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Extramedullary foramen magnum tumors and their surgical management: An experience with 29 cases

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ABSTRACT

Introduction: Surgical management of foramen magnum (FM) tumors is challenging by virtue of their location and vital neurovascular relationships. The ideal approach to anterior/anterolateral tumors continue to evoke controversy even in the modern era. In this article, we present and discuss our experience in the surgical management of these tumors.

Materials and Methods: This retrospective study includes 29 consecutive patients (mean age 36.6 years, M: F = 2.63:1) of extramedullary tumors at the surgical foramen magnum, operated at our center, between 2007 and 2012.

Results: Their mean duration of symptoms was 14. 6 months. A majority of the patients presented with motor symptoms (quadri/paraparesis, *n* = 21, 72.4%), neck pain with/without suboccipital radiation (*n* = 16, 55.2%) and sensory symptoms like tingling/numbness (*n* = 16, 55.2%). There were nine extradural (31%) and 20 intradural tumors (69%). Most of the tumors were located posterolateral to the neuraxis (*n* = 13, 44.8%). Nerve sheath tumors (*n* = 11, 38%) and meningiomas (*n* = 5, 17.2%) were the most commonly encountered histologies in our series. The standard posterior approach was the most frequently employed surgical approach (*n* = 20, 69%). Operative mortality and morbidity were 3.4 and 18.9%, respectively. At a mean follow-up of 27.3 months, 13 out of the 18 available patients improved.

Conclusion: A majority of the foramen magnum tumors are amenable to excision via the standard posterior approach. Small anterior dural-based meningiomas/recurrent tumors may require a lateral approach like the far lateral approach.

Key words: Extramedullary, foramen magnum tumors, pathology, surgical approach

Introduction

Foramen magnum (FM) lesions are relatively rare, accounting for 1.1-3.8% of all spinal cord and brain tumors.[1,2] Nonetheless, they have long intrigued neurosurgeons by virtue of their pathological diversity, bizarre clinical presentations, and the difficulties encountered in their surgical excision.

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Over the years, various surgical approaches for these tumors have been described. These include the standard midline suboccipital approach (with its modifications), lateral approaches with or without partial condyle resection, and the vertebral artery transposition, anterior transoral, and lateral transcervical approaches.[1-8] However, the optimal surgical approach, especially in anterior/anterolateral tumors, still remains debatable.

In this article, we present our experience in the surgical management of extramedullary foramen magnum tumors from a tertiary care teaching neurosurgical center in northern India and review some of the their salient clinical- and management-related issues.

Material and Methods

In this retrospective study, 29 consecutive patients (from June 2007 to August 2012) with extramedullary (extradural and intradural extramedullary) foramen magnum tumors (having their major bulk (>50%) located in the surgical foramen magnum[6,9]) were analyzed.

Their medical records were reviewed and the demographical, clinical, neuroradiological, and treatment related data were collected. The histopathological slides were reviewed by an experienced neuropathologist (SJ) and the diagnosis was reconfirmed. We classified these tumors as anterior, anterolateral, posterolateral, and posteriorly located tumors, on the basis of the findings on axial magnetic resonance imaging (MRI), at the level of the tumor. The anterior tumors were located on either side of the midline, anterior to the spinal cord; anterolateral tumors were located predominantly between the anterior midline and the plane of the dentate ligament; and posterolateral tumors were those that were located posterior to the spinal cord, but were off the posterior midline to one side. Those tumors located behind the neuraxis, near the midline, were labeled as purely posterior tumors. This classification was the basis for selection of the operative approach.

We used three surgical routes. In the anterior route, the transoral approach was used for dealing with the ventral extradural pathology and trans- ventricular endoscopic-decompression of the masswas carried out for an intradural ventrally located arachnoid cyst that was extending as high as the interpeduncular cistern. The lateral approaches included the standard far lateral and the extreme lateral approaches – the technical details of which have been extensively described in the literature. The posterior approach was used most frequently in our series. The posterior approach we used is briefly described below:

We preferred a Concorde position and used a Sugita head holder for head fixation. We used the classical midline incision, starting superiorly at the inion to the bulky palpable spinous process of C2, inferiorly. The muscles were divided along the avascular midline plane till we reached the occiput and spinous process of C2. Subsequently, the suboccipital region and C1 posterior arch were exposed subperiosteally. If the tumor was lateral or anterolateral, this exposure was typically on the side where the tumor was more bulky. The medial limit of bone removal was typically kept at the midline and laterally we went just medial to the condyle (occipital exposure) and the medial end of the sulcus arteriosus (C1 exposure). We rarely exposed the condyles and usually did not mobilize the vertebral artery. The C2 lamina was rarely exposed, unless the tumor extent demanded it. When it was complete, the lamina was removed up to the lateral masses. Suboccipital craniectomy did not extend up to the inferior margin of the transverse sinus, unlike the standard posterior fossa exposures. The dura was usually opened (vertically over the cervical region extending superolaterally on the cerebellar dura), but we preferred a paramedical duratomy. We used a T-extension of the lateral dural flap to aid the exposure. Additional exposure was obtained by division of the dentate ligament and sometimes dorsal root of the C1 (when present) and rarely C2. Tilting of the table to the other side and maneuvering of the microscope further improved visualization of the tumor. The standard microsurgical techniques were used for tumor resection, except when the tumor could be easily mobilized away from the neuraxis, like nerve sheath tumors, and initial internal decompression was carried out (either with suction or with a Cavitron Ultrasonic Surgical Aspirator (CUSA)), before separating the tumor from the nervous tissue, preserving the arachnoid layer. The dura was closed (in intradural tumors) in a water-tight fashion and layered wound closure was done. We did not perform intraoperative neurophysiological monitoring in our patients because of the lack of that facility in our center.

The extent of the excision was labeled as total if no visual tumor residue was present after excision; subtotal if some part of the tumor was deliberately left behind (>50% tumor removed); and biopsy if only <50% of the tumor was removed. Postoperative contrast-enhanced MRI, whenever done, was used to confirm the extent of excision. Follow-up data was obtained from re-admissions, Outpatient Department (OPD) visits, and telephonic/postal interviews. The follow-up statuses were recorded in terms of the patients' symptoms and ability to carry out their daily activities, and were labeled as 'improved', 'same' or 'worse,' as compared to their preoperative statuses.

Results

A total of 29 patients with extramedullary FM tumors, with a mean age of 36.6 years (range 1-72 years) were analyzed. The study included twenty-one males and eight females. The mean duration of the first symptom before clinical diagnosis was 14.6 months (range: fifteen days to eight years). Their clinical features are summarized in Table 1. The most common clinical presentation was motor complaints (n = 21, 72.4%). This was in the form of asymmetrical quadriparesis (n = 18, 62%) or paraparesis (n = 3, 10%). Neck pain with/without suboccipital radiation and sensory symptoms like tingling/ numbness (n = 16 each, 55.2%) were the next most frequent presentations; 31% of the patients (n = 9) presented with bowel-bladder disturbances. Two patients (6.9%) had associated neurofibromatosis-1 and both of them had intradural neurofibromas. As far as the location of the tumors was concerned, the majority (n = 20, 69%) were located intradurally. Of these, six (20.7%) had an additional extradural component. Nine (31.03%) patients had purely extradural tumors. On axial MRI sections through the tumor, a majority of the tumors were located posterolaterally (n = 13, 44.8%). Only three (10.3%) patients had tumors entirely behind the dentate ligaments. Thirteen (44.8%) patients had tumors located anteriorly or anterolaterally, with respect to the neuraxis. Histopathologically, nerve sheath tumors (neurofibromas and schwannomas) were most frequently encountered in our series (n = 11, 39%), followed by meningiomas (n = 5, 17.2%). Table 2 depicts the various histologies encountered in the

Table 1: Summary of the various clinical presentations of 29 foramen magnum tumors encountered in our series

Clinical features	Frequency (*n*)	Percentage
Neck pain	16	55.2
Motor symptoms	21	72.4
Sensory symptoms	16	55.2
Lower cranial nerve dysfunction	5	17.2
Bladder-bowel disturbances	9	31
Respiratory complaints	3	10.3

Table 2: Summary of the various histologies encountered in our series

Histologies	Frequency (*n*)	Percentage
Neurofibroma	4	13.8
Schwannoma	7	24.1
Meningioma	5	17.2
Neuroenteric cyst	2	6.9
Arachnoid cyst	2	6.9
Metastasis	2	6.9
Melanoma	1	3.4
Solitary fibrous tumor	1	3.4
Chordoma	1	3.4
Aneurysmal bone cyst	1	3.4
Malignant meningeal tumor	1	3.4
Epitheloid osteoblastoma	1	3.4
Neurocysticercosis	1	3.4

Table 3: The distribution of various histopathologies in relation to the horizontal section of the foramen magnum

Relation to neuraxis	Frequency (*n*) (percentage)	Histologies
Anterior	6 (20.7)	Neuroenteric cyst (2), meningioma (1), arachnoid cyst (1), solitary fibrous tumor (1), aneurysmal bone cyst (1)
Anterolateral	7 (24.1)	Neurofibroma (2), Schwanoma (1), meningioma (2), metastasis (1), malignant meningeal tumor (1)
Posterolateral	13 (44.8)	Neurofibromas (2), Schwanoma (6), meningioma (1), melanoma (1), chordoma (1), osteoblastoma (1), metastasis (1)
Posterior	3 (10.3)	Meningioma (1), arachnoid cyst (1), neurocysticercosis (1)

current series. The location of these pathologies in relation to the cross-section of the cord is summarized in Table 3. A majority of the tumors measured more than 3 cm in maximum dimension ($n = 20$, 69%). Associated hydrocephalus was observed in one patient (2.8%). Vertebral artery encasement was present in three (8.3%) patients. Excision of the tumor was attempted in 27 (93.1%) patients, whereas, only diagnostic biopsy was performed in two (6.9%) patients. Of the 27 surgical excisions, complete excision was obtained in 15 (51.7%), whereas, 12 (41.4%) patients underwent subtotal excision.The most common operative approach used in our series was the standard posterior approach ($n = 20$, 69%). This approach was used for removing not only lateral and purely posterior tumors, but anterolateral tumors as well. A majority of the lesions approached posteriorly were intradural in location, however, certain extradural tumors like neurofibromas ($n = 2$, 6.9%), chordoma arising from the posterolateral element of C1 ($n = 1$, 3.4%), and malignant meningeal tumor ($n = 1$, 3.4%) were also removed via this approach. The anterior approaches used were the endoscopic transventricular approach and biopsy ($n = 1$) and the transoral approach ($n = 2$). In one of the transoral approaches, the additional posterior approach was also taken and the tumor (aneurysmal bone cyst) was decompressed. This was followed by posterior fusion with occipito-C2/3 lateral mass fixation. The lateral approaches were used in six patients (20.7%). Of them, the far lateral approach was used in four (13.8%) and the extreme lateral approach was used in two (6.9%). Tumors treated by the far lateral approach included one case each of osteoblastoma, meningioma, neuroenteric cyst, and solitary fibrous tumors, respectively. Figure 1a-c shows an anterolaterally located foramen magnum meningioma, which was T2 hypointense, showing homogenous bright enhancement with dural tail and displacement of the neuraxis posteriorly and toward the left. This patient was operated by a right-sided far lateral approach and Simpson grade 2 excision of the tumor was achieved [Figure 1d and e]. On the other hand, the extreme lateral approach was used in treating metastasis ($n = 1$) and anterior extradural meningioma ($n = 1$). Table 4 depicts the various surgical approaches employed in the present series.

Operative mortality was 3.4% ($n = 1$). This patient had an anterolaterally located foramen magnum meningioma, which was operated via the standard posterior approach. Postoperatively, he developed lower cranial nerve dysfunction and aspiration pneumonitis and died. Complications following surgery were encountered in five (18.9%) patients. The complications are summarized in Table 5. Cerebrospinal fluid (CSF) leaks and meningitis were managed conservatively and all the patients improved. No vertebral artery related complications were seen. Three (13.5%) patients had worsening of power in the limbs after surgery, which gradually improved to preoperative levels at the time of discharge in two of them, while no improvement was observed in the third patient. Lower cranial nerve dysfunction worsened in two patients and one patient had fresh lower cranial nerve dysfunction after surgery. These patients were managed with nasogastric tube feeding initially, which could be removed in two of them in six weeks' time. In one patient, nasogastric feeding was continued for three months. One patient was discharged with tracheostomy, which was decannulated within three months of discharge. Eighteen patients were

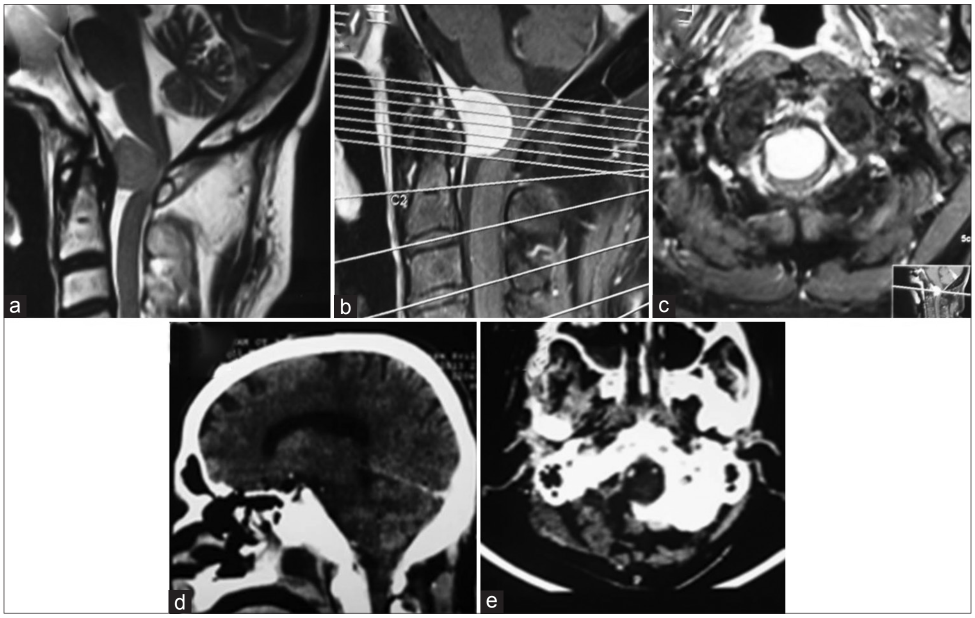

Figure 1: (a) T2 sagittal image shows a ventrally located iso- to slightly hyperintense mass at the foramen magnum without any cord intensity changes. On contrast administration, it shows homogenous and uniform enhancement on contrast administration with a dural tail sign; (b) Axial contrast MRI shows a ventrolateral location of the mass with displacement of the cord posterolaterally toward the left; (c) Figure 1d and e show postoperative CT images with complete excision of the mass and bony exposure done during surgery

Table 4: Various pathological substrates encountered in our series with respect to the cross section of the neuraxis and the approaches used to address them

Location with respect to cross-section of neuraxis	Pathology	Approach used
Anterior (*n*=6)	Neuroenteric cyst (*n*=2)	Posterior approach (*n*=1), Far lateral approach (*n*=1)
	Meningioma (*n*=1)	Extreme lateral approach
	Solitary fibrous tumor (*n*=1)	Far lateral approach
	Arachnoid cyst (*n*=1)	Anterior (endoscopic transventricular)
	Aneurysmal bone cyst (*n*=1)	Anterior (Transoral)
Anterolateral (*n*=7)	Nerve sheath tumor (*n*=3)	Posterior approach
	Meningioma (*n*=2)	Posterior approach (*n*=1), Far lateral approach (*n*=1)
	Metastasis (*n*=1)	Anterior (Transoral)
	Malignant meningeal tumor (*n*=1)	Posterior approach
Posterolateral (*n*=13)	Nerve sheath tumor (*n*=8)	Posterior approach
	Meningioma (*n*=1)	Posterior approach
	Melanoma (*n*=1)	Posterior approach
	Chordoma (*n*=1)	Posterior approach
	Osteoblastoma (*n*=1)	Far lateral approach
	Metastasis (*n*=1)	Extreme lateral approach
Posterior (*n*=3)	Meningioma (*n*=1)	Posterior approach
	Arachnoid cyst (*n*=1)	Posterior approach
	Neurocysticercosis (*n*=1)	Posterior approach

available for follow-up (mean duration 27.3 months, range 3-60 months). At the last follow-up, three patients had died. The diagnoses in these three patients were metastasis ($n = 2$) and ventrally located solitary fibrous tumor ($n = 1$). The patients with metastases died after one and three months of surgery, respectively, probably from widespread metastases. The patient with the solitary fibrous tumor was operated using the far lateral approach. He improved after surgery and went back home. He died after one year of surgery, apparently from some cardiac cause. Thirteen (44.8%) patients had significant improvements in their neurological status and were back to their normal activities; one (3.4%) patient had no improvement in his symptoms, whereas, another patient (3.4%) had worsening of the neurological status after surgery.

Radiotherapy was advised for five patients (17%). These patients had metastasis ($n = 2$), Chordoma ($n = 1$), malignant meningeal tumor ($n = 1$), and aggressive osteoblastoma ($n = 1$), respectively. However, the course could be completed only in the latter three cases, as both patients with metastasis died before completion of the same. No patients received chemotherapy.

Discussion

Foramen magnum tumors are rare and have long been intriguing. The nonspecific and often bizarre symptomatology,[10,11] combined with the technical difficulties encountered in their surgical excisions, make these tumors difficult propositions for neurosurgeons around the world. In recent times, development of MRI, improvements in neuroanesthesia, and various advances in microneurosurgery, including skull-base exposures, have significantly reduced the morbidity and

Table 5: Summary of the operative complications encountered in our series

Complications	Frequency (*n*)	Percentage
CSF leak	4	13.5
Meningitis	1	5.4
Fresh neurological deficits	3	10.3

CSF – Cerebrospinal fluid

mortality associated with their surgical excisions. Although surgical approaches to the foramen magnum are aplenty,[1-8] an optimal surgical approach, especially for anteriorly located tumors, still remains largely debatable.

Anatomical considerations

A sound anatomical knowledge of the FM is very important for properly understanding the various clinical and management-related aspects of these tumors. From a surgical point of view, the foramen magnum is defined by the following landmarks: [6,9]

Anterior border: From the lower third of the clivus to the upper edge of the body of C2

Lateral borders: From the jugular tubercles to the upper aspect of the C2 laminae

Posterior border: From the anterior edge of the squamous occipital bone (posterior rim of FM) to the C2 spinous process.

This region lodges the neuraxial transition from the brainstem to the cervical spinal cord, the suboccipital segment (V3) of the vertebral artery (VA) and its branches, the upper two cervical nerve roots, and the lower cranial nerves. On either side, lies the jugular foramen, with its contents. A combination of the wider sagittal diameters of the first two cervical vertebrae and decreased girth of the spinal cord compared to the subaxial cervical cord enlargement, accounts for a generous buffer subarachnoid space in this area.[10]

Another important aspect of this craniovertebral junction area is that it is designed to provide mobility at the cost of stability. The assembly of various ligaments and lateral joints with their capsules provide a strong contribution to the biomechanical stability of this area. Of note, the otherwise strong investing capsule of the occipito-atlantal lateral joint is weaker inferomedially and very strong on the outer aspect.[11] Posteroinferior condylar resection, not exceeding 50%, is thus usually not associated with any instability of the occipitocervical region.[12] Similar to the occipital condyles, the C1/2 facet joints are relatively weakly supported by the capsule on the medial aspect, biomechanical stability being mostly provided by the lateral pillars.[10]

Clinical presentation

Foramen magnum tumors are known to have protean manifestations, which rarely, if ever, allow proper clinical localization and diagnosis. The symptomatology observed in our series was similar to other reports[10,11,13-15] and only endorsed their non-specific nature. Of all the symptoms, lower cranial nerve dysfunction and respiratory impairments appeared to have a negative effect on the postoperative outcome in our series. The clinical diagnosis of FM tumors is often characteristically delayed.[6,15] Even in this era of MR imaging, the mean length of symptoms, prior to diagnosis, may be as delayed as 30.8 months.[8] A host of different factors, apart from the nonspecific symptomatology, like a generous subarachnoid space at FM and generally slow growing nature of these lesions, are implicated in this delay. In addition, in a country like ours, delayed referral to tertiary centers and ignorance of the initial symptoms are observed in a good number of patients. Although most of the FM tumors present late, a subgroup of them with malignant and inflammatory lesions may, however, manifest very early in their course. There exists another group of patients, who remain asymptomatic for long until they present with sudden onset of rapidly progressive myelopathy, often with severe lower cranial nerve signs and respiratory embarrassment. This is because the symptoms usually do not occur until the physiological limits of the buffer space are totally exhausted.[10] Thereafter, seemingly trivial factors can precipitate a sudden onset of myelopathy. Such an acute presentation was observed in one of our patients with a neuroenteric cyst. The radiological findings in this case initially led us to consider the possibility of intratumoral hemorrhage [Figure 2a-e] as an explanation for such an acute onset of symptoms, which was however, not substantiated at the time of surgery.

Role of neuroimaging

The role of neuroimaging in the evaluation and management of FM tumors cannot be overemphasized. With advantages of excellent soft tissue images in multiple planes and lack of image degradation by bony artifact, MRI is clearly the investigation of choice for these lesions.[16] Additionally MR angiography provides a noninvasive means of demonstrating the vascular anatomy, collateral circulation, and the effect of the tumor on the vertebral arteries (VAs). An MRI shows the exact location, extent, and relationship of the tumor with surrounding neurovascular structures. Appearance of the tumor on T2WI indicates tumor consistency and has great surgical implications. Most importantly, T2 hyperintensity involving the neuraxis adjoining the tumor indicates pial invasion and loss of the arachnoid plane. This information guides the surgeon that any attempt to dissect along the cord-tumor interface will result in cord/brainstem damage. In such cases, the surgeon usually resorts to subtotal removal leaving an intact capsule along the interface. MRI is, however, poor in assessing bony anatomy and often overestimates the available surgical corridor.[17] Hence, a computed tomography (CT) scan, with a bone window, remains the imaging of choice for delineating the osseous surgical corridor as well as detecting calcifications and hyperostosis. A CT scan of the craniovertebral junction provides

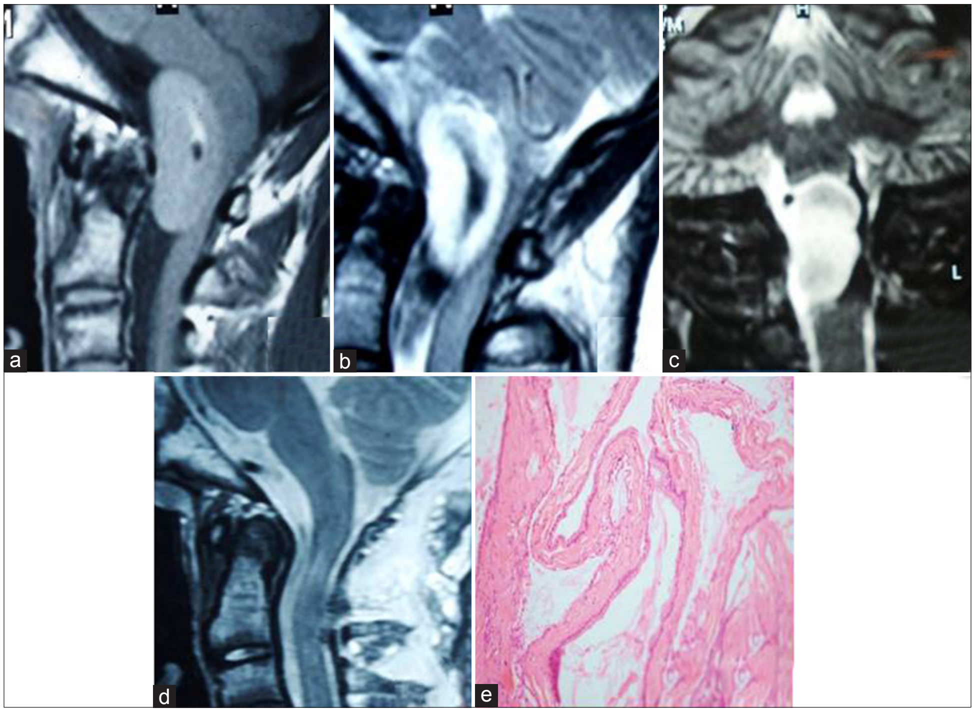

Figure 2: Sagittal (a and b) and coronal (c) MRI sections show a T1 hypointense and T2 hyperintense anteriorly placed intradural foramen magnum mass, with altered intensity at the center. Postoperative sagittal T2WI shows complete tumor excision; (d) The H and E stained section (200X) shows a cyst wall, lined by a columnar and focal mucinous epithelium resting on the subepithelial fibrocollagenous tissue (e) Suggestive of a neuroenteric cyst

information regarding the bony anatomy as well as any associated anomalies. In extradural destructive lesions, it gives information regarding the stability of the area. One can measure the distance between the posteromedial aspect of the occipital condyles and lateral margin of the neuraxis to determine the surgical corridor in case of ventrally located lesions. If this surgical corridor is either large (>2 cm) or adequate (1-2 cm), then there is hardly any need for intraoperative condylar resection. On the other hand, if the corridor is narrow (<1 cm), then one has to drill the posteromedial aspect of the condyle to access the tumor, without unduly retracting the cord.[16] Such information helps the surgical team prepare for the likely need for additional bony work and the prospect of some sort of instrumented stabilization after tumor resection. Hence, for proper preoperative assessment, a combination of both CT and MRI of the FM area is essential.[16,17]

Angiography remains another important neuroimaging tool in FM tumors. Angiography helps in determining the vascularity of the tumor and allows preoperative embolization of the feeders.[18] In addition, for better visualization of the vertebral artery (VA) encasement (with or without balloon occlusion of the same, when required), angiography allows demonstration of the posterior inferior cerebellar artery (PICA), which may sometimes have an extracranial origin.[16,17] Usually, an angiographic study is accomplished with MR angiography, and this modality has largely replaced conventional angiograms. In addition to revealing the vascular (arterial as well as venous) anatomy, including displacements/encasements/tumor blood supply, an angiogram helps to determine the dominant vertebral artery, which has significant surgical considerations. The surgical approach for lesions abutting vertebral arteries is usually from the side of the non-dominant artery and unlike its non-dominant counterpart, the dominant vertebral artery has to be preserved at any cost, during surgery. Moreover, we believe, an angiogram is useful in extradural bony lesions around the course of the vessel. Such information may avoid inadvertent arterial injuries during surgery of these tumors. However, in a recent review, Sohn and Chunghave opined against a catheter angiogram stating that their risks outweighed their benefits.[19]

Histopathological spectrum

A large variety of tumor histologies were encountered in our series, similar to the observations made by other authors.[15,20,21] Nerve sheath tumors and meningiomas were the most common histologies encountered in our current series ($n = 16$, 55.1%), a fact noted by other authors as well.[15,20,21] In addition, we encountered two (6.9%) cases of extradural metastasis. This was relatively higher when one considered that metastatic involvement of the clivus and foramen magnum was stated to be extremely rare.[22] In addition, we encountered some unusual cases like a solitary fibrous tumor and epithelioid osteoblastoma, which further added to the well-known pathological diversity of FM tumors.

Surgical treatment

Controversy surrounds the optimal surgical approach to the FM tumors, especially those located ventrally.[16] The ideal approach should provide direct access to the mass and preserve not only the neurological functions, but also the biomechanical stability of this highly dynamic area. Over the years, numerous surgical approaches to the FM have been developed. Selection of a particular approach depends, foremost, amongst other things, on the location of the tumor with respect to the neuraxis. Broadly the approaches include the anterior, lateral, and posterior approaches.

Posterior and posterolaterally located FM tumors are relatively straightforward as far as the selection of a surgical approach is concerned. The standard posterior approach is easy to perform, less time consuming, and neurosurgeons are highly familiar with it. Craniovertebral junction instability is usually not a concern. Subtle additional maneuvers like medial facetectomy (one-third), lateral T-extension to duratomy placed in the paramedical location, denticulate ligament division, and tilting the operating table by 15 to 30 degrees to the contralateral side, facilitate tumor excision with minimal cord retraction.[23] Moreover, to further widen the corridor, the dorsal roots of C1/2 nerves can be cut with relative impunity. Although some hypoesthesia in the suboccipital area generally accompanies C2 nerve division,[24] the consequences are usually negligible, and paradoxically, it may be beneficial if the patient has had distressing suboccipital pain preoperatively. This has been the most commonly used approach in our series (n = 20, 69%).

The optimal surgical approach to the anterior and anterolaterally located FM lesions is highly debatable.[1-8,25-29] These tumors can be approached anteriorly, laterally or even posteriorly. The transoral route, as noted by Guidetti *et al.*,[25] is theoretically the best approach for ventral FM tumors. However, numerous drawbacks have seen this approach fall out of favor in recent times, especially for intradural lesions. As the tumor is encountered first in this approach, the tumor-cord/brainstem surface is approached blindly. There are problems of CSF leak and meningitis. In addition, velopharyngeal insufficiency and limited lateral access, leading to incomplete excisions and instability, requiring an additional fusion procedure, are the other major shortcomings of the transoral route.[29] The anterior approach was used for two extradural lesions (metastasis (n = 1) and aneurysmal bone cyst (n = 1)) and one intradural lesion (ventral foramen magnum arachnoid cyst). It included a transoral (n = 2) and endoscopic transventricular approach (n = 1). Even as one patient with extradural metastasis underwent transoral biopsy of the mass, the other patient with aneurysmal bone cyst underwent a combined ventral and dorsal decompression, with posterior stabilization by lateral mass plates. In the third patient, with an anteriorly located intradural arachnoid cyst, with associated hydrocephalus, we performed endoscopic transventricular (third ventricle) decompression, and biopsy of the cyst along with endoscopic third ventriculostomy.

The lateral approaches to FM include the anterolateral or the extreme lateral approach (ELA) and the posterolateral or the far lateral approach (FLA). There is, however, a lack of consensus as far as the nomenclature is concerned. These approaches are technically demanding and not all neurosurgeons are well conversant with these approaches. The lateral approaches have developed in parallel with other skull-based techniques and rely on additional bony work in order to minimize neural retraction. Even as FLA is directed behind the sternocleidomastoid muscle and vertebral artery, the ELA is essentially a trajectory undertaken beneath the anterior border of the muscle and in front of the vertebral artery. These two approaches provide different angles of view of the foramen magnum from a lateral perspective. The main difference lies in the extent of the bony drilling and manipulation of the vertebral artery.[16,28] As the bone work in ELA is much more extensive and the vertebral artery needs to be transposed medially, occipitocervical instability and potential injury to the vertebral artery are higher with ELA. Most authors agree that FLA provides adequate access to the anterolateral and large anteriorly located tumors[16,20,27], without the disadvantages of ELA. A small and firm, anteriorly located intradural lesion, like a meningioma causing minimal displacement of the neuraxis, is however, a different scenario, and as noted by Babu *et al.*,[30] ELA becomes an absolute necessity in this situation. Fortunately, such tumors are rare. In an excellent review of foramen magnum meningiomas, Bruneau and George[27] have concluded that the benefits of ELA in terms of surgical exposure is often negated by the risks associated with vertebral artery transposition, eleventh nerve handling, and condylar drilling. Sekhar and Sen[4] first described this approach for ventral foramen magnum lesions and reported 60% complete excision in their series. They concluded that this approach is ideal in dead anterior tumors, recurrent tumors, and anterior extradural meningiomas. Bartalanffy *et al.*[5] used ELA in 19 patients with ventral/ventrolateral foramen magnum meningiomas and reported complete (grade 1 and 2) excision in 100% of the cases with minimal morbidity. There were no recurrences in their series even after five years. In a previously published study from our center,[31] ELA was used in seven patients with various extra- and intradural pathologies. Total excision was obtained in five patients. There were no deaths, no patient needed occipitocervical fusion, and complications were noted in three patients (CSF leak, lower cranial nerve dysfunction). FLA is an extension of the posterior suboccipital approach, wherein, the vertebral artery is mobilized laterally so that the additional length of the C1 posterior arch can be removed for lateral exposure. Many authors[20,32] have found this approach helpful for anterior intradural lesions. In a recent review, it has been reported that gross total resection with this approach ranges from 70 - 100%[32] In a recent meta-analysis involving all reported cases of FLA so far, Komotar *et al*.[33] found that 80.6% of the patients improved, 6.7% remained stable, and 9% worsened postoperatively. The most common complication of FLA has been stated to be lower cranial nerve dysfunction followed by CSF fistula. Although iatrogenic instability of the craniovertebral junction is less likely compared to ELA, as high as 66% of the patients, who had the far lateral approach, are reported to have undergone condylar resection. Mortality with this approach in modern days is, however, very low (<3%)[32]

Use of a predominantly posterior approach for anterior/anterolateral tumors, has generated a lot of interest over the years. Early results were, however, very poor until Goel *et al.*[27] reported excellent results in their series. Thereafter, many others have reiterated the same.[19,28,29,34] They argue that sizeable anterior/anterolateral tumors often create a sufficient corridor by pushing the cord posteromedially, aiding their removal by the posterior approach. Goel *et al.*[27] used this approach in 17 patients with ventral and ventrolateral meningiomas. They extended the bony exposure laterally (lateral suboccipital approach) on the side where the tumor was 'pointing'. They reported partial condylar drilling in two patients. They reported 82% complete resection and only one complication in the form of worsening of lower cranial nerve function. In a recent study, Sohn and Chung,[19] using a pure midline approach, have shown complete excision of 11 ventral foramen magnum meningiomas. They encountered two complications (pseudomeningocele and lower cranial nerve dysfunction). The average size of the tumors in their series was 2.5 cm indicating that even small ventral tumors can be removed via the posterior approach. Gupta *et al.*[29] have also found this approach useful in anterior/anterolateral tumors (13 complete excisions out of 27 patients). The major advantages of the posterior approach include technical ease, practically no need for condyle/facet drilling, decreased chances of vertebral artery injury/instability, faster recovery and decreased morbidity. The disadvantages include a limited corridor, inability to detach the dural blood supply first in the highly vascular tumors, and the increased retraction needed in recurrent/en plaque tumors.[19,27,34] We operated six anterolateral tumors via the posterior approach. We did not perform condylar drilling in any of them. Of these, there were three patients with nerve sheath tumors, one patient each with a neuroenteric cyst, anterolateral meningioma, and malignant meningeal tumor. In cases of neuroenteric cysts, the consistency of the tumor helped in tumor delivery, whereas, in the remaining patients, the natural corridor was created by the tumor itself. However, a number of publications have pointed out higher rates of mortality and morbidity associated with this approach, when dealing with anterior or anterolateral FM lesions.[30,34]

We agree that there have not been many cases of anterior/anterolateral meningiomas in our series. These ventral meningiomas with broad dural attachment are definitely a much more challenging proposition, compared to nerve sheath tumors and neuroenteric cysts. The limited number of patients in our study is a major limitation in drawing any inference regarding the optimal surgical approach. However, at the same time, we do not believe that each and every such case needs one of the lateral approaches. Unless it is a small, ventrally located midline meningioma or a recurrent tumor, we would prefer a modified posterior midline approach from the side of the maximum bulk of the tumor. This is because of the fact that an enlarging tumor usually displaces the neuraxis in such a way that at least an adequate surgical corridor becomes available in a majority of the patients.

No matter which surgical approach is used, the basic principles of microsurgical tumor excision remain the same. Especially in ventrally located tumors, it is important to avoid any kind of retraction of the neuraxis. A generous internal decompression of the tumor is very essential, especially in meningiomas. Either suction decompression or ultrasonic aspiration (CUSA), depending on the texture of the tumor, can be employed. In this regard, the appearance of the tumor on a T2-weighted image (iso- to hypointense, indicating a fibrous tumor, and heterogeneous hyperintense indicating a softer interior) can be of some predictive importance. Although CUSA is very effective, we have found suction decompression and piecemeal decompression using micro scissors very useful. Adequate internal decompression gradually widens the working space and allows easy separation of the capsule from the nervous tissue. Preservation and separation of the arachnoid from the surface of the tumor is vital. It ensures easy separation of the cranial nerves, vessels, and very importantly, the brainstem. Copious saline irrigation, meticulous attention to hemostasis, and minimal use of bipolar coagulation, especially over the tumor surface, are of utmost importance in this regard.

Intraoperative neurophysiological monitoring is an essential adjunct in surgeries around the foramen magnum, especially for intradural procedures. Monitoring of the brainstem (with brainstem auditory evoked potential and somatosensory evoked potential) and lower cranial nerves (electromyography of the ninth, eleventh, and twelfth nerve innervated muscles) are the recommended monitoring practices in such surgeries. The purported advantages include identification of structures, guidance during surgery, and importantly avoidance of complications by responding to the warning signals immediately.[19] These monitoring practices require specific neuroanesthetic considerations and acquaintance with the technicalities of the procedure, both on the part of the surgeon and the theater personnel. Their use is, however, often subject to surgeon prejudice and preference. In the present series, neurophysiological monitoring has not been used in any of the patients, because of the lack of this facility at our center during that time.

The role of radiotherapy/chemotherapy in these tumors is primarily determined by the histopathogical diagnosis and totality of the excision. For obvious reasons, in malignant histologies, it becomes imperative to administer adjuvant therapy. Some authors[32,35] have reported their results of Gamma Knife radiosurgery (GKS) in foramen magnum meningiomas. GKS has been advocated for elderly patients, who have medical contraindications for surgery, recurrent, residual tumors, and in patients refusing surgery. Primary GKS, instead of surgery, has been reported, to treat small meningiomas in this location. Studies indicate that GKS at best halts tumor growth and seldom brings down the volume. The long-term results of GKS in FMM are, however, awaited. We do not have gamma knife services at our center. As only three patients completed radiotherapy (none with meningioma) and no patient received chemotherapy in the present series, it is difficult for us to state whether the adjuvant therapy affects the final outcome in these tumors.

Thirteen out of 29 patients (44%) showed significant clinical improvement in the present series. Although this appears relatively low, one has to consider that 11 patients were not available for follow-up. Hence, among those who turned up for follow-up or were able to be contacted, a majority (13 out of 18) showed improvement. This was in spite of the fact that nearly half of our patients underwent subtotal excisions. Our results indicated that most of these patients tended to do well in the long run and every effort had to be made to reduce the immediate postoperative morbidity and mortality. In this regard, neuraxial decompression by the least morbid route, rather than radiological complete tumor excision, should be the goal of surgery. Studies have indicated that the extent of excision depends more often on the tumor consistency, vertebral artery encasement, and tumor invasiveness rather than the approach used.[15,27]

Conclusion

Foramen magnum tumors are more frequent in males. Most patients present with a long history of motor complaints followed by neck pain and sensory symptoms. A majority of the tumors are located intradurally and situated posterolateral to the neuraxis. Although a diverse group of tumors occur in this region, nerve sheath tumors and meningiomas account for most such cases. A surgical approach to these tumors is usually guided by the tumor location. Most tumors can be approached by the conventional posterior approach with subtle modifications, however, a far lateral approach is preferable for anterior or anterolaterally situated tumors. Surgical morbidity and mortality are low in experienced hands. Most patients do well in the long run.

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