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ORIGINAL ARTICLE

Spinal tumors: Trends from Northern India

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ABSTRACT

Introduction and Methods: We retrospectively analyzed 111 patients with spinal tumors operated over a period of 9 years to observe the relative frequency of different lesions, their clinical profile, functional outcome and prognostic factors. 30/111 (27%) were extradural, 40/111 (36.1%) were intradural extramedullary (IDEM) and 41/111 (36.9%) were intramedullary spinal cord tumors (IMSCTs). Mean age at surgery was 30.81 years (range 1–73 years). The average preoperative duration of symptoms was 16.17 months (15 days to 15 years). Major diagnoses were ependymomas and astrocytomas in IMSCT group, schwanommas and neurofibromas in IDEM group, and metastasis, lymphoma in extradural group. The common clinical features were motor weakness in 78/111 (70.27%), sensory loss in 55/111 (49.54%), pain 46/111 (41.44%), and sphincter involvement in 47/111 (42.43%) cases.

Results: Totally, 88/111 (79.27%) patients had improvement in their functional status, 17/111 (15.31%) remained same, and 6/111 (5.4%) were worse at time of their last follow-up. The mean follow-up was 15.64 months (1.5 m^{-10} years). Totally, 59 out of 79 patients, who were dependent initially, were ambulatory with or without the aid. Most common complication was persistent pain in 10/111 (9%) patients and nonimprovement of bladder/bowel symptoms in 7/111 (6.3%). One patient died 3 months after surgery.

Conclusions: (1) Congenital malformative tumors like epidermoids/dermoids (unrelated to spina bifida) occur more frequently, whereas the incidence of spinal meningioma is less in developing countries than western populations. (2) The incidence of intramedullary tumors approaches to that of IDEM tumors. Intramedullary tumors present at a younger age in developing countries. (3) Rare histological variants like primitive neuroectodermal tumors should also be considered for histological differential diagnosis of spinal tumors. (4) Preoperative neurologic status is the most important factor related to outcome in spinal tumors.

Key words: Microsurgical treatment, prognostic factors, spinal tumors

Introduction

There are few organs in the human body in which neoplastic disease occurs in a more benign form, and the results of surgery are more brilliant than in the spinal cord and its membranes. At the same time, there is no organ in which total restoration of function following the removal of the neoplasm is so completely dependent on an early diagnosis.[1]

Benign and malignant neoplasms can arise from intraspinal structures such as meninges, spinal cord, nerve roots, blood vessels and other tissues. These are 10 times less frequent than intracranial tumors with majority of them being benign. According to their location, spinal tumors are conveniently classified as extradural and intradural, although some can be both inside and outside the dura. Intradural tumors can be intramedullary (intramedullary spinal cord tumor [IMSCT]) or extramedullary (intradural extramedullary [IDEM]).[2]

Space occupying lesions in the spinal canal cause compression of the structures with resultant neurological deficits. Rapidly growing lesions cause severe loss of function as there is no time for the spinal cord to adjust itself. The presence of a tumor interferes with the normal movements of the cord, which occur during movements of the spinal column. Such impairment contributes to cord damage. In long standing tumors, there may be gliosis in the spinal cord due to ischemia and recovery may be incomplete despite complete removal of the tumor. Initial assessment of the patient with a primary spinal tumor requires meticulous application of common clinical tools including a detailed history and clinical examination. Clinical presentation usually relates to pain, varying degrees of motor sensory deficits as well as bowel bladder symptoms in

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some, which may be due to either mass effect or neurological compression.[3] In this study, we analyzed the patients of spinal tumors surgically managed by us, to see the trend of spinal tumors in northern India. The cases were evaluated with regard to the pathological diagnosis, preoperative medical history, clinical symptoms, surgical treatment, outcome, recurrence and prognostic factors.

Materials and Methods

This was a retrospective study of 111 consecutive patients with spinal tumors who were treated surgically by single surgeon (senior author) at Department of Neurosurgery, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow, India, from 2003 to 2011. This is one of the largest tertiary care referral institutes in north India.

Inclusion criterion

- Primary and secondary tumors of the spinal canal.
- Tumor like conditions like eiosinophillic granuloma.

Histiocytosis X, aneurysmal bone cyst.

Exclusion criterion

- Vascular malformations,
- Infective pathologies including potts spine
- The patients with inadequate record, or
- Follow-up <1.5 months after surgery were excluded from the study.

Each patient's medical history, findings of the physical examination performed in an out/in-patient department, radiological examination records were investigated. We used modified McCormicks grade to evaluate the neurological and functional status of patients [Table 1]. All patients were investigated by contrast magnetic resonance imaging of the whole spine. Appropriate radiology was obtained during follow-up, as and when required, depending upon the clinical situation or to assess the extent of resection and recurrences. Statistical analysis was performed by using Fischer exact and Chi-square tests.

Observations

There were a total of 111 patients who fulfilled the inclusion criterion. Out of 111 patients, there were 30 patients with extradural tumors, 40 had IDEM, and 41 had intramedullary tumors. There were 80 (72.08%) males. Mean age at time of surgery was 30.7 years. There were 34 (30.63%) patients in pediatric (<18 years) age group. The demographic profile of patients is represented in Table 2.

Histopathological diagnoses

We observed a variety of benign and malignant lesions. Overall 34/111 (30.63%) were neuroepithelial tumors (ependymomas + astrocytomas) followed by nerve sheath tumors 25/111 (22.53%). The dermoid/epidermoids were present in 11 (9.9%), and meningiomas were present in 7 (6.3%) cases only. The single most frequent histopathological diagnosis was ependymoma (22; 19.81%) followed by schwanomma (14; 12.61%) and astrocytomas (11; 9.9%). Various diagnoses in terms of their location are presented in Table 3.

Location

the thoracic region of spinal canal was most frequently involved (38; 34.2%) followed by cervical 21 (18.91%), thoracolumbar (15; 13.5%) and lumbar (12; 10.8%) region. Two patients had multiple lesions both affecting the extradural compartment. One patient had an intramedullary lesion involving the cervical cord as well as medulla (cervical cord > medulla) [Table 4].

Clinical profile

The common clinical symptom was motor weakness (78 cases; 70.27%) followed by sensory loss and pain. Sphincter disturbances were present in 47 patients (42.34%). The weakness in the majority was spastic, however, hypotonia was observed in 10 patients (9.1%) as shown in Table 5.

Table 1: Modified McCormick scale

Grade	Modified McCormick scale
I	Intact neurologically, normal ambulation, minimal dysesthesia
II	Mild motor or sensory deficit, functional independence
III	Moderate deficit, limitation of function, independent with external aid
IV	Severe motor or sensory deficit, limited function, dependent
V	Paraplegia or quadriplegia, even w/flickering movement

Table 2: Demographic profile of patients

	Extradural	IDEM	IMSCT	Total
Number of patients (%)	30 (27)	40 (36.1)	41 (36.9)	111
Mean age at surgery (range)	30.77 years (1-73 years)	35.88 years (11-62 years)	25.79 years (3-63 years)	30.81 years (1-73 years)
Age <18 years	9	9	16	34 (30.63%)
Gender (male/female)	25/5	26/14	29/12	80 (72.08%) males 31 (27.92%) females
Mean preoperative duration of symptoms	4.9 months (0.5-48 months)	18.73 months (1-120 months)	24.89 months (9 days to 180 months)	16.17 months (9 days to 180 months)
Mean follow-up after surgery	7.5 months (1.5-60 months)	16.72 months (1.5-66 months)	22.7 months (4-120 months)	15.64 months (1.5-120 months)

IDEM – Intradural extramedullary; IMSCT – Intramedullary spinal cord tumor

Table 3: Histological diagnosis of cases

Extradural (*n*=30)	IDEM (*n*=40)	IMSCT (*n*=41)	Total (*n*=111)
Metastasis-4	Schwannomma-12	Ependymoma-17	Ependymoma-22
Lymphoma-3	Neurofibroma-8	Low grade glioma-9	Schwannomma-14
Plasmacytoma-3	Meningiomma-7	High grade glioma-2	Gliomas-11
ABC-2	Ependymomma-5	Epidermoid-4	Neurofibroma-8
Mesenchymal tumor-2	Epidermoid-3	Dermoid-4	Meningioma-7
Chondrosarcoma-2		Lipoma-2	Epidermoid-7
Lipoma	Enterogenous cyst-1	Ganglioglioma-1	Dermoid-4
Mix germ cell tumor	Arachnoid cyst-1	IM	Lipoma-3
Hitiocytosis X	Neurenteric cyst-1	Schwanomma-1	
Eiosinophilic granuloma	Paraganglioma-1	Hemangioblastoma-1	
Ewings sarcoma			
Chordoma			
Malignat peripheral nerve sheath tumor*			
Myeloma			
Hemangioma			
Schwanomma*			
Giant cell tumor			
Osteoclastoma			
PNET 1 each			
Inconclusive-1	Inconclusive-1		

*These were without intradural extension. PNET – Primitive neuroectodermal tumor; IDEM – Intradural extramedullary; IMSCT – Intramedullary spinal cord tumor; ABC – Aneurysmal bone cyst; IM – Intramedullary

Table 4: Location of lesions

Location of tumor	Total
Cervicomedullary	1 (IM)
Upper cervical	11 (extradural-2, IDEM-5, IM-4)
Lower cervical	9 (extradural-1, IDEM-4, IM-4)
Cervico-thoracic	10 (extradural-1, IDEM-2, IM-7)
Thoracic	38 (extradural-9, IDEM-19, IM-10)
Thoracolumbar	15 (extradural-3, IDEM-4, IM-8)
Lumbar	12 (extradural-1, IDEM-5, IM-6)
Lumbosacral	6 (extradural-4, IDEM-1, IM-1)
Sacraococcgeal	7 (extradural-7)
Multiple	2 (extradural-2)

IDEM – Intradural extramedullary; IM – Intramedullary

Results

A posterior approach using standard microsurgical techniques was performed in all cases, and this was irrespective of the location of a tumor. We did not require instrumentation in any case. Total excision was achieved in 57 (51.35%) [Table 6]. Only biopsy was performed in two cases.

Outcome

We defined "good outcome" as the improvement in patient's preoperative modified McCormick score, at the time of last follow-up. Those who had an improvement of ≥2 grades were labeled as having "significant improvement". The patients who either remained same or showed a deterioration of modified McCormick score were considered "poor outcome". In our series, 88/111 (79.27%) patients had a good outcome at time of last follow-up. All these patients were ambulatory with or without a walking aid (M. McCormick score ≤3). Totally, 79/111 (71.17%) patients were dependent preoperatively (unable to walk even with external aid; M. McCormick Grade 4 and 5). Totally, 59 out of these 79 patients were independent with or without external aid at last follow-up [Table 7].

Complications

Persistent pain was present in 10 (9.1%) patients at follow-up. Bladder and bowel symptoms were not relieved in (7; 6.3%). Six (5.41%) patients deteriorated. There were 2 (1.8%) patients with wound dehiscence; one had cerebrospinal fluid leak (0.9%). Both of them could be managed conservatively. There was one mortality in intramedullary group-this patient, who presented with spinal shock and respiratory depression, failed to have any improvement after surgery. He became ventilator dependent and died 3 months after surgery [Table 8].

Recurrences

In this series, "recurrence" has been assessed on the basis of new symptoms related to tumor growth or tumor regrowth on routine radiological assessment. Two tumors in extradural group (a chondrosarcoma and a malignant peripheral nerve sheath tumor) showed recurrence, 9 months and 5 years after surgery, respectively. Three patients having IDEM tumors (meningioma; 2 years postoperatively, meningioma; 3 years, and a myxopapillary ependymoma; 8 months postoperatively) had recurrences. Two patients in intramedullary group (an anaplastic ependymoma at

20 months, and a conus dermoid at 10 years follow-up) had recurrences.

Discussion

Primary spinal cord tumors account for 4–10% of all central nervous system tumors and are characterized based on their location as intramedullary (IMSCT), IDEM, and extradural.[4] Totally, 2/3 of all spinal tumors are said to be IDEM and 10% IMSCT[5] but we had nearly equal incidence of IDEM (40/111; 36.1%) and IMSCTs (41/111; 36.9%) in our series. This difference may be due to tertiary referral at our institute or this may represent the epidemiological trend of a developing country.

Table 5: Clinical features

Symptoms/signs	Number (%)
Motor weakness	78 (70.27)
Sensory loss	55 (49.54)
Pain	46 (41.44)
Bladder involvement	38 (34.23)
Bowel involvement	9 (8.1)
Parasthesias	20 (18)
Hypotonia	10 (9.1)
Wasting	3 (2.7)
Local tenderness	2 (1.8)
Band like sensation	3 (2.7)
Scoliosis	2 (1.8)
Foot deformities (w/o spinal dysraphism)	1 (0.9)
Spinal shock	1 (0.9)
Hemifacial spasm*	1 (0.9)

*Hemifacial spasm was observed in a cervical lesion extending to medulla

Table 6: Extent of resection

	Extent of resection (%)
Total excision (complete removal)	57 (51.35)
Near total excision (>90%) (small part of tumor adherent to vital structures was left behind)	22 (19.81)
Subtotal excision (40-60%) (decompression)	30 (27.02)
Biopsy	2 (1.8)

The literature indicates that in western populations, the primary spinal tumors occur more frequently in females, whereas Asian studies show a slight male preponderance.[6,7] We had a male to female ratio of 2.5:1 among our patients. Similar male to female ratio has been reported by other studies from India.[8,9] We observed that all histopathological subtypes, except meningioma, were more common in males as compared to females (in this series, 6/7 patients with meningioma were females). The female preponderence of meningioma is a well-known entity[6,10-12] and our study also supports it.

The mean age of presentation of patients with IDEM tumors in our study was 35.88 (range, 11–62 years), those having IMSCT was 25.79 (3–63 years) and in extradural tumors was 30.77 (1–73 years). The mean age of IMSCTs in our series is less as compared to that of the western world[5] but was comparable to Indian reports.[8,9] This is due to the presence of significant number of pediatric patients in this subgroup (16/41; 39%).

Astrocytomas and ependymomas represent the most common intramedullary neoplasms. Ependymomas are the most frequent IMSCTs in adults while astrocytomas are more frequent in the pediatric population.[8,13] Other intramedullary lesions include dermoid, epidermoids, hemangioblastomas (HGBs), lipomas, gangliogliomas, lymphomas and metastasis etc., Congenital malformative tumors like dermoids and epidermoids (not associated with spina bifida) account for only 5–8% of intramedullary lesions in western populations. However, these were more common (23%) in our series of pediatric intramedullary mass lesions[14] depicting a difference of their incidence in a developing country. In this mix population series, we had 8/41 (19.51%) dermoids and epidermoids in IMSCT group, which is a higher percentage than western countries.

The most common IDEM spinal cord tumors are nerve sheath tumors (schwannomas/neurofibromas) followed by meningiomas. Fewer frequent subtypes include epidermoids,

Table 7: Outcome

	Extradural	IDEM	IMSCT	Total (%)
Good outcome				
Significant improvement (increase in modified McCormick score ≥2 from preoperative)	9	23	12	44 (39.96)
Improvement (increase in modified McCormick score by 1 grade from preoperative)	11	14	19	44 (39.96)
Poor outcome				
Same as preoperative	8	2	7	17 (15.31)
Deteriorated	2	1	3	6 (5.41)
Functionally dependent patients preoperatively (modified McCormick score 4/5)	20	29	30	79
Patients (from above-mentioned group) who became independent postoperatively (modified McCormick score ≤3)	9	27	23	59
Mean preoperative McCormick score	3.86	3.9	3.78	3.84
Postoperative	3.0	2.25	2.70	2.65

IDEM – Intradural extramedullary; IMSCT – Intramedullary spinal cord tumor

dermoid, developmental cysts, paraganglioma, lipoma, spinal nerve sheath myxoma, etc.[15] We had 20/40 (50%) nerve sheath tumors and 7/40 (17.5%) meningiomas in IDEM group. This compares favorably with existing literature.

The common extradural tumors include metastasis,[5] lymphomas, and various benign (osteoid osteoma, osteoblastoma, giant cell tumor, aneurismal bone cyst, hemangioma), and malignant (chordoma, chondrosarcoma, osteogenic sarcoma, Ewing's sarcoma, plasmacytoma) bony tumors. We had similar representation of these entities in their subgroups in our study. We also had rare tumors like primitive neuroectodermal tumor (PNET) and malignant peripheral nerve sheath tumor in this subgroup [Table 3].

Overall, in our series of spinal tumors, the most common histological subtypes were neuroepithelial tumors (ependymoma + astrocytomas) −35/111; 31.53%, followed by nerve sheath tumors (25/111; 22.52%). The nonneoplastic malformative tumors (dermoids/epidermoids) were present in 11 (9.9%) and meningiommas in (7/111; 6.3%) cases only. When compared with studies from western countries, we observed a comparable frequency of nerve sheath tumors as well as of the neuroepithelial tumors in our series, whereas the incidence of meningioma was low (6.3%) as compared to that reported by western authors (16–46%).[10,11,16-18] However, the latter was similar to report by studies from Japan[6] to China[7] (11–15%). This compares favorably with epidemiological studies that Asian populations have a relatively less incidence of spinal meningioma than western countries.

Our series represents single handed experience of the senior author. Histology and location of the tumor affect the extent of surgical resection. Ependymomas often can be surgically resected. However, astrocytomas infiltrate the spinal cord, and complete resection is rare. The extent of resection is dependent largely on presence/absence of a cleavage plain observed intraoperatively. Presence of an associated syrinx in IMSCTs favors the resectability of the tumor, up to some extent.[19] IDEM tumors (represented by schwannomas, neurofibromas, and meningiomas in majority) are usually amenable to surgical resection. In extradural pathologies, benign lesions are managed by intralesional (e.g., osteoid osteoma) or marginal excision. Those lesions with a tendency for local recurrence (e.g., aggressive osteoblastoma, and giant cell tumor) are best excised with a wide margin. For the most part, resectable malignant lesions require a radical margin by en bloc excision. We could excise 33/40 (82%) of IDEM tumors totally. In IMSCT group, we were able to excise 13/41 (31.17%) lesions totally while near total excision were achieved in 16/41 (39%). Subtotal excision was performed in 11/41 (26.82%), and only biopsy was possible in one case. These rates are comparable to resection rates reported by others.[20-22]

Radiotherapy is reserved for malignant variants and recurrent gliomas, whereas chemotherapy is administered for recurrent primary spinal cord tumors without surgical or radiotherapy options.[5] Totally, 31 of our patients received postoperative radiotherapy (20 in IMSCT Group, 8 in extradural Group and 3 in IDEM group).

We evaluated the effect of different preoperative factors on outcome of patients [Table 9]. Age and gender had no effect on outcome ($P = 0.864$ and $P = 0.27$ respectively). We found a statistically significant effect of the preoperative neurologic status on the postoperative outcome of patients. The patients with a good preoperative neurologic status (modified McCormick score <3) had a better overall outcome than those having a preoperative score of 4 or 5 ($P = 0.04$). This is similar to observations made by other studies.[8,19,23]

A delay in diagnosis or management of patients with mass lesions causing compression of neuronal tracts in the spinal cord may result in residual deficits and poor outcome. Dutch

Table 8: Complications

	n (%)
Persistant pain	10 (9.1)
Nonimprovement of bladder or bowel symptoms	7 (6.3)
Neurological deterioration	6 (5.41)
Retrograde ejaculation	1 (0.9)
Kyphoscoliosis	1 (0.9)
Wound dehiscence	2 (1.8)
Persistant paraesthesias	2 (1.8)
CSF leak	1 (0.9)
Recurrence	7 (2 extradural, 3-IDEM, 2-IM group)
Death	1 (0.9)

CSF – Cerebrospinal fluid; IDEM – Intradural extramedullary; IM – Intramedullary

Table 9: Prognostic factors

Parameter	Number of patients	Good outcome	Poor outcome	*P*
Age				
<50 years	93	74	19	0.84
>50 years	18	14	4	
Gender				
Male	80	59	21	0.26
Female	31	26	5	
Preoperative neurologic status				
Modified McCormick score ≤3	32	29	3	0.047*
Modified McCormick score 4 or 5	79	59	20	
Length of involved segment				
≤3	76	63	13	0.25
≥4	35	25	10	
Preoperative duration of symptoms				
≤12 months	82	66	16	0.59
>12 months	29	22	7	

*P< 0.05 is significant

investigators[24] collected a series of 108 patients with both intradural and extradural tumors. They found that 35% of patients were diagnosed more than 2 years after the onset of symptoms. Delayed presentation is one of the main factors leading to poor neurological grade at time of surgery/intervention. This affects the postoperative outcome. The average preoperative duration of symptoms in our series was 16.17 months, with a range from 15 days to 15 years and a significant number of our patients were in poor neurological status at presentation (79/111; 71.17%). The long-standing compressive pathologies cause profound and irreversible neuronal degeneration due to destructive changes such as ischemic necrosis and neuronal loss in gray matter as well as demylenation in white matter and in posterior and lateral white columns.[25] In our series, the patients with a short preoperative duration of symptoms had a slightly better outcome in terms of functional status, when compared to patients, who were symptomatic for a period >12 months (66/82; 81.5% vs. 22/29; 75.7%). However, this difference was not statistically significant ($P = 0.5$).

We tried to observe the effect made by longitudinal extent of the tumor on the surgical outcome. The longitudinal extent of tumor is, in turn, related to be histological diagnosis (for example glial tumors frequently extend over several spinal cord levels and especially, astrocytomas show an infiltrative growth pattern but others like HGBs and schwanommas usually have limited extent). Our results show that the longitudinal extent of tumor did not have a statistically significant effect on long-term outcome after surgery ($P = 0.25$). This compares to results shown by other groups.[26]

The reported rate of recurrences in spinal tumors is 7.2%, and IDEM masses tend to recur more commonly.[27] We observed 7/111 (6.3%) recurrences, out of which 4/7 (57.1%) were from IDEM group.

A recent study, based on NIS database (2003–2010) in US has reported the overall in-hospital mortality rate of 0.46% in operated spinal tumors.[28] We had one (0.9%) mortality in a patient with an intramedullary tumor.

The various series of spinal tumors had a good functional outcome in 15–90%[29-34] of patients. In our series, a total of 79.27% patients were mobile at last follow-up. This compares favorably with another series.

In our series 59 patients out of 79, who were dependent preoperatively, became independent with or without the help of a walking aid regardless of prognostic factors. Similar observations have been made by other authors.[27] We recommend that surgical intervention if indicated, should be considered for spinal tumors regardless of the prognostic factors. Even patients with poor neurological grade preoperatively may improve in their functional status.

Conclusions

The congenital malformative tumors (unassociated with spina bifida) occur more commonly in developing the world while the incidence of spinal meningioma is less. The incidence of intramedullary tumors approaches to that of IDEM tumors. Intramedullary tumors present at a younger age in developing countries. Rare histological variants like PNETs should also be considered for histological differential diagnosis of spinal tumors. Delayed presentation is the main reason for poor preoperative neurologic status, leading to poor outcome in the management of spinal tumors. A high level of suspicion and acknowledging the classical symptoms of cord compression are the most important factors in shortening the time to diagnosis of spinal tumors.

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