture of a GCT is an osteolytic lesion. Both CT and MRI are essential for tumor staging and management. Although CT is superior to MRI in outlining tumor extent and bony destruction of the skull base, MRI is currently the best imaging modality for GCT because of its superior contrast resolution and multiplanar imaging capabilities that allow accurate tumor delineation. The GCT shows low intensity on T1 and heterogeneous high intensity on T2-weighted images. Gadolinium enhancement reveals areas of hypervascularity and enhancement with a very heterogeneous signal pattern.

The surgical approach depends on the position and extent of the tumor. Since most GCTs are found involving the temporal bone and the infratemporal fossa, the ITFA types B and D provide the ideal approach and exposure to these tumors with a blind-sac closure of the EAC. For tumors involving the MCF with or without dural invasion, a MCF craniotomy must be performed. For additional

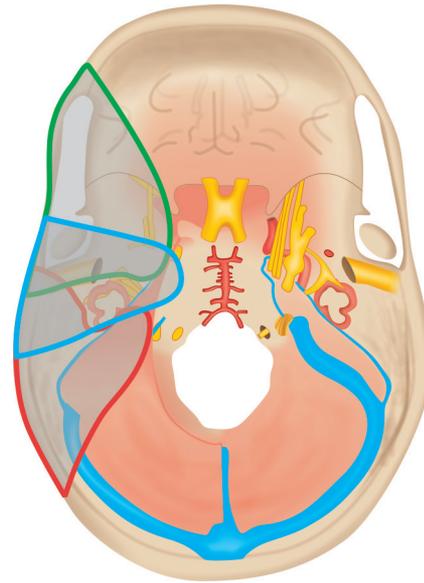
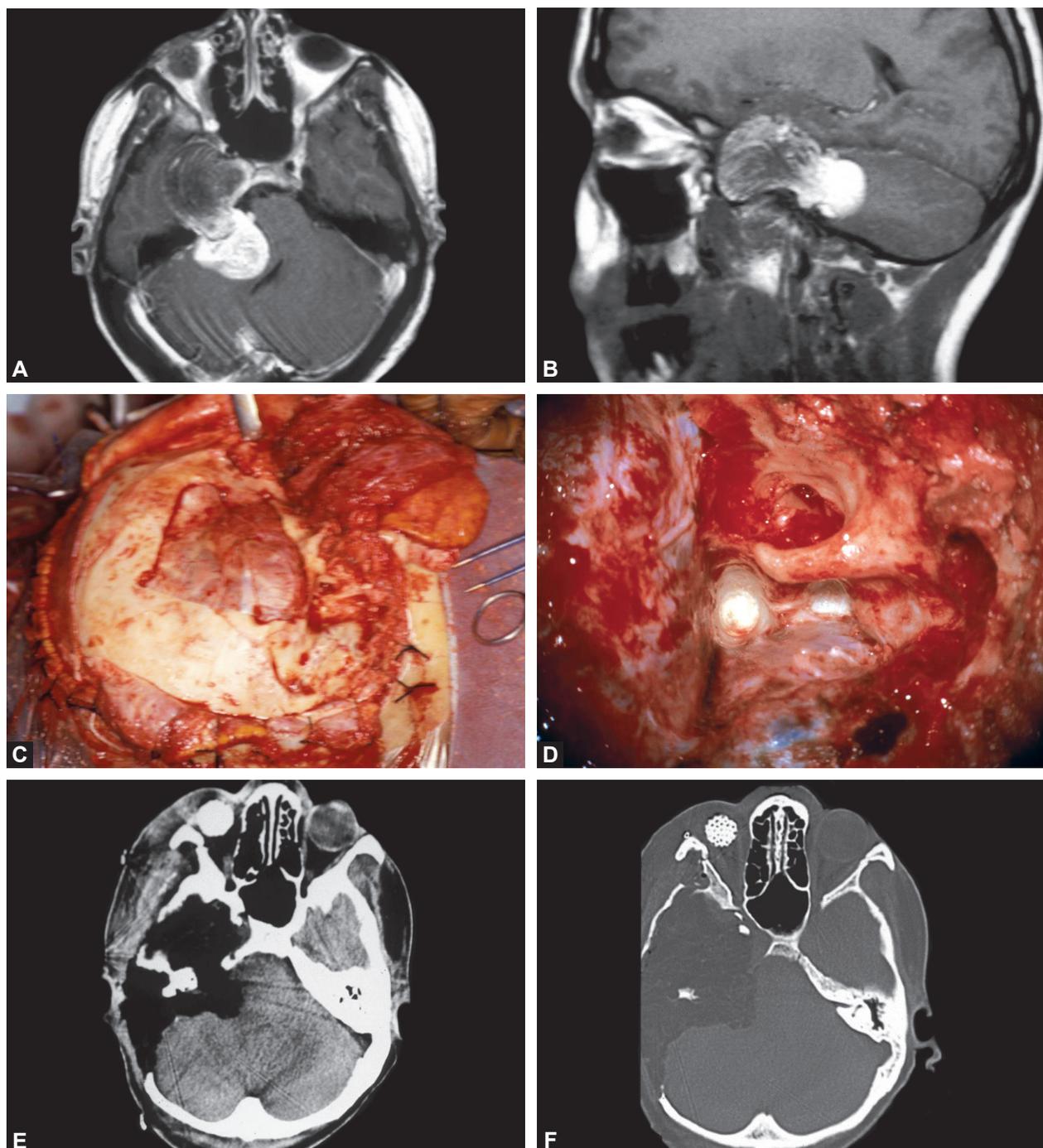


Fig. 44.37: Schematic diagram of the different approaches, radical and infratemporal, to lesions of the Meckel's cave.

exposure any of the skull base procedures may be added. Posterior extensions into the PCF though rare can be aggressive and extremely challenging to treat.

CONCLUSION

The PA is a complex area surrounded by vital neurovascular structures and the brain. Pathologies of this area are a diagnostic and surgical challenge. A variety of diseases, especially neoplasms, affect the PA and the skull base. Surgery is the treatment of choice for all neoplastic lesions. In the past, attempts at surgical excision of lesions of this area were fraught with serious complications and morbidities. However, in the era of modern skull base surgery, this is no longer the case due to the evolution of excellent surgical approaches aided by advances in neuroradiology, neuroanesthesia, and microsurgery. Modern imaging studies have also greatly increased the ability to diagnose lesions, especially in view of the often vague symptomatology associated with these lesions. Tumors involving the temporal ICA are no longer deemed inaccessible or inoperable due to the development of appropriate surgical approaches and the technique of stenting. In quaternary referral skull base centers across the world, PA as well as other skull base pathologies are routinely performed with minimal morbidities or complications.



Figs. 44.38A to F: Huge dermoid of the Meckel's cave area. (A) Axial and (B) sagittal view of T2-weighted MRI with gadolinium enhancement showing tumor involving the posterior fossa and the petrous apex. (C and D) Translabirithine approach with transapical, subtemporal orbitozygomatic extension. (E and F) Postoperative computed tomography scan showing complete tumor removal.

REFERENCES

1. Sanna M, Saleh E, Khrais T, et al. Atlas of Microsurgery of the Lateral Skull Base. Stuttgart: Georg Thieme Verlag; 2008.
2. Connor SE, Leung R, Natas S. Imaging of the petrous apex: a pictorial review. *Br J Radiol.* 2008;81:427-35.
3. Chapman PR, Shah R, Cure JK, et al. Petrous apex lesions: pictorial review. *Am J Roentgenol.* 2011;196:WS26-37 Quiz S40-23.
4. Razek AA, Huang BY. Lesions of the petrous apex: classification and findings at CT and MR imaging. *Radiographics.* 2012;32:151-73.

5. Ozgen B, Oguz KK, Atas A, et al. Complete labyrinthine aplasia: clinical and radiologic findings with review of the literature. *Am J Neuroradiol.* 2009;30:774-80.
6. Saini J, Kesavadas C, Thomas B, et al. Aberrant petrous internal carotid artery with cochlear anomaly-an unusual association. *Surg Radiol Anat.* 2008;30:453-7.
7. Ng M, Niparko JK. Osseous sarcoidosis presenting as a destructive petrous apex lesion. *Am J Otolaryngol.* 2002;23:241-5.
8. Prasad SC, Piccirillo E, Nuseir A, et al. Giant cell tumors of the skull base: case series and current concepts. *Audiol Neurotol.* 2014;19:12-21.
9. Cece H, Yildiz S, Iynen I, et al. A rare case of petrous apex osteoma. *J Pak Med Assoc.* 2012;62:608-9.
10. Agheshio N, Shimono T, Goto T, et al. Imaging appearance of petrous apex dermoid cysts containing little or no fat. *Jap J Radiol.* 2013;31:133-7.
11. Terao K, Cureoglu S, Schachern PA, et al. Pathologic correlations of otologic symptoms in acute lymphocytic leukemia. *Am J Otolaryngol.* 2011;32:13-18.
12. Paparella MM, Berlinger NT, Oda M, et al. Otolological manifestations of leukemia. *Laryngoscope.* 1973;83:1510-26.
13. Burgesser MV, Basquiera AL, Diller A. [Unusual presentation of plasma cell myeloma]. *Medicina* 2012;72:251-4.
14. Verma R, Malone S, Canil C, et al. Primary skull-based yolk-sac tumour: case report and review of central nervous system germ cell tumours. *J Neurooncol.* 2011;101:129-34.
15. Barnes DE, Welling DB, Lucas JG. Melanoma of the petrous apex of the temporal bone. *Ann Otol Rhinol Laryngol.* 1997;106:519-21.
16. Sun GH, Myer CM 3rd. Otolaryngologic manifestations of Maffucci's syndrome. *Int J Ped Otorhinolaryngol.* 2009;73:1015-8.
17. Palacios E, Valvassori G, D'Antonio M. Enchondroma of the petrous bone and parasellar area in Maffucci's syndrome. *Ear Nose Throat J.* 2001;80:75.
18. Cushing SL, Ishak G, Perkins JA, et al. Gorham-stout syndrome of the petrous apex causing chronic cerebrospinal fluid leak. *Otol Neurotol.* 2010;31:789-92.
19. Evans DA, Baugh RF, Gildsford JR, et al. Lymphangiomatosis of skull manifesting with recurrent meningitis and cerebrospinal fluid otorrhea. *Otolaryngol Head Neck Surg.* 1990;103:642-6.
20. Noel G, Feuvret L, Calugaru V, et al. Chondrosarcomas of the base of the skull in Ollier's disease or Maffucci's syndrome-three case reports and review of the literature. *Acta Oncol.* 2004;43:705-10.
21. Ahmed SK, Lee WC, Irving RM, et al. Is Ollier's disease an understaging of Maffucci's syndrome? *J Laryngol Otol.* 1999;113:861-4.
22. Jackler RK, Cho M. A new theory to explain the genesis of petrous apex cholesterol granuloma. *Otol Neurotol.* 2003;24:96-106; discussion 106.
23. Hoa M, House JW, Linthicum FH, Jr. Petrous apex cholesterol granuloma: maintenance of drainage pathway, the histopathology of surgical management and histopathologic evidence for the exposed marrow theory. *Otol Neurotol.* 2012;33:1059-65.
24. Selman Y, Wood JW, Telischi FF, et al. Development of cholesterol granuloma in a temporal bone petrous apex previously containing marrow exposed to air cells. *Otol Neurotol.* 2013;34:958-60.
25. Sanna M, Dispenza F, Mathur N, et al. Otoneurological management of petrous apex cholesterol granuloma. *Am J Otolaryngol.* 2009;30:407-14.
26. Sanna M, Zini C, Gamoletti R, et al. Petrous bone cholesteatoma. *Skull Base Surg.* 1993;3:201-13.
27. Sanna M, Pandya Y, Mancini F, et al. Petrous bone cholesteatoma: classification, management and review of the literature. *Audiol Neurotol.* 2011;16:124-36.
28. House WF, Hitselberger WE, Horn KL. The middle fossa transpetrous approach to the anterior-superior cerebello-pontine angle. *Am J Otol.* 1986;7:1-4.
29. Sanna M, Piazza P, De Donato G, et al. Combined endovascular-surgical management of the internal carotid artery in complex tympanojugular paragangliomas. *Skull Base.* 2009;19:26-42.
30. Sanna M, Khrais T, Menozi R, et al. Surgical removal of jugular paragangliomas after stenting of the intratemporal internal carotid artery: a preliminary report. *Laryngoscope.* 2006;116:742-6.
31. Sanna M, Khrais T, Menozi R, et al. Surgical removal of jugular paragangliomas after stenting of the intratemporal internal carotid artery: a preliminary report. *Laryngoscope.* 2006;116:742-6.
32. Sanna M, Piazza P, Shin SH, et al. Microsurgery of Skull Base Paragangliomas. Stuttgart: Georg Thieme Verlag KG; 2013.
33. Chernov M, DeMonte F. Skull base tumors. In: Levin V (Ed). *Cancer in the Nervous System*, 2nd edition. New York: Oxford University Press; 2002. pp. 300-19.
34. Kirazli T, Oner K, Bilgen C, et al. Facial nerve neuroma: clinical, diagnostic, and surgical features. *Skull Base.* 2004;14:115-20.
35. Bacciu A, Nusier A, Lauda L, et al. Are the current treatment strategies for facial nerve schwannoma appropriate also for complex cases? *Audiol Neurotol.* 2013;18:184-91.
36. Sanna M, Khrais T, Mancini F, et al. *The Facial Nerve in Temporal Bone and Lateral Skull Base Microsurgery.* Stuttgart: Georg Thieme Verlag; 2006.