Intradural extramedullary spinal cord tumors: a retrospective study at tertiary referral hospital

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Abstract: *Introduction*: Intradural extramedullary (IDEM) spinal cord tumours account two thirds of all intraspinal tumours. The objective of this study was to determine short- and long-term outcomes of surgical patients with IDEM spinal cord tumours, and to see clinical features that could be helpful in management of patients with these lesions (operated by single senior surgeononly). *Methods*: A retrospective review of 201 operative IDEM spinal cord tumours cases between 1993 and 2014 was performed. Outcomes were scored at one month and at mean follow-up of 8.5 months postoperatively. In addition, patient demographics, tumour types and locations were also collected. Statistical analysis was conducted utilizing Chi-square and Student's t-tests. *Results*: There were 93 men and 108 women (mean age 48 yrs, range 5 -87 yrs). Men presented at a younger age than women (42 vs 51 yrs, P<0.02). 165 (82.08 %) patients presented with severe radiculopathy and myelopathy. The 36 (17.91 %) had symptoms of radiculo -pathy. Mean duration of symptoms prior to diagnosis was 11 months. Schwannomas (113 patients) had the longest mean duration of symptoms (14.9 months), followed by meningiomas (68 patients, 8.4 months), and ependymomas (20 patients, 2 months). Hundred and eighty nine (94%) of patients demonstrated significant improvement at one-month and 186 (92%) at 6-month mean follow-up. Only 39/201 (19.4%) patients had residual focal deficits on long term follow-up. *Conclusions*: Surgery for IDEM should be expected to produce significant and dramatic improvement in most of patients. Demographic, tumor-specific and anatomic considerations will be clinically useful while managing IDEM.

Key words: Intradural extramedullary tumors (IDEM), Spinal cord, Surgical outcomes

Introduction

Spinal tumors account for only approximately 5-15% of the nervous system neoplasms (13, 18). Intradural extramedullary (IDEM) spinal cord tumours account for about 60% of the intraspinal tumors, (17) and include schwannomas (1, 6) (30%; incidence rate, 0.3-0.4 cases annually per 100,000 people), meningiomas (6, 22) (25%; incidence

rate, 0.32 cases annually per 100,000 people), neurofibromas, teratomas, lipomas, and metastatic tumors.

The primary aim of this study was to examine surgical outcomes of IDEM spinal cord tumours in a large retrospective cohort of patients operated by single surgeon. Secondary objectives includes examination of clinical data related to demographics, symptoms, tumor location and type that could be helpful in clinical decision making while managing these tumours.

Materials and methods

In this retrospective study of 201 surgical patients who underwent surgery for IDEM spinal cord tumours between January 1993 and November 2014 were included. Parameters recorded include patients demographics, symptoms (severity and duration), tumor characteristics (anatomic and pathologic), postoperative follow-up and surgical outcomes.

Surgical intervention was indicated by a combination of presenting symptoms (radiculopathy and/or myelopathy) and radiographic findings of MRI. The neuroimaging procedure of choice was contrast-enhanced MRI.

Surgical outcomes were scored at 1 month and then at the mean follow-up period. The mean follow-up was calculated from the interval between surgery and the last complete clinical examination in the patient chart (in this study, 8.5 months). Patient records were carefully reviewed and surgical outcomes were scored strictly according to the modified criteria of Odom, et al (Table 1) (16).

TABLE I

Excellent	Complete relief of pain and other symptoms, return to full activity.
Good	Partial relief of pain and other symptoms, return to full activity.
Fair	Improvement with persistent limitation of activities.
Poor	No improvement or further deterioration.

Descriptive statistics, Chi-square test and Student's t-test were utilized for data analysis. Statistical significance was set at alpha = 0.05.

Note: other symptoms - paresthesias, paresis, sensory loss

Results

In this study there were 93 men and 108 women (mean age 48 yrs, range 5 -87 yrs). Overall men presented at a younger age than women (42 vs 51 yrs, $P<0.02$). The age distribution was bimodal with a major peak around 41 yrs and a minor peak around 70 yrs. The second peak around age 70 represents predominantly because of older meningioma patients, whereas the peak around age 41 due to schwannoma and ependymoma patients.

There were three primary tumor types: schwannomas (113/201), meningiomas (68/201), and myxopapillary ependymomas (20/201) (Table 2A). Schwannomas were noted to be more common in men, meningiomas more common in women, and ependymomas distributed equally among men and women (Table 2A). Patients with meningiomas tends to be older (57 yrs) than those patients with schwannomas (44 yrs) and ependymomas (40yrs, $P<0.01$) (Table 2B).

Tumor locations varied between the three tumour subtypes in a predictable fashion (Table 2C) schwannomas were distributed

fairly evenly among the three anatomic regions (cervical, thoracic, and lumbosacral), meningiomas common in cervical and thoracic regions and ependymomas were commonly localized to the lumbar region. The ratio of Schwannomas to Meningiomas to Ependymomas was approximately 4:2:1 in this study (actual ratio 113:68:20).

The mean duration of symptoms before diagnosis was 11 months. Schwannomas had a statistically longer duration of symptoms (14.9 months) than meningiomas (8.4 months, $P<0.05$) for cervical and thoracic tumors. Ependymomas had shorter duration of symptoms (2.0 months) than schwannomas (10.8 months, $P<0.05$) for lumbosacral tumors.

Results of one month follow up

Excellent	Good	Fair	Poor
75 (37.3%)	114 (56.7%)	9 (4.5%)	3 (1.5%)

Results of Six months follow up

Excellent	Good	Fair	Poor
123 (61.2%)	66 (32.8%)	12 (6%)	

Three patients had multiple IDEM, all of which were schwannomas. Two of these patients were NF-II positive. Four patients had recurrence of their IDEM spinal cord tumours. Three of these were schwannomas and one was an ependymoma. Two patients with recurrent dumbbell-shaped schwannomas within the soft tissues of the neck underwent surgical treatment with acceptable results (one patient returned with new symptoms 5 years after initial tumor excision while another patient presented 15 years after the initial surgery). One patient with recurrent ependymoma at 4 years following initial surgery underwent a successful course of radiotherapy, and one patient with schwannoma underwent non-operative semi-annual observation for a localized radiographic recurrence.

While no metastases were noted during the 8.5 month mean follow-up period (range 1.5 to 30 months), this study did not specifically examine whether metastatic disease did appear at a later time. One mortality was noted in this series, in case of large cervical C1-C2 schwannoma due to respiratory complications, and the only complications included a superficial wound infection.

TABLE 2

Histologic, demographic, and anatomic considerations for IDEM spinal cord tumours

A. Distribution of tumours by histologic type and patient gender in this series

Patient gender	**Total**	**Men**	**Women**
Schwannoma	113 (56.21%)	69	44
Meningeoma	68 (33.83%)	14	54
Ependymoma	20 (9.9%)	10	10
Total	201	93	108

B. Tumour incidence by age group

Age	**<40**	**40-60**	**>60**
Schwannoma	47	47	19
Meningeoma	4	39	25
Ependymoma	10	4	6
Total	61	90	50

C. Tumour incidence by anatomic location

Location	**Cervical**	**Thoracic**	**Lumbar**
Schwannoma	31	38	44
Meningeoma	19	49	0
Ependymoma	0	0	20
Total	50	87	64

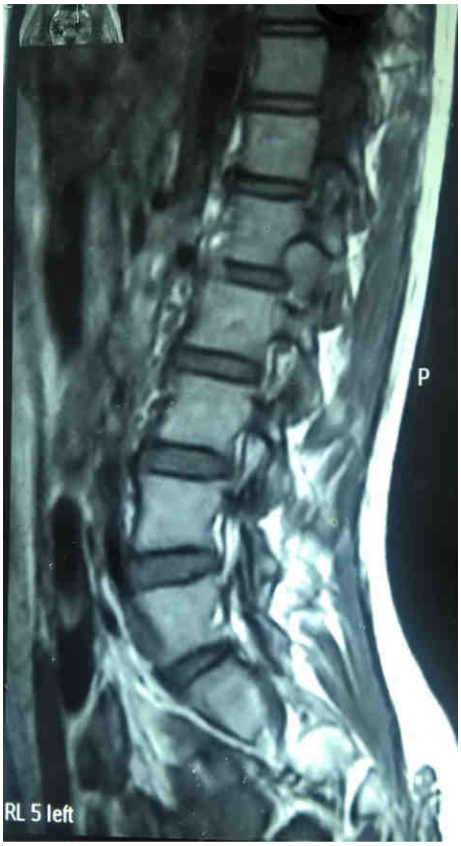

Figure 1 - T1W MRI sagittal view showing L1-2 neurofibroma

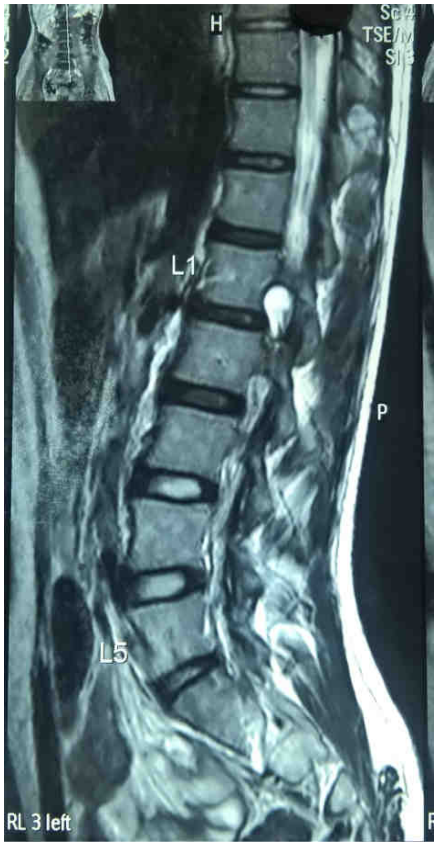

Figure 2 - T2W MRI sagittal view showing L1-2 neurofibroma

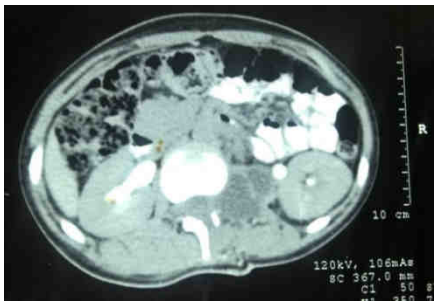

Figure 3 - CT abdomen showing left L1-2 neurofibroma

Discussion

Intradural extramedullary tumors account for two-thirds of primary spinal tumors (12). Most intradural extramedullary tumors are benign, and they exhibit no specific symptoms. Radicular pain and worsening sensory and motor loss are common manifestations. Therefore, most of the patients are wrongly diagnosed with cervical spondylopathy or intervertebral disk herniation. MRI is very crucial to confirm the diagnosis of intradural extramedullary tumors. Once the diagnosis is confirmed, the best treatment for nonmalignant intradural extramedullary tumor is surgery. The goal of surgery is complete surgical resection while preserving spinal stability, without worsening the preoperative neurological status.

All the processes of conventional surgical tumor resection are carried out without using microscope, which may lead to a greater likelihood of incomplete tumor resection, as well as more damage to the spinal cord and vessels surrounding the spinal canal. With the improve ement of medical devices, surgeons are increasingly using microscopy to perform surgical tumor resection. MIS can provide a clearer visual operative field and more delicate operative maneuvers, which can avoid the damage to the spinal cord and peripheral nerves as far as possible, reduce intraoperative blood loss and postoperative complications, and increase the rate of complete removal of tumors. It was reported that MIS allowed removal of the tumor with minimal impairment from cutting of nerve fibers at the nerve root (14). No Poor results were noted in this study at the mean 8.5 month follow-up. Our findings are similar with those of other authors, with majority of clinical improvement noted either immediately or within 6 months of the operative intervention, with less notable clinical change after this initial period (13, 19). Other studies reported that duration of preoperative symptoms appears to correlate with postoperative improvement, and that successful complete microsurgical tumor excision is of utmost importance (11, 13, 21). In terms of mortality, our result correlate well with other authors, with a reported range between 0-4.4% (2, 4, 13).

Approximately 20% of patients in our study had residual focal deficits, none of which were disabling. Other authors report similar outcome among patients with similarities to our patient sample, with significant improvement in 62-88% of cases and clinical worsening in only a minority of patients (1-5%) (13).

The demographics in our study are similar to those in previous studies (13, 19). We found that schwannomas affected younger male patients, meningiomas tended to occur in women and older patients. Patients with myxopapillary ependymomas were younger than patients with schwannomas.

The reported frequencies of schwannomas among IDEM spinal cord tumours vary from 43% to 67% in other studies as copared to 56.21% in our series (7, 10, 13, 19). Schwannomas tend to produce localized pain, radiculopathy, and cauda equina syndrome. Most report schwannomas to be solitary, with a 2.5% malignancy rate, which carries a poor prognosis (8). In our study, schwannomas

were the only group of tumors without a predominant location of occurrence, and constituted 75% of recurrent tumors.

In this study, patients with meningiomas were older than those with other tumor types. Findings in this series agree with the literature in that meningiomas are the second most common IDEM, with approximately 80% localized to the thoracic region (72.05% in this study) (8, 9, 11). Between 75% and 85% of patients with meningiomas are women (79.41% in this study), who tend to be older than patients with schwannomas or ependymomas (8, 11, 19). Consistent with previous reports, we found meningiomas to be more aggressive in younger patients, with the higher incidence of myelopathy likely due to predilection of meningiomas for the thoracic region (5). Others describe higher operative morbidity associated with IDEM spinal cord tumours located in the thoracic region 19. Meningiomas tend to produce the "dural tail" sign on MRI scans in sagittal, axial, and coronal planes, and it is recommended that all three planes of visualization be used 20. We follow this recommendation because as many as 10% of meningiomas can be both intradural and extradural (12).

Myxopapillary ependymomas constitute 9.9 % of IDEM spinal cord tumours in this study, and although in this study ependymomas had equal distribution among men and women, another series reported twice as many men as women (24). In this study, the mean age of the ependymoma group was lower than the mean age of patients with meningioma or schwannoma. Symptomatically, ependymomas tended to produce cauda equina syndrome, localized pain, and radiculopathy.

A 4:2:1 ratio of schwannomas to meningiomas to ependymomas was observed in this series. Although not specifically described by others, similar ratio of tumor types can be noted in previous reports (10, 13, 19).

Limitations of this study include its retrospective nature, lack of patient follow up data beyond the 8.5-month mean follow up period, lack of complete data on recurrences or metastases beyond the end of the study period, and the drawbacks of the Odom's modified criteria as a measurement tool for morbidity. The Odom's modified scale is a very 'rough' instrument of outcome assessment and does not have the capacity to truly delineate actual morbidity. The Odom's scale was chosen for this study because of standard and uniform reporting of this scale on our patient charts. Although the use of scales such as SF-36 or Nurick grading would greatly enhance this study, the long period of this study as well as the difficulty of assigning these scales in a retrospective fashion to every patient in this study, make their applicability impractical (3, 15).

Conclusions

Surgery for IDEM spinal cord tumours, with goal of complete tumor removal, is a safe and effective option. At the 8.5 month mean follow-up, majority of patients had complete or near complete relief of symptoms and return to full activity Schwannomas and ependymomas were the only histologic types to recur. Demographic, tumor-specific and anatomic considerations may be clinically useful when approaching IDEM spinal cord tumours.

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