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Original Research Article

Institutional experience of surgical management of intramedullary spinal cord tumours

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ABSTRACT

Background: Intramedullary spinal cord tumors are rare entity accounting for only 5-6% of all central nervous system tumors. Much literature is not available regarding these tumors owing to their rarity. Present study includes 24 cases of intramedullary spinal cord tumors with pain and progressive weakness. Radiological evaluation showed various intramedullary lesions like ependymoma, astrocytoma, epidermoid etc. spreading over various levels of spinal cord like cervicomedullary, cervicodorsal and dorsal cords.
Methods: The study included all the cases admitted with intramedullary spinal cord tumors in neurosurgery ward in King George hospital, Visakhapatnam, Andhra Pradesh, India during a period of three years from 2014 to 2016. Clinical profile of the patients was analyzed for the clinical presentation, age and sex distribution, histopathological study, pre-and postoperative neurological status, complications and functional outcome. All the patients are followed for a period of 6months to 3years.
Results: Of the 24 cases, most common age group was second (7 patients) and third (7 patients) decades which is upto 29.16% each. Males (16 patients) are more affected than females in 66.66%. Ependymoma is the most common tumor seen in 41.66% (10 patients) followed by astrocytoma in 33.33% (8 patients). The surgical technique, extent of resection, pre and postoperative neurological status and functional outcome are discussed.
Conclusions: Intramedullary tumours occur commonly among males in the 2nd to 4th decades. Among the intramedullary tumours ependymoma from the commonest lesion subtypes. Most common location is cervical cord segment. Ependymomas have a good plane of cleavage and are thus amenable to radical excision. Patients with good Mc Cormicks grade in pre-operative stage are more amenable for total or near total excision. Prognostic factors affecting outcome are the preoperative neurological status, the plane of cleavage, the extent of resection, the nature and subtype of the lesion. With the improvement in microsurgical techniques and novel adjunctive like MRI, CUSA and intraoperative neurophysiological monitoring, surgery for intramedullary lesions can be carried out with acceptable morbidity and mortality.

Keywords: Astrocytoma, CUSA, Ependymoma, Intramedullary spinal cord tumours, Microsurgical techniques, Midline myelotomy

INTRODUCTION

Intramedullary spinal cord tumours are rare. Intramedullary spinal cord tumors (IMSCTs) comprise 5% to 6% of all central nervous system (CNS) tumors and one third of primary spinal tumors.[1] The most frequent of these tumors are of glial origin, astrocytomas and ependymomas comprising the majority of them. Among intradural spinal tumors, intramedullary neoplasms account for only 8% to 10%.[2] The most common intramedullary lesions are spinal ependymoma (60%) and spinal astrocytoma (10%-20%), but others include hemangioblastoma (3%-8%), cavernous malformation, metastases, and lipoma.[2] Spinal

astrocytoma seems to occur in childhood or young adulthood, whereas ependymoma can occur throughout life but most commonly in the early to middle adult years. Much less attention seem to be paid to intramedullary spinal cord tumors because of their overall rarity. The long-term results depend on their varying natural histories and the surgical approach. Historically, treatment of these tumors consisted of biopsy, bone decompression, and/or radiation therapy. This approach resulted in only moderate improvements in slowing disease progression. Modern advances in microsurgical techniques have allowed for increasingly aggressive resection of these tumors, often permitting complete resection and resulting in increased long-term survival and improved quality of life.[3] Early attempts at removal of intramedullary spinal cord tumours were associated with prohibitive operative morbidity and mortality.[4] Surgery was primarily employed for diagnosis (frequently by visual inspection alone), cyst aspiration and duroplasty.[4] For the next several decades, improved microsurgical techniques with total resection and radiation therapy evolved as the major treatment modality for these tumours.

Charles Elsberg in 1911 was the first person to perform surgery for intramedullary astrocytoma in two stages i.e. myelotomy followed a week later by removal of extruded tumor. But, the first successful complete removal of intramedullary tumor was done by Greenwood in 1967. First largest surgical series on intramedullary tumors with a clinical grading system was developed by Mc Cormick in 1990.

In 1954, Greenwoods utilization of bipolar cautery and Ioupe magnification enabled complete removal of 9 intramedullary ependymomas.[5] Since then, advances in microsurgery, pharmacology, and neurosurgical anesthesia have remarkably improved the prognosis, especially the functional outcome of surgery.[6] The two most innovative surgical techniques developed were the operating microscope and the Cavitron ultrasonic surgical aspirator (CUSA). With better imaging with MRI and further refinement of microsurgical techniques, and the continued use of surgical adjuvants like CUSA, intra operative ultrasound, electrophysiological monitoring and radical resection of well-defined spinal neoplasms like ependymomas, hemangioblastomas, epidermoids and dermoid cyst have become possible with less complications.[4]

The present study was undertaken to retrospectively analyze our institutional experience in the surgical management of intramedullary spinal cord tumours. It was our aim to study the clinical and radiological features, the histological subtypes, operative findings including resectability, postoperative complications and procedure or disease related morbidity and mortality. The functional outcome of these patients was determined in relation to the tumour subtypes and the extent of resection.

Aim of the study was to study the clinical profile, outcome of surgical intervention and the histological diagnoses of all intramedullary lesions operated in the study period, to evaluate and quantify the preoperative, post-operative status of the patients in terms of a functional scale and also during the follow up period and to assess the surgical outcome in relation to type, location of lesion and extent of resection.

METHODS

Present study consists of 24 cases of IMSCTs which attended neurosurgery OPD with various clinical manifestations ranging from mild pain without any neurological deficits to progressive neurological deficits with or without involvement of bowel and bladder who were managed surgically. Diagnosis, clinical profile, surgical treatment, histopathology, results and follow up were discussed.

Both retrospective and prospective study carried out in Department of Neurosurgery, King George Hospital, Visakhapatnam, Andhra Pradesh, India. Case files of patients who were registered, diagnosed and treated for IMSCTs from 2014-2016 were retrieved. The clinical symptoms, neurological status and radiological data in surgical group were noted. They were compared with postoperative period and in follow up period.

The work up patients included routine investigations like complete blood picture, ESR, chest X ray. Metastatic workup done in doubtful cases. Contrast MRI of spine is the main imaging modality in the present study.

MRI of patients showed various intramedullary lesions like ependymoma, astrocytoma, ganglioglioma, epidermiod etc. spreading over varying spinal cord segments. The above study was analyzed.

RESULTS

Out of the 24 patients who were included in the present study most of the patients belonged to 15-50 years who were manual labourers by occupation. Most of the patients belong to second and third decades (Table 1).

Table 1: Age distribution of IMSCTs.

Age	No. of patients	Percentage
0-10	1	4.16%
11-20	7	29.16%
21-30	7	29.16%
31-40	4	16.66%
41-50	4	16.66%
51-60	1	4.16%
Total	24	

Total duration of symptoms ranged from 6 months to 2 years. Patients age group ranged from 4 to 60 years. Out

of the 24 patients who were included in the study 16 were males (66.66%) and females were 8 (33.33%) (Table 2).

Table 2: Sex distribution of IMSCTs.

Sex	No. of patients	Percentage
Male	16	66.66%
Female	8	33.33%
Total	24	

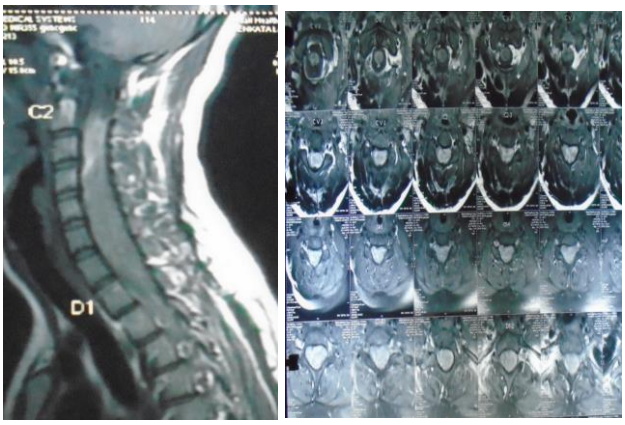

Figure 1: Case 1- MRI of a case of astrocytoma.

Table 3: Clinical presentation of IMSCTs.

Presentation	No. of patients	%
Pain	14	58.33
Weakness	20	83.33
Parasthesias	12	50
Dissociative sensory loss	4	16.66
Gait disturbances	2	8.33
Bladder involvement	1	4.16

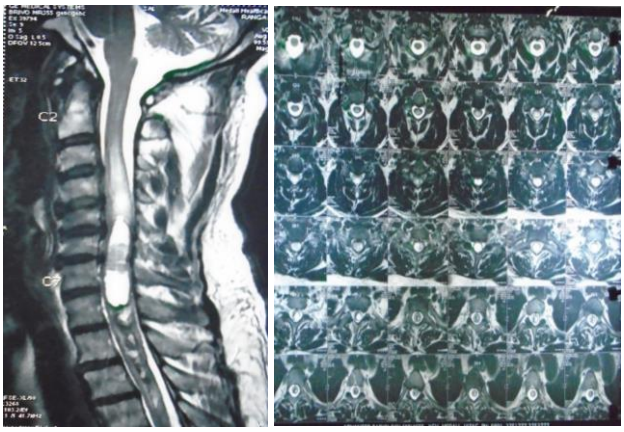

Figure 2: Case 2- MRI of a case of ependymoma.

Clinical manifestations include (Table 3). Radiological findings depicted: case1 (Figure 1), case 2 (Figure 2). Number of patients according to Mc Cormick's grading (Table 4). The site of the lesion on imaging is given in Table 5.

Table 4: Preoperative McCormic's grading of IMSCTs.

McCormic grade	No. of patients	%
Grade 1	6	25
Grade 2	12	50
Grade 3	3	12.5
Grade 4	2	8.3
Grade 5	1	4.1
Total	24	

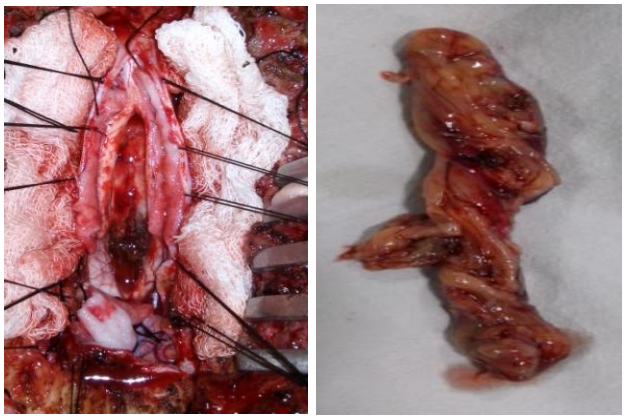

Figure 3: Operative specimen- case 1.

All the 24 patients were operated by laminectomy and excision of tumour, may it be complete or partial resection. Extent of resection is given in Table 6.

Table 5: Site of lesion of IMSCTs.

Site	No. of patients	%
Cervicomedullary	1	4.16
Cervical	10	41.66
Cervicodorsal	8	33.33
Dorsal	4	16.66
Dorsolumbar	1	4.16
Total	24	

Table 6: Extent of resection of IMSCTs.

Extent of resection	No. of patients	%
Complete	16	66.66
Partial	7	29.16
Biopsy	1	4.16

The goals of surgery for intramedullary tumour are gross total resection and preservation of neurologic function.[4,7-12] Gross resection is usually sufficient to achieve long-term tumor control or cure in these low-grade lesions.[7] Although the presence of a syrinx may improve the chances of a gross total resection by facilitating surgical manipulation of the tumor, it cannot be used as an independent predictor of outcome.[7,13,14] Gross total removal is the treatment of choice in these cases for optimum disease control.[4,7-12]

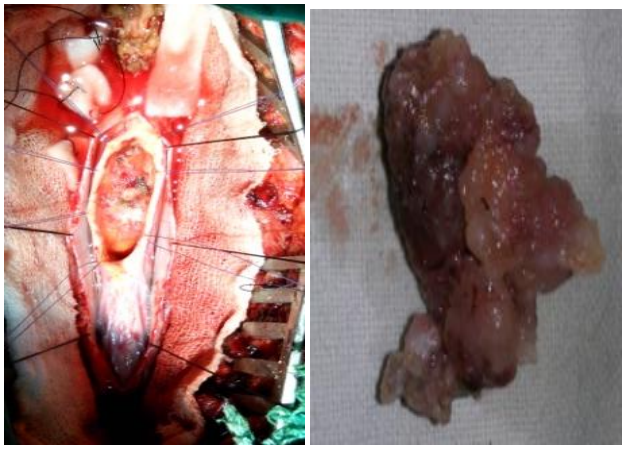

Figure 4: Operative specimen-case 2.

Table 7: Histopathology of IMSCTs.

Histopathology	No. of patients	Percent
Ependymoma	10	41.66
Astrocytoma	8	33.33
Ganglioglioma	2	8.33
Epidermoid	1	4.16
Mucinous cyst	1	4.16
Meningioma	1	4.16
Metastasis	1	4.16

Table 8: Post-operative outcome of IMSCTs in relation to extent of resection.

Extent of resection/ post op status	Improved	Static	Deteriorated
Complete	1	15	0
Partial	1	5	1
Biopsy	0	1	0
Total	2 (8.33%)	21 (87.50%)	1 (4.16%)

Table 9: Post-operative outcome of IMSCTs in relation to tumor type.

Tumour subtype/status	Improved	Static	Deteriorated
Astrocytoma	1	6	1
Ependymoma	1	9	0
Ganglioglioma	0	2	0
Epidermoid	0	1	0
Mucinous cyst	0	1	0
Meningioma	0	1	0
Metastasis	0	1	0
Total	2 (8.33%)	21 (87.50%)	1 (4.16%)

Average length of hospital stay was 8-21 days postoperatively. The intra operative finding along with the operative specimen was shown in Figure 3 and Figure 4.

Table 10: Complications.

Complication	No. of patients	%
Neurological deterioration	2	8.33
Parasthesias	2	8.33
Wound infection	1	4.16
Wound dehiscence	1	4.16
CSF leak	1	4.16
Meningitis	0	0
Death	1	4.16

Table 11: Comparative study of the present study with study carried out in AIIMS 2003.

Incidence of IMSCTs	AIIMS 2003	Present study
Ependymoma	33%	41.66%
Astrocytoma	28%	33.33%
Epidermoid	8%	4.16%

Histopathologic diagnosis is depicted in Table 7. The neurological outcome is discussed in Table 8 and Table 9 respectively. Complications are discussed in Table 10. Present study is compared with the study carried out in All India institute is presented in Table 11.

DISCUSSION

IMSCTs are rare entity with slight increased incidence in male patients in our study. Most common age incidence is second and third decade. Pain and weakness were the most common symptoms. 75% patients were in McCormick grade-1 and grade-2 pre-operatively. Ependymoma is the most common tumor followed by astrocytoma. Cervical cord is the most common site of occurrence of IMSCTs. Ependymomas with good plane of cleavage are amenable to complete resection. Complete resection of IMSCTs is associated with good neurological and functional outcome. Respiratory complications like aspiration pneumonia is the most common cause of death in these patients.

The advent of modern microsurgical techniques has led to a significant change in the approach to IMSCTs. The pioneering work of Epstein and colleagues.[15,16] and the legacy of detailed neurosurgical studies over the last few decades have dramatically improved the outcome in patients harboring these tumors. In cases of IMSCTs one should not wait for the onset of clinical deterioration but rather institute treatment as soon as possible. The earlier the diagnosis and the more radical the resection of an IMSCT, the greater the likelihood of preserving the patient's neurological function.[17] The present study was undertaken to analyze our institutional experience in the surgical management of intramedullary tumours. It was our aim to evaluate the clinical and radiological features,

the histological subtypes, operative findings including resectability, postoperative complications and procedure or disease related morbidity and mortality. The functional outcome of these patients was determined in relation to the tumour subtypes and the extent of resection and preoperative functional grade. The differences between various intramedullary tumors in terms of age incidence, clinical presentation, radiological characteristics, surgical aspects and outcome have all been described in the literature.[9,10,18] The present study also noted similar differences and was tabulated in the earlier section which confirm the fact that ependymomas are mostly low grade neoplasms, have a good tumor-cord interface, seen more commonly in the 3rd to 4th decades , are amenable to total excision in most cases and have a good functional outcome.[10,19] Epstein et al, in their series of 38 patients with ependymomas have noted the male preponderance and a mean age of occurrence of 37 years.[8] In contrast, astrocytomas are seen to be more heterogenous as far as the histological grade is concerned with low and high grade lesions. They tend to occur in a slightly younger population, many lesions have a poor plane of cleavage from the surrounding cord, less amenable to radical resection and have a less favorable outcome. Astrocytomas in the present study were mostly of low grade variants and the same has been validated in other series.[12,20] The present study records a total excision rate of 66.66% in 16 pts and partial excision rate of 29.16% in 7 pts and biopsy in 1 patients (4.16%). Outcome in the present study was correlated with various prognostic factors elucidated in the literature and includes preoperative functional grade, histological subtype, histological grade, and extent of resection. Present analysis of the functional grade of the patients preoperatively, at discharge and at follow up reveals that better the preoperative functional grade, better is the outcome.

There were 18 (75%) out of 24 patients in grade 1 and 2 preoperatively. This number rose to 21 (87.5%) at follow up. This indicates that the benefits of surgery for intramedullary tumors are more "prophylactic" as propounded by Cristante and yields better results in a patient with minimal neurological deficits.[4,10,11,14,15,18] An argument has been made for radical excision of intramedullary lesions by various authors.[10,18-20] Chandy et al advocated radical excision of all intramedullary lesions, provided there is a good plane of cleavage.[18] Cohen et al advise radical surgery even for a high grade lesion, citing that the sampling error of the tissue to determine true histology is better with larger volume of tissue. They also advocate radical resection for cytoreductive debulking and pain control.[20] Extent of resection has shown to affect outcome in ependymomas in other series as well.[4,8,11,18] S Nair et al in 1997 advocated although total resection of ependymomas have become a procedure with good functional results in most hands, a radical resection can be achieved with long term stabilization of neurological deficits in majority of astrocytomas.[23]

The present study substantiates this, based on our observation that patients who underwent either a total or near total resection improved or remained the same in the postoperative period. In present study, out of 24 patients on follow up, 16 patients underwent total excision, 7 patients underwent partial resection. 23 patients (95.83%) improved or were neurologically stable at follow up compared to their preoperative status.

Among the factors influencing outcome, apart from the extent of resection as previously mentioned, the histology was also found to be important, as seen by other studies.[10,11,14,18] In 1994 Cristante and Herrmann reported that the outcome was aggravated, unchanged, and improved in 31, 55, and 17% of 69 patients intramedullary tumors respectively.[11]

In present study, patients with a histological diagnosis of astrocytoma, 12.5% improved, 75% were neurologically stable while 12.5% showed worsening in their functional grade. 10% of patients with ependymomas showed improvement when compared to their pre-operative status while 90% remained static and none of the patients showed worsening in neurological status. Intramedullary lesions arising in the thoracic cord have been found to have a less favorable outcome by various authors.[11,15,21] Hoshimaru et al have postulated that the tenuous blood supply of the thoracic cord may explain its higher susceptibility to operative injury during tumor removal.[22] Another reason is that thoracic lesions cause minor and vague symptoms and their long standing nature causes cord atrophy and arachnoid scarring which that particular segment of the cord tolerates poorly.[21]

In the present study out of 24 patients there were 10 (41.66%) patients harboring tumors in the cervical cord and 4 (16.66%) patients in thoracic cord and 8 (33.33%) were in the cervicodorsal region. Out of the 24 patients during follow up, 1 (10%) patient with cervical cord and 1 (12.5%) patient with dorsal cord lesions showed improvement.

In present study 8.33% (2 patients) patients showed improvement in their functional grades when compared to pre-operative status; in 4.16% (1 pt) outcome was not better while 87.5% (21 pts) were unchanged.

There have been various advances in diagnosis, surgical adjuncts like electrophysiological monitoring and advanced microsurgical techniques which help us evaluate, plan and execute better treatment for patients with spinal cord intramedullary tumors. Recent advances in adjuvant therapies including Cyber knife stereotactic frameless radiosurgery, improved drug delivery systems and gene-based therapies have also revolutionized the approach to these difficult tumors and their long-term management. A healthy trend towards aggressive surgery followed by equally aggressive rehabilitation programs along with appropriate adjuvant therapy will probably aid

in preserving and improving the neurological status of patients with this functionally devastating disease.

CONCLUSION

Intramedullary tumours occur commonly among males in the 2nd to 4th decades. Among the intramedullary tumours ependymoma form the commonest lesion subtypes. Most common location is cervical cord segment. Ependymomas have a good plane of cleavage and are thus amenable to radical excision. Patients with good Mc Cormicks grade in pre-operative stage are more amenable for total or near total excision. Prognostic factors affecting outcome are the preoperative neurological status, the plane of cleavage, the extent of resection, the nature and subtype of the lesion. With the improvement in microsurgical techniques and novel adjunctives like MRI, CUSA and intraoperative neurophysiological monitoring, surgery for intramedullary lesions can be carried out with acceptable morbidity and mortality

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