Skull Base Tumours

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Abstract: Skull base tumours pose a significant challenge to the neurosurgeon. Skull base tumours can be benign or malignant. Primary skull base neoplasms can originate from the meninges, cranial nerves, bones of the cranial vault and skull base, cavernous sinus, or from the orbit and its contents. Tumours of the nose, paranasal sinuses, ears, or the infratemporal fossa can extend to the skull base and cranial cavity. Common skull base tumours are chondrosarcoma, chordoma, schwannoma, adenocarcinoma, and metastatic tumours. They can affect patients at almost any age. The clinical presentation depends on the site, size, and on the age of the patient. The most common presentation includes headache, nasal bleeding, hearing loss, cranial nerve palsy, and focal neuro-deficit/s. Head CT scan and MRI with contrast are enough to diagnose most of the tumours of the skull base. Surgery is the primary modality of management. Surgical approaches differ according to size, site, patient age, nature of neoplasm, and operator experience and expertise as well. Prognosis depends on histological type, extent of resection, and postoperative therapy (where needed) as well. In this chapter, we will briefly discuss the surgical management of all common skull base tumours, along with a short orientation of the surgical approaches used for the removal of skull base tumours.

Abbreviations

CP	cerebellopontine	ICP	intracranial pressure
CPA	cerebellopontine angle	JNA	juvenile nasopharyngeal angiofibroma
CS	cavernous sinus	MRI	magnetic resonance imaging
CSM	cavernous sinus meningioma	SRS	stereotactic radiotherapy
CT	computed tomography	TS	trigeminal schwannoma
IAC	internal acoustic canal	VS	vestibular schwannoma

1. Introduction

Because of their diverse histology, tumours of the base of the skull can be difficult to diagnose and treat before surgery. The precise determination of which tumours are regarded as tumours of the base of the skull is a controversial matter. Any tumour involving or adjacent to the base of the skull would fall under the broadest definition, which would include numerous tumours of the posterior fossa and cavernous sinus as well as pituitary tumours. However, a more specific definition would only include tumours that originate from the connective tissues and bones that make up the base of the skull. Practically speaking, it is occasionally impossible to discern between lesions that originate in the cranial cavity, the upper neck, or the paranasal sinuses and those that subsequently damage bone and cartilage tissues. Common skull base tumours are chondrosarcoma, chordoma, schwannoma, adenocarcinoma, and metastatic tumours. They can affect patients at almost any age. The most common presentation includes headache, aural/nasal bleeding, hearing loss, cranial nerve palsy, and focal neuro-deficit/s. Head CT scan and MRI of the head with contrast are enough to diagnose most tumours of the skull base. Surgery (endoscopic/microscopic) is the primary modality of management. Surgical approaches differ according to size, site, patient age, nature of neoplasm, and operator experience and expertise as well. Prognosis depends on histological type, extent of resection, and postoperative therapy (where needed) as well.

2. Esthesioneuroblastoma

2.1. Clinical Considerations

Esthesioneuroblastoma, also called olfactory neuroblastoma, generally begins as a tumour in the upper part of the nasal cavity and may grow or extend into the sinus, eyes, and brain. It can also spread to the cervical lymph nodes and the parotid glands. In advanced cases, it can also spread to other parts of the brain and body, such as the lungs, liver, bone marrow, bones, and skin. It is believed to originate from sensory neuroepithelial cells, also known as neuroectodermal olfactory cells (Fiani et al. 2019).

It can present at any age in both adult male and female patients. It is considered a relatively rare skull base tumour and constitutes only 3–5% of all tumours in the nasal cavity.

Patients may usually present with anosmia, epistaxis, nasal congestion, sinus infection, nasal obstruction, and difficulty breathing. The lesion may also cause eye pain, proptosis, ophthalmoplegia, diplopia, loss of vision, facial pain, facial numbness, rhinorrhoea, headache, nausea, vomiting, seizure, and, rarely, dental problems. The clinical features depend on the site of the pathology. The most commonly used staging systems are the modified Kadish staging, Dulguerov classification, and Haymes histopathological grading system (Fiani et al. 2019; Kumar 2015).

2.2. Investigative Considerations

Any patient with persistent nasal symptoms should undergo a thorough evaluation by an ear, nose, and throat specialist. Flexible endoscopy may reveal a fleshy, reddish-yellow mass filling the nasal cavity covered by the mucosa, with possible ulceration. CT scan of the skull base, paranasal sinus, and neck with and without contrast will reveal a hyperdense mass and heterogenous contrast enhancement. CT can also reveal bony destruction of the lamina papyracea, cribriform plate, and sphenoid sinus wall. MRI is the best neuroimaging modality to detect tumour extension and consistency. A cerebral angiogram is sometimes required to see carotid encasement by the tumour (Fiani et al. 2019; Kumar 2015).

2.3. Treatment Considerations

The various treatment modalities include a transcranial approach, endoscopic endonasal surgery, a craniofacial approach, stereotactic radiation therapy, chemotherapy, and palliative therapy. The choice of modality of treatment depends on tumour location and on its extension. It is a slow-growing but malignant tumour with high recurrence rates (Fiani et al. 2019; Kumar 2015).

3. Juvenile Nasopharyngeal Angiofibroma

3.1. Clinical Considerations

Juvenile nasopharyngeal angiofibroma (JNA) is a rare, benign tumour of the nasopharynx. It is aggressive locally and causes bone resorption by invading the surrounding tissue (Figure 1). A total of 0.5% of head and neck tumours are caused by it. There is a definite masculine predominance among young people. The Indian subcontinent has a higher tumour prevalence than the West.

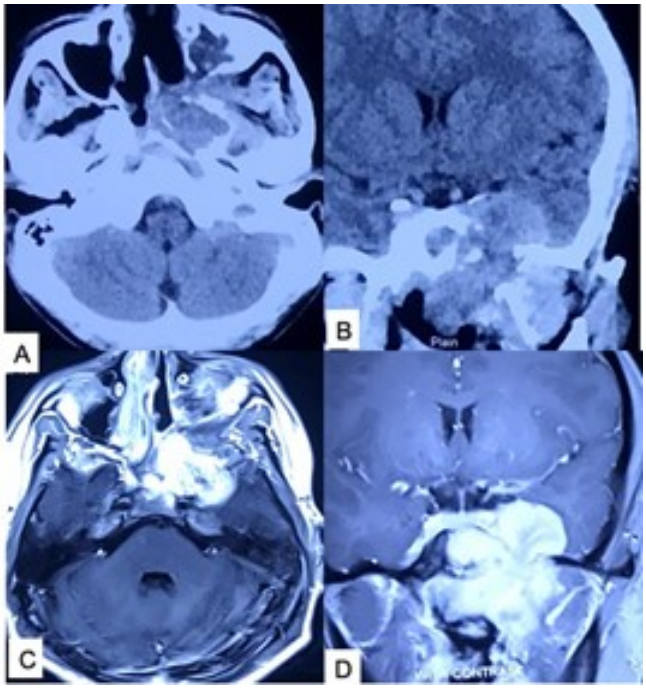

Figure 1. A case of juvenile nasopharyngeal angiofibroma in a 14-year-old boy. Contrast CT, axial and coronal view (**A**,**B**), and contrast MRI, axial and coronal view (**C**,**D**), show left-sided juvenile nasopharyngeal angiofibroma. Source: Figure by authors.

The tumour begins in the nasal cavity's lateral wall, near to the superior border of the sphenopalatine foramen. The growth begins in the submucosa of the nasopharynx's floor, progresses to the nasal septum, and grows into the posterior region of the nose, eventually obstructing the airway. Continuous growth involves the sphenoidal sinus, nasal fossa and middle turbinate, and pterygopalatine fossa, as well as the posterior wall of the maxillary sinus. Eventually, the tumour may invade the infratemporal fossa and the middle cranial fossa.

JNA typically manifests as a unilateral, painless nasal blockage that worsens with time. There may also be epistaxis, rhinorrhoea, and face discomfort. There may be facial deformities, proptosis, altered visual acuity, and impaired eustachian tube function. Cranial nerve palsy can result from an invasion of the intracranial space. In the nose and nasopharynx, a hard and friable mass may be found during a clinical examination (Martins et al. 2013; Garça et al. 2010).

3.2. Investigative Considerations

The precise location, extent, and relationship of the tumour to nearby structures like blood vessels and nerves can be determined using a CT scan, MRI (Figure 1), and angiography. Pathognomonic for JNA is the antral sign or Holman–Miller sign (forward bowing of the maxillary posterior wall). Nasal endoscopy clearly delineates the lesion and can be used to perform a biopsy for histopathological confirmation.

JNA is categorized into three types based on its radiological and clinical characteristics. Lesions classified as type 1 are primarily localized to the nasopharynx, paranasal sinus, nasal cavity, or pterygopalatine fossa. Type 2 JNAs are those with intact dura mater and little anterior and/or middle cranial fossa expansion that extend into the infratemporal fossa, buccal area, or orbital cavity. Type 3 is a giant, calabash-like tumour lobe in the middle cranial fossa (Martins et al. 2013; Garça et al. 2010).

3.3. Treatment Considerations

The preferred treatment is angiography followed by preoperative embolization and surgical resection. The supply of these tumours is usually via the external carotid artery (ascending pharyngeal artery, internal maxillary artery, and palatine artery) and less commonly the internal carotid artery (sphenoidal branch and ophthalmic artery).

Treatment is primarily surgical. The different surgical approach modalities include the transpalatine approach, infratemporal approach, transpalatine plus sublabial approach, middle fossa approach, maxillary swing approach or facial translocation, transmaxillary approach, and extended endonasal endoscopic approach.

These are benign tumours, and metastasis usually does not occur, but they are highly vascular and grow rapidly locally (Martins et al. 2013; Garça et al. 2010).

4. Clival Chordoma

4.1. Clinical Considerations

Chordoma is a rare tumour, presumably originating from remnants of the primitive notochord. The sacrococcygeal region accounts for about 50% of chordoma development, the spheno-occipital region for 35%, and the vertebrae for 15%. The upper and middle clivus and the spheno-occipital synchondrosis are all areas where skull base chordomas can be found. They are regarded as low-grade malignancies that grow slowly, but because of local bone infiltration, extension into nearby soft tissue, a high risk of recurrence, and occasional metastasis, they behave more aggressively.

No age group is immune to chordoma, but it is usually seen in adults (30–70 years). Sacrococcygeal is the commonest location, and chordoma is the commonest primary malignant sacral tumour.

The clinical presentation depends on the extension of the tumour. Clival chordomas are classified as upper, middle, lower, or of the craniovertebral junction. Sometimes they are classified as basisphenoidal or basioccipital, depending on whether they arise above or below the spheno-occipital synchondrosis. They may present with diplopia due to abducens palsy, visual loss, pituitary encephalopathy, chiasmal syndrome, cavernous sinus syndrome, nasopharyngeal mass, multiple cranial nerve involvement, brainstem sign, cerebellopontine angle involvement, hydrocephalus, and lower cranial nerve palsy (Chugh et al. 2007; Walcott et al. 2012; Tamura et al. 2015).

4.2. Investigative Considerations

On CT scan, clival chordomas are usually isodense on non-contrast-enhancing. CT also shows bony destruction and areas of calcification. MRI usually shows a lobulated tumour which is hypointense in T1WI and hyperintense in T2 WI, with variable contrast enhancement (Figure 2).

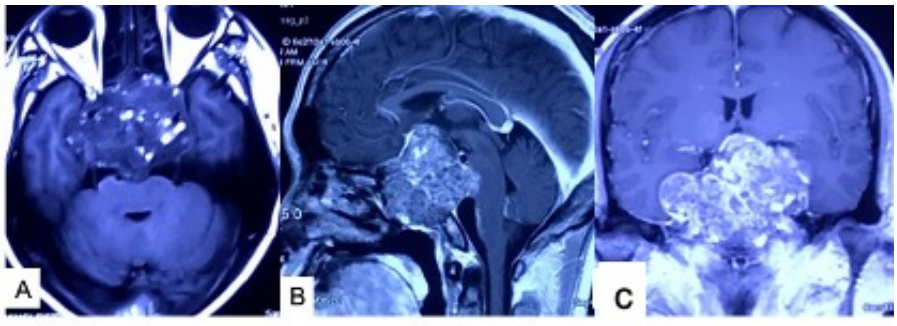

Figure 2. MRI of brain; (**A**) axial, (**B**) sagittal, and (**C**) coronal images showing large clival chordoma. Source: Figure by authors.

4.3. Treatment Considerations

Radical surgical removal is the primary treatment choice. But by the time these tumours are diagnosed, they will have reached a considerable size and invaded many critical structures, which makes surgical access very difficult. There are different anterior and lateral approaches to deal with these lesions depending on their extension. The extended subfrontal, trans-sphenoidal, endoscopic endonasal, transfacial, transoral, and transmandibular circumglossal retropharyngeal are among some of the anterior approaches. The lateral approaches include frontotemporo-zygomatic, preauricular subtemporal and infratemporal, presigmoid combined supratentorial and infratentorial, and also extreme lateral transcondylar.

Radiotherapy, i.e., C12, can be used for residual tumour. There are some restrictions on the utilization of radiation therapy for chordoma treatment because of its close vicinity to the optic nerve, chiasm, brainstem, and pituitary gland (Chugh et al. 2007; Walcott et al. 2012; Tamura et al. 2015).

5. Trigeminal Schwannoma

5.1. Clinical Considerations

Only 0.2–0.4% of all intracranial tumours are trigeminal schwannomas (TSs). The Gasserian ganglion is where they mostly manifest. Compared to acoustic neuroma, they are less common. Patients are usually middle-aged, commonly in their third or fourth decade of life, on first presentation. There is a relationship with neurofibromatosis type 2, just like there is with other schwannomas (Chowdhury et al. 2014; Agarwal 2015).

They can be preganglionic (cisternal), ganglionic (trigeminal ganglion), or postganglionic. Preganglionic TSs are confined to the prepontine cistern and cerebellopontine cistern. Ganglionic TSs are confined to Meckel's cave and they are the commonest. Postganglionic TSs are only found in the cavernous sinus or through the appropriate foramina at the base of the skull. Classically, the ophthalmology division is involved.

Women are somewhat more likely to develop TS. Patients typically have facial pain, most frequently conventional trigeminal neuralgia or atypical facial pain, when they first present with trigeminal nerve dysfunction. Trigeminal neuralgia is a sudden, typically unilateral, intense, fleeting, stabbing, recurrent pain that affects one or more trigeminal nerve branches. Numbness or a burning feeling are additional frequent clinical characteristics. Long-standing lesions can also cause motor symptoms such difficulties chewing, jaw deviation, and masseter and temporalis muscle weakness (Chowdhury et al. 2014; Agarwal 2015).

In 1955, Jefferson created the first classification scheme and divided TS into three categories (Jefferson 1955). Type A tumours arise from the Gasserian ganglion in the middle cranial fossa. Type B tumours develop from the roots of the trigeminal nerve roots in the posterior cranial fossa. The middle and posterior cranial fossa are both occupied by type C tumours, often known as hourglass tumours. Tumours with extracranial extension were introduced as a fourth categorization, type D, by some writers. Six types of TSs were proposed by Yoshida and Kawase (Yoshida and Kawase 1999) in their classification of TSs. Trigeminal nerve tumours of type P come from

the root of the trigeminal nerve. Type M tumours arise from the Gasserian ganglion or the peripheral branch at the lateral wall of the cavernous sinus. Extracranial peripheral branches of trigeminal nerves are the source of type E tumours. Combinations of P, M, and E tumours are indicated as types MP, ME, and MPE.

5.2. Investigative Considerations

Conventional X-ray delineates a bony erosion of the petrous apex. The margins are usually smooth without sclerosis. Large tumours may cause erosion of the sella turcica and of the clinoid process as well as the widening of superior orbital fissure. CT scan with bone window reveals all bony changes and iso- to hyperdense lesions with contrast enhancement. These tumours appear as hypointense on T1WI and hyperintense on T2WI MRI, with intense contrast enhancement (Figures 3 and 4). Cerebral angiography sometimes demonstrates an enlarged feeder artery and vessel encasement or displacement.

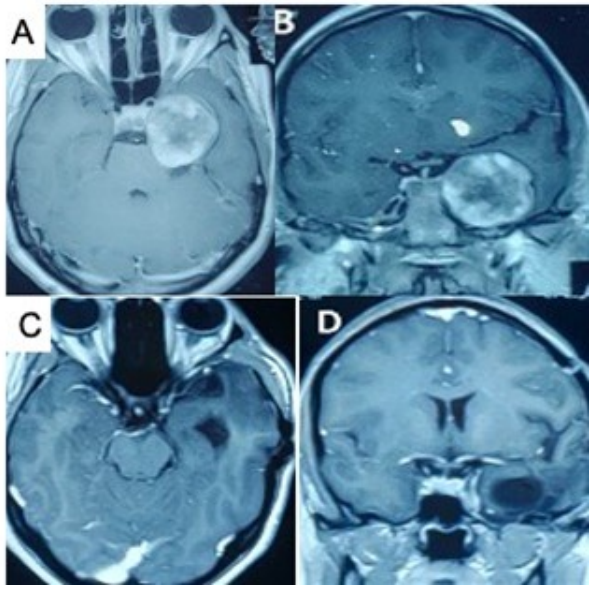

Figure 3. Contrast MRI of brain; pre- (**A**,**B**) and postoperative (**C**,**D**) images of a case of trigeminal schwannoma. Source: Figure by authors.

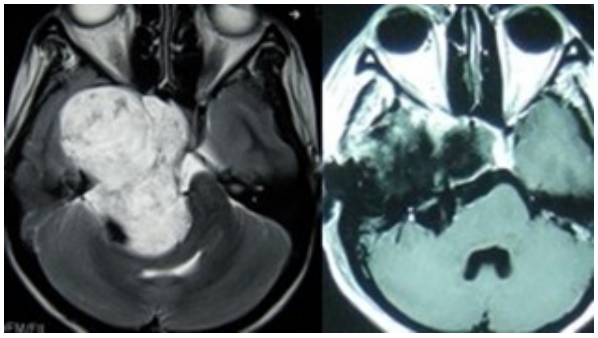

Figure 4. Contrast MRI of brain showing preoperative image of right trigeminal schwannoma occupying both middle and posterior cranial fossa on left-hand side and postoperative image of the same case on right-hand side. Source: Figure by authors.

5.3. Treatment Considerations

The site and size of the tumour determines the appropriate surgical approach (Figures 3 and 4). The commonest approaches are a middle fossa, retrosigmoid, presigmoid, and combined approach. Smaller tumours or residual tumours may be subjected to stereotactic radiosurgery (SRS) (Chowdhury et al. 2014; Agarwal 2015; Jefferson 1955; Yoshida and Kawase 1999).

6. Tumours of the Cerebellopontine Angle (CPA) and Vestibular Schwannoma

6.1. Clinical Considerations

The most frequent location for posterior fossa tumours is the cerebellopontine angle. Acoustic neuromas or vestibular schwannomas (VSs) make up 80% of all intracranial tumours that are found in this area, which in turn represent around 10% of all intracranial tumours. Meningiomas, arachnoid cysts, epidermoids, dermoids, lipomas, various cranial nerve schwannomas, and metastases are further tumours in this area (Carlson and Link 2021).

The petrosal surface of the cerebellum is folded around the lateral side of the pons and around the middle cerebellar peduncle to create the V-shaped CPA or fissure. The middle cerebellar peduncle serves as the floor of this space. The facial, cochlear, superior, and inferior divisions of the vestibular nerve are the four nerves that make up the internal acoustic meatus. Most frequently, VS is caused by the vestibular nerve (80%). In about 65–75% of cases, the inferior branch of the vestibular nerve is the origin of the tumour (Carlson and Link 2021; Komatsuzaki and Tsunoda 2001).

The junctional (Obersteiner–Redlich) zone where central and peripheral myelin converge is where VS originates from, although recent data point to a relationship with the sensory ganglia of the vestibular nerve in the internal auditory canal. The tumour initially grows within the canal and thereafter extrudes into the CPA. Depending on the direction of growth of the tumour, the facial nerve may run in a different direction: anterior to the tumour in about 70% of cases, superior in 10%, posterior in 7%, and inferior in 13% of cases (Carlson and Link 2021; Komatsuzaki and Tsunoda 2001; Xenellis and Linthicum 2003; Roosli et al. 2012; Tryggvason et al. 2012).

VS is the most common of CPA tumours. It is a benign, slow-growing tumour with WHO grade 1. The annual growth rate of VS is 1 to 10 mm. The highest incidence is in patients between their fourth and sixth decade of life. VS developing in patients with neurofibromatosis type 2 tends to present earlier. Most VS cases are unilateral and sporadic in nature. Bilateral VS is a hallmark of NF-II. Iodizing radiation is considered a risk factor.

The signs and symptoms of VS are those due to the involvement of cranial nerve VIII itself, as well as those due to involvement of adjacent cranial nerves (VII, V, IX, and X), the cerebellum, and the brainstem. Compression of the fourth ventricle may also produce features of raised ICP. The commonest symptoms of VS are unilateral progressive sensorineural hearing loss, tinnitus, unsteadiness, headache, facial numbness, and diplopia. Usually, they are slow-growing and present insidiously, but they can have acute presentation when there is haemorrhage within the tumour or due to the rapid expansion of a cyst (Carlson and Link 2021; Komatsuzaki and Tsunoda 2001; Xenellis and Linthicum 2003).

6.2. Investigative Considerations

A neuro-otological work-up is necessary to evaluate these tumours. Clinically, the Rinne test and Waber test are conducted to determine the type of deafness. Other specialized testing includes pure-tone audiometry, impedance audiometry, speech discrimination, and auditory evoked response monitoring.

Plain X-ray of the mastoid (Towne view) is still useful as a first step to see the enlargement and/or erosion of the porus acusticus and the internal acoustic canal (IAC). The transorbital projection (Caldwell view) depicts the canal and the meatus in their actual form and size in one single film without interference from other natural artifacts. CT scan with contrast can easily detect a tumour with bone window to demonstrate the classical widening of the internal acoustic meatus. Brain MRI depicts the tumour details, its consistency, and its relationship with critical neurovascular structures (Figure 5). On T1WI, they are isointense, and on T2WI and heterogenous contrast enhancement, they are slightly hyperintense. Due to their containment in the IAC and growth in the extracanalicular region, they can have a recognizable ice cream cone appearance (Carlson and Link 2021; Tryggvason et al. 2012). The Hannover classification (Table 1) is a dependable grading system for grading the size of vestibular schwannomas (Atchley et al. 2022) and can be used in counselling and in the evaluation of postoperative facial nerve and hearing outcomes.

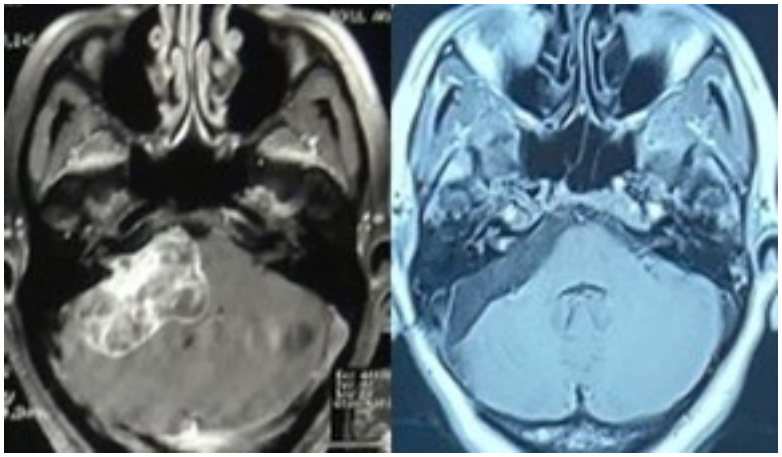

Figure 5. Preoperative and postoperative MRI of a patient with right-sided acoustic schwannoma. Source: Figure by authors.

Table 1. The Hannover classification of vestibular schwannomas.

Grade	Tumour
T1	Purely intracanalicular
T2	Intrameatal or extrameatal
T3A	Filling the cerebellopontine cistern
T3B	Reaching the brainstem
T4A	Compression of brainstem
T4B	Compression of brainstem with dislocation of fourth ventricle

Source: Authors' compilation based on data from Atchley et al. (2022).

6.3. Treatment Considerations

These tumours are very slow-growing. So, if the lesion does not produce significant symptoms, conservative treatment is an option. The therapeutic goals have changed from radical tumour removal to tumour control and to the preservation of neurological function. There are mainly three operative approaches to VS. They are the retromastoid retrosigmoid suboccipital approach, the middle fossa approach, and the translabyrinthine approach. Other special approaches are the transcanal approach, the suboccipital translabyrinthine approach, and the endoscopic approach. Many small tumours (less than 3 cm) are treated by radiosurgery. Gamma Knife surgery has also been used for residual tumour (Carlson and Link 2021; Koos et al. 1993).

Different strategies can be used in resection. The advantage of the retromastoid method is that it preserves hearing, although there is a chance of incomplete resection. This method involves making an incision behind the ear region and mastoid bone. Depending on the size of the tumour, hearing may be preserved using the middle cranial fossa method, which involves making an incision anterior to the ear. Hearing loss is an inevitable outcome of the translabyrinthine method, which passes through the inner ear. This method can be appropriate for individuals who do not have any functional hearing. In general, individuals with large tumours (larger than 4 cm), recurrent tumours following radiation therapy, compression of the brainstem, cranial neuropathy, and hydrocephalus should undergo surgical resection (Carlson and Link 2021).

The retrosigmoid approach is the most versatile as it offers excellent visualization of the CP angle, brainstem, and the IAC (Figure 5). Neuromonitoring for facial nerve and brainstem function and auditory evoked response monitoring for hearing function during the surgery provide tremendous improvement and reduce postoperative morbidity (Carlson and Link 2021).

After exposure, the tumour is debulked with suction and ultrasonic aspiration. The superior and inferior vestibular nerves are exposed and sectioned following meatal drilling to identify the facial nerve. The tumour is dissected away from the facial nerve and cerebellum using neuromonitoring.

Vestibular schwannomas that are surgically removed have a high chance of resection and low risks of recurrence. For the best results, a skilled surgeon and careful patient selection are essential. When 1000 vestibular schwannoma resections were reviewed, the results showed a 98% complete resection rate, a 68% hearing preservation rate, and a 1.1% mortality rate (Samii and Matthies 1997). With total resection, the local

recurrence rate is 0–2%. There is around a 30% probability of regrowth if only subtotal resection is feasible (Carlson and Link 2021).

Complications include postoperative CSF leak (9–13%), headache, CN V and VII neuropathies, hearing loss, hydrocephalus, haematoma, aseptic meningitis (2–4%), and hemiparesis (Samii and Matthies 1997).

The management strategy of vestibular schwannoma in NF2 includes microsurgery with or without adjuvant radiation with the aim of preserving hearing and facial function. When choosing a course of treatment, it is critical to consider the needs of the patient, the features of the tumour, and the resources available to the healthcare facility. Treatment options for medium- or small-sized vestibular schwannomas include radiation, surgery, and/or wait-and-scan. There has been a shift in recent years toward the use of planned SRS subsequent to planned subtotal resection. It appears that the late adverse effect of inducing a secondary neoplasm following radiotherapy has very little danger, and novel therapeutic alternatives based on drugs are emerging (Yao et al. 2020).

7. Glomus Tumour

7.1. Clinical Considerations

Glomus tumours are also referred to as chemodectomas or paragangliomas. They are benign neuroendocrine tumours that originate from glomus cells and vegetative nervous cells. They can be encountered in many areas of the body. The temporal bone, close to the jugular foramen, is one of the commonest sites. These are the most common tumours that form in the jugular foramen.

Glomus tumours of the cranial base can be broadly categorized into three anatomical descriptions: glomus tympanicum, which is confined to the middle ear (arises from Jacobson's nerve—inferior tympanic branch of cranial nerve IX); glomus jugulare, which is confined to the jugular foramen (arises from the adventitia of the jugular bulb along Arnold's nerve—auricular branch of cranial nerve X or the jugular fossa course of Jacobson's nerve); and glomus jugulotympanicum, which involves both the jugular foramen and the middle ear.

They are slow-growing, abundantly vascular tumours. They are usually benign; however, they may be locally aggressive and extend into the adjacent petrous bone, cerebellopontine angle, and sometimes the neck. They may be asymptomatic when small in size, but large tumours produce pulsatile tinnitus, conductive hearing loss, otalgia, aural fullness, vertigo, dysphagia and/or dysphonia due to lower cranial nerve palsy, facial palsy, Horner's syndrome, and diplopia. Various syndromes, like Vernet's syndrome, Collet–Sicard syndrome, and Villaret's syndrome, are associated with these lesions. Otoscopy classically reveals a red mass (Jayashankar and Sankhla 2015; Kirollos et al. 2019).

7.2. Investigative Considerations

Combined CT and MRI are the investigations of choice for the diagnosis and staging of the disease (Figures 6 and 7). CT delineates the bone invasion, which usually has irregular edges. T1WI and T2WI MRI characteristically show mixed signal intensity, described as a salt-and-pepper appearance, reflecting hypervascular signal voids together with focal areas of signal intensity due to intratumoural blood products. Contrast shows avid contrast uptake and demonstrates a smooth tumour contour. Catheter angiography shows a dense, hypervascular tumour blush, often with early venous filling. The two most common staging classifications are the Glasscock–Jackson classification and the Fisch classification (Jayashankar and Sankhla 2015; Kirollos et al. 2019).

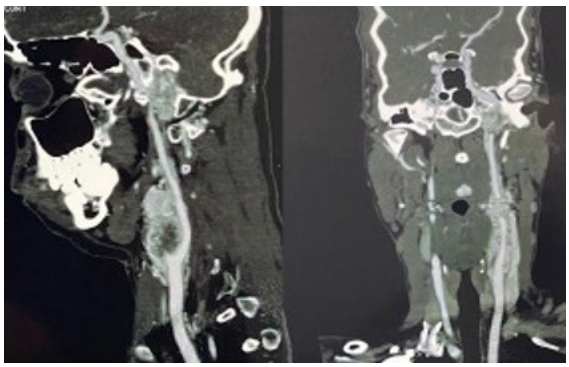

Figure 6. CT and CTA of brain and neck showing left-sided glomus jugulare associated with carotid body tumour. Source: Figure by authhors.

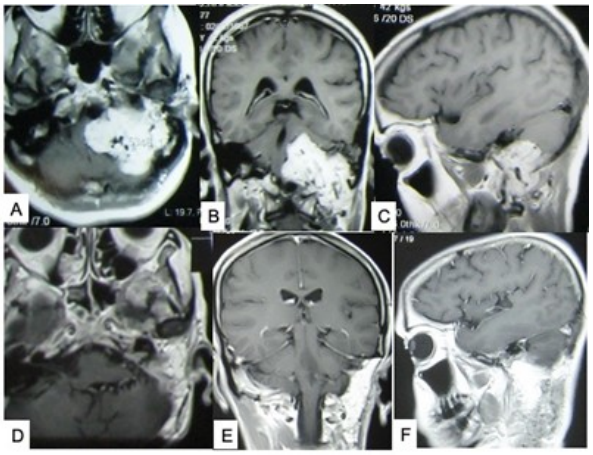

Figure 7. Contrast MRI of head. (**A**–**C**) Show left-sided large glomus jugulare; (**D**–**F**) show excision of the tumour. Source: Figure by authors.

7.3. Treatment Considerations

The optimal management of these tumours is challenging and requires a personalized approach administered by an experienced multidisciplinary team. Because of comorbidities, a watch, wait, and rescan policy is often the first line of management. Larger tumours, fast-growing small tumours, and secretory tumours may require treatment with surgery and/or radiotherapy (Jayashankar and Sankhla 2015; Kirollos et al. 2019).

8. Cavernous Sinus Lesion

8.1. Clinical Considerations

The cavernous sinus is a venous plexus located laterally at both sides of the sella turcica which houses important neurovascular structures. For neurosurgeons, the cavernous sinus (CS) region has traditionally presented a difficulty because of its intricate architecture and unique location in the anterolateral skull base. Dolenc's anatomical study and surgical experiences ultimately led to the development of a logical surgical strategy and to the definition of the various relationships between the lesion and the neurovascular structures. Yasargil writes, "There is no doubt that this type of microsurgical anatomical study is a new step in the 100-year history of neurosurgery" in the preface to Dolenc's book (Dolenc and Yasargil 1989). Ali Krisht et al. described the transcavernous microsurgical approach (Krisht et al. 2022).

Pathological findings in the cavernous sinus are diverse and include intrinsic and extrinsic lesions, like vascular, traumatic, inflammatory, congenital, or neoplastic lesions. The primary sites of cavernous sinus metastatic tumours are the breast, prostate, and lung. Primary intracranial tumours include meningioma, haemangioma (Figure 8), neurofibroma, chondroma, and lymphoma. Sometimes, localized tumour spread may occur from nasopharyngeal and pituitary growths.

Cavernous sinus lesions produce symptoms like ophthalmoplegia, chemosis, proptosis, Horner's syndrome, and trigeminal nerve lesions. Depending on the extension of the tumour, it can cause cavernous sinus syndrome or orbital apex syndrome (Chowdhury et al. 2012; Chowdhury and Haque 2017; Al-Mefty and Smith 1988).

8.2. Investigative Considerations

CT scan as well as MRI of the brain delineate the location, extension, and involvement of the cranial nerves and of the internal carotid artery.

8.3. Treatment Considerations

The approach to these tumours is very difficult because of their complex neurovascular structures relationship. The approach can be transcranial or endonasal endoscopic depending on the position of the pathology. In some lesions, SRS is useful (Chowdhury et al. 2012; Chowdhury and Haque 2017; Al-Mefty and Smith 1988).

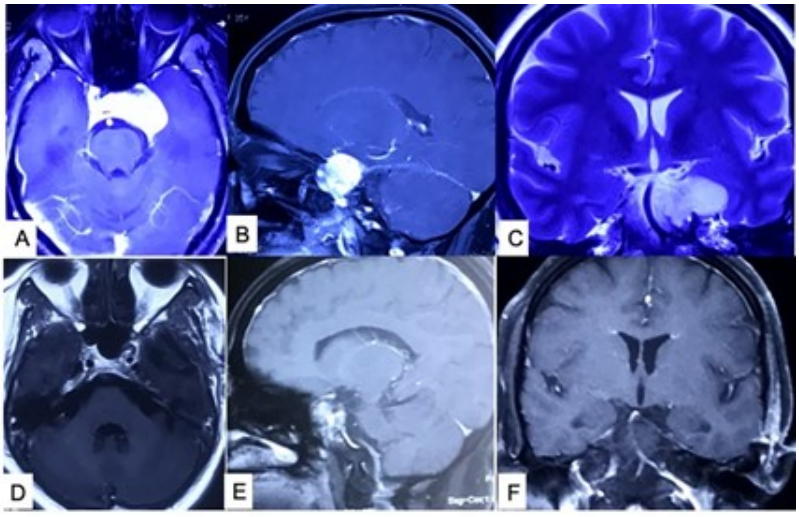

Figure 8. MRI of head. (**A**–**C**) Preoperative axial, sagittal, and coronal images showing left cavernous haemangioma. (**D**–**F**) Postoperative images showing excision of tumour. Source: Figure by authors.

8.3.1. Cavernous Sinus Meningioma

Cavernous sinus meningioma (CSM) is the commonest primary cavernous sinus (CS) lesion. The majority of these lesions affect women in their third or fourth decade of life. Any one of the three anatomical presentations of meningiomas can be referred to as a cavernous sinus meningioma. Rarely, a meningioma develops and remains inside the boundaries of the CS proper; other meningiomas mostly originate outside the CS proper and may sporadically penetrate the CS lateral wall; eventually, the majority will spread to encompass both the extracavernous compartments and the CS proper (Raheja and Couldwell 2020).

Clinically, the patient may present with headache, diplopia, visual impairment, (third to sixth) cranial nerve palsy, facial pain, and features of cavernous sinus syndrome. Imaging includes contrast MRI and CT scan of the head and skull base as well as an angiogram.

Today, neurosurgeons face challenges in managing cavernous sinus meningiomas (CSMs) due to inadequate understanding of their natural history, early involvement of critical neurovascular structures, lack of clear tissue planes with normal surrounding structures, and high rate of aggressive surgery-related morbidity. The neurosurgical community's preferred approach has changed over the past few decades from aggressive microsurgical resection to maximal safe resection and the possible application of adjuvant radiotherapy (Raheja and Couldwell 2020). Nowadays, many prefer biopsy and stereotactic radiosurgery (SRS).

8.3.2. Cavernous Sinus Haemangioma

About 3% of benign cavernous sinus masses are cavernous sinus haemangiomas (CSHs), which are more frequent in middle-aged women. Usually slow-growing groups of vascular channels with thin walls, these masses have the ability to affect nearby neurovascular systems mass-wise. These masses can be categorized pathologically into three main categories: mixed, mulberry-like, and sponge-like. The majority of those affected by CSHs are middle-aged women. The symptoms might vary, but headache, altered vision, and cranial nerve palsies, brought on by the mass effect, are common (Noblett et al. 2018).

When it comes to the workup of patients with CSH, MRI is crucial. On T1-weighted imaging, these lesions usually show hypo- or isointensity, and on T2-weighted images, hyperintensity. Resection surgery is the cornerstone of CSH treatment. However, when a tumour is found in the cavernous sinus, up to 40% of tumours show significant intraoperative bleeding; therefore, the decision to perform total surgical excision must be carefully evaluated against the risks of neurovascular injury. Patients who have limited subtotal resection or inoperable lesions may benefit from stereotactic radiosurgery. It has been demonstrated that Gamma Knife radiosurgery (GKS) can considerably reduce tumour size and volume, in addition to relieving some of the neurological symptoms related to CSH (Noblett et al. 2018).

9. Orbital Tumour

There are seven bones that contribute to the bony orbit: 1. pars orbitalis of the frontal bone; 2. lacrimal bone; 3. lamina papyracea of the ethmoid bone; 4. orbital process of the zygomatic bone; 5. orbital surface of the maxillary bone; 6. orbital process of the palatine bone; 7. greater and lesser wings and body of the sphenoid bone. The orbits are bony structures of the skull that house the globe, extraocular muscles, nerves, blood vessels, lacrimal apparatus, and adipose tissue. Common orbital tumours are lymphoma, metastases, lacrimal gland tumours, rhabdomyosarcoma, retinoblastoma, optic nerve glioma, optic nerve sheath meningioma, schwannoma/neurofibroma, choristoma, dermoid, epidermoid, teratoma, haemangioma/cavernoma, lymphangioma, orbital pseudotumour, and orbital sarcoidosis.

9.1. Clinical Considerations

Orbital tumours are rare. Tumours may arise from the globe, from the bony orbit, or from any content within the orbit. The commonest malignant orbital tumours in the adult population are metastatic tumours.

Orbital tumours produce symptoms and signs either due to the compression, infiltration, or infarction of orbital structures. Patients with orbital tumours may present with proptosis, diplopia due to external or internal ophthalmoplegia, visual disturbance, eye pain, chemosis, and sensory impairment. Intraconal tumours usually produce axial proptosis; extraconal lesion push the eye out of the lesion. Proptosis may be caused by lesions located outside the orbit, like cavernous sinus lesions. Pulsatile proptosis usually occurs due to a vascular lesion within the orbit or due to a carotid cavernous fistula. Visual impairment may occur in the form of loss of vision—either complete, incomplete, or transient—or visual field defects. Compression of the optic nerve may cause either primary optic atrophy, as in optic nerve glioma and optic nerve sheath meningioma, or optic nerve disc swelling. Usually, inflammatory and malignant lesions produce orbital pain (Chagla 2012; Mercandetti 2019).

9.2. Investigative Considerations

The assessment of orbital tumours requires formal visual assessment, which includes visual acuity, visual field analysis, colour vision, and fundoscopy. Plain X-ray of the orbit may reveal bony erosion, sclerosis, or calcification. CT scan of the orbit provides the localization of the tumour, bony changes, calcification, and enhancement of the pathology. Contrast-enhanced MRI is the investigation of choice as it shows soft-tissue structures well (Figure 9). Angiography is sometimes required for vascular lesions like meningioma, haemangiopericytoma, dural AVM, or carotid cavernous fistula. Sometimes, ultrasonography may be helpful in detecting lesions in the anterior segment of retrobulbar tumours (Chagla 2012; Mercandetti 2019).

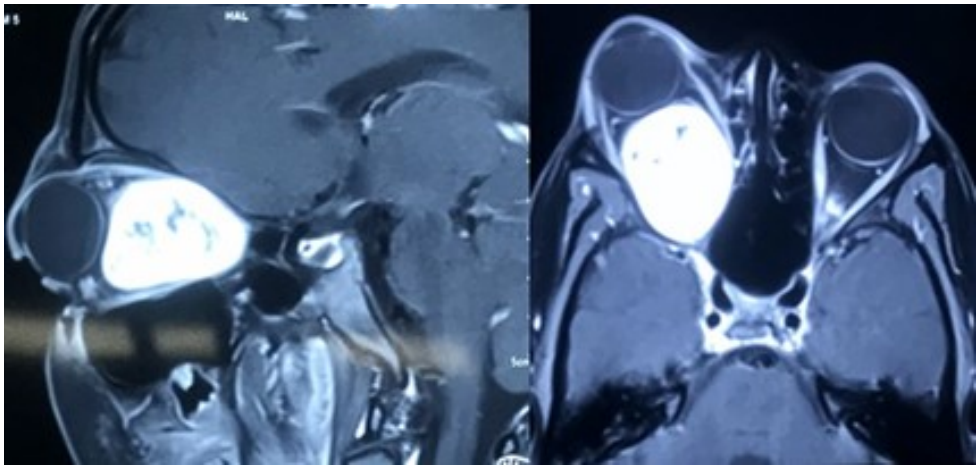

Figure 9. Contrast MRI of brain and orbit showing huge right-sided retrobulbar schwannoma. Source: Figure by authors.

9.3. Treatment Considerations

The management of orbital tumours involves clinicians from a variety of surgical disciplines, including neurosurgeons, ophthalmologists, ENT specialists, maxillofacial surgeons, and skull base endoscopic surgeons. The surgical approaches can be transcranial, orbital, and endoscopic. The cranium provides a safe access to the

orbit; hence, it is imperative for neurosurgeons to have knowledge on the orbit. There are different ways to approach it transcranially, like a subfrontal approach with superior osteotomy, fronto-orbito-zygomatic craniotomy, fronto-orbito-zygomaticotemporal craniotomy, or lateral orbitotomy. Transcranial approaches are best for lateral, superior, and posteriorly located tumours. The endoscopic approach is best for medially located lesions medial to the optic nerve (Chagla 2012; Mercandetti 2019).

10. Surgical Approaches to the Skull Base

10.1. Pterional Craniotomy

Pterional craniotomy (Figure 10) is a very important daily neurosurgical practice, which is why it requires excellent execution knowledge. Aneurysms of the anterior circulation, basilar apex, the proximal segment of the superior cerebellar and posterior cerebral arteries, arteriovenous malformations and cavernous haemangiomas of the basal forebrain, anterior and middle skull base tumours, gliomas of the frontal, parietal, and temporal opercula, insula, mediobasal temporal region, cerebral peduncles, interpeduncular fossa, and orbital lesions are treated in the pterional trans-sylvian corridor. In order to increase surgical freedom and lower the risk of complications associated with the approach, an overview of the fundamental methods and variations of the pterional approach are described here.

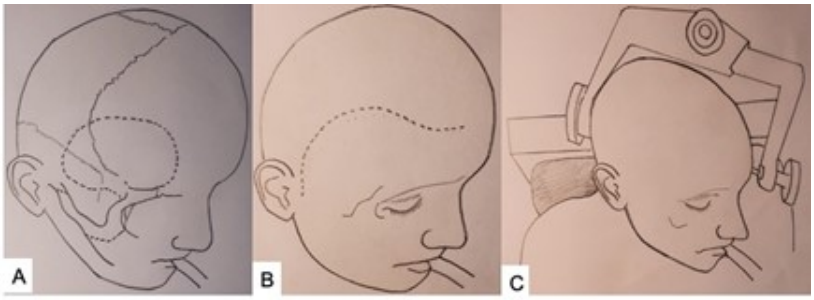

Figure 10. Hand drawings showing (**A**) removal of bone in pterional craniotomy; (**B**) incision mark for pterional craniotomy; (**C**) head positioning and fixation in pterional craniotomy. Source: Figure by authors.

Positioning: To enable the best possible venous outflow, the patient is put supine with their head elevated above the level of the heart by 30° and secured to a Mayfield–Kees or Sugita skull clamp. Yasargil, in 1976, advocated for an extension of about 20° in his classic description. The malar eminence is to be the highest point of the horizon. It is advised to rotate the head contralaterally, with a range of 15°–45° depending on the particular neurovascular target (Yaşargil 1984; Yaşargil et al. 1987).

Incision: Local subcutaneous infiltration of lignocaine and adrenaline diluted in normal saline is useful to reduce pain and enable easier detachment of the skin from the subcutaneous layers. The skin incision starts 1 cm in front of the tragus, anteriorly to the superficial temporal artery and auriculotemporal nerve. The incision curves upward, behind the hairline, to reach the midline. The skin flap is divided by the temporalis muscle and reflected forward. To avoid damaging the frontal branch of the facial nerve, the superficial layer of the temporal fascia and the fat pad within which the nerve courses are separated from the deep layer (interfascial technique).

Two different ways can be followed. In the subfascial technique, an incision into the superficial and deep layers of the superficial temporal fascia is made. In the submuscular technique, an incision into the deep temporal fascia, subperiosteal blunt dissection of the temporalis muscle, and forward reflection of the myocutaneous flap are performed.

The submuscular technique is quicker and poses less risk of intraoperative damage to the frontal branch of the facial nerve, which is why this is mostly practiced. Regardless of the method, the temporalis muscle needs to be cut above the posterior root of the zygoma and subperiosteally detached in a manner that is retrograde, going superior-to-posterior and back-to-front, as described by Oikawa et al. Electrocauterization must be avoided to preserve the anatomical continuity of the deep fascia along with blood supply from the internal maxillary artery. To avoid functional and cosmetic issues as well as atrophy of the temporalis muscle, it is imperative to preserve the deep fascia (Zabramski et al. 1998; Coscarella et al. 2000). Recognizing skull sutures can also be aided by a compulsive subperiosteal dissection of the muscle. The fronto-zygomatic suture and the superior aspect of the posterior root of the zygomatic process of the temporal bone should always be visible.

Craniotomy: The MacCarty keyhole, which is drilled 5 mm behind the point where the fronto-zygomatic, spheno-zygomatic, and frontosphenoidal sutures intersect, allows for access to the dura of the anterior cranial fossa and periorbita (Shimizu et al. 2005). To access the anterior cranial fossa, the first burr hole can be created slightly above the MacCarty keyhole when treating the pathology, which does not require the exposure of the orbit and its contents. The second burr hole is to be made at the level of the temporal squama, above the posterior root of the zygoma.

The extensive drilling of the lateral portion of the larger sphenoid wing until the SOF is a crucial step in the pterional approach. This makes the sphenoidal part of the sylvian fissure, which serves as the entrance to the entire anterior and middle skull base (Figure 11), fully visible. The dural flap can be reflected with the aid of skeletonization and partial expansion of the SOF. It is advised to use a drill to thin the orbital roof in order to create a working corridor and line of sight as close to the anterior cerebral fossa as feasible. This approach is termed the "extended pterional approach". These adjustments allow for unhindered surgical approaches to subfrontal and parasellar targets. Osteotomy along the orbital roof has some of the same advantages as orbito-zygomatic craniotomy, but it is a more effective operation with fewer cosmetic deformities.

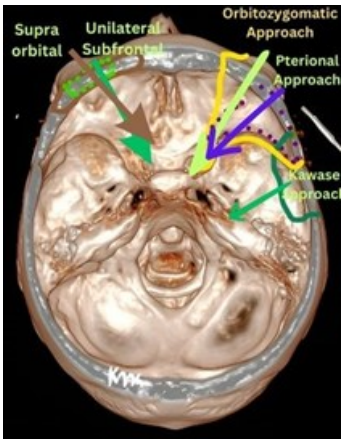

Figure 11. Skull base showing approaches to bony corridor of anterior and middle fossa. Source: Figure by authors.

10.2. Orbito-zygomatic Craniotomy

Skull base approaches were revolutionized in the 1990s, when OZ (Figure 12) began to be the mainstay of the skull base for accessing the sellar and parasellar regions, and complex skull base techniques began to be used. However, there are a few drawbacks and cosmetic concerns to this method. These include diplopia, exophthalmos, enophthalmos, forehead hypaesthesia and dysaesthesia, frontal muscular weakness, and persistent periorbital and eyelid oedema, which can be avoided by deploying the extended pterional approach when possible.

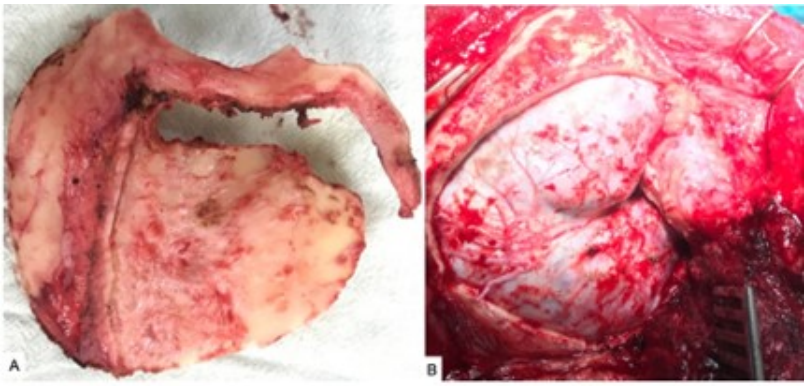

Figure 12. (**A**) Right-sided one-piece OZ craniotomy bone flap; (**B**) right-sided OZ craniotomy after bone flap removal. Source: Figure by authors.

The cranio-orbito-zygomatic (COZ) approach is an extension of the pterional approach, involving the adjunct of orbito-zygomatic (OZ) osteotomy to allow for wider exposure of the anterior and middle skull base and upper

retroclival region. It provides advantages when treating giant aneurysms of the anterior communicating artery (ACoA) and distal basilar artery, tuberculum sellae, large anterior clinoidal and spheno-orbital meningiomas, large craniopharyngiomas, giant pituitary adenomas, cavernous haemangiomas of the hypothalamus, and crus cerebri of the midbrain.

Placement and positioning: This is fairly similar to the pterional approach described above. A 30° rotation of the head makes the longer axis of the anterior clinoid process perpendicular to the floor, while at 45°, the surgical view of the subfrontal area is maximized.

Skin incision and soft-tissue dissection: The skin incision is made in a curvilinear fashion behind the hairline and goes from 1 cm anterior to the tragus to the contralateral midpupillary line. The dissection of soft tissue is fairly similar to the pterional approach described above.

Craniotomy: The COZ approach can be executed in a one-piece, two-piece, or three-piece technique.

In the one-piece COZ approach, the MacCarty keyhole is positioned 5 mm behind the junction between the fronto-zygomatic, spheno-zygomatic, and frontosphenoidal sutures. Two further burr holes are placed in the temporal squama, above the posterior root of the zygoma and on the superior temporal line, respectively. The first cut involves the posterior root of the zygoma and is carried out with a reciprocating saw. The second cut connects the keyhole to the inferior orbital fissure (IOF). The third cut starts at the level of the lateral orbital wall and is advanced across the malar eminence. The fourth cut is made at the intraorbital side and crosses the superior orbital rim and the orbital roof until the lateral aspect of the SOF. The keyhole, temporal, and frontal burr holes are interconnected with the craniotome. The COZ bone flap is then fractured by inserting and gently rotating a periosteal elevator into the groove made by the cut of the superior orbital rim. The temporalis muscle and the galea-pericranium flap are reflected downward. The tent stitches of the galea displace the eyeball slightly inferiorly, flattening the exposure of the anterior cranial fossa by a few millimetres. Drilling of the temporal squama can adequately expose the middle fossa, whereas drilling of the lesser sphenoid wing, along with an extra- or intradural anterior clinoidectomy, expands the surgical access to most targets related to this approach.

The two-piece COZ approach can be executed through two different techniques. In the Zabramski technique, after the pterional craniotomy, subtraction of the OZ bar is realized through six cuts. As in the one-piece technique, the first cut is made at the level of the posterior root of the zygoma. The second cut involves the malar eminence, from lateral to medial. The third cut is carried out across the lateral orbital wall, from medial to lateral. The fourth cut is intracranial, involves the orbital roof and the superior orbital rim, and is executed starting from the lateral end of the SOF, taking care to preserve the underlying periorbita. The fifth and sixth cuts connect the SOF and the IOF, thus freeing the lateral orbital wall. The fifth cut starts at the level of the IOF and ends at the level of the anterior part of the middle fossa. The sixth cut moves from the IOF toward the fifth cut. The OZ bar is then detached from the masseter muscle (Zabramski et al. 1998).

In the Al-Mefty technique, the orbitopterional two-piece COZ approach includes two zygomatic cuts involving the anterior and posterior root of the zygoma, respectively. The zygomatic arch is mobilized inferiorly without detachment of the masseter muscle, thus avoiding the risk of postoperative masticatory imbalance. The other steps are those of the one-piece variant of the COZ approach, where all the cuts are carried out extracranially (Al-Mefty 1987).

The three-piece COZ approach consists of a combination of both two-piece variants. It involves zygomatic osteotomy without detachment of the masseter, inferior mobilization of the zygomatic arch, pterional bone flap, and orbital osteotomy as separate pieces entailing the superolateral orbital rim and the orbital roof.

After opening the dura, the COZ approach allows for four different corridors: (1) subfrontal, (2) trans-sylvian, (3) pretemporal, and (4) subtemporal. The trans-sylvian and pretemporal corridors are related to four well-defined deep windows to the infratentorial region through the opening of the Liliequist membrane. The deep windows are as follows: (1) optic–carotid, (2) carotid–oculomotor, (3) supracarotid, and (4) oculomotor–tentorial.

10.3. Subtemporal Approach

A surgical pathway to several lesions located in the middle cerebral fossa was made available via temporal craniotomies. By the 1960s, Drake's groundbreaking work in surgically treating over 1700 posterior circulation and basilar aneurysms had brought the subtemporal technique into a thriving phase. To access the perimesencephalic and midclival regions, various variants of the subtemporal routes were created, including the subtemporal keyhole approach, expanded exposure with zygomatic excision, and removal of the petrous apex (Drake 1965).

Steps of approach: After anaesthesia, lumbar drain should strongly be considered based on the pathology for which surgery is being undertaken. The head is rotated to the contralateral side, aligning the anterior–posterior axis parallel with the floor. The vertex is slightly tilted down toward the floor for efficient intraoperative viewing trajectory as the base of the middle cranial fossa inclines upward steeply. Operating as close to the base of the middle cranial fossa of the skull as possible is the goal of the subtemporal approach. As a result, the skin incision should be made so that the temporal squama is exposed, at the very least, to the zygomatic arch root. An inverted U-shaped skin incision is often performed 1–2 cm anterior to the tragus, starting at the level of the zygomatic arch root. The incision is made around the ear, curving anteriorly, upwards, and finally backward across the pinna. Alternatively, for a minimized temporal bone flap, a linear skin incision can be made from the inferior rim of the zygomatic arch, approximately one finger's width, anterior to the external auditory canal.

The skin flap is then reflected inferiorly, and the temporalis muscle is divided and reflected as well, exposing the zygomatic process of the temporal bone and the root of the zygomatic arch. A 4–5 cm-sized craniotomy at the exposed temporal squama is performed, placing the inferior border of the bone flap as low as possible. If mastoid air cells are inadvertently opened, careful closure by bone waxing or temporal muscle flap should be completed. The inferior overhanging edge of the temporal bone needs to be drilled down flush with the base of the middle cranial fossa (Yasargil et al. 1976; Kawase et al. 1991; Hernesniemi et al. 2005).

10.4. Kawase Approach

The Kawase approach, first described by Kawase et al. in 1985, has added the step of removal of the bones in the petrous apex to the standard subtemporal approach. As the Kawase approach and the extended Kawase approach significantly expanded the exposure range in the upper, middle, and partial inferior regions of the clivus, this approach is used extensively. The approach overcomes the obstructions of the petrosal range and extends the region exposed in the posterior cranial fossa without bringing more retraction of the temporal lobe and Labei's vein, due to which many lesions in the petrosal apex and the upper and middle clivus, including meningioma, chordoma, basilar trunk aneurysm, prepontine epidermoid, trigeminal schwannoma, and pontine cavernoma, could be satisfactorily exposed and safely resected. The gains in the exposure volume and area are more when the manipulation angle is less than 135° (Kawase et al. 1985).

When using the traditional Kawase method, the drilling range of the bone is restricted to the petrosal apex, with a rhomboid shape by the view from the lateral–superior direction. The posterior margin of the trigeminal nerve anteriorly, the petrosal ridge medially, the greater superficial petrosal nerve or petrosal segment of the internal carotid artery laterally, and the arcuate eminence posteriorly define the boundaries of the rhomboid. Restricted is the deep boundary of debone, which is also the boundary between clivus and the petrosal apex. It would be possible to expand the Kawase method by drilling bones outside of the IPS. It could expand the deboning range to the upper and middle clivus and the jugular tubercle (JT).

10.5. Transpetrosal Approaches to the Posterior Fossa

Different degrees of resection of the petrous temporal bone provide for varying degrees of access to posterior fossa lesions. The many variations of transpetrosal techniques can be roughly divided into anterior and posterior groups, despite the nomenclature often being unclear. While the techniques in the anterior group are variations of the fundamental middle fossa approach, the posterior transpetrosal procedures comprise the retrolabyrinthine, translabyrinthine, and transcochlear approaches. The petroclival region and the cerebellopontine angle can potentially be exposed from both the anterior and posterior approaches. The posterior techniques entail varying degrees of petrous bone resection and are based on standard mastoidectomy. This results in progressively increased exposure anteriorly, but in the translabyrinthine approach, hearing is lost, and in the transcochlear approach, hearing and facial strength are lost. On the other hand, the middle fossa techniques entail varying degrees of medial petrous bone excision while sparing the lateral petrous bone. The goal of any middle fossa method is to protect hearing. In order to expose the posterior fossa, extensions of the middle fossa techniques entail the resection of bone within the Kawase rhomboid and division of the tentorium (Tummala et al. 2005).

10.6. Retrosigmoid Approach

In Figure 13 surgical approaches to the bony corridor of the posterior skull base are shown. The retrosigmoid approach (Figure 14) is the workhorse for posterior fossa surgery because of its advantage in providing a versatile

corridor to tackle different types of lesions in and around the cerebellopontine angle. Bony anatomical landmarks are helpful in localizing the venous sinuses and planning the craniotomy. Extensions of the approach include, among others, the transmastoid, supracerebellar, far lateral, jugular foramen, and perimeatal approaches. The retrosigmoid approach applies to a broad range of pathologies and, with its extensions, can provide adequate exposure, obviating the need for extensive and complicated approaches.

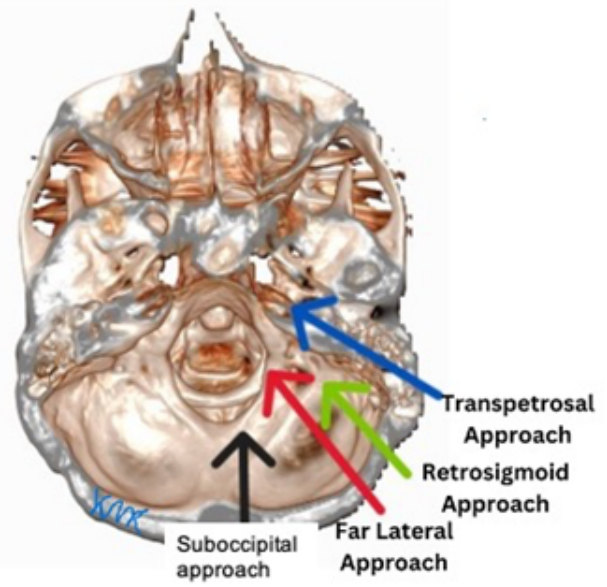

Figure 13. Skull base showing the approaches to the bony corridor of the posterior skull base. Source: Figure by authors.

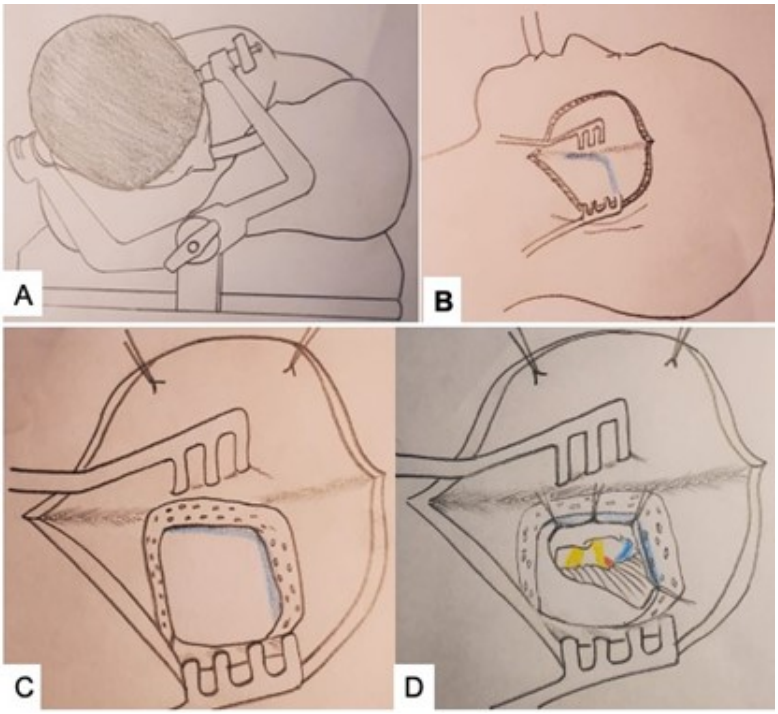

Figure 14. Hand drawings of retrosigmoid craniotomy. (**A**) Positioning and head fixation; (**B**) skin incision and exposure; (**C**) craniotomy; (**D**) after dural opening**.** Source: Figure by authors.

The retrosigmoid or lateral suboccipital approach was popularized by Woolsey and Krause in the early 1900s. After several modifications, the lateral suboccipital approach evolved into what is now called the retrosigmoid approach. This approach provides optimal access to the cerebellopontine and cerebellomedullary cisterns. The retrosigmoid approach uses a lateral suboccipital craniotomy combined with a partial mastoidectomy to enter the dorsolateral aspect of the posterior fossa. This is the most widely used approach for vestibular schwannoma and

other lesions which require the exposure of the brainstem and cranial nerves. It is also used for aneurysms of the anterior inferior cerebellar artery, the posterior inferior cerebellar artery, and basilar trunk. Microvascular decompression of the trigeminal nerve can also be treated through this approach. Small-to-medium-size tumours of the internal auditory canal can be excised with the preservation of hearing (Elhammady et al. 2012).

These patients can be positioned in a sitting, supine, or lateral decubitus position, with lateral decubitus being the most common. Care should be taken to preserve the lesser occipital and greater auricular nerves to reduce postoperative headache and dysaesthesia. Hearing preservation is one of the main advantages of the retrosigmoid approach to resection in vestibular schwannoma treatment, as opposed to the translabyrinthine approach, in which the inner ear structures are sacrificed. Postoperative complications include retraction injury to the cerebellum, venous sinus injury, damage to the cranial nerves and brainstem, as well as postoperative CSF leaks. In particular, the mastoid emissary vein should be located during craniotomy exposure because it can cause substantial bleeding and be the source of an air embolism. Injury to the vertebral artery can also occur during osteotomy of the lower portion of the exposure at the occipital bone.

The RS approach may be carried out with the patient in the supine, lateral, park-bench, sitting, or semi-sitting position. While every surgeon may have different preferences according to the specific case, this section will describe the operation in the semilateral position. The patient is positioned supine on the operating table with the operative side up. The ipsilateral shoulder is bolstered to rotate the patient's torso into a semilateral position, and the patient should be well secured onto the operating table. The head is rotated about 70-80 degrees to the contralateral side, flexed slightly anteriorly and towards the floor to open the angle between the occiput and the neck, and secured in place with a Mayfield clamp pin fixation.

In the RS approach, the single-pin arm is positioned just above the ipsilateral superior temporal line, behind the hairline and anterior to the EAM, avoiding the forehead, and the double-pin arm is positioned just superior and posterior to the contralateral superior nuchal line, avoiding the suboccipital muscles. To widen the angle even more and create more workspace, the ipsilateral shoulder could also be taped down. In the surgical field, the mastoid tip ought to be the highest point. Patients with medium-sized to large tumours may benefit from the placement of a lumbar drain at this time to aid in the draining of cerebrospinal fluid (CSF).

Incision of the skin: The common variations of skin incisions for the RS approach are the linear, lazy S-shaped, C-shaped, and curvilinear inverted U-shaped incisions. After using the zygoma–inion line and mastoid tip–asterion line to approximate the TS, SS, and TSSJ, a C-shaped incision is drawn from 2 cm superior to the middle of the pinna to 1 cm medial to the mastoid tip with the apex of the curve 5 cm posterior to the postauricular crease. The incision may be extended inferiorly depending on the case.

The skin is incised over the region of the temporalis muscle. The incision continues towards the mastoid tip, making sure to cut through subcutaneous adipose tissue, which may decrease the risk of greater auricular nerve and lesser occipital nerve (LON) damage. Special attention needs to be paid to the superior half of the incision to visualize and preserve the LON because it is more superficial and enters the region at the posterior edge of the subcutaneous tissue (approximately at the 4 o'clock position for the left side and at the 8 o'clock position for the right side). The skin flap is then elevated with the fascia of the posterior auricular muscle and the sternocleidomastoid muscle and retracted anteriorly over the mastoid process.

Dissection of the muscles: Traditionally, muscle dissection in the RS approach has been described as following the skin incision. The underlying muscles can be incised in line with the skin to create a single myocutaneous flap and retracted anteriorly in a C-shaped skin incision, divided in line with the skin and retracted anteriorly and posteriorly in a slightly curved skin incision, or a single myocutaneous flap based along the suboccipital muscles can be reflected inferiorly in an inverted U-shaped skin incision.

Craniotomy: Following the retraction of soft tissues, a craniectomy or craniotomy may be performed. The number of burr holes depends on the patient's underlying pathology and on their age. Typically, more burr holes or a craniectomy should be employed for patients over 60 years old because the dura is more attached to the cranial surface. The craniotomy may be limited to just the edge of the TS and SS in the standard RS approach or further extended to expose the sinuses with a limited posterior mastoidectomy in the extended approach.

In the standard RS approach, the most important burr hole, which should expose the junction of posterior fossa dura and the margins of the adjacent transverse sinus and sigmoid sinus, is conventionally placed over the asterion or slightly below it; however, it is also argued that the Teranishi technique (placing the burr hole 6.5mm inferior and 6.5mm anterior to the asterion) or the Ribas technique (10 mm anterior to the asterion with

the superior edge of burr hole adjacent to the parietomastoid suture) are the most suitable approaches for the placement of the initial burr hole in the standard and extended RS approach, respectively.

When performing the craniotomy, it is preferable to preserve its shape by starting and completing the cut from the outermost edge of the burr hole to keep the perimeter of the craniotomy as flush as possible. Any exposed mastoid air cells are sealed with bone wax or fibrin glue to prevent postoperative CSF leakage.

10.7. Suboccipital Craniotomy

Suboccipital craniotomy and craniectomy are performed by removing the caudal portion of the occipital bone. Wide exposure to approach the posterior fossa was performed early in the disease course to avoid brainstem compression during surgery and to allow for posterior fossa decompression. Suboccipital craniectomy is the standard treatment for Chiari 1 malformation. The posterior fossa is the deepest of the three cranial fossae, containing a complex anatomy, including the cerebellar hemispheres and vermis, brainstem, cranial nerves, and vasculature. Neurophysiological monitoring is used during risky procedures. Access to the posterior fossa can be gained through the suboccipital approach, allowing for the treatment of the cerebellar hemispheres, cerebellar tonsils and vermis, medulla, and fourth ventricle. In addition, lesions of the craniocervical junction and foramen magnum can be accessed. Patients with Chiari 1 malformation also undergo resection of the C1 posterior arch (Perneczky and Reisch 2008).

Patients can be operated in either the prone, lateral decubitus, or sitting position. The prone position has a lower risk of venous air embolism, but blood may pool within the operative bed, limiting visibility. There is also more pressure placed on the face. The lateral position carries a decreased risk of venous air embolism than the sitting position, but the upper cerebellar hemisphere may fall into the surgical field and interfere with the approach. The sitting position carries a higher risk of cardiopulmonary instability, venous air embolism, and rapid CSF leak, which can result in brain herniation. In cases where opening of the dura is necessary, duraplasty is performed using the nuchal ligament, pericranium from the occipital bone, or a dural substitute. Surgery in the posterior fossa has been reported to carry a complication rate as high as 32%. CSF leaks are the most common surgical complication in the posterior fossa. Cerebellar mutism is a rare complication, occurring in less than 1% of patients and manifesting as slow or frank mutism (Ngwenya et al. 2012). It is transient and thought to be secondary to oedema or ischaemia to the dentate nucleus or pathways of the dentatorubrothalamic tract.

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