

Skull base bony lesions: Management nuances; a retrospective analysis from a Tertiary Care Centre

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ABSTRACT

Background: Skull base lesions are not uncommon, but their management has been challenging for surgeons. There is large no of bony tumors at the skull base which has not been studied in detail as a group. These tumors are difficult not only because of their location but also due to their variability in the involvement of important local structure. Through this retrospective analysis from a Tertiary Care Centre, we are summarizing the details of skull base bony lesions and its management nuances.

Materials and Methods: The histopathologically, radiologically, and surgically proven cases of skull base bony tumors or lesions involving bone were analyzed from the neurosurgery, neuropathology record of our Tertiary Care Institute from January 2009 to January 2014. All available preoperative and postoperative details were noted from their case files. The extent of excision was ascertained from operation records and postoperative magnetic resonance imaging if available.

Results: We have surgically managed 41 cases of skull base bony tumors. It includes 11 patients of anterior skull base, 13 middle skull base, and 17 posterior skull base bony tumors. The most common bony tumor was chordoma 15 (36.6%), followed by fibrous dysplasia 5 (12.2%), chondrosarcoma (12.2%), and ewings sarcoma-peripheral primitive neuroectodermal tumor (EWS-pPNET) five cases (12.2%) each. There were more malignant lesions ($n = 29, 70.7\%$) at skull base than benign ($n = 12, 29.3\%$) lesions. The surgical approach employed depended on location of tumor and pathology. Total mortality was 8 (20%) of whom 5 patients were of histological proven EWS-pPNET.

Conclusions: Bony skull base lesion consists of wide variety of lesions, and requires multispecialty management. The complex lesions required tailored approaches surgery of these lesions. With the advent of microsurgical and endoscopic techniques, and use of navigation better outcomes are being seen, but these lesions require further study for development of proper management plan.

Key words: Bony tumors, operative approach, radiotherapy, skull base

Introduction

Bony tumors involving the skull base are rare entities, and^[1,2] their management nuances are very sparsely described. These tumors are difficult not only because of their location but also due to their variability in the involvement of

vital neurovascular structures. The primary skull tumors account for 0.8% of all bone tumors, and primary skull base lesions are even more infrequent.^[3] The skull base is of cartilaginous origin as compared to the membranous origin of vault of skull, thus influencing the pathologies affecting this of the region. According to the epicenter of the lesions, they can be broadly divided into three; anterior, middle, and posterior skull base lesions. Multidisciplinary approaches for these tumors provide feasible platform

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to achieve the goal of maximal safe resection with decompression of vital neurovascular structure without adding any further morbidity/mortality. In the present era of advanced microneurosurgery and endoscopic-assisted skull base surgeries, the management policies and outcome of this heterogeneous subgroup has to be redefined. In this retrospective analysis from a Tertiary Care Centre, we are summarizing the details of our experience with skull base bony lesions and their management nuances.

Materials and Methods

This retrospective study involved 41 cases of skull base bony lesions operated in our Tertiary Care Centre between January 2009 and January 2014. All the cases are biopsy proven, and their data collected from neurooncology registration of our institute.

The lesions were divided into groups of anterior, middle, and posterior skull base on the basis of their boundaries; anterior skull base posteriorly limited by lesser wing of sphenoid and at midline by chiasmatic sulcus. The margin between middle and posterior fossae is formed by the petrous ridges joined by the dorsum sellae and posterior clinoid processes.^[4] These lesions were further divided into midline and lateral region of skull base by imaginary line passing through clinoidal process and occipital condyles. The middle portion covers the ethmoid and cribriform plate in anterior fossa, sella, and parasellar area in middle fossa and clivus with foramen magnum in posterior fossa [Figure 1]. Apart from demographic profile; clinical features, radiological parameters (size, location, and extent of the lesion in terms of skull base territories), operative techniques and approaches with intraoperative and postoperative complications were recorded in data collection. Mean follow-up was 22.36 months (range 6–55 months). Ethical Committee Board of our institute evaluate and provide clearance for this study under project “clinical, radiological, and prognostic indicators

of nongliomatous nonmeningiomas lesions of the brain” (IEC: 2015-94-IMP-86).

Results

We have surgically managed 41 cases of skull base bony tumors. It includes 11 patients of anterior, 13 middle, and 17 posterior skull base tumors. The most common bony tumor was chordoma 15 (36.6%), followed by fibrous dysplasia 5 (12.2%) among all bony tumors. Plasmacytoma and osteosarcoma were relatively rare, one case each (2.4%). A total of 12 varied pathologies were encountered in our series as described in Table 1. The age ranged from 5 to 51 years with mean age of 24.5 years. There were 24 males and 17 female patients with M: F ratio of nearly 3:2.

Bony tumors in skull base territories: Demographic profile and clinical features

Anterior skull base

Among the variable bony tumors (11 cases) encountered in the anterior skull base, the most common pathology was fibrous dysplasia ($n = 4$) closely followed by the ewings sarcoma-peripheral primitive neuroectodermal tumor (EWS-pPNET) ($n = 3$). The laterally located lesions were more common; 10 cases (90.9%) and were mostly benign (seven benign, three malignant). Only one case had a medial located lesion which was malignant [Table 2]. The most common clinical presentations included local cosmetic disfigurement of the frontal bone ($n = 3$), proptosis ($n = 4$) or both ($n = 2$). The age group involved was 5–37 years (mean age: 18.7).

Middle skull base

Of the 13 middle skull base lesions, chondrosarcoma was the most common pathology encountered followed by other varied rare pathologies [Table 3]. There was nearly equal distribution of malignant and benign lesions (7:5). The most common presentation was temporal swelling and 6th nerve paresis. Sex ratio was nearly equal, and age ranged from 10 to 51 years (mean age: 24.9). The benign lesions were more

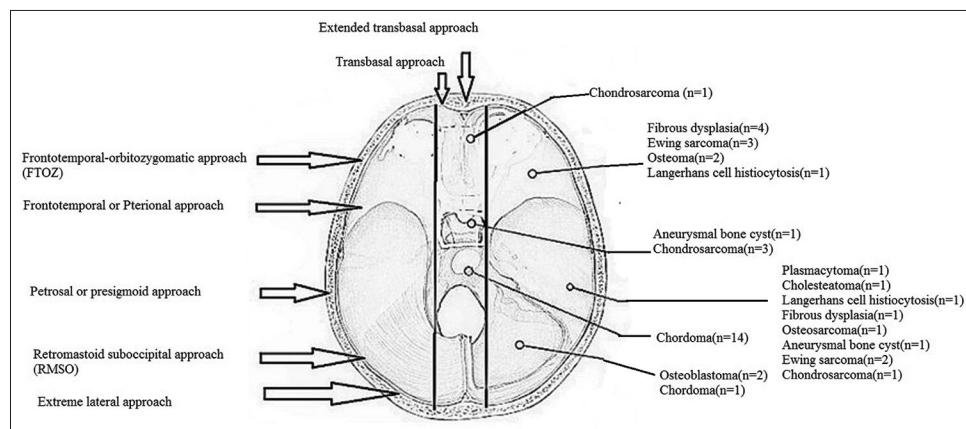


Figure 1: Line diagram showing division of skull base into median and lateral along with varied pathology encountered in our series and different types of approaches used

common in the second and third decade of life (2 in each decade out of 5). No age predilection was present among the malignant tumors. Benign lesions were present in the lateral region while malignant lesions were nearly equal in distribution (benign: 4 lateral, 1 medial; malignant: 5 lateral, 3 medial).

Posterior skull base

The posterior cranial fossa base was the most common site for skull base bony tumors ($n = 17$) among the three skull base territories. They consisted of only two pathologic types: Chordoma ($n = 15$) and Osteoblastoma ($n = 2$). The chordomas were central in location while osteoblastomas were lateral. The Chordomas had a strong male predilection. The most common presenting complaints were hearing loss, lower cranial nerve involvement, diplopia, and long tract signs [Table 4].

Biological aggressiveness

There were more malignant lesions ($n = 29$, 70.7%) at skull base than benign ($n = 12$, 29.3%) lesions. The malignant lesions were more common in posterior skull base (all lesions were malignant), (anterior - 4, middle - 8, posterior skull

base - 17) and includes chordoma, osteoblastoma, and chondrosarcoma [Figure 2]. The male:female ratio among malignant lesions was 1.5:1 with mean age of 25.8 years. Similarly, the benign lesions were more common in anterior skull base (anterior - 7, middle - 5) with 1.4:1 male:female ratio with mean age of 21.5 years. The most common benign pathologies were osteoma and fibrous dysplasia.

Surgical approach

The surgical approach employed depended on the tumor location and histopathology. Overall, the pterional ($n = 9$) and frontotemporal orbitozygomatic (FTOZ) ($n = 7$) approaches were the most common ones. Anterior skull base lesions were approached by transbasal approach for medial lesion ($n = 1$), frontolateral or frontotemporal craniotomy for lateral lesion (seven patients) and simple drilling of the involved outer table of frontal bone was done in two cases.

Tumors of the medial middle skull base were approached via extended transbasal ($n = 1$), endoscopic transsphenoidal ($n = 2$), and fronto temporal craniotomy ($n = 1$) with near total excision in all the four cases. The lateral lesions were approached through frontotemporal craniotomy alone ($n = 3$) or combined with orbito-zygomatic osteotomy ($n = 4$). Subtemporal and anterior transpetrosal (kawase) approaches and infratemporal approach of Fisch each was used in one patient, achieving total, near-total and subtotal excision in two, six and one case, respectively.

In posterior fossa tumors were approached by endoscopic transsphenoidal route ($n = 1$), FTOZ ($n = 2$), extended transbasal approach ($n = 2$), transmastoid presigmoid approach ($n = 2$), middle fossa half and half approach ($n = 1$), and frontotemporal craniotomy alone ($n = 2$) or combined with orbitozygomatic osteotomy and/or subtemporal approach ($n = 3$) for midline lesions achieving total resection in two, near total excision in 11 patients. The three lateral located lesions were approached

Table 1: Summary of total skull base bony tumors in our study

Histological types	Skull base bony tumors	n (total, n=41)
Benign	Fibrous dysplasia	5
	Aneurysmal bone cyst	2
	Langhans cell histiocytosis	2
	Osteoma	2
	Cholesteatoma	1
Malignant	Chordoma	15
	Chondrosarcoma	5
	Ewing sarcoma	5
	Osteoblastoma	2
	Osteosarcoma	1
	Plasmacytoma	1

Table 2: Clinical characteristics and treatment of anterior skull base bony tumors in our study (total, n=11)

Skull base territories	Bony tumors	Age	Male/female	Location	Clinical presentation	Surgical approach and extent of removal	Adjuvant therapy RT/CT	Follow-up
Anterior skull base	Median Chondrosarcoma ($n=1$)	25	0:1	Sinonasal and orbital	Proptosis, decreased vision	Basifrontal approach with orbital decompression (NT=1)	RT	2 years, improve
	Lateral Fibrous dysplasia ($n=4$)	27.2	3:1	Left frontal, orbital and sphenoid extension	Swelling over forehead, proptosis, vision loss	Frontal craniotomy with shaving of frontal bone and orbital roof (GT=1, NT=2, ST=1)	-	2 years, improve
	Ewing sarcoma ($n=3$)	9.6	1:2	Frontal base with orbital extension	Headache, proptosis	Frontotemporal craniotomy with orbitozygomatic osteotomy (GT=2, NT=1)	RT + CT	2-3 years, died
	Osteoma ($n=2$)	26	1:1	Fronto-orbital	Swelling over forehead	Excision with reconstruction of supra-orbital ridge (NT=1)	-	2 years, died
	Langerhans cell histiocytosis ($n=1$)	5	1:0	Frontal with orbital involvement	Swelling over forehead, headache	Complete removal of osteolytic bone (GT=1)	-	1.5 years, improve

GT – Gross total; NT – Near total; ST – Subtotal; RT – Radiotherapy; CT – Chemotherapy

Table 3: Clinical characteristics and treatment of middle skull base bony tumors in our study (total, n=13)

Skull base territories	Bony tumors	Age	Male/female	Location	Clinical presentation	Surgical approach and extent of removal	Adjuvant therapy	Follow-up
Middle skull base								
Median	Aneurysmal bone cyst (n=1)	17	0:1	Sella with suprasellar extension	Headache, ptosis, diplopia	Extended basifrontal approach (NT=1)	-	1 year, improve
	Chondrosarcoma (n=3)	30	2:1	Clival, sellar, sphenoid and petrous bone extension	Headache, ptosis, diplopia, hormonal imbalance	Endoscopic endo-nasal approach, + open extradural subtemporal approach (NT=3)	RT	2 years, died
Lateral	Plasmacytoma (n=1)	51	1:0	Medial 2/3 petrous bone	Diplopia, headache, hearing loss	FTOZ with presigmoid petrosal translabyrinthine approach (ST=1)	RT	1 year, improve
	Cholesteatoma (n=1)	23	1:0	Petrous bone	Discharging sinus, headache, hearing loss	Extradural sub temporal middle fossa approach (GT=1)	-	2 years, improve
	Langerhans cell histiocytosis (n=1)	10	1:0	Petrous bone	Hoarseness, facial deviation	Extended middle fossa approach with anterior petrosectomy (kawase approach (NT=1)	-	Lost follow-up
	Fibrous dysplasia (n=1)	23	0:1	Frontotemporal with extension of orbit and sphenoid bone	Swelling over forehead and temporal region	Fronto temporal craniotomy with decompression (GT=1)	-	2 years, improve
	Osteosarcoma (n=1)	23	1:0	Right temporal bone with temporal lobe involvement with middle ear extension	Swelling in temporal region, headache altered sensorium	Extradural sub temporal approach (NT=1)	RT	Lost follow-up
	Aneurysmal bone cyst (n=1)	19	0:1	Fronto-temporo-sphenoid bone	Swelling in temporal region, headache	FTOZ with excision of lesion (NT=1)	-	1 year, improve
	Ewing sarcoma (n=2)	11.5	1:1	Sphenoid wing to infratemporal fossa	Headache, vomiting proptosis	FTOZ with excision of lesion (NT=2)	RT	died
	Chondrosarcoma (n=1)	45	0:1	Left petrous bone	Hearing loss, mass protruding from ear	Infratemporal (Fisch) fossa approach (NT=1)	RT	8 months, improve

GT – Gross total; NT – Near total; ST – Subtotal; RT – Radiotherapy; CT – Chemotherapy; FTOZ – Fronto-temporo-orbito-zygotomy

Table 4: Clinical characteristics and treatment of posterior skull base bony tumors in our study (total, n=17)

Skull base territories	Bony tumors	Age	Male/female	Location	Clinical presentation	Surgical approach and extent of removal	Adjuvant therapy	Follow-up
Posterior clival base								
Median	Chordoma (n=14)	27.8	9:5	Clivus, petrosphenoid, one with infratemporal ext	Diplopia, multiple cranial nerve involvement, hemiparesis	Endoscopic endonasal approach (11), extended frontobasal approach (2), preauricular transzygomatic approach + half and half (1) approach (GT=2, NT=11, ST=1)	RT	5-died 1-improve
Lateral	Osteoblastoma (n=2)	35	1:1	Petro-occipital with middle ear extension, basiocciput	Hearing loss, Swelling at angle of mandible, mastoid, hoarseness	Combined far lateral and parapharyngeal approach, RMSO with translabyrinthine approach (NT=2)	RT	1 year, same
	Chordoma (n=1)	24	1:0	Left jugular foramen	Headache, vomiting, hearing loss	Postauricular transmastoid presigmoid approach (GT=1)	RT	1.5 years, improved

GT – Gross total; NT – Near total; ST – Subtotal; RT – Radiotherapy; CT – Chemotherapy; RMSO – Retromastoid suboccipital approach

by transmastoid presigmoid approach, retromastoid suboccipital approach and far lateral approach each in one case, with total resection in one and near total excision in two patients [Table 4].

Postoperative complication and long-term outcome

The outcome was defined by biological behavior of the tumor, its location, and surgical approach after considering all the important factors which significantly determine its outcome.

Benign lesions of ant skull base had only wound-related complication such as swelling ($n = 3$) and infection ($n = 2$) requiring bone flap removal in one and implant removal in the other one. Four patients with posterior and middle skull base lesion operated via endoscopic approach experienced postoperative cerebrospinal fluid (CSF) leakage which was managed by lumboperitoneal shunt in two and temporary lumbar drainage in the other two. Another major complication was cranial nerve deficit which occurred in

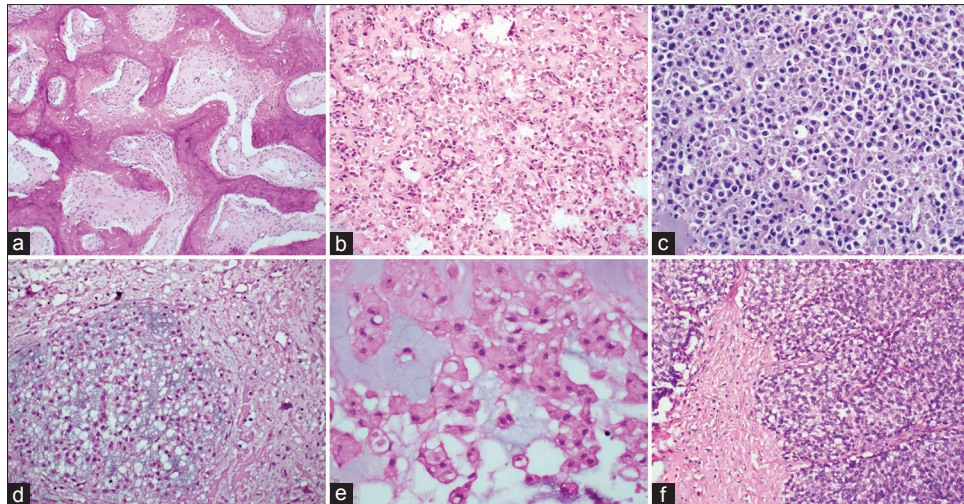


Figure 2: Pathology of skull base lesions (H and E); fibrous dysplasia (a), proliferation of fibroblastic cell along with interspersed irregular bony trabeculae, chondrosarcoma (b), sheets of large round to polygonal cells having enlarged vesicular pleomorphic nuclei, plasmacytoma (c), sheets of atypical plasma cells, chordoma (d and e), lobules of large polygonal cells with eosinophilic cytoplasm floating in myxoid background, with physaliphorous cells, Ewing's sarcoma (f), round to irregular hyperchromatic nuclei, inconspicuous nucleoli and scant cytoplasm with increased mitotic activity

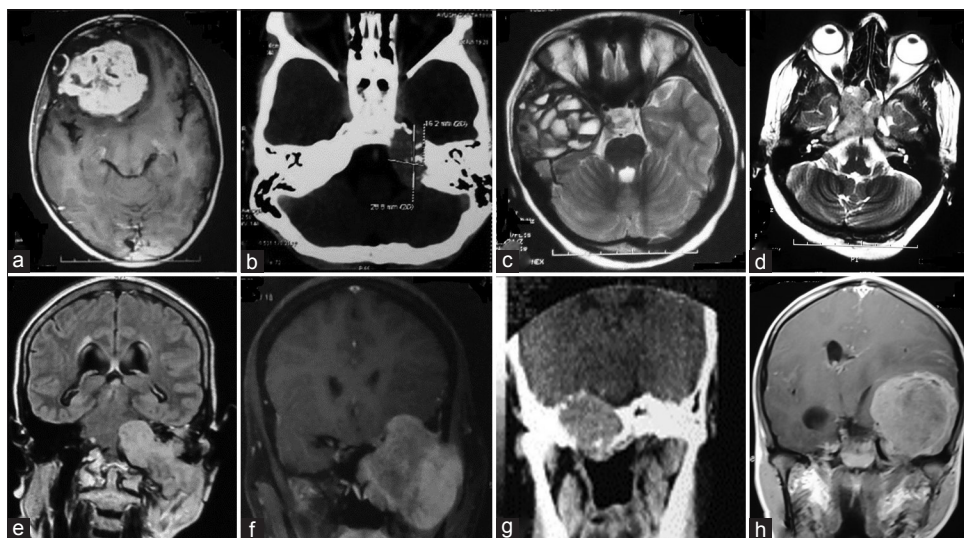


Figure 3: Varied location and radiological presentation of lesions at skull base (a) anterior skull base chondrosarcoma, (b) petrous apex Langerhans cell histiocytosis, (c) temporal bone aneurysmal bone cyst, (d) clival chondrosarcoma, (e) chondrosarcoma extending from petrous to C2, (f) middle skull base chordoma, (g) petrous bone plasmacytoma, (h) middle skull base Ewing's sarcoma

five patients (oculomotor in three and facial in two patients). One patient had decreased consciousness and hemiparesis after the surgery; who expired early after the operation. During the follow-up period, 8 (20%) patients deceased, of whom five suffered from EWS-pNET. No recurrence/regrowth and new clinical symptoms were noted in patients with benign lesions during the mean follow-up of 27.3 months (10–55 months).

Discussion

Variable clinical presentation of skull base bony lesions

The bony lesions of the skull base usually present with complaint of local pain and swelling. These lesions present

with symptoms due to the expansion of the skull bones, compression, or infiltration of the adjacent brain, cranial nerves, and paranasal sinuses [Figure 3]. The clinical features can be classified according to the location of the tumor [Table 5].

Management nuances of skull base surgical approach

In our series, we have collaborated with neuro-ophthalmologist, neuro-otologist, and plastic surgeons in 11 cases for treatment of these lesions. During planning for the surgical removal of the skull base bony tumors, determination of the exact site of origin, direction and nature of local spread (invasion versus displacement), extent of dural and basal bone involvement,

Table 5: Various skull base syndromes in different territories of skull base

Location	Syndrome		
Anterior skull base	Median	Olfactory syndrome	
	Lateral	Orbital syndrome	
		Forehead deformity	
Middle skull base	Median	Anterior	Pituitary disturbances
		Posterior	Cavernous syndrome
	Lateral	Anterior	Gasserian ganglion syndrome
		Posterior	Temporal deformity
		Posterior	Petrous syndrome
Posterior skull base	Median	Bilateral cranial nerve syndrome	
	Lateral	Upper	Cerebellopontine angle syndrome
		Lower	Jugular foramen syndrome
		Lower	Occipital condyle syndrome

presence or absence of an arachnoid plane between the tumor and brain, and an estimate of the tumor vascularity and consistency are the essential prerequisites for optimal outcome.

Anterior skull base approaches

Anterior skull base lesions can be approached via intracranial, extracranial, or combined approaches. As orbital involvement with proptosis was seen in majority of laterally situated anterior skull base bone lesions in our series orbital decompression was considered as a surgical goal in these patients.

Intracranial approaches often include a basifrontal or extended basifrontal approach. Basifrontal approach after bifrontal or unifrontal (frontolateral) craniotomy provides exposure of bony lesions situated in anterior skull base, paranasal sinus, and orbit for superior decompression. Extensive manipulation of the frontal sinuses and supraorbital rim must be avoided as much as possible. Whenever required resection of superior half of frontal sinus can provide further huge exposure of anterior skull base. The advantage of transcranial approach (transfrontal sinus approaches) to transsphenoidal approach in such cases is that the intracranial extension of tumor can be removed and the dissection around the optic nerve and the carotid artery can be done under direct vision.^[5] Transbasal approach is a transcranial extradural anterior approach to the midline skull base that offers the advantage of allowing tumor removal with dural repair and reconstruction of skull base in one stage.^[6,7] We have taken this approach in one patient with chondrosarcoma, without any significant complication. Since most of the bony lesions in the lateral part of anterior skull base are benign tumors such as osteoma, fibrous dysplasia and Langerhans cell histiocytosis, direct excision of the lesion by drilling of the involved bone is usually sufficient.

Extended transbasal approach provides adequate midline exposure for medial anterior cranial fossa lesions as well as

for the sphenoclivus lesion. It combines a bifrontal craniotomy with an orbitonasal or orbitonasooethmoidal osteotomy, and even a sphenoidectomy to provide a wide access to the midline lesions of anterior, middle, and posterior skull base.^[8,9] Further addition of orbitozygomatic osteotomy enables access to lesions extending more laterally along the temporal base and the greater wing of sphenoid.^[10,11] This approach was utilized in seven patients with anterior and middle skull base with complications of ophthalmoplegia and altered sensorium each in one. Demerits of this approach include extensive soft tissue dissection, technically demanding, prolonged operative time, the potential for the CSF leak, olfactory denervation. We have utilized the approach in two patients with clival region tumors achieving complete tumor excision and complication of CSF rhinorrhea in one. Alternatively, there are few extracranial transfacial approaches very rarely used for selected anterior skull base lesions extending to paranasal sinuses. We have used midfacial approach for staged removal of a large EWS-pPNET with extension into the infratemporal fossa in one case.

Middle and posterior skull base

Approaches can be divided into midline, and lateral skull base approaches based on the extent of lesion. Midline lesions involve sellar/suprasellar, and clival tumors and lateral lesions involve temporal base and infratemporal region and lateral petroclival lesion. As bony lesions of middle and posterior skull base usually arise from petrous and/or clivus bones, they can be approached through endoscopic approach for midline lesions and lateral/posterior skull base approaches for lateral petroclival lesions. Nonetheless, personal preferences and experience and the nature of lesion and goal of surgery are most crucial factors that can affect the decision making regarding the surgical approaches.

For midline skull base lesions endoscopic transnasal approaches are usually preferred to open surgical approaches. Among the lateral skull base approaches an extradural subtemporal approach is indicated when the major portion of the tumor is in the middle fossa or the region of cavernous sinus. Nevertheless, the petrous ridge is an obstructing hump in the approach to clival or petroclival region from subtemporal route. In these cases, the subtemporal approach is supplemented with anterior transpetrous approaches, to provide better exposure of the tumors extension to cerebellopontine angle and petroclival region. Most of the lesion situated in the petroclival region can be approached and safely resected by conventional neurosurgical approaches such as the subtemporal and retrosigmoid or combined subtemporal and retrosigmoid approaches. However, some tumors are hidden behind bony protuberance and in the angle between petrous bone and the clivus,^[12] and are approached through transpetrosal route.^[13,14]

Posterior approaches to the skull base include the extreme or far lateral approach, and retrosigmoid/suboccipital craniotomy

or craniectomy. An extreme lateral approach exposes the lower third of the clivus and cerebellopontine angle. Retrosigmoid craniotomy is used to approach lesions of the cerebellopontine angle and the petrous surface of the temporal bone.

Skull base malignancies near the jugular foramen or temporal bone may be approached through a postauricular incision and transjugular approach and/or petrosectomy. As far as surgical approaches to clival chordoma is concern a modified lateral subtemporal with or without transpetrous apex approach is most suitable.

Endoscopic surgery

Endoscopic neurosurgery offers a minimally invasive route to specific lesions and is increasingly being implemented as a tool for the biopsy and removal of these lesions. The endoscope can help to access many lesions of the skull base with minimal manipulation of neurovascular structures and decreased brain retraction.^[15,16] These approaches use either transnasal, transsphenoidal, transthemoidal, or transmaxillary corridors.^[17]

Disadvantages of endoscopic surgery compared with more traditional minimally invasive techniques include difficulty in three-dimensional vision due to the lack of binocular vision, increased risk of CSF leak and difficulty to control bleeding in the event of a hemorrhagic complication. We have used this approach for biopsy in two patients and tumor removal in three patients with satisfactory results, and we advocate the concept of endoscopic-assisted microneurosurgery for these complex lesions.

Neuronavigation

In bony skull base lesions, the osseous bony landmarks are distorted or destroyed, thus, surgery of frontobasal or clival tumors could prove challenging due to loss of anatomical orientation. In these settings, neuronavigation may become a useful guiding tool.^[18-21] Neuronavigation was used in one case of chordoma with extended transbasal approach, achieving complete excision.

Outcome and prognosis

Because skull base tumors can be any one of many unrelated tumors, outcome and prognosis vary widely. Adjuvant radiotherapy also plays a substantial role.^[22-24] Benign tumors, such as osteomas, can be resected with minimal mortality and morbidity. However, malignant lesions have significant morbidity with considerable mortality. Chordomas have a 5-year survival of 67.6%, and 20-year survival of only 13.1%,^[25] Ewing's sarcoma group (EWS-pPNET) have a 5-year survival of 50%,^[26] while chondrosarcomas have a 5-year survival of 89%.^[27] In our series, eight patients (20%) expired during 2 years follow-up, of whom five were of EWS-pPNET group.

Conclusions

Bony skull base lesions consist of wide varieties of tumors and require multispecialty management for optimal outcome.

Surgical approach should be individualized for each patient according to tumor location and histopathology. Application of microsurgical and endoscopic techniques, combined with neuronavigation help achieve better outcomes.

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Conflicts of interest

There are no conflicts of interest.

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