

Clinico-radiological profile and nuances in the management of cervicomedullary junction intramedullary tumors

Anup P. Nair, Anant Mehrotra, Kuntal Kanti Das, Arun K. Srivastava, Rabi Narayan Sahu, Raj Kumar

Department of Neurosurgery, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow, Uttar Pradesh, India

ABSTRACT

Background: Cervicomedullary junction (CMJ) intramedullary tumors comprise of tumors that often pose a surgical challenge even in the present era. Though classified under brainstem glioma CMJ tumors are well amenable for surgical resection and have a good outcome. Various factors are involved in the outcome of these patients following surgery and a proper pre-operative assessment is often required to reduce the morbidity and mortality.

Materials and Methods: Patients admitted in the Department of Neurosurgery with a diagnosis of CMJ intramedullary tumors from January 2001 to January 2010 were included in the study. Patients were analyzed retrospectively regarding their symptomatology, clinical findings, radiology and outcome after surgery. All patients underwent pre-operative magnetic resonance imaging (MRI) and post-operatively all were managed in the neurosurgery intensive care unit for days to weeks or as dictated by the clinical condition of the patient.

Results: A total of 32 patients were included in the present study. The number of males was 21 (65.6%) and females were 11 (34.4%) respectively. The mean age of presentation was 22.97 ± 9.8 years and the mean duration of pre-operative symptoms was 13.3 ± 12.9 months. The tumor had extension from the CMJ into the cervical region in 17 (53.1%) and into the medullary region in 14 (43.8%) patients. Tumor decompression was done in 9 (28.1%) patients and gross near total excision done in 23 (71.87%) patients.

Conclusions: Cervicomedullary tumors are a subset of tumors quite distinct from the usual brainstem tumors. Patients having predominant cervical involvement present early and have less post-operative deficits. Those with predominantly more medullary involvement present late, hence have a much more morbid outcome. Though closely related to vital neural structures, surgery forms the mainstay of treatment. Adequate pre-operative planning and preparation of the patient along with intense post-operative monitoring and ventilatory assistance as and when required helps in a good surgical outcome.

Key words: Cervicomedullary junction, ependymoma, pilocytic astrocytoma, tracheostomy

Introduction

Cervicomedullary junction (CMJ), as the name implies is the region where the brainstem continues as the spinal cord. A lesion located in this region affects either the

brainstem or cervical cord or both depending on its extent and pathology. Involvement of brainstem is manifested as cranial nerve palsies, decreased respiratory drive, long tract signs and hydrocephalus if there is obstruction of the fourth ventricle.^[1] The cervical cord involvement is associated with long tract signs, features of myelopathy, Horner's syndrome and facial hypoesthesia. Intramedullary lesions in this region can obstruct the flow of cerebrospinal fluid (CSF) and lead to the formation of syringobulbia and syringomyelia.^[2-4] Intramedullary tumors account for 20-30% of intradural tumors in adults and up to 50% of intradural tumors in children. Glial tumors such as astrocytomas and ependymomas represent 80% of intramedullary tumors.^[1] These tumors push vital neural structures (tracts and nuclei) that come in their way as they grow cranially. Majority of these tumors appear to represent cervical spinal cord tumors where rostral growth into the medulla has been blocked by the decussating white matter tracts.^[5,6] As a result most of the tumors are diverted

Access this article online	
Quick Response Code:	Website: www.asianjns.org
	DOI: 10.4103/1793-5482.131060

Address for correspondence:

Prof. Raj Kumar, Department of Neurosurgery, Trauma Centre, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow - 226 014, Uttar Pradesh, India.
E-mail: rajkumar1959@gmail.com

dorsally and enters the fourth ventricle. This often leads to obstruction of the CSF outflow from the fourth ventricle thereby leading to hydrocephalus and features of raised intracranial pressure. Rarely malignant tumors may infiltrate through the decussating fibers to involve the medulla. In this case patient may present with features of lower cranial nerve palsies in addition to the symptoms referable to upper cervical cord involvement. Surgery appears to be the most appropriate treatment option; however complete surgical excision is often difficult due to the absence of a tumor-parenchymal interface.^[7] With modern surgical techniques and instruments it has now become possible to excise these lesions with significant reduction in morbidity and mortality. Here we have tried to analyze the outcome of cervicomedullary tumors taking a planned pre-operative preparation and excision according to our department protocol.

Materials and Methods

Between 2001 and 2010, 32 cases of intramedullary CMJ tumors were operated at the Sanjay Gandhi Post Graduate Institute of Medical Sciences, Lucknow. The pre-operative clinical and radiological data along with intra and post-operative intensive care monitoring data were collected and reviewed. These patients were managed according to department protocol. The histopathological diagnosis were also collected and analyzed. Inclusion criteria was defined as:

- Intramedullary tumors having their bulk situated in the CMJ with cervical, medullary or fourth ventricular extension.
- Patients who had a minimum follow-up of 6 months. The records were assessed for patient's age at diagnosis, gender, chief complaints, pre-operative symptoms and signs, duration of pre-operative symptoms and any specific intra-operative events such as bradycardia or hypotension. Patients were assessed pre-operatively using the Kumar and Kalra's scoring system that included motor power, gait, sensory involvement, sphincter involvement, spasticity and respiratory difficulty on a scale ranging

from 1 to 5. The maximum score described was 30 and the grading was done as follows: Grade I: 25-30; Grade II: 19-24; Grade III: 13-18; Grade IV: <13 [Table 1].

In the pre-operative period special emphasis was given to improve patient's respiratory reserve especially if they were compromised. Patients were advised to stop smoking and start incentive spirometry right from the outpatient department. Once they were admitted bedside breath holding time (BHT) and single breath count (SBC) was noted. Those with a BHT of less than 15 and with a SBC of less than 20 were considered as those with poor respiratory reserve. Pulmonary function tests were also done in all patients and those with altered forced expiratory volume (FEV1) and forced vital capacity (FVC) were also given special consideration. The patients with compromised breathing parameters underwent chest physiotherapy and nebulization with bronchodilators from the day of admission. Breathing exercises along with steroids (dexamethasone) to take care of the edema surrounding the tumor was also initiated in the pre-operative period. Patients who did not improve significantly even after such intense physiotherapy was kept under close scrutiny in the post-operative period. In the post-operative period if they required ventilatory assistance for more than 24 h, elective tracheostomy was done. Patients who had dysphagia and absent cough reflex were prepared for Ryle's tube feeding or percutaneous feeding gastrostomy. A detailed and informed consent was taken in the pre-operative period for tracheostomy and Ryles tube feeding that may be required in the process of weaning.

All patients underwent pre-operative MRI of the CMJ with Gadolinium contrast images. On contrast images special focus was given to identify any leptomeningeal enhancement as it indicated the malignant potential of the tumor.

Patients were compared in the pre-operative and post-operative period and were analyzed. All patients underwent surgical excision and the specimen was sent for histopathological specimen. Surgery was carried out via a cervical laminectomy

Table 1: Kumar and Kalra scoring system

	1	2	3	4	5	Total score
Motor power	Contraction without movement or plegia	Movement with gravity eliminated	Movement against gravity	Movement against gravity and some resistance	Normal power	5
Gait	Wheel chair bound or bedridden	Restricted mobility despite aid	Mobility using aid	Slight disturbance no aid required	Normal	5
Sensory involvement	Total loss of function	Restriction of function of daily living	Significant(>25%) but no dysfunction of daily living	Insignificant	No sensory loss	5
Sphincteric involvement	Retention requiring indwelling catheter	Occasional clean intermittent catheterization	Hesitancy with residual urine not requiring catheter	Hesitancy but no residual urine	Normal	5
Spasticity	Affected part rigid in flexion or extension	Passive movement difficult	Passive movements easy	Slight increase, a catch felt	No increase in tone	5
Respiratory difficulty	Requires assisted respiration	Dyspnea at rest	Dyspnea at mild exertion	Dyspnea at moderate exertion, unable to do active work	Normal	5

and suboccipital craniectomy with the patient in the prone position. This provided complete exposure of the tumor from its upper to lower extent. Post-operatively patients were managed in the intensive care unit and were analyzed as to the need for tracheostomy, Ryles tube feeding, percutaneous endoscopic gastrostomy (PEG) and ventilator support as mentioned earlier. Tracheal toilet and chest physiotherapy were continued actively in the post-operative period. Ventilator dependent patients were gradually weaned and in case of sepsis antibiotics were started according to culture and sensitivity reports. Regular follow-up at 6 weeks, 6 months, 12 months and then yearly was done.

Results

The minimum age was 8 years and maximum 43 with a mean of 22.97 ± 9.8 years. Males constituted 65.6% ($n = 21$) and females 34.4% ($n = 11$) of the total number of patients. 13 (40.6%) patients belonged to the pediatric (<18 years) age group. The mean duration was 13.9 months with a minimum of 2 months and a maximum of 56 months. The most common clinical presentation was limb weakness and was present in 25 (78.1%) patient. Other presenting features included sub-occipital pain in 22 (68.8%) patients, abnormal sensations ($n = 18$, 56.3%), bowel and bladder involvement in nine patients (28.1%), respiratory problems in 15 patients (46.9%), vomiting ($n = 15$, 46.9%), swallowing difficulty ($n = 14$, 43.8%), nasal regurgitation ($n = 14$, 43.8%) and headache ($n = 11$, 34.4%) respectively. Pre-operative shunting was done in three patients (9.4%) as there was blockage of CSF flow in the region of the fourth ventricle due to extension of the tumor into the fourth ventricle. These patients had hydrocephalus on pre-operative imaging and patients were drowsy at admission. Patient had improvement in neurological status in terms of conscious level after shunt procedure. Ninth and tenth cranial nerves were involved in 14 (43.8%) patients. Eleventh nerve involvement was present in 10 (31.3%) patients and twelfth nerve was involved in 7 (21.9%) patients. Cerebellar signs were present in 7 (21.9%) patients and 5 (15.6%) had Horners syndrome. Posterior column sensations were impaired in 10 (31.3%) patients [Table 2].

Pre-operatively patients were graded according to the Kumar and Kalra scoring system. 4 (12.5%) patients came under Grade I, 17 (53.1%) were Grade II, 9 (28.1%) were Grade III and 2 (6.3%) were Grade IV. Radiology revealed tumor localized within the CMJ or extending into medulla, cervical spinal cord or into the fourth ventricle.

The lesion was confined to the CVJ in 1 (3.1%). There was predominant extension into the medulla in 14 (43.8%) patients and into the cervical region in 17 (53.1%). In one patient the tumor (3.1%) was extending from the medulla to the thoracic region [Table 3]. There was extension into the fourth ventricle in 7 (21.9%) patients, of which three patients

Table 2: Symptoms and signs

Symptoms and signs	N=32 (%)
Suboccipital pain/neck pain	22 (68.8)
Headache	11 (34.4)
Vomiting	15 (46.9)
Tingling and paresthesias	18 (56.3)
Limb weakness	25 (78.1)
Bowel and bladder involvement	9 (28.1)
Respiratory difficulty	15 (46.9)
Dysphagia	14 (43.8)
Nasal regurgitation	14 (43.8)
IX and X	14 (43.8)
Spinal accessory nerve	10 (31.3)
Hypoglossal nerve	7 (21.9)
Horner's syndrome	5 (15.6)
Cerebellar involvement	7 (21.9)

Table 3: Radiological extent of tumor

Radiological extent	N=32 (%)
Cervicomedullary junction	1 (3.1)
Medullary extension	14 (43.8)
Cervical extension	17 (53.1)
Medulla to thoracic region	1 (3.1)
Fourth ventricular extension	7 (21.9)

required pre-operative shunting due to hydrocephalus (9.35%). There was evidence of syrinx on pre-operative imaging in 10 (31.25%) patients.

All patients underwent surgery in the prone position where midline suboccipital craniectomy and laminectomy was done. The cervical laminectomy was extended to the level corresponding with the radiology and if required was extended into the thoracic region as was in one of our case. Post-operatively, patients were managed in the neurosurgery intensive care unit. Intra-operatively, 12 (37.5%) patients experienced episodic bradycardia and hypotension while dealing with the tumor in the cervicomedullary region. As soon as bradycardia occurred, surgery was stopped and heart rate was reverted back to normal with atropine. Once normalized, surgery was continued. If bradycardia continued then surgery was stopped for a while and further decompression was done at a site distant from the present one. We would then return to the present site after giving adequate time for the vital parameters to get settled. Tumor decompression was done in 9 (28.1%) patients and in 23 (71.9%) patients we could achieve a gross near total tumor excision. The completeness of tumor excision was based on the surgeon's intra-operative impression. Patients where tumor decompression was done were subjected to post-operative radiotherapy.

A total of 12 (37.5%) patients were tracheostomized in the post-operative period. 15 (46.9%) patients had poor respiratory

reserve pre-operatively of which eight did not improve even after intense breathing exercise. These eight patients underwent elective tracheostomy within 24 h after surgery as weaning was not possible in them. Four patients (who did not belong to the pre-operative poor respiratory reserve group) underwent tracheostomy within the 1st week of surgery as it was difficult to wean them off the ventilator.

At follow-up of 12 months, 9 (75%) of the tracheostomized patients could be decannulated. 2 (16.7%) of them died due to septicemia following aspiration pneumonitis and 1 (8.3%) died at his residence probably due to tracheostomy tube blockade.

10 (31.3%) patients were ventilator dependent in the post-operative period. These patients underwent regular tracheal toileting and intense chest physiotherapy following which they eight of them could be weaned over a period of 2 months and the remaining two could be weaned over a period of 4 and 6 months.

15 (46.9%) patients required Ryle's tube feeding in the post-operative period, out of which four were later converted to PEG. At 12 months follow-up, nine of the fifteen patients could tolerate oral feeds and discontinued Ryles tube feeds. PEG was removed at 6 months follow-up in all four patients and could tolerate orally well. Out of the remaining two, one began to tolerate orally after 24 months. One patient was lost to follow-up.

Biopsy was pilocytic astrocytoma in 22 (68.8%) patients. Others included ependymoma in 5 (15.6%) patients, hemangioblastoma and fibrillary astrocytoma in 2 (6.2%) patients each and glioneuronal tumor in one patient [3.1%, Figure 1]. One patient whose biopsy was reported a hemangioblastoma was a case of Von-Hippel Lindau syndrome with cerebellar hemangioblastomas and renal cortical cysts. Patients were regularly followed-up at 6 weeks, 6 months, 12 months and then yearly. At 6 months follow-up, Kumar and Kalra scoring system was reapplied in these patients and their results were analyzed and compared with the pre-operative grades. It was found that 20 (62.5%) patients fell in Grade II, 8 (25%) in Grade I, 3 (9.4%) in Grade III and 1 (3.1%) patient came under Grade IV [Table 4]. Post-operative CSF leak was present in three patients (9.3%). Three (9.3%) patients developed recurrence at a follow-up of 36, 24 and 18 months. Their biopsies were fibrillary astrocytoma in one patient and the rest two were pilocytic astrocytoma. The patient whose biopsy was fibrillary astrocytoma had received radiotherapy and developed recurrence after 36 months. The remaining two patients had developed recurrence after 18 and 24 months. All except the patient of fibrillary astrocytoma had undergone gross near total excision the 1st time. The patient with fibrillary astrocytoma underwent near total excision and portion of tumor attached to brainstem had to be left behind. The 2nd time all patients underwent total excision and are on regular follow-up.

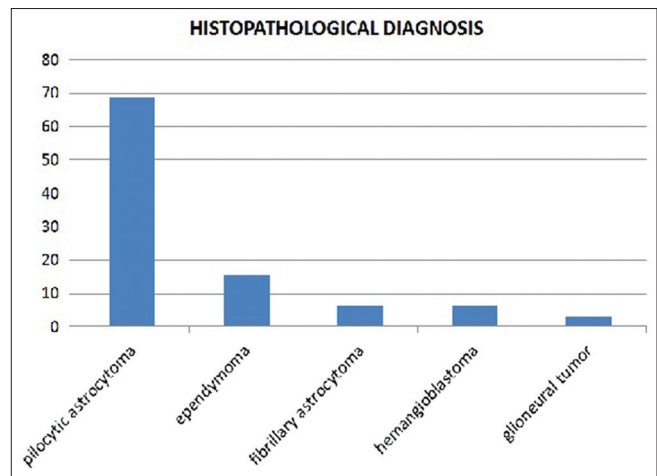


Figure 1: Graphic representation of histopathological diagnosis

Table 4: Comparison of pre-operative and post-operative Kumar and Kalra scoring system

K and K score	Grade I	Grade 2	Grade 3	Grade 4
Pre-operative	4 (12.5)	17 (53.1)	9 (28.1)	2 (6.3)
Post-operative	8 (25.8)	20 (64.5)	3 (9.6)	1 (3.2)

Discussion

Tumors of the CMJ were initially considered a part of brainstem glioma and hence like brainstem glioma they too were considered surgically inaccessible and were associated with a poor prognosis.^[8,9] In 1987, Epstein and Wisoff classified them into five categories namely, diffuse, focal, exophytic, cystic and cervicomedullary.^[10] Tumors of the cervicomedullary region are different from the rest in that they have a long duration of symptoms, low grade pathology in most patients, are amenable for surgical excision and have a good outcome.^[11,12] However much of these studies were done in the pediatric population. In the current study the mean age of presentation was 22.97 years and 13 patients (40.7%) belonged to the pediatric category. In children, brainstem gliomas constitute 10% of brain tumors, whereas in adults they contribute 2% of glioma. However our series had a significant larger number of adult cases (60%).

CMJ represents a transition zone where the cranial compartment continues into the spinal canal. Important structures lying in its close vicinity are the cardiorespiratory center and origin of lower cranial nerves and their infranuclear portions. The presence of pyramidal, spinothalamic tract and posterior column in this region add to its complexity.^[13,14] Therefore lesions affecting the CMJ present have a diverse spectrum of clinical presentation. In our series limb weakness was the most common presentation and was seen in 25 (78.1%) patients. Lower cranial nerve involvement in the form of dysphagia was present in 9 (28.1%) patients. The mean duration of presentation was 13.34 months. This was slightly less when compared to the reviews from literature where a mean duration

of 24 months have been reported.^[10] The early presentation in our study may be due to the fact that majority of our patients had limb weakness as the presenting symptom unlike other studies where cranial nerve palsies was the dominant symptom. This can also be explained by the fact that in our patients the extension into the cervical region was more in number and much earlier than the medullary involvement [Figure 2a-e]. It is difficult to determine the exact epicenter of a CMJ tumor and hence we have included all tumors involving the CMJ with extension into either medulla or cervical region. It is found that children who only have a medullary extension usually present late as they are often investigated for complaints pertaining to respiratory, metabolic and gastrointestinal system [Figure 3a-c]. Their symptoms are due to involvement of lower cranial nerves; however they are picked up late and hence a delay in the diagnosis occurs.^[14] Those with cervical extension present with features of limb weakness and myelopathy and the diagnosis is often straight forward.

Tumors extending into the medulla and CMJ often have symptoms of respiratory compromise such as a decrease in BHT, SBC and altered FEV1 and FVC values.^[14] This is mainly due to the long standing compression on the respiratory center in the medulla. In our series, of the fourteen patients who had medullary involvement eleven had compromised

pulmonary function tests and decreased SBC and BHT. This is often important as it helps us to pre-operatively prepare the patients respiratory compliance by incentive spirometry and active chest physiotherapy. It also helps in reducing the incidence of ventilator support and tracheostomies. Adequate breathing exercises with steroids were started in these patients right from the time of admission. This was done mainly to take care of the edema surrounding the tumor. Patients who did not improve despite respiratory exercises and physiotherapy were planned for elective tracheostomy in the immediate post-operative period. Hence the pre-operative impression of a low respiratory reserve indeed helped our neurosurgical team to prepare the patient well for surgery. Patients who had difficulty in weaning off from the ventilator in the post-operative period were tracheostomised and were subjected to adequate tracheal toileting and physiotherapy. Rath *et al.* compared pre-operative and post-operative pulmonary function tests in patients with craniovertebral junction anomalies.^[15] He showed that there was no improvement in the post-operative period and to the contrary there was deterioration in vital capacity on the 1st post-operative day. It has been postulated that sudden release of spinal cord compression causes acute paracentral microhemorrhages in the CMJ and this may be a cause for deterioration of pulmonary function.^[16,17] Tumiati and workers have proposed that presence of syringomyelia and syringobulbia may also be a cause for altered respiratory drive.^[18]

MRI plays an important role in differentiating a benign from a malignant lesion where Cohen *et al.* have shown that leptomeningeal spread during the clinical course of the disease favors a malignant behavior.^[19] In our study, there was evidence of leptomeningeal enhancement in one patient and his biopsy was Grade III ependymoma [Figure 2b]. We had a case of Von Hippel Lindau disease in our series who had features of cerebellar and CMJ hemangioblastoma [Figure 4a-c].

Though CMJ intramedullary tumors were once considered surgically inaccessible, surgical excision is presently the treatment of choice. Surgical excision is possible due to the fact that 90% of these tumors are low grade. However, there are certain facts that the operating surgeon should

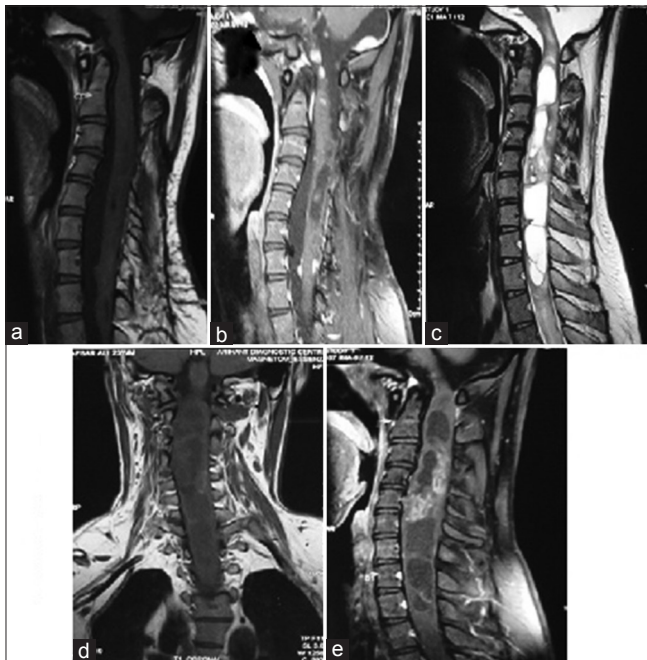


Figure 2: (a) Magnetic resonance imaging T1WI of the cervicomedullary junction with cervical spine showing ill-defined intramedullary tumor extending from the cervicomedullary junction (CMJ) to D2 level. The tumor is causing diffuse cord expansion (b) Contrast images showing heterogeneous contrast enhancement with multiple areas of leptomeningeal enhancement (c) T2WI showing multiple hyperintense areas within the tumor with cord edema and cord expansion (d and e) coronal T1WI and contrast images showing diffuse cord expansion from CMJ to D2 level

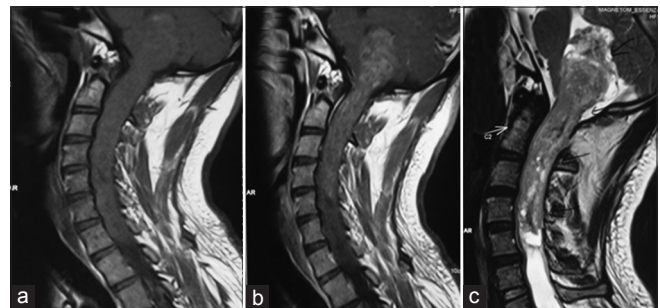


Figure 3: (a-c) T1WI, contrast T1WI and T2WI images of a tumor extending from the medulla up to C7 level with heterogeneous contrast enhancement and evidence of syrinx formation below the lesion

keep in mind while operating the same. Intra-operatively, there is always a high chance of bradycardia and one has to be cautious about this. Monitoring of cardiovascular and respiratory parameters is advocated to detect changes in brainstem function that might result from surgical handling.^[19] The nucleus tractus solitarius (NTS) and the reticular formation situated in the caudal dorsal medulla is responsible for this.^[20,21] Even light stimulation of these structures by bipolar forceps during surgery results in acute hypotension and bradycardia. The middle and the caudal regions of the NTS have been reported to represent the sites where afferents mediating cardiovascular and respiratory reflexes make their primary synapse.^[22,23]

We experienced intra-operative episodic bradycardia in 12 (37.5%) patients but these were managed successfully. What we would like to stress is that recurrent bradycardia may be an indication for compromising with a tumor decompression rather than an aggressive total tumor excision.

Radical surgical excision is the preferred treatment modality for tumors located in the CMJ.^[24] However if the tumor is closely attached to the brainstem or the lower cranial nerves, then a decompression rather than total excision would be better to lessen the morbidity of the patient. For this reason, nine of our patients underwent tumor decompression. Radical excision was avoided in these patients either due to recurrent intra-operative mishaps such as bradycardia or hypotension or due to their close proximity to the caudal dorsal medulla. Absence of a plane between the tumor and the normal tissue added to the difficulty in achieving a total excision in cases where it could not be achieved [Figures 5a,b and 6a-e]. Patients who had a residual tumor were subjected to radiotherapy.

We applied the Kumar and Kalra scoring system pre- and post-operatively and analyzed the results. Grades I and II were considered favorable grades and it was noticed that the number of patients increased to 28 from 21 in the



Figure 4: (a) Computed tomography head of a patient presenting with features of posterior fossa tumor and the biopsy was hemangioblastoma. This patient developed another lesion after 1 year in the cervicomedullary junction (b and c) T1WI and T2WI showing diffuse involvement of cord and the biopsy was hemangioblastoma. This patient had renal cortical cysts and polycythemia and was diagnosed as a case of Von Hippel Lindau disease

post-operative period. Grades III and IV were unfavorable grades and there was a reduction from 11 to 4 in this group. This showed that once the compressing factor was removed from the CMJ, there was significant improvement in various aspects such as respiration, sensorimotor deficits, gait and spasticity.

Cranial nerve deficits are variable and depend on the pre-operative involvement and surgical approach adopted.^[25,26] Damage to the lower cranial nerves leads to dysphagia, absent gag and cough reflexes.^[27-29] These individuals are at risk for frequent micro aspirations and hence when indicated tracheostomy with PEG/Ryles tube feeding is recommended. Post-operative respiratory compromise is a serious issue and was observed in twelve of our patients. Tracheostomy was done in all of them; however at a mean follow-up of 1 year nine could be decannulated. The role of radiotherapy in CMJ intramedullary tumors is not well established. Their role in diffuse brainstem high grade glioma is only marginal. Promising results have been obtained when combined with chemotherapy.

It is quiet unusual to find structural complications related to spinal instability in the post-operative follow-up of these patients. Epstein has reported that such complications are common in children less than 3 years of age and is unusual in adult population.^[1]

Conclusion

It seems that CMJ intramedullary tumors be classified as a separate entity distinct from brainstem tumors. Though they are closely related to vital anatomical structures, they can be surgically excised with acceptable morbidity and mortality provided pre-operative evaluation for breathing parameters

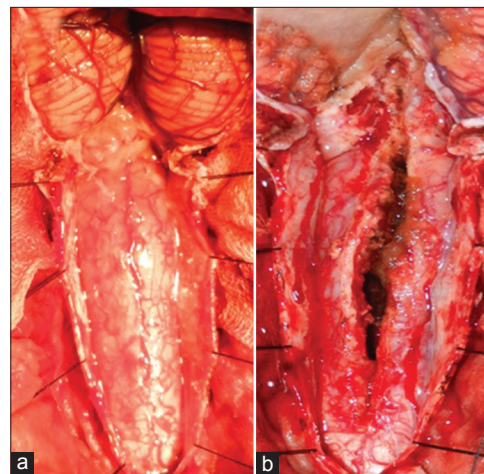


Figure 5: (a) Intra-operative photograph showing intramedullary lesion situated in the medulla and the cervical cord. The tonsils appear to be pushed and separated apart and the cord is expanded (b) Intra-operative photograph showing tumor excision and the decompressed cord after excision

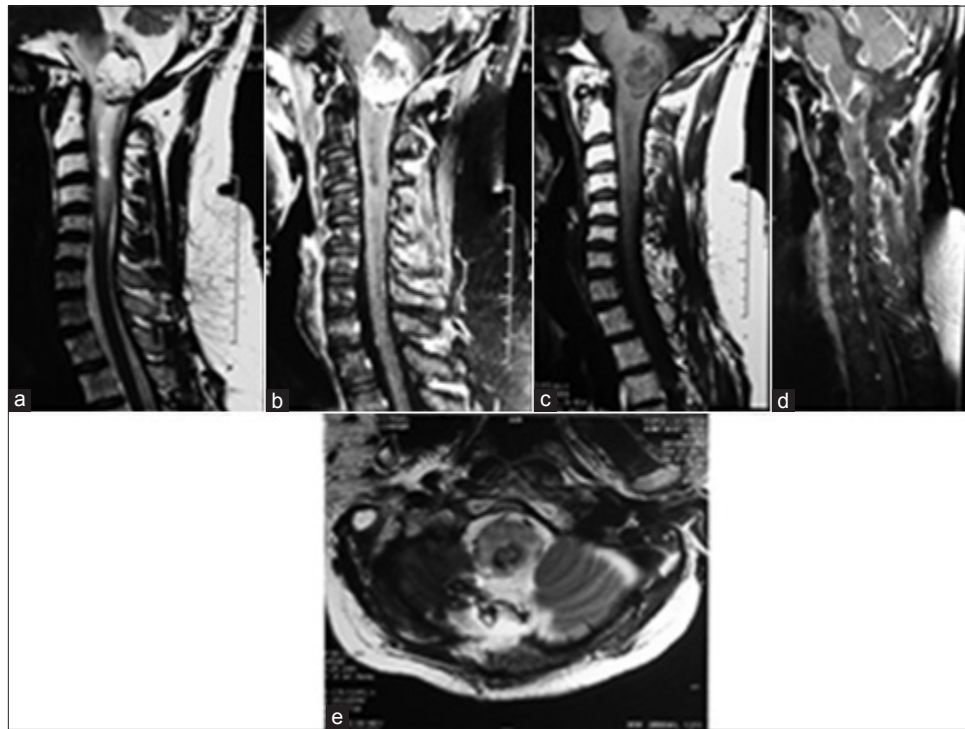


Figure 6: (a-c) Magnetic resonance images of a heterointense lesion that is contrast enhancing with areas of necrosis situated in the lower medulla and extending into the cervicomedullary junction. The whole of the medulla appears to be enlarged and there is evidence of associated edema extending into the cervical cord (d and e) Post-operative magnetic resonance images of the patient showing almost complete excision of the tumor with slight residual tumor in the region of the brainstem

are adequate. This is due to the fact that majority of them are low grade tumors. It has been shown that tumors having a medullary component present late and are associated with a worse outcome when compared to those with cervical extension. Tumors with cervical component present early and have a better prognosis. Significant medullary involvement increases the need for post-operative tracheostomy and Ryles tube feeding. However with active physiotherapy and chest expansion exercises, it is often possible to recover patients from these morbid conditions. Intraoperative bradycardia and hypotension may occur during surgery and whether they affect the outcome of surgery is not yet established. Certain amount of tumor needs to be left behind if there is intractable bradycardia and hypotension. CMJ intramedullary tumors, though benign in majority, pose significant difficulties in surgical management due to its intimate relation to vital structures. However with proper pre-operative planning and good surgical acumen they can be excised with negligible morbidity.

References


- Epstein F, Wisoff J. Intra-axial tumors of the cervicomedullary junction. *J Neurosurg* 1987;67:483-7.
- Heffez DS, Zinreich SJ, Long DM. Surgical resection of intrinsic brain stem lesions: An overview. *Neurosurgery* 1990;27:789-97.
- Hoffman HJ, Becker L, Craven MA. A clinically and pathologically distinct group of benign brain stem gliomas. *Neurosurgery* 1980;7:243-8.
- Epstein F, Wisoff JH. Intrinsic brainstem tumors in childhood: Surgical indications. *J Neurooncol* 1988;6:309-17.
- Ahyai A, Woerner U, Markakis E. Surgical treatment of intramedullary tumors (spinal cord and medulla oblongata). *Analysis of 16 cases. Neurosurg Rev* 1990;13:45-52.
- Allen JC, Lassooff SJ. Outcome after surgery for intramedullary spinal cord tumors. *Neurosurgery* 1990;26:1091.
- Abott R, Ragheb J, Epstein FJ. Brainstem tumors: Surgical indications. In: Cheek WR, editor. *Pediatric Neurosurgery: Surgery of the Developing Nervous System*. 3rd ed. Philadelphia: Saunders; 1994. p. 374-82.
- Alvisi C, Cerisoli M, Maccheroni ME. Long-term results of surgically treated brainstem gliomas. *Acta Neurochir (Wien)* 1985;76:12-7.
- Bricolo A, Turazzi S, Cristofori L, Talacchi A. Direct surgery for brainstem tumours. *Acta Neurochir Suppl (Wien)* 1991;53:148-58.
- Guillermo JS, Monjour A, Taillandier L, Devaux B, Varlet P, Haie-Meder C, *et al.* Brainstem gliomas in adults: Prognostic factors and classification. *Brain* 2001;124:2528-39.
- Smith RR, Zimmerman RA, Packer RJ, Hackney DB, Bilaniuk LT, Sutton LN, *et al.* Pediatric brainstem glioma. Post-radiation clinical and MR follow-up. *Neuroradiology* 1990;32:265-71.
- Fulton DS, Levin VA, Wara WM, Edwards MS, Wilson CB. Chemotherapy of pediatric brain-stem tumors. *J Neurosurg* 1981;54:721-5.
- Robertson PL, Allen JC, Abbott IR, Miller DC, Fidel J, Epstein FJ. Cervicomedullary tumors in children: A distinct subset of brainstem gliomas. *Neurology* 1994;44:1798-803.
- Robertson PL, Muraszko KM, Brunberg JA, Axtell RA, Dauser RC, Turrisi AT. Pediatric midbrain tumors: A benign subgroup of brainstem gliomas. *Pediatr Neurosurg* 1995;22:65-73.
- Rath GP, Bithal PK, Guleria R, Chaturvedi A, Kale SS, Gupta V, *et al.* A comparative study between preoperative and postoperative pulmonary functions and diaphragmatic movements in congenital craniovertebral junction anomalies. *J Neurosurg Anesthesiol* 2006;18:256-61.
- Albright AL, Price RA, Guthkelch AN. Brain stem gliomas of children. A clinicopathological study. *Cancer* 1983;52:2313-9.

17. Kumar R, Kalra SK, Mahapatra AK. A clinical scoring system for neurological assessment of high cervical myelopathy: Measurements in pediatric patients with congenital atlantoaxial dislocations. *Neurosurgery* 2007;61:987-93.
18. Tumiatì B, Casoli P. Syringomyelia in a patient with rheumatoid subluxation of the cervical spine. *J Rheumatol* 1991;18:1403-5.
19. Cohen AR, Wisoff JH, Allen JC, Epstein F. Malignant astrocytomas of the spinal cord. *J Neurosurg* 1989;70:50-4.
20. Endo T, Sato K, Takahashi T, Kato M. Acute hypotension and bradycardia by medulla oblongata compression in spinal surgery. *J Neurosurg Anesthesiol* 2001;13:310-3.
21. Lawrence AJ, Jarrott B. Neurochemical modulation of cardiovascular control in the nucleus tractus solitarius. *Prog Neurobiol* 1996;48:21-53.
22. Andresen MC, Kunze DL. Nucleus tractus solitarius – Gateway to neural circulatory control. *Annu Rev Physiol* 1994;56:93-116.
23. Horiuchi J, Potts PD, Polson JW, Dampney RA. Distribution of neurons projecting to the rostral ventrolateral medullary pressor region that are activated by sustained hypotension. *Neuroscience* 1999;89:1319-29.
24. Deletis V, Sala F, Morota N. Intraoperative neurophysiological monitoring and mapping during brain stem surgery: A modern approach. *Oper Tech Neurosurg* 2000;3:109-13.
25. Bowers DC, Krause TP, Aronson LJ, Barzi A, Burger PC, Carson BS, *et al.* Second surgery for recurrent pilocytic astrocytoma in children. *Pediatr Neurosurg* 2001;34:229-34.
26. Allen J. Brain stem glioma. *Neurol Neurosurg Updates Series* 1983;4:2-7.
27. Ammirati M, Bernardo A, Musumeci A, Bricolo A. Comparison of different infratentorial-supracerebellar approaches to the posterior and middle incisural space: A cadaveric study. *J Neurosurg* 2002;97:922-8.
28. Behnke J, Christen HJ, Brück W, Markakis E. Intra-axial endophytic tumors in the pons and/or medulla oblongata. I. Symptoms, neuroradiological findings and histopathology in 30 children. *Childs Nerv Syst* 1997;13:122-34.
29. Mehta VS, Chandra PS, Singh PK, Garg A, Rath GK. Surgical considerations for ‘intrinsic’ brainstem gliomas: Proposal of a modification in classification. *Neurol India* 2009;57:274-81.

How to cite this article: Nair AP, Mehrotra A, Das KK, Srivastava AK, Sahu RN, Kumar R. Clinico-radiological profile and nuances in the management of cervicomedullary junction intramedullary tumors. *Asian J Neurosurg* 2014;9:21-8.

Source of Support: Nil, **Conflict of Interest:** None declared.

Announcement



Download
iPhone, iPad
application

FREE

A free application to browse and search the journal’s content is now available for iPhone/iPad. The application provides “Table of Contents” of the latest issues, which are stored on the device for future offline browsing. Internet connection is required to access the back issues and search facility. The application is Compatible with iPhone, iPod touch, and iPad and Requires iOS 3.1 or later. The application can be downloaded from <http://itunes.apple.com/us/app/medknow-journals/id458064375?ls=1&mt=8>. For suggestions and comments do write back to us.