Previous literature on spinal tumors reveals that 55% of spinal tumors are extradurally located. 14 90% of extradural spinal tumors are metastases and they are usually from lung, breast, prostate and kidney. 31 10% of extradural spinal tumors are primary and they are neurofibromas, Ewing's sarcoma, angiolipoma, aneurysmal bone cyst. Primary extradural spinal tumors can be benign like neurofibromas, meningioma,
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osteoma, giant cell tumor, vertebral hemangioma, aneurysmal bone cyst, angiolipoma etc or malignant like osteosarcoma, Ewing's sarcoma, chondrosarcoma, plasmacytoma/ multiple myeloma etc. ³

These patients with extradural spinal masses usually present with back pain, radicular pain, motor and sensory deficit, sphincters dysfunction and/or combination of one of them. ¹⁴

Choice of investigation is MRI with Gadolinium enhancement, isotope bone scan and search for primary lesion if there is suspicious of secondary. Treatment ranged from biopsy to total excision of tumor with or without instrumentation (depending on stability of spines) followed by radio and chemotherapy if lesions are malignant.

Results

Out of 124 cases of spinal tumors that we had operated over a period of 12 years, 36 (29.1%) cases turned out to be extradural spinal tumors.

These 36 cases of extradural spinal tumors are the matter of discussion in this study. This is a retrospective study of 36 cases having extradural spinal tumors undergone gross total surgical excision over a period of 12 years, between May 1999 and December 2012 in our institute. There were 20 male and 16 female and age ranged from 10 to 80 years. On admission, 30 patients suffered from back pain/neck pain and 15 patients with radicular pain, 18 patients presented with sensory motor deficit and 15 patients had sphincters dysfunction. Regarding spinal level of involvement, 13 patients had thoracic, 9 had cervical, 3 had cervico thoracic, 7 had lumbar, 2 had thoracolumbar and 2 had sacral extradural masses.

All patients, whether they had benign or malignant lesions, underwent gross total excision followed by instrumentation in 5 patients who had unstable spine after tumor resection (Figure1 A, B; 2 A, B and 3).

Histology revealed 2 metastatic lesions and 34 primary spinal tumors (Table 1). Among primary extradural spinal tumors 22 were benign and 12 were malignant. Primary benign lesions were neurofibromas (16), granulomas (3), angiolipoma (2) and aneurysmal bone cyst (1) (Table 1).

Primary malignant lesions were Ewing's sarcoma (7), chordoma (2), plasmacytoma (1), Rabdomyosarcoma (1) and neuroblastoma (1) (Table 2).

Figure 1: Preoperative CT Scan of lumbosacral spines A) axial and B) reformatted sagittal views depicted huge extradural sacral mass with erosion of surrounding bones

Figure 2: A) and B), Preoperative MRI of lumbosacral spines showing large extradural sacral mass suggestive of Chordoma

Figure 3: Postoperative plain X-Ray of lumbosacral spine after having en block excision of chordoma showed normal alignment of spine with posterior instrumentation.
Two metastatic epidural masses were adeno and renal cell carcinomas (Table 3). 10 patients who harbored primary malignant and 2 metastatic lesions were subjected for a course of radio and chemotherapy. Two patients with chordomas were not sent for radiotherapy believing that these lesions are radio resistant and in fact they both had en block resection. Post operative complication included wound infection in 5 patients, which were treated with a course of systemic antibiotics.

Despite gross total excision, 15 patients came for follow up with features of recurrence of tumors. 3 patients had recurrent neurofibromas which required second surgeries. Other 12 patients had recurrent malignant masses for which they were referred to oncology department for repeated course of radio and chemotherapy.

There was no surgery related mortality, however, we observed 11 deaths during first three years of follow up who had malignant lesions.

Functional outcome was assessed by McCormick’s grading scale. Pre operatively 6 patients had grade I, 12 had grade II, 10 had grade III and 8 had grade IV. After the surgical treatment, 15 patients had grade I, 10 had grade II, 3 had grade III and 8 had grade IV (Table 4).

Nine patients’ neurological status improved to grade I from grade II and III. Neurology of 7 patients improved to grade II from grade III. Neurological status of 8 patients of grade IV remained same after surgical treatment.

<table>
<thead>
<tr>
<th>Pathology</th>
<th>No of cases</th>
<th>Extent of resection</th>
<th>Instrumentation</th>
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<tbody>
<tr>
<td>Neurofibroma</td>
<td>16</td>
<td>total</td>
<td>2</td>
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<tr>
<td>Granulomas</td>
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<td>-</td>
</tr>
<tr>
<td>Angiolipoma</td>
<td>2</td>
<td>Total</td>
<td>-</td>
</tr>
<tr>
<td>Aneurysmal bone cyst</td>
<td>1</td>
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<th>Pathology</th>
<th>No of cases</th>
<th>Extent of resection</th>
<th>Instrumentation</th>
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<tbody>
<tr>
<td>Ewing’s sarcoma</td>
<td>7</td>
<td>Gross total</td>
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<tr>
<td>Chordoma</td>
<td>2</td>
<td>En block</td>
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<tr>
<td>Plasmacytoma</td>
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<td>Rhabdomyosarcoma</td>
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<td>Neuroblastoma</td>
<td>1</td>
<td>Gross total</td>
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<table>
<thead>
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<th>Pathology</th>
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<td>Adenocarcinoma</td>
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<tr>
<td>Renal cell Carcinoma</td>
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<td>Gross total</td>
<td>-</td>
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<th>Preoperative neurology</th>
<th>McCormick’s grade</th>
<th>Postoperative neurology</th>
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<tr>
<td>6</td>
<td>I = normal neurology</td>
<td>15</td>
</tr>
<tr>
<td>12</td>
<td>II = mild to moderate deficit</td>
<td>10</td>
</tr>
<tr>
<td>10</td>
<td>III = moderate to severe deficit, walks with support</td>
<td>3</td>
</tr>
<tr>
<td>8</td>
<td>IV = severe deficit, wheelchair bound</td>
<td>8</td>
</tr>
</tbody>
</table>

Two metastatic epidural masses were adeno and renal cell carcinomas (Table 3). 10 patients who harbored primary malignant and 2 metastatic lesions were subjected for a course of radio and chemotherapy. Two patients with chordomas were not sent for radiotherapy believing that these lesions are radio resistant and in fact they both had en block resection. Post operative complication included wound infection in 5 patients, which were treated with a course of systemic antibiotics.

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Discussion

Spinal tumors are a common cause of back pain, sensory motor deficit and sphincters dysfunction. About 55% of spinal tumors are extradural, 35% intradural extra medullary and about 10% intramedullary.

More than 90% of extradural spinal tumors are metastatic and only 10% are primary extradural spinal tumors. Primary lesions are further divided into benign and malignant. Benign lesions are neurofibromas, meningiomas, osteomas, osteochondromas, chondromas, osteoclastomas, aneurysmal bone cysts, haemangimas, giant cell tumors, angiolipomas and granulomas. Benign tumors such as neurofibromas and meningiomas are generally intradural extramedullary but occasionally have an extradural component or may be limited to the extradural space. Benign extradural tumors are usually present in young adults with constant back pain, scoliosis and less often with neurological deficit.

Malignant primary extradural spinal tumors are multiple myeloma, plasmacytoma, osteosarcoma, chondrosarcoma, Ewing’s sarcoma, rabdomyosarcoma and neuroblastoma. Malignant primary tumors present in middle age with back pain or spinal deformity and neurological deficits are more common.

The spinal column is the most common site of skeletal metastases in patients with cancer. Up to 70% of cancer patients are found to have spinal metastasis on autopsy. Most common primary include breast, lung, prostate, kidney etc. The thoracic, lumbar and cervical spines are commonly affected. Extradural metastases are often reported.

CT scan demonstrates well the degree of bone involvement and also helps with pre operative planning for instrumentation. MRI is preferred imaging modality. MRI demonstrates the extent of disease including paraspinal extensions and neural tissue involvement. Bone scan can detect local and distant bone pathology but has decreased specificity.

Following individual extradural spinal tumors that were commonly encountered in our study are discussed below with review of relevant literature.

Primary benign extradural spinal tumors

Neurofibromas

They are benign tumor. They are both intradural extramedullary in about 70% of cases, and remaining 30% are equally divided between a strictly extradural location and a dumbbell. Half of these lesions are located in thoracic spine and next half in cervical followed by lumbar spines. Common clinical presentations are back pain with or without radiculopathy, progressive neurological deficit and bowel bladder involvement in some cases. MRI is the choice of investigation. Since they are benign lesions total surgical excision should be the aim of treatment. Prognosis has been excellent after the surgical resection, however, some recurrences have been reported in the literature.

Angiolipoma

Spinal angiolipomas are rare benign extradural spinal tumors. They contain mature lipomatous elements and proliferating vessels. They account for only 0.14%-1.2% of the spinal tumors and about 3% of extradural spinal tumors. Angiolipoma is benign and noninfiltrating lesion, however, few cases of infiltrating angiolipomas have been described in literature. These tumors occur in young and middle age and female are mainly affected. Most angiolipomas are located in thoracic level and arise in the posterior extradural space.

Pure lumbar localization is extremely rare.

The histopathogenesis of angiolipoma is unknown. They probably arise from abnormal primitive pluripotential mesenchymal cell that can differentiate into lipomatous, angiomatos or mixed tissue. Some authors suggested angiolipomas to be true hematomas. Patients with angiolipomas most commonly have long standing pain and then develop progressive neurologic symptoms secondary to spinal cord compression. Similarly to other vascular lesions female patients with those lesions may have onset or deterioration during pregnancy.

MRI is the imaging modality of choice in detecting angiolipomas. Angiolipomas appear as hyperintense lesions on T1 weighted images. Gadolinium enhancement is due to the vascularity of these lesions. Gadolinium infusions with fat saturation sequences are useful in the study of those lesions.

These lesions are benign lesions and total excision should be the aim of treatment. Surgical outcome are excellent and there has been no recurrence reported in literature.

Tuberculoma

The spinal column is involved in less than 1% of all cases of tuberculosis. The typical picture of the spinal tuberculosis is of destruction of the vertebral bodies with involvement of the adjacent discs, however, in some instances cord involvement may be secondary to epidural tuberculosis. This epidural tuberculoma, atypical tuberculosis, may present as compression of the spinal cord or cauda equina with features ranging from paresthesia, radiculopathy, neurological deficit and loss of sphincters function. Extradural spinal tuberculomas are difficult to identify on plain radiograph. MRI has been used to diagnose epidural granulomas. Final diagnosis has to be made by histopathological examination.
Aneurysmal Bone Cyst

They are benign, expansile, osteolytic cystic lesions that usually arise in long bones, but up to 30% are seen in spine. They account for nearly 15\% of primary spine tumors. They are commonly located in posterior elements of thoracic and lumbar spine. They are commonly found in young adults with presentation of gradual progressive pain, palpable mass and possibly progressive neurological deficits. CT reveals typical expansile, lytic lesions with a so called eggshell layer of a cortical bone surrounding the lesion. On MRI, they have a heterogeneous appearance on both T\(_1\) weighted and T\(_2\) weighted images, with multiple fluid filled interfaces representing loculations within the lesion. Treatment is gross total excision with instrumentation, if the spine is unstable. Intra-lesional resection with or without post operative radiation is associated with higher recurrence rate.

Primary malignant extradural spinal tumors

Ewing's Sarcoma

This is a small, round, blue cell tumor of unknown origin. Young patients aged 10 to 30 are most commonly affected. It exists in two different clinicopathological entities; osseous Ewing’s Sarcoma and extra osseous Ewing’s Sarcoma. Most common site of occurrence is thoracic followed by cervical and lumbar spines. The main clinical presentation is back pain, but up to 60\% of patients have neurological deficit. MRI is the diagnostic imaging study of choice. Best treatment is aggressive surgical resection followed by radiation and chemotherapy. The local control approaches 100\% and long term survival is 86\%.

Chordomas

These tumors arise from remnant of notochord. They are either found in clivus (40\%) or Sacrum (60\%), peak incidence is 5\(^{th}\) to 6\(^{th}\) decade of life. Clinical presentations are gradual onset of back pain, numbness and motor weakness and feeling of rectal fullness or incontinence. On plain X-ray, chordomas appear as destructive lesions and can have an associated soft tissue mass. CT scan shows both the osseous and soft tissue components of the tumor and is helpful in assessing neural foraminal involvement. MRI reveals enhancements of lesion and has low to intermediate signal intensity on T\(_1\) weighted images and very high signal intensity on T\(_2\) weighted images.

Extradural Spinal Tumor

When possible en block resection should be the aim of surgical treatment and if not possible, marginal excision along with pseudo capsule with postoperative radiotherapy have been advised with mixed results.

Plasmacytoma

Plasmacytomas are B-cell lymphocyte neoplasm and they are malignant. They can affect either vertebral bodies or posterior elements. Plasmacytomas are very rare entity. In about 5\% of patients with multiple myeloma the disease can be diagnosed in the setting of a solitary tumor (plasmacytoma) of the bone. 50\% of plasmacytomas eventually convert into multiple myelomas after some duration. They are commonly found in middle and old age. Clinical presentations are usually local back pain with or without radicular pain and neurological deficit. Plain X-ray and CT scan may be the choice of preliminary investigation, however, MRI would be the best option. Most commonly involved levels are thoracic and cervical spine. Complete resection should be attempted if possible, and spine should be stabilized when indicated. Radiotherapy can be used as adjuvant treatment after surgical resection. Pathological fracture can be seen in some cases of plasmacytoma for which vertebroplasty and kyphoplasty have been tried. Median survival period of patients harboring a solitary plasmacytoma exceeds 60 months.

Neuroblastomas

They are rare and primarily a tumor of infancy and childhood and very rare in adult population. It originates from migrating neural crest cells destined for the adrenal medulla and sympathetic nervous system, thus, primary epidural spinal neuroblastoma is very rare and only few cases have been reported in the literature. The presentations are back pain, radicular pain, and neurological deficit and may be associated with sphincters dysfunction. MRI with Gadolinium enhancement is the choice of investigation. Primary lesions in the other part of the body should be searched before confirming it as a primary lesion. Histology will make the final diagnosis along with immunohistochemistry. Patients with neurological deficit require prompt surgical excision followed by radio and chemotherapy. Despite combined therapies neuroblastoma has still a poor prognosis for survival.

Rhabdomyosarcoma

Primary spinal epidural rhabdomyosarcoma is an extremely rare tumor and only few cases have been reported. It is a highly aggressive and rapidly growing neoplasm of skeletal muscle origin that occasionally appears in the vertebral column and spinal epidural
Metastatic extradural spinal tumors

About 1.3 million new cases are diagnosed annually in USA with about two third of these patients developing metastatic lesions. The most frequently involved organs are lung, liver and bone. The spine is the most common site for skeletal metastasis. The most common primary site for spinal metastasis includes breast, lung, prostate and kidney. The studies have shown that the majority of spinal metastasis involve lumbar followed by thoracic and cervical spines, however, thoracic metastatic lesion become more symptomatic earlier than cervical and lumbar. More than 95% of spinal metastatic lesions are extradural, 5% intradural extramedullary and less than 0.5% intramedullary, rarely encountered.

Their usual clinical presentations are neck pain or back pain, radicular pain, neurological deficit and may have bowel bladder symptoms. A cancer patient with new onset of neck or back pain has spinal metastasis until proven otherwise. MRI has revolutionized the diagnosis of spinal extradural metastatic lesions which require complete examination (T1, T2 and T1 weighted image with gadolinium enhancement).

Treatment of symptomatic spinal metastasis is undertaken to relieve pain and to preserve or restore neurological function. Palliation is the reasonable objective in the vast majority of cases. Surgery for spinal extradural metastatic lesion must provide both decompression of the neural elements as well as stabilization of the spinal column if required. If not medically contraindicated, steroids are recommended for any patient with neurological deficits suspected or confirmed to have extradural spinal metastasis.

Radiotherapy has been the primary therapy for managing metastatic spinal extradural mass, however, surgical decompression followed by spinal instrumentation for stabilization has also proven effective. Although among primary benign extradural spinal tumors osteoma, osteoblastoma, osteochondroma, giant cell tumors, vertebral haemangiomas, aneurysmal bone cyst, neurofibroma, meningioma have been described as primary benign spinal tumors, however, in our series neurofibromas, granulomas, angioliopomas were common benign lesions. Angioliopomas are very rare and only few cases have been reported in literature. Surprisingly, there were two angioliopomas in our series.

In our series, Ewing’s Sarcoma was most common primary malignant extradural spinal pathology followed by chordoma, plasmacytoma, rhabdomyosarcoma and neuroblastoma, where as in literature, osteosarcoma, Ewing’s Sarcoma, chondrosarcoma, plasmacytoma, multiple myeloma were described as common spinal extradural malignant pathologies. Our department, as being a national neurosurgical referral centre, receives many referrals of patients harboring secondaries to spines, but we operated on only two cases. These two patients were young, had single lesion with incomplete neurology and primary lesions were disappeared after treatment. According to protocol of our institute we do not operate on those patients who have multiple secondaries in spine, old age with comorbidities, medically unfit for general anaesthesia, complete loss of neurological function and if primary lesion is still active. We did gross total excision of all tumors whether they are benign, malignant, or metastatic lesions. Five patients required posterior stabilization after the excision of extradural spinal masses, they were chordomas (2), neurofibromas (2) and aneurysmal bone cyst (1). Functional outcome was excellent in those patients who were operated for primary benign lesions.

Three patients, out of 16 patients with extradural metastasis, they were chordomas (2), neurofibromas (2) and aneurysmal bone cyst (1). Functional outcome was not satisfactory in patients with malignant pathology and their neurology remained static despite total resection of lesions. There has been no evidence of recurrence in patients with angioliopomas, granulomas and aneurysmal bone cyst till to date. Three patients with extradural spinal granulomas received 18 months’ course of anti-tuberculosis drugs.

Functional outcome was not satisfactory in patients with malignant pathology and their neurology remained static despite total resection of lesions. There has been no evidence of recurrence in patients with chordomas. All the patients with Ewing’s Sarcoma, rhabdomyosarcoma, and neuroblastoma despite gross total resection followed by radio and chemotherapy, died within 3 years’ followed up due to local recurrence and distant metastases. Two patients having metastasis to spine also died within 20 months of diagnosis and treatment.
Conclusion

Total excision should be the aim of all extradural spinal tumors whether they are benign or malignant. Benign extradural spinal tumors show excellent result after total surgical excision. Malignant lesions, whether primary or secondary, have poor prognosis despite all modalities of treatment.

References


Extradural Spinal Tumor