Patterns of Spinal Tumors in Nepal: a Clinico-radiological Study

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The objective of this study was to analyze the patterns of spinal tumors in a tertiary care hospital in Nepal and to describe their radiological features and pathological diagnosis.

A total of 77 patients with spinal tumors who underwent surgery at our institution from March 1996 to December 2003 were analyzed. There were 19 cervical, 40 thoracic and 18 lumbosacral lesions. Among the 19 cervical, there were 15 (79%) intradural extramedullary, and 3 (15.8%) extradural lesions and one (5.2%) intramedullary lesion in the operated group. Among intradural extramedullary lesions, neurofibromas (12) were the commonest, followed by schwannoma (2). Only one meningioma was found. Among the 3 extradural lesions, one was hemangioma, one was chondromyxoma and one rhabdomyosarcoma

In the thoracic group 27 (67.5%) out of 40 were intradural extramedullary lesions and 12(30%) were extradural lesions. Among intradural extramedullary lesions, neurofibromas were the commonest (10 in 40 i.e. 25%), followed by meningiomas (9(36%))and schwannomas (4(16%)). Neurilemoma was the least common neoplasm (2(8%)). The histopathology reports were inconclusive in 2 patients. Among extradural lesions, metastases (6 out of 12) were the commonest. Only one (2.5%) intramedullary lesion (astrocytoma) was operated.

In the lumbar region, extradural lesions were most common comprising 50% (9 out of 18) in which metastasis topped the list (5 out of 9) followed by 2 lymphoma and 2 giant cell tumor. In the 5 intradural extramedullary lesions, 2 were neurofibromas, 2 were meningiomas and only one was arachnoid cyst. Ependymoma was the only intramedullary lesion at lumbar level, detected in 2 patients. The histopathology was inconclusive in 2 patients.

The most common level for spinal tumor was thoracic followed by cervical and lumbar. Both at cervical and thoracic regions intradural extramedullary lesions were common, whereas extradural lesions were more common in the lumbar region.

Key Words: CT, MRI, neurofibroma, spinal tumors

Spinal tumors are commonly encountered entities in the neurosurgical practice. They may arise from the neural tissue, the meninges, surrounding bone and soft tissues, from embryonal rests or as metastases. Based on the location, they are classified as extradural, intradural/ extramedullary, and intramedullary tumors. Extradural lesions constitute the lesions of the osseous spine, epidural space and paraspinous soft tissue. Intradural extramedullary lesions are located under the dura matter but outside the spinal cord whereas intramedullary lesions are lesions of the spinal cord.

The objective of this study was to analyze the patterns of spinal tumors in a tertiary care center in Nepal and to describe their radiological features with pathological diagnosis.

Materials and Methods

Our institution is a tertiary level teaching hospital providing neurosurgical services since 1995. A total of 77 patients were operated for spinal tumors at the hospital in the 7-year period (March 1996- December 2003). All patients had different radiological investigations before surgery.

Level	Number	Age range (years)	Male	Female
Cervical	19	7 - 79	16	3
Thoracic	40	16 - 65	25	15
Lumbar	18	15 - 69	12	6

Table 1. Distribution of lesions and patients' age range and gender distribution in 77 patients.

In the earlier part of the study most of the patients were operated based on the myelographic findings whereas in the later part almost all had either computerized tomography (CT) scan or magnetic resonance imaging (MRI) scan before surgery.

We retrospectively analyzed the prospectively acquired data on these 77 patients in terms of level and location of the lesions, and describe the radiological features.

Results

Table 1 shows the age and sex distribution in the series of 77 patients. Spinal tumors were detected mainly in the adult patients and there was a strong predilection for female sex.

Cervical region

Among the 19 lesions, there were 15 (79%) intradural extramedullary and 3 (15.8%) extradural lesions and 1 (5.2%) intramedullary lesion in the operated group at the cervical level **(Table 2)**. Hemangioma, chondromyxoma and rhabdomyosarcoma constituted the extradural lesions. Among the intradural extramedullary lesions, neurofibroma was the commonest lesion followed by schwannoma and meningioma. The intramedullary lesion was an ependymoma **(Figure 1)**.

Location	Pathology	Number	Percentage (of total)
Extradural (15.8%)	Hemangioma	1	1.3
	Chondromyxoma	1	1.3
	Rhabdomyosarcoma	1	1.3
Intradural extramedullary (79%)	Neurofibroma	12	15.6
	Schwannoma	2	2.6
	Meningioma	1	1.3
Intramedullary (5.2%)	Ependymoma	1	1.3

Table 2. Location of cervical lesions in a series of 19 patients.

Thoracic region

As shown in **Table 3**, there were a total of 40 thoracic lesions. Extradural lesions comprised 30% of the total. Twenty-seven (67.5%) were intradural extramedullary lesions. There was only one intramedullary lesion in this series.

Among extradural lesions, metastases were the commonest followed by hemangioma and lymphangioma. In 2 patients the histopathology was inconclusive. Neurofibroma was the commonest among intradural extramedullary lesions comprising 13% of the total spinal lesions, followed by meningioma, schwannoma and neurilemoma.There was only one intramedullary lesion (astrocytoma) at the thoracic region. **Figure 2** shows the typical example of intradural extramedullary lesion typical of meningioma.

Lumbar region

A total of 18 lesions were present at the lumbar level with extradural lesions representing 9 (50%) of them. Metastases **(Figure 3)** were the commonest extradural lesions followed by lymphoma and giant cell tumor. Among 5 intradural extramedullary tumors neurofibroma and meningioma were the commonest (2 of 5 each) and one lesion was arachnoid cyst. Ependymoma was the only intramedullary lesion at the lumbar level. In 2 patients neither the location nor the histopathology of the tumor was specified.

Location	Pathology	Number (of total)	Percentage
Extradural (30%)	Metastasis	6	7.8
	Hemangioma	2	2.6
	Lymphoma	2	2.6
	Pathology inconclusive	2	2.6
	Neurofibroma	10	13
Intradural extramedullary (67.5%)	Meningioma	9	11.7
	Schwannoma	4	5.2
	Neurilemoma	2	2.6
	Pathology inconclusive	2	2.6
Intramedullary (2.5%)	Astrocytoma	1	1.3

Table 3. Distribution of thoracic lesions in the consecutive series of 40 patients.

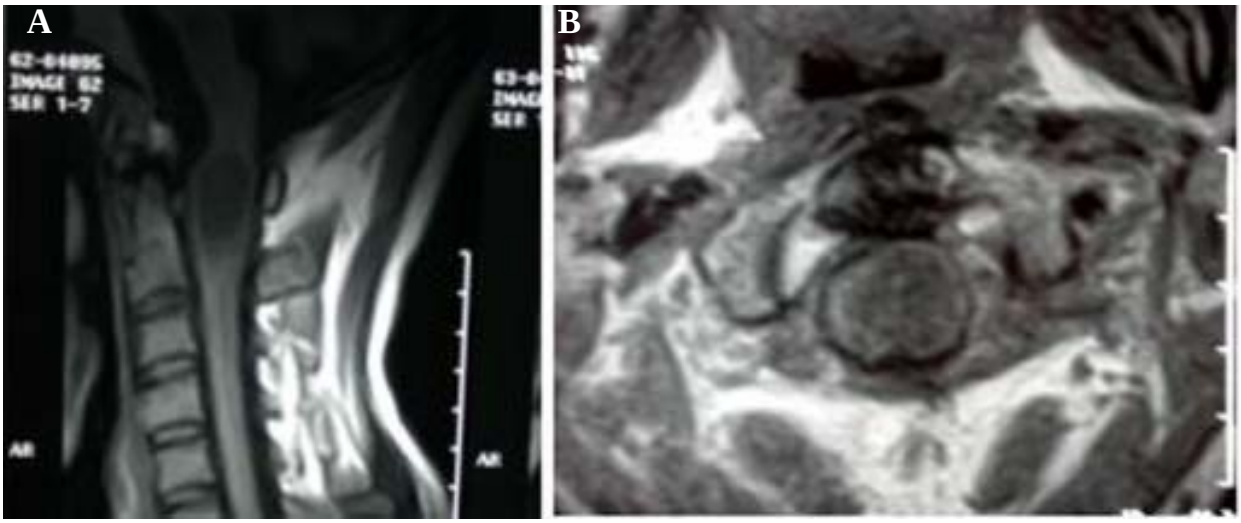

Figure 1. MRI of a 40-year-old woman with progressive quadriparesis. Sagittal T1- (A) and axial T1- (B) weighted MR scans show diffuse expansion of the cord at the cervical level characteristic of an intramedullary lesion most likely an ependymoma.

Discussion

Spinal tumors comprise a wide spectrum of diseases due to different level, and location in the spinal canal as well as the vast neoplastic conditions that can potentially involve the spine and the spinal cord. Accurate preoperative diagnosis helps in the correct decision making for the optimal surgical management of the patient. Sparing the intramedullary lesions, the majority of the extradural and extramedullary lesions are asymptomatic for a prolonged period of time and by the time the patients seek a clinician's help the tumor attains significant size. With the advent of modern imaging technologies such as CT scan and MRI, it has been easier to diagnose these lesions preoperatively accurately. From the radiological point of view, these tumors are traditionally classified into three categories:

Location	Pathology	Number (of total)	Percentage
Extradural (50%)	Metastasis	5	6.5
	Lymphoma	2	2.6
	Giant cell tumor	2	2.6
Intradural extramedullary (27.8%)	Neurofibroma	2	2.6
	Meningioma	2	2.6
	Arachnoid cyst	1	1.3
Intramedullary (11.1%)	Ependymoma	2	2.6
Location and type of tumor not specified		2	2.6

Table 4. Distribution of lumbar lesions in aseries of 18 patients.

Extradural Lesions

These lesions are located outside the thecal sac. They arise from osseous spine, epidural space and paraspinous soft tissues.[23] General imaging hallmarks of extradural tumors are focal displacement of the thecal sac and its contents away from the mass.[23] Myelography shows extrinsic compression of the thecal sac. If there is a complete block the interface between the mass and contrast column is poorly defined with feathered appearance at the level of obstruction.[29] The imaging features of individual lesions are described below.

Metastasis

The most common extradural tumor is metastasis and usually the primary tumors are in the breast, lung and prostate.[6] Other tumors in this location are lymphoma, melanoma, and renal cell carcinoma.[6] In children the primary sites are mostly Ewing's sarcoma of bone and neuroblastoma followed by osteogenic sarcoma, rhabdomyosarcoma, Hodgkin's disease, soft tissue sarcoma, and germ cell tumor.[14] In our series metastases were the commonest lesions. On plain X-ray, most metastases are osteolytic, and the most common finding is pedicle

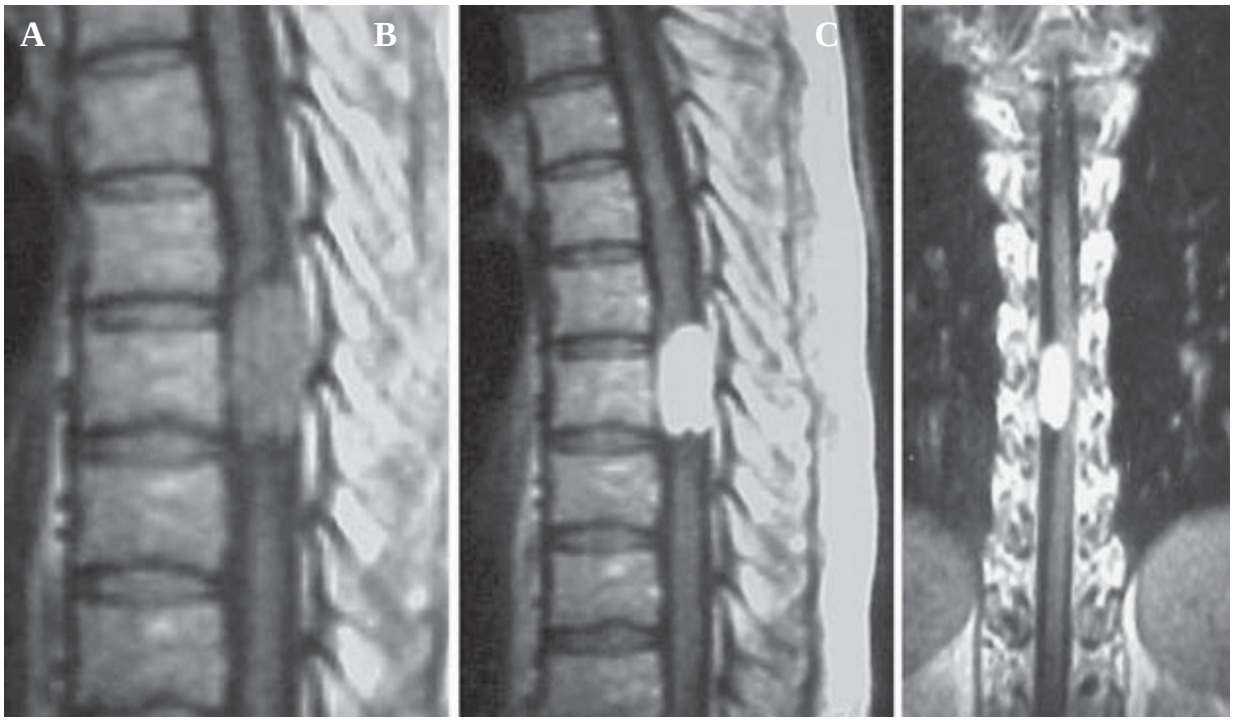

Figure 2. MRI Sagittal pre- (A), post-contrast (B) and coronal post contrast (C) T1-weighted scans of this 43-year-old gentleman with progressive paraparesis. The radiological diagnosis was a thoracic meningioma, which was subsequently confirmed histopathologically after surgical resection.

destruction. Other findings include multifocal lytic vertebral body lesions, pathological compression fractures and paraspinal soft tissue mass. On occasion there may be indistinct posterior body margin.[22] CT scan distinguishes the lytic from the blastic lesions. However, to define the extent of the lesion, intrathecal contrast is required.[8] MRI has been the investigation of choice in patients with suspected spinal tumors. In addition to superior tumor resolution, it has the added advantage of delineating the epidural and paraspinous soft tissue involvement as well as cord compression. Four patterns of metastatic disease have been described in MRI:

a. Focal lytic lesion is the most common abnormality and shows low signal characteristics on T1 and high signal characteristics on T2 weighted (W) sequences.[13]
b. Focal sclerotic lesions are hypointense on both T1- and T2W sequences.
c. Diffuse inhomogenous lesions
d. Diffuse homogenous lesions show low signal characteristics on T1W and high signal characteristics on T2W sequences.[1]

MRI also helps to distinguish pathological fracture of vertebra, which shows marrow as low signal characteristics on T1 and high signal characteristics on T2W sequences, from benign compression fracture where the marrow signal is isointense with normal vertebra.[4]

Lymphoma

Lymphoma can involve the spine and epidural soft tissues. In the spine, non-Hodgkin's lymphoma (NHL) is more common than Hodgkin's lymphoma. Imaging findings are nonspecific. NHL can cause bone destruction and hyperostosis,[26] along with spinal cord compression.[25] Epidural extension is better delineated on MRI which shows low signal characteristics on T_1 and inhomogenous hyperintensity on T2 W images.[26]

Hemangioma

Vertebral hemangiomas are slow growing benign neoplasms of capillary, cavernous or venous origin. They vary from predominantly fatty lesions to hemangiomas comprising largely vascular stroma with little or no adipose tissue.[15] These lesions are mostly asymptomatic and discovered incidentally but sometimes progressive neurological deficit or symptoms of acute cord compression can occur.[11]

As per imaging, plain radiographs show lytic foci with honeycomb trabeculation or thick vertical striations. Non-contrast CT scans show a lucent lesion with typical ' polka-dot' densities in the medullary space representing the trabeculae. They range in size from small focal to large lesions involving the entire vertebral body. Myelography or CT- myelography show an extradural mass. Angiographic findings vary from normal to intense hypervascular stain on selective segmental spinal angiogram.[15]

MRI findings also vary. They appear well-defined round vertebral lesions with high signal in both T_1 and T2 W images representing the stroma surrounding foci of very low signal intensity on T1W sequence, representing the trabeculae. These lesions enhance with contrast and may have an extradural soft tissue component as well.[15]

Figure 3. Sagittal T1- (A) and T2- (B) weighted MRI scans of 70-year-old gentleman with a known carcinoma of lung who presented with rapid paraparesis. Pathological compression fracture with thecal sac compression is evident. Metastatic lung carcinoma.

Giant Cell tumor

These tumors are lytic, expansile, locally aggressive primary benign lesions extending to the cortex but rarely beyond the periosteum.[8] Plain radiographs and CT scans show a lytic, expansile, destructive sacral or vertebral mass. Angiography demonstrates hypervascularity. MRI scans show a mixed signal, multicompartmental cystic mass with evidence of blood degradation products.[3]

Other lesions such as chondromydoma, rhabdomyosarcoma and ectopic ossifications are uncommon in the region of the spine. The patients are symptomatic due to their extraosseous extension, especially in the epidural space, sometimes with features of cord compression. The imaging findings are nonspecific and diagnosis is confirmed histopathologically.

Intradural Extramedullary Lesions

The tumors in this group arise inside the dura but outside the spinal cord. The tissues of origin are nerve roots, leptomeninges and CSF spaces.[23] The general imaging features show the lesion displacing the spinal cord along with enlargement of ipsilateral subarachonoid space. Myelogram of these lesions show classic intradural filling defect outlined by sharp meniscus of contrast, deviation of the spinal cord, and enlarged subarachnoid spaces.[23] Other imaging features of individual lesions are described below.

Nerve Sheath Tumor

Two main types are schwannoma/ neurilemoma and neurofibroma. Ganglioneuroma is relatively rare.

Most nerve sheath tumors arise from dorsal sensory roots. Depending on their origin along the root, these can be intradural, extradural or combined intra- and extradural 'dumbbell' lesions.[26] Multiple lesions are common in neurofibromatosis. Plain radiographs show pedicle erosion and enlarged neural foramina. Paraspinal soft tissue masses are common with 'dumbbell' and extradural lesions. Kyphoscoliosis, ribbon ribs and posterior vertebral body scalloping may be seen in neurofibromatosis.[8,19] Myelography shows typical features of an intradural extramedullary lesion (vide supra). Large tumors can cause complete block of contrast. Non-contrast CT scans show bony changes with hypo- or slight hyperdensity. Calcifications and hemorrhage are rare.[8,12] On MRI, 75% of these tumors are isointense to cord on T1W images. On T2W sequences these tumors are hyperintense.[12,19,16] A " target" appearance with hyperintense rim and hypointense center is often seen on T2W and contrast enhanced sequences in neurofibroma. Sometimes schwannoma may show a cystic component, necrosis or hemorrhage.[35] Schwannoma and neurofibroma cannot be reliably distinguished on MR scans.[35]

Meningioma

Most spinal meningiomas are benign; the aggressive ones such as hemangiopericytoma being rare, [8] and most of them are solitary. They are the second most common spinal tumors.

The majority of the lesions are intradural but sometimes "dumbbell or extradural lesions do occur and they are mostly lateral to the spinal cord.[32] Plain films are usually

normal. Myelography shows features of typical intradural extramedullary lesions (vide supra). Non-contrast CT scans may show extradural or dumbbell mass that is iso- or moderately hyperdense (compared to muscle). Intradural lesions require intrathecal or intravenous contrast for adequate delineation. On MRI, most meningiomas are isointense with spinal cord on both T1 and T2W sequences.[32] On contrast administration relatively homogenous enhancement is seen.[17] Most spinal meningiomas have a broad-based dural attachment (dural tail sign).[16] Occasionally densely calcified meningiomas are profoundly hypointense on MR scans and show minimal contrast enhancement.[23]

Arachnoid Cyst

Intradural arachnoid cysts are rare. Their etiology is unclear. Most of them arise near the septum posticum, and are located posterior to the spinal cord. Most of them communicate with the subarachnoid space and opacify following intrathecal contrast injection. CT myelography typically shows compression and anterior displacement of the spinal cord. Signal intensity on MRI is that of CSF; hence it might be missed on MRI unless there is displacement and flattening of the cord.

Metastasis is one of the rare tumors in intradural extramedullary location.

Intramedullary Tumors

These are tumors of the spinal cord. Most are malignant and are gliomas. Most of the cord gliomas are ependymomas and low-grade astrocytomas. Other primary spinal cord tumors such as hemangioblastoma are rare. Spinal cord lipomas are considered congenital in origin. General imaging features show diffuse multisegmental cord enlargement with reduced subarachnoid space at the level of the lesion.[23]

Ependymoma

Ependymomas arise from ependymal cells lining the central canal or its remnants, or from cells of the ventriculus terminalis in the filum terminale.[20] They often attain large size. Cystic degeneration and hemorrhage are common.[18,21] Ependymomas occur most often in the conus medullaris and filum terminale.[26] Cervical cord is the commonest site for these tumors.[18] Plain films show widened canal or bone destruction.[28] With large lesions the posterior margin of vertebral bodies will be scalloped and neural foramina may be widened. Myelography reveals non-specific cord widening. Multisegmental lesions are common.[18] Small conus medullaris and filum terminale lesions are seen as well-delineated intradural masses with a contrast 'meniscus' around the lesion. CT scan also shows only non-specific canal widening. MR imaging shows a widened cord or a filum terminale mass. Most ependymomas are isointense to cord on T1W sequences.[24] However, mixed signal is seen when there is a cyst formation, necrosis or hemorrhage. T2 W sequences show hyperintense lesion. Hypointensity at the tumor margin is highly suggestive of ependymoma.[21] Strong but inhomogenous contrast enhancement is usual.[24]

Astrocytoma

Most spinal cord astrocytomas are low-grade tumors.[8] They diffusely expand the cord. Intratumoral cyst formation is common and associated syrinxes are frequent.[23] The cervical cord is the most common site and multisegmental involvement is the rule. Plain films are often normal or show only mild scoliosis, and a widened canal. Increased interpedicular distances may be present.[8] Myelogram and CT scans show nonspecific cord and canal expansion. On MR imaging, they are iso- or slightly hypointense on T1 and hyperintense on T2W sequences and show contrast enhancement.[24,34] Tumor, syrinx and cysts can be delineated.[31]

The differentiating points between ependymomas and astrocytomas are that ependymomas often are hemorrhagic and typically more sharply delineated than astrocytomas. But there are no specific patterns to reliably differentiate between these two tumors.[10]

Conclusions

Though retrospective in nature, our study described a large series of patients with spinal tumors at a tertiary care center in Nepal. Spinal tumors represent important spinal lesions that can be addressed surgically. With the advent of new generation CT and MR scans, these lesions are detected more frequently. Accurate radiological description of these lesions is essential in order to institute appropriate treatment in a timely manner.

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