

Intradural Extramedullary Spinal Cord Tumors: Surgical Outcome in a Newly Developed Tertiary Care Hospital

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Abstract

Background: Intradural extramedullary tumors constitute nearly 65% of all spinal tumors. Over the years there has been dramatic improvement in diagnostic and therapeutic modalities especially the improvised surgical techniques.

Materials and Methods: Outcome of 23 patients was analyzed with respect to symptom improvement following surgery, at 1 month and later at 6 months.

Results: Overall, nearly 75% patients showed excellent results as per Frankel grading system, while as 8% and 17% had shown good and fair response, respectively.

Keywords: Intradural extramedullary, Spinal tumors, Tertiary hospital

INTRODUCTION

Primary spinal cord tumors constitute 2–4% of all central nervous system neoplasms. They are characterized based on their location as extradural, Intradural extramedullary (IDEM), and intramedullary tumors.^[1] IDEM tumors are rare central nervous system tumors that are found only in 0.3 out of 100,000 patients each year.^[2,3] [Figure 1].

The most common IDEM tumors are derived from sheath cells covering the spinal nerve roots (schwannoma and neurofibroma) or meningeal cells located along the spinal cord surface (meningiomas). Myxopapillary ependymomas are extramedullary tumors arising from the conus medullaris and filum terminalis. Less common

IDEM lesions include hemangiopericytomas, lipomas, paragangliomas, epidermoid cysts, arachnoid cysts, and dermoid cysts.^[4]

Patients with IDEM tumors often present with symptoms of spinal cord compression.^[5] Local or radicular pain is the most common presenting symptom and had the highest incidence in tumors located in the region of lumbar spine.^[3] Pain is followed by motor deficits, sensory loss and last but not the least is the sphincter dysfunction.^[6]

Magnetic resonance imaging (MRI) is the investigation of choice for diagnosis of IDEM tumors. It is not only preferred method for detail radiological assessment of tumors but also can even suggest histological subtype.^[7] Magnetic resonance angiography or spinal angiogram can be beneficial if the tumor has a vascular component.^[4]

Surgical excision is the best treatment modality in almost all patients with symptomatic IDEM lesions. Gamma knife is reserved for cases in which surgery could not be done due to various risks.^[8] The results of surgical excision of IDEM tumors have improved in past few decades

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due to development of more precise diagnostic tools such as computed tomography, MRI for understanding the anatomical structures, and with the advancement of surgical instruments and techniques especially with the use of high-resolution intraoperative microscopes.^[9]

The aim of this study was to impress on the excellent surgical outcome of IDEM tumors when performed under high-resolution microscope by young neurosurgeons in their initial learning curve of their surgical experience.

MATERIALS AND METHODS

This retrospective study was conducted in the Department of Neurosurgery of a newly started tertiary care Super Speciality Hospital of Government Medical College, Jammu, Jammu and Kashmir, India. All the patients were admitted to neurosurgery department of this hospital. Patients were evaluated for clinical symptoms and duration of symptoms, neurological examination and radiological evaluation by contrast MRI of the spine was done. All the patients operated with the diagnosis of IDEM lesions were included in the study.

A total of 23 patients included in this study were operated in 2 years with effect from July 1 2014 to June 30 2016. All the patients were operated by performing laminectomy with excision of tumor through posterior approach alone irrespective of location and position of tumor. After a midline incision and performing laminectomy, longitudinal incision was made in the dura, tumor detached and removed.

The outcome of all the operated patients were compared, based on the preoperative “Frankel grade” [Table 2] and sphincter disturbance with that of post-operative Frankel

grade and sphincter control at 1 month and then at mean follow-up period of 6 months.

RESULTS

We in our department have operated 23 patients with pre-operative diagnosis of IDEM spinal cord tumors in a period of 2 years. Out of 23 patients, 13 (56.52%) were male, and 10 (43.48%) were female. All of them were clinically evaluated preoperatively and assessed based on Frankel grading system and for Sphincter control. All the patients were operated by performing a midline skin incision, laminectomy with excision of tumor through posterior approach alone irrespective of location and position of tumor. The post-operative period was uneventful in all the cases. None of the patient required posterior stabilization.

The most common site of lesions on MRI in our study was dorsal in 11 (47.86%) followed by cervical in 4 (17.39%), dorsolumbar in 4 (17.39%), lumbar in 3 (13.04%), and Cervicodorsal junction in 1 (4.34%) patient [Table 1]. Pain, local or radicular was seen in almost all the patients admitted with a diagnosis of IDEM tumors. Myelopathic features were seen in 11 (47.82%), and Sphincter control (bowel/bladder) was lost in 11 (47.82%) patients especially in conus medullaris and cauda equine tumors.

Histopathology reports have confirmed the diagnosis of schwannoma in 10 (43.48%), meningioma in 4 (17.39%), neurofibroma in 3 (13.04%), ependymal cyst in 2 (8.69%), lipoma in 2 (8.69%), Arachnoid cyst in 1 (4.34%), and ganglioneuroma in 1 (4.34%) patients.

All the patients were put on IV antibiotics for initial 5 days and were discharged after removal of stitches on 7–8th post-operative days. Local wound infection and cerebrospinal fluid leak was seen in 2 patients, and both were managed conservatively. We followed all the patients for initially at 1 month and later at 6 months for post-operative recovery of motor/sensory deficits based on Frankel grading and for the sphincter control.

We have observed that 17 (74%) out of 23 operated patients had shown overall excellent outcome, 2 (8%) patient had shown good, and 4 (18%) patients had shown fair outcome [Figure 2].

Excellent result means Frankel E with normal sphincter control, good outcome we have taken as partial recovery and Fair grade as clinical insignificant recovery. 9 out of 10 patients with the diagnosis of schwannoma showed excellent results, and all the 4 patients with the diagnosis of meningioma, clinically improved on 6 months follow-up.



Figure 1: Magnetic resonance images of intradural extramedullary at cervical region

DISCUSSION

Primary spinal cord tumors constitute 2–4% of all central nervous tumor neoplasm and they are characterized based on their location as extradural, IDEM, and intramedullary.^[1] Patients with spinal cord tumors can present with radiculopathy, myelopathy, neck pain, back pain and maybe the sphincter involvement.^[6]

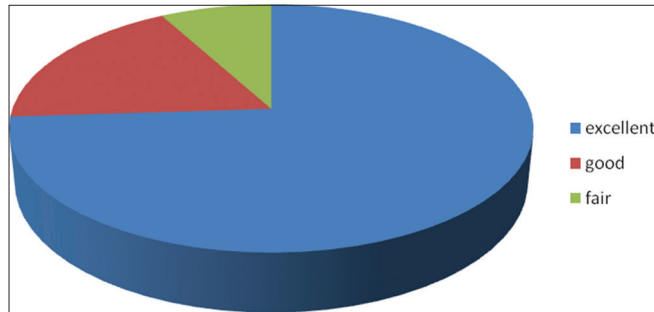


Figure 2: Pie chart depicting response rates

We are presenting 23 cases of IDEM spinal cord tumors which were operated in our department over a period of 2 years with effect from July 1 2014 to June 30 2016. These patients were followed up initially on monthly follow-up and later at 6 months to see the sensory and motor recovery and of course the sphincter recovery in those patients who presented with sphincter involvement initially.

IDEM tumors commonly seen are schwannomas, neurofibromas, and meningiomas. Less common tumors include metastatic deposit, paragangliomas, lipomas, nerve sheath myxomas, and vascular tumors.^[3]

In our study out of 23 operated patients with pre-operative diagnosis of IDEM tumors, 10 were Schwannomas, 4 were Meningiomas, 3 were Neurofibromas, 2 were Myxopapillary Ependymoma, 2 were Lipomas, and 1 Arachnoid cyst [Table 1], as confirmed on histopathology report.

Table 1: Patient characteristics and treatment response

Age/sex	Chief complaint	Pre-operative diagnosis	Pre-operative Frankel Level	Level	Pre-operative sphincter control	Procedure done	Post-operative Frankel	Post-operative Sphincter control	Result
42/F	MW, RS	Meningioma	C	D6	Present	LE	E	Present	Excellent
50/F	MW, RS, SD	Meningioma	B	D8-D9	Absent	LE	E	Present	Excellent
65/M	RS	Schwannoma	D	C2	Present	LE	E	Present	Excellent
32/F	MW, RS, SD	Ependymal cyst	B	D12-L3	Absent	LE	D	Absent	Good
26/M	RS, SD	Schwannoma	D	L2	Absent	LE	E	Present	Excellent
45/F	MW	Schwannoma	C	D6	Present	LE	E	Present	Excellent
22/M	MW, SD	Arachnoid cyst	B	D8	Absent	LE	E	Present	Excellent
33/F	RS	Schwannoma	D	D7	Present	LE	E	Present	Excellent
52/F	MW, RS, SD	Ependymal cyst	B	D12-L2	Absent	LE	C	Absent	Fair
50/M	RS, SD	Schwannoma	D	D7-D9	Absent	LE	E	Present	Excellent
55/M	RS	Schwannoma	D	L5-S1	Present	LE	E	Present	Excellent
54/M	MW, RS, SD	Ganglioneuroma	B	D12-L1	Absent	LE+Fixation	B	Absent	Fair
22/M	RS	Schwannoma	D	C2-C3	Present	LE	E	Present	Excellent
65/M	MW, RS, SD	Schwannoma	C	D4-D5	Present	LE	E	Present	Excellent
45/M	MW, RS	Meningioma	C	C5-C6	Present	LE	E	Present	Excellent
60/M	RS, SD	Lipoma	D	L3-L5	Absent	LE	E	Present	Excellent
36/F	MW, RS, SD	Schwannoma	C	D9-D10	Absent	LE	D	Absent	Fair
23/F	RS, SD	Lipoma	D	D12-L5	Absent	LE	D	Absent	Fair
50/F	RS	Meningioma	D	C7-D1	Present	LE	E	Present	Excellent
43/M	RS	Neurofibroma	D	C5-C6	Present	LE	E	Present	Excellent
49/F	RS	Neurofibroma	E	D7	Present	LE	E	Present	Excellent
51/M	MW, RS, SD	Neurofibroma	C	D9	Absent	LE	D	Absent	Good
40/M	RS	Schwannoma	E	D3-D4	Present	LE	E	Present	Excellent

MW: Motor weakness, RS: Radicular symptoms, SD: Sphincter disturbance

Table 2: Frankel grading system*

Grade A	Complete neurological injury	No motor or sensory function detected below level of lesion
Grade B	Preserved sensation only	No motor function below level of lesion, some sensory function below level of lesion detected
Grade C	Preserved motor, non-functional	Some voluntary motor function preserved below level of lesion but too weak to serve any useful purpose, sensation may or may not be preserved
Grade D	Preserved motor, functional	Functionally useful voluntary motor function below level of injury is preserved
Grade E	Normal motor function	Normal motor and sensory function below level of lesion, abnormal reflexes may persist

*van Middendorp JJ, Goss B, Urquhart S, *et al.* Diagnosis and prognosis of traumatic spinal cord injury. *Global Spine J* 2011;1:1-8

Schwannomas

Schwannomas are nerve sheath tumors that arise from dorsal nerve root. They are considered as benign, but the malignant schwannomas do exist. These neoplasm histologically arise from myelin-producing Schwann cells and have two types of cells called Antoni A and Antoni B which contribute to their imaging characteristics.^[10]

Patients with schwannoma present in 4–6th decade of life. High risk of malignant schwannomas is seen in patients with a diagnosis of neurofibromatosis Type II that too in young age. Patients with schwannomas are usually asymptomatic as the lesions are found incidentally on MRI. Many patients present with mild sensory symptoms such as shooting pain and paresthesias, while few may present with motor deficits.^[5,6]

MRI is the imaging method of choice to evaluate the lesion. On MRI, schwannoma appear as solid tumors in the dorsal sensory root region, which may displace spinal cord, conus medullaris, or filum terminale.^[11] Schwannomas appear isointense on T1W images as hyperintense on T2W images.

In our study out of 23 operated patients, 10 were histologically diagnosed as schwannomas. Out of these 10 patients, 6 were male, and 4 were female. As per location, 2 patients had cervical, 6 patients had dorsal, and 2 patients had lesion in lumbar spine. Common clinical presentation in our study was pain with radicular symptoms (RS). All the patients underwent midline skin incision, laminectomy with total excision. Our results were consistent with most of the studies in the literature,^[7-10] as in our series of 9 out of 10 operated patients they showed excellent outcome after a mean 6 months of follow-up. One young female patient who had a lesion at D9-D10 showed fair outcome as there was no significant improvement in terms of motor and bladder recovery.

Neurofibroma

Neurofibromas are benign tumors that arise from peripheral sensory nerves. There are two types of neurofibromas one is called solitary and other one as plexiform. In contrast to schwannomas, neurofibromas encase nerve roots rather than displacing them. Pain and paresthesias are the most common presenting symptoms. Type I neurofibromatosis (NF 1) may have multiple neurofibromas that may increase in number and size with increasing age of patient. On MRI imaging, neurofibromas appear as round or fusiform tumors that are isointense on T1W images and hyperintense on T2W/FLAIR images. The neurofibromas intensely enhance on contrast.^[7] Total surgical resection is primary treatment. To obtain this, the ventral and the dorsal roots are commonly sacrificed; however, the resection of nerve root is usually not associated with pronounced post-operative

motor or sensory deficits.^[2] Tumor recurrence is <5% and might be associated with subtotal tumor resection.^[8] We operated total of 3 patients with histologically proven neurofibromas. One patient was male and two were female. One patient had tumor in cervical region and 2 in dorsal. Out of 3 operated patients, 2 showed excellent outcome whereas one had shown good outcome on mean follow-up.

Meningiomas

Spinal meningiomas constitute up to 46% of spinal neoplasm and are common IDEM lesions. Thoracic is the most common site (80%).^[12] Out of four cases we operated for meningiomas, 3 were dorsal, and one cervical, the case with cervical meningiomas was having NF-2. Majority of meningiomas are detected between fifth and seventh decade of life.^[5] Meningioma arise from arachnoid cap cells embedded in dura near the nerve root sleeve, reflecting their predominant lateral location and meningeal attachment. Meningiomas may arise occasionally from dura or pia.^[5]

We have operated 4 patients, out of which one was male (25%) and three female (75%). In the literature available, meningiomas are common in female patients,^[12] even in our study 3 out of 4 were female. Posterior laminectomy provides adequate exposure for spinal meningiomas in most of the cases. Unilateral laminectomy with facetectomy can be used for ventral tumors. Costotransversectomy or lateral extra cavity approach may be utilized for ventral thoracic tumors. We have excised all the four meningiomas by posterior midline laminectomy with complete excision of tumors in all the 4 cases. All patients showed excellent (Frankel E) outcome.

Ependymal Cyst

Filum ependymomas have been classified as intramedullary lesions by virtue of the neuroectodermal derivative of filum, but as per anatomical and surgical prospective it is grouped under IDEM tumors.^[13] Myxopapillary ependymomas account for roughly 40–50% of spinal ependymomas.^[13] Myxopapillary ependymomas arise in the filum terminalis and account for more than 80% of ependymomas found in the cauda equina.^[5] They are benign well-circumscribed tumors. On MRI they appear as well-circumscribed hypointense lesion on T1, hyperintense on T2 and homogenous enhancement on gadolinium contrast. Ependymal rosettes and perivascular pseudorosettes with characteristic deposition of myxoid material around blood vessels are characteristic of histopathology in ependymomas.

Total excision of ependymal cyst is feasible if the nerve roots in the cauda equine are not entrapped within the tumour.^[14] We operated two patients with the diagnosis of

ependymal cyst, and both of these patients were having lesions at conus medullaris. Sphincter involvement was seen in both of these female patients. Even after 6 months of regular follow-up in both the patients, there was only some improvement of motor power in one patient but no improvement in sphincter control in both the patients

Lipoma of Cord

Lipoma of spinal cord is commonly seen by pediatric age group. They are rare tumors of spinal cord and cause symptoms secondary to compression (pain) or due to compressive myelopathy (motor deficits). Intramedullary lipomas involve the cervical and dorsal cord. IDEM lipomas are usually located in the lower thoracic and lumbosacral levels.^[15] Conus medullaris is the common site of lipomas and may present as lipomyelomeningocele. It is the most common form of the fatty masses in the spine and can be divided into dorsal, caudal, and transitional forms. These lesions are manifestation of occult spinal dysraphism and a common cause of tethered cord syndrome (TCS).^[16] Timely surgical intervention prevents significant neurological deficits. We operated one male and one female patient with a diagnosis of lipoma of cord. None of these patients have associated TCS or spinal bifida. One has lesion from L3 to L5 levels and the other one from D12 to L5 spinal levels. Both of patients were having Frankel D before surgery and on follow-up only one patient showed a bit neurological improvement.

Paranglioma

Parangliomas are derived from autonomic-nervous-system paraganglion cells and less common in CNS. Spinal paragangliomas are generally non secreting sympathetic neoplasms which tend to occur in fourth to fifth decade of life and show a male predominance. Intradural paragangliomas are most commonly located in cauda equine and lumbar region.^[17-19] On MRI paragangliomas are characteristically hypervascular, and after contrast, these tumors show typical salt and pepper pattern.^[20]

We have operated only one patient with dorsolumbar paraganglioma which was confirmed on histopathology. 54-year-old male patient presented with motor weakness, left-sided radicular pain and bladder involvement. The lesion was located at D12-L1 spinal level. The lesion was eroding the D12 pedicle on the left side, so excision of tumor with fixation D11-L1 spine was done. Patient has not shown any clinical improvement in the motor deficit and sphincter control even after 6 month follow-up.

Arachnoid Cyst

Intradural spinal arachnoid cysts are the rare causes of spinal cord compression. Basically congenital collections of CSF contained within the arachnoid membrane and subarachnoid space. As the cyst expands, it causes progressive

compression of spinal cord. Usual age of presentation is adolescents and young adults more commonly seen in males. Commonly seen in mid and lower thoracic spine. Patients present with spastic quadriplegia or paraparesis. Other symptoms such as backache, radiculopathy, sensory impairment, and sphincter disturbance may occur. MRI is an investigation of choice. Laminectomy with excision of cyst is the treatment in small arachnoid cysts, and cystoperitoneal shunt placement is required in cysts involving multiple spinal segments. We operated only one young male patient with MRI documented arachnoid cyst at D8 level. Patients pre-operative Frankel has improved from B to E with excellent outcome. We did laminectomy and excision of arachnoid cyst.

CONCLUSION

1. Spinal cord tumors are uncommon cause of back pain, cervical pain, RS, myelopathy, and sphincter (bowel/bladder) dysfunction.
2. 60% of spinal cord tumors are extradural and 30% are intradural.
3. IDEM tumors constitute 65 % of all primary intra spinal tumors.
4. Pre-operative diagnosis on MRI, detailed clinical assessment and accurate surgical plan is always associated with excellent outcome.
5. All the patients with diagnosis of IDEM tumors can be operated by performing laminectomy with excision of tumor through posterior approach alone irrespective of location and position of tumor.
6. Use of intraoperative microscope is always associated with excellent outcome.
7. Hence, patients with IDEM tumors can safely be operated even in rural areas and newly developed hospitals like ours, provided you have a good operating microscope.

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