Functional Outcome of Intradural Extramedullary Spinal Tumors after Surgical Resection

Intradural extramedullary (IDEM) spinal tumors account about 40% of all intraspinal tumors and mainly represented by nerve sheath tumors and meningiomas. These two tumors represent about 55% of IDEM tumors and other rare tumors are dermoid, epidermoid, lipomas, metastatic tumors, paragangliomas etc.

Technical advances in imaging techniques, MRI and surgical procedures have brought about excellent clinical results of IDEM tumors after surgery in last two decades. However, a small percentage of patients still present poor postoperative neurological outcome due to delayed in diagnosis and surgical intervention, severity of preoperative neurological deficits and adverse pathology.

The aim of this study is to analyze and discuss about the surgical outcome of 65 IDEM tumors operated in twelve years’ period.

This is a retrospective study of 65 patients who were operated for IDEM tumors, between 1999 and 2012 in Department of Neurosurgery. One patient who had IDEM arteriovenous malformation was excluded from the study.

Neurological outcome was scaled by McCormick’s grading. Follow up period ranged for 5 years to 17 years.

After the clinical evaluation, all the patients suspected of having spinal tumors were subjected for MRI with Gadolinium enhancement of presumed level of spine based on neurological findings. Total excision of all IDEM tumors was performed using operating microscope. No intraoperative neurophysiological monitoring was used.

There were 40 male and 25 female and age ranged from 10 to 80 years. Most common IDEM tumors were nerve sheath tumors (44), meningiomas (13), hydatid cyst (2), dermoid/epidermoid (2), arachnoid cyst (2) and neurenteric cyst (2). Common levels involved
were thoracic, cervical and lumbar spines. Total tumor excision was performed in all cases. Post operative complications rate was 12.3% (7) and common complications included were CSF Leak (5), wound infection (2), meningitis (1).

There was no surgery related mortality. Postoperatively 60 patients had improved neurological status, 5 patients had stable neurology. There was no postoperative neurological deterioration.

On followed up period 2 patients showed features of recurrence of tumor in 5 years’ period and underwent resurgical treatment. Those two patients with recurrent tumors were nerve sheath tumors.

Majority of IDEM tumors are benign and total cure is possible in almost all cases if tumor is excised totally. Excellent neurological recovery has been observed in more than 95% of cases.

Key Words: functional outcome, intradural extramedullary tumors, spinal tumors, surgical excision

Less than 15% of all CNS tumors are spinal tumors. 55% of spinal tumors are extradural, 40% are IDEM and remaining 5% are intramedullary. Among IDEM tumors, meningiomas and nerve sheath tumors (Schwannomas and Neurofibromas) are most common. Other rare IDEM spinal tumors are lipomas, dermoid, epidermoid, paragangliomas, hemangiopericytomas, metastatic tumors, teratomas, arachnoid cyst and neurenteric cysts.

Before the introduction of MRI, myelography or CT myelography was the choice of investigation to diagnose spinal tumors. MRI gradually replaced the myelography from last 30 years, because MRI was noninvasive and does offer superior soft tissue delineation. First recorded resection of IDEM mass was performed by Sir Victor Horsley in 1888 in a 42 year old patient. The lesion was originally classified as fibromyxoma, but was probably a degenerated schwannoma. Treatment of IDEM tumors is microsurgical excision, however, radiosurgery like Gamma knife, and cyber knife have been tried recently.

Materials and Methods

This is a retrospective study of 65 patients who harbored IDEM spinal tumors and undergone surgical resection between May 1999 and December 2012 in Department of Neurosurgery, National Academy of Medical Sciences, Bir Hospital, Kathmandu, Nepal. Data were retrieved from OT registers, patient’s files and follow up records. MRI was the diagnostic tool in all cases. Total excision of tumors were carried out in all cases via classical laminectomy under operating microscope. Level of the lesions were confirmed by C-arm after positioning the patient on OT table. There were no use of intraoperative neurophysiological monitoring and USG. All patients were subjected for postoperative MRI of spines after three months of surgical intervention to assess the extent of excision or whenever indicated. Preoperative and postoperative neurological status were measured by McCormick grading scale. Patients were followed up in three weeks, three months, six months and each year.

Results

Out of 124 spinal tumors operated in our institute over a period of 12 years, between May 1999 and December 2012, 66 cases were finally diagnosed as IDEM masses. Among 66 cases, 65 cases were tumors and 1 case was AVM. We excluded AVM from this study and total number of IDEM tumors in this study group is 65 which were being studied.

There were 40 male and 25 female and age ranged from 10 to 80 years.
On admission, 41 patients had local pain (neck/back), 39 had radicular pain, 37 had motor deficit, 34 had sensory deficit, 8 had sphincters dysfunction and 6 patients had torticollis and scoliosis (Table 1).

<table>
<thead>
<tr>
<th>Clinical presentations</th>
<th>No.</th>
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<tbody>
<tr>
<td>Local pain (Neck/back)</td>
<td>41</td>
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<tr>
<td>Radicular pain</td>
<td>39</td>
</tr>
<tr>
<td>Motor deficit</td>
<td>37</td>
</tr>
<tr>
<td>Sensory deficit</td>
<td>34</td>
</tr>
<tr>
<td>Sphincters dysfunction</td>
<td>8</td>
</tr>
<tr>
<td>Torticollis/scoliosis</td>
<td>6</td>
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Table 1: Symptomatology of patients with IDEM spinal Tumors

<table>
<thead>
<tr>
<th>Level of Spine</th>
<th>No. of Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cervical</td>
<td>17</td>
</tr>
<tr>
<td>Cervicothoracic</td>
<td>2</td>
</tr>
<tr>
<td>Thoracic</td>
<td>28</td>
</tr>
<tr>
<td>Thoracolumbar</td>
<td>3</td>
</tr>
<tr>
<td>Lumbar</td>
<td>16</td>
</tr>
<tr>
<td>Total</td>
<td>65</td>
</tr>
</tbody>
</table>

Table 2: Level of IDEM spinal tumors

<table>
<thead>
<tr>
<th>Types of tumors</th>
<th>No. of Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nerve Sheath tumors (Schwannomas, Neurofibromas)</td>
<td>44</td>
</tr>
<tr>
<td>Meningiomas</td>
<td>13</td>
</tr>
<tr>
<td>Neurenteric Cyst</td>
<td>2</td>
</tr>
<tr>
<td>Arachnoid Cyst</td>
<td>2</td>
</tr>
<tr>
<td>Dermoid epidermoid</td>
<td>2</td>
</tr>
<tr>
<td>Hydatid Cyst</td>
<td>2</td>
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<tr>
<td>Total</td>
<td>65</td>
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</table>

Table 3: Types of IDEM spinal tumors

<table>
<thead>
<tr>
<th>McCormick’s grade</th>
<th>Preop neurological status of patients</th>
<th>Postop neurological status of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>38</td>
<td>60</td>
</tr>
<tr>
<td>II</td>
<td>9</td>
<td>4</td>
</tr>
<tr>
<td>III</td>
<td>10</td>
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<tr>
<td>IV</td>
<td>7</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>65</td>
<td>65</td>
</tr>
</tbody>
</table>

Table 4: Functional neurological outcome (McCormick’s grading)

All 65 patients suspected of having spinal lesions on clinical ground underwent MRI of spines with Gadolinium enhancement (Figure 1A, B). 17 patients had IDEM mass at cervical level, 2 at cervicothoracic level, 28 at thoracic level, 3 at thoracolumbar level, and 16 at lumbar level and there was no mass at Sacral level (Table 2).

Of 65, 63 patients had total tumor excision and 2 patients had gross total excision. Surgical resection was performed using operating microscope with classical laminectomy (Figure 2A & B).

Among 65 patients who had histologically proven IDEM tumors, 44 (67.6%) were nerve sheath tumors (neurofibromas and schwannomas), 13 (20%) meningioma, 2 (3%) neurenteric cysts, 2(3%) arachnoid cyst, 2 (3%) dermoid/epidermoid and 2 (3%) Hydatid cysts (Table 3). The recurrence occurred in 2 (4.2%) patients and they were neurofibromas which required second surgery. No patient received postoperative radio and chemotherapy. Functional neurological outcome was assessed in term of McCormick’s grading scale. All patients were subjected for MRI of spines after 3 months postoperatively to observe the extent of excision of tumors (Figure 1B).

Before surgery 38 patients (58.5%) had McCormick grade I, 9(13.8%) had grade II, 10 (15.4%) had grade III and 8 (12.3%) had grade IV. On followed up period, 60 (92.3%) improved to grade I, 4(6.1%) to grade II and 1(3%) had grade III and IV (Table 4). Functional outcome of IDEM tumors after total surgical resection is excellent. More than 95% of patients, in our
series, had achieved near to normal neurological function after surgical intervention.

Discussion

First recorded resection of IDEM spinal tumors was carried out by Sir Victor Horsley in 1888 in a 42 year old patient. The lesion was originally classified as fibromyxoma, but was probably a degenerated Schwannoma.

The most common primary intradural extramedullary (IDEM) tumors are nerve sheath tumors (Schwannomas and Neurofibromas) arising from sheath cells of spinal nerve roots and meningioma from arachnoid cap cells. Lipoma and dermoid/epidermoid are less common, hemangiopericytomas, paragangliomas and metastatic lesions are very rare.

These patients with IDEM tumors mostly present with features of spinal cord and or root compression. Local pain and or radicular pain are common presenting feature. Paresthesia and numbness are other common presentation. Motor and sensory deficit are other common neurological findings. Motor deficit ranged from group of muscles weakness to mono, hemi, para and quadriplegia. Sphincters dysfunction can develop earlier if there is involvement of Cauda equina. Local examination of spines may reveal spinal deformities like torticollis, scoliosis, kyphosis or loss of lordosis. Skin stigmata like dermal sinus, tuft of hairs, lipoma, hyperpigmentation or portwine stain can be found if there is underlying lipomas or epidermoid/dermoid.

While some authors reported that pedicle erosion, vertebral body erosion, neural foraminal widening, and scoliosis were found on plain radiographs of spines in about 38-56% of patients with IDEM Spinal tumors. So, plain x-rays of spines may provide us some clues about the underlying pathology but MRI is best imaging modality with regard to this tumor in assessing the size, shape and in anatomical relations with adjacent structures, particularly with dura matter and spinal cord allowing best treatment guidelines and surgical approaches.

Nerve sheath tumors are isointense lesion on T1W1 and a hypointense lesion on T2W2 images. Contrast enhancement is a common finding. On MRI, spinal IDEM meningiomas have a hypo to isointense signal on T1W1 and hyperintense signal on T2W2 images. Gadolinium contrast gives strong homogenous enhancement and offers dural tail appearance on dural attachment.

MRI findings of dermoid and epidermoid are variable, often, the distinction between these two tumors by imaging are difficult. MRI shows hypo to hyperintense signal on T1W1 images and iso to hyperintense signal on T2W2 images. These are usually minimal enhancement with contrast.

Microscopic total excision while preserving and improving neurologic function is the usual goal of surgery. Intraoperative neurophysiological monitoring, if available, is helpful to reduce the postoperative morbidity. Intraoperative use of C-arm is very useful to precise localization of lesion. Some neurosurgeons are also using intraoperative USG to localize tumors. The operative approach is planned based on clinical, neurologic and radiological evaluation. Approaches are based on location, size and extension of tumors and other parameters.
Excessive removal of bony structures and ligaments may result in spinal instability. While performing laminectomies care should be taken to preserve facets, its capsules and intertransverse ligaments to avoid post operative spinal instability and deformity. Sometimes only hemilaminectomy may suffice for IDEM tumor resection.4,5,18,26,29

Surgical results of IDEM tumors are excellent and even long lasting preoperative neurologic deficit may be improved and reversed postoperatively. Although surgical excision remains primary treatment option for most of IDEM tumors, radiosurgery offers an alternative therapeutic modality, especially for recurrent and residual lesions or when surgery is contraindicated.8,11,17 Previous studies have demonstrated to have short term clinical benefit for IDEM lesions, however, long term efficacy is not known.11,17

Most common postoperative complications after IDEM tumor resection are CSF leak, pseudomeningocele and wound infection.1,3,18,31 Less common complications are spinal instability and neurological deficit.

Recurrence of IDEM tumors after total excision is common and depends on extent of surgical excision, type and histological character of lesions. Recurrence for radically resected nerve sheath spinal tumors was reported to be 10 to 28% 2,3,10,18 after 5 to 15 years respectively where as recurrence of meningiomas is about 3-7%.3,18,31

Some of IDEM spinal tumors are summarized as follows:

**Nerve sheath tumors**

a) Schwannomas: They are benign, slow growing, encapsulated tumors arising from the peripheral nerves (mainly sensory). They are the most common spinal IDEM tumors arising in posterolateral location of spines. They are more common in lumbar and cervical level than in thoracic level. They are common in adult than in children and equally occur in male and female. Peak incidence is in forth to fifth decade of life. Occasionally multiple spinal schwannomas are seen with Neurofibromatosis type II. Histologically schwannomas comprise spindle-shaped Schwann cells arranged in either compact occasionally pellisading cells pattern (Antoni A) or loosely organized hypocellular areas (Antoni B) pattern.18,31

The main treatment of schwannomas is total surgical excision and sometimes subtotal resection might be an option if the tumor is attached to vital structures like spinal cord or vertebral arteries. During resection one or two ventral or dorsal roots are usually sacrificed, however, division of these nerve roots do not produce profound postoperative neurological deficit. Radio or chemotherapy is usually reserved for those tumors that have malignant characters. Tumor recurrence is less than 5% and might have a high association with subtotal tumor removal.

b) Neurofibromas: These are indolent, slow growing tumors arising from ventral roots, occurring either sporadically or within the setting of Neurofibromatosis type I. 3,4,18,31 Neurofibromas are less common than schwannomas. Histologically it comprises not only Schwann cells but also fibroblast, axons, mast cells, collagen and perineural-like cells. Malignant neurofibromas can occur but rare and more than 50% of them are associated with neurofibromatosis type I. Total surgical removal of neurofibromas is usually curative.

**Meningiomas**

They represent 25-45% of all IDEM tumors.12,23,25 These tumors are more common in women with middle and old age. Thoracic level is the most common location. Most of these tumors are benign and slow growing arising from arachnoid cap cells. There is a wide spectrum of histological varieties i.e. meningothelial, psammatous, fibrous and transitional. The aim is total excision which can be attainable in more than 90% of cases. The recurrence rate is 3-7%. Radiotherapy could be considered after subtotal resection or recurrence of these tumors.

**Dermoid/epidermoid**

These are congenital tumors arising from heterotopic ectodermal-cell implantation into the neural tube early in embryogenic development.18,31 They are rare spinal pathology. They are commonly located in Lumbosacral region and rarely in thoracic level. They usually become symptomatic in first two decades of life. MRI picture of both these lesions are variable, often, the distinction between those tumors by imaging are difficult. MRI shows hypo to hyperintense signal on T1W1 images and iso to hyperintense signal on T2W2 images. There is usually minimal enhancement with contrast. Although total resection of the tumor should always be a goal, but often, adhesion of tumor capsule with neural tissues can prevent aggressive approach. Recurrence of these tumors is rare and malignant transformation is also rare.

**Lipomas**

Lipomas are congenital, benign and uncommon IDEM tumors. They are usually associated with spinal dysraphism in about one third of cases and common in children. Mostly located in thoracic and lumbosacral regions. On MRI, Lipomas have a hyperintensity signal on T1W1 images and hypointensity on T2W2 images. Asymptomatic Lipomas should be left alone. Surgical resection should be recommended for symptomatic one and subtotal excision is usually undertaken to prevent postoperative neurological deficit. Intraoperative neurophysiological
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monitoring is often helpful to differentiate between mass and spinal cord or neural elements. 

Metastatic tumors

IDEM spinal metastatic lesions are extremely rare. These tumors are usually the result of drop metastasis like adenocarcinoma, melanoma, lymphoma or intracranial neoplasms like ependymoma, medulloblastoma and glioma. They are usually found at thoracic levels. These lesions are usually a signal of advanced widespread progression of systemic malignancy, so aggressive resection of these lesions is often declined. Treatment is always palliative to relieve symptoms.

Paragangliomas

These are uncommon IDEM lesions originated from automatic nervous system paraganglion cells. They are usually non-secreting, sympathetic tumors usually located at thoracolumbar region and predominantly occur in male. On MRI those tumors present as a well circumscribed mass with a hypointense to isointense signal on T1W1 images and a hyperintense on T2W2 images. And after administration of contrast they look like heterogenous salt and pepper pattern of enhancement.

Histological pictures display highly vascularized tumor bed containing round and polygonal cells grouped in clusters called “Zellballen”. Aim of treatment should be total excision and recurrence rate is less than 5%. Preoperative screening for a hyper adrenergic state is necessary to prevent hypertensive crisis during tumor removal, though catecholamine secreting spinal paragangliomas are rare.

Neurenteric cyst

Neurenteric cysts account for 0.7% to 1.3% of spinal axis tumors. This tumor is also called endodermal cyst. These lesions result from the inappropriate partitioning of the embryonic notochordal plate and presumptive endoderm during the third week of human development. Heterotopic nests of epithelium reminiscent of gastrointestinal and respiratory tissue lead to eventual formation of compressive cystic lesions of the paediatric and adult spine. Approximately 90% of neurenteric cysts are located in IDEM compartment. While the remaining 10% are divided between extradural and intramedullary. On MRI, neurenteric cysts are isointense on T1W1 sequence and hyperintense on T2W2 sequence and there is no uptake of contrast. This cyst usually occurs in the 2nd and 3rd decade of life with male female ratio of 2:1. Majority of neurenteric cysts are located ventral to the spinal cord and found at thoracic and lumbar regions. Surgical resection is the first line of treatment with the goal of gross total excision. Appropriate surgical approach depends on location of cysts. Reports of recurrence have ranged from 0 to 37%.

Hemangiopericytoma (HPS)

Primary intradural extramedullary HPC are very rare and only few cases have been reported in literature. HPC arises from pericytes. Most of the spinal HPC are extradural and intramedullary HPC has never been reported. These tumors are very vascular, high recurrence rate and can be metastase to other parts of the body. On MRI, they are isointense signal on T1W1 and hyperintense on T2W2 signals after the contrast injection they are brilliantly enhanced. Treatment of choice is gross total excision followed by local radiation. Despite these two modalities of treatment, prognosis of disease remains same. The recurrence rate is more than 50% in one year.

Teratomas

Spinal IDEM teratomas are very rare. More common in children than in adults and often associated with spinal dysraphism. They are usually located at thoracolumbar level. Histological pictures showed all three germinal layers i.e ectoderm, mesoderm and endoderm. Primary treatment modality for symptomatic patients is total surgical resection, however, adhesions of wall of teratoma to the surrounding neural parenchyma can make difficult to resect totally. Teratomas are classified as mature, immature and malignant and prognosis depends on its histological character.

The results and functional outcome of IDEM tumors after surgical excision in our series are similar to the results of other published series except some dissimilarities which are being discussed here. Nerve sheath tumors and meningiomas were common IDEM tumors in our series which is same as of other series, however, other rare tumors like hemangiopericytomas, lipomas, paragangliomas and metastatic lesions which were described in other previous series were not found in our series. We do often receive IDEM spinal metastatic lesions but most of the time we decline to operate on those patients who come to us in a terminal stage due to advancement of primary malignancy. We had additional rare two IDEM masses like hydatid cyst and arachnoid cyst in our series which were not mentioned in previous published series.

These days although there is increasing trend of performing hemilaminectomy for the excision of IDEM masses believing that this procedure is less invasive and prevents instability of spines. In our center we routinely do standard laminectomy to resect IDEM tumors and have not found a single patient having unstable spines on regular follow up of 5 to 17 years. Published.
series also did not reveal any difference between the outcome of patients who underwent hemilaminectomy and laminectomy for IDEM tumor excision. Postoperative complication rate in our series was 12% which is similar to published series. More than 95% of our patients had improved neurological status after surgical excision and this functional outcome is similar with the outcome of published international series. Regarding recurrence of tumor we had two patients who were reoperated for recurrent neurofibromas in five years' period. So recurrence rate in our series for neurofibromas is 4.5% which is comparable with other series.

We did not recommend radiotherapy and chemotherapy to our any patient although there is recent trend to advocate radiosurgery for patients with subtotal excision and recurrent tumors.

Conclusion

Majority of IDEM tumors are benign and total cure is possible if excised totally. Excellent neurological recoveries have been observed after surgical excision.

References

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