INTRODUCTION

The incidence of spinal schwannomas varies between 0.3-0.4 cases /100,000 persons per year (9). Spinal schwannoma constitutes approximately 25% of the intradural tumours (18). Intramedullary schwannomas constitute approximately 1.1% of all the spinal schwannoma and 0.3% of all intramedullary tumors (6,12). They are generally associated with neurofibromatosis types 1 and 2 (6). Intramedullary spinal schwannomas most commonly occur in the cervical spine (61%) and are less common in the thoracic (29%) and lumbar (10%) spine (2). In particular, intramedullary schwannomas in the conus medullaris are extremely rare when no relationship of schwannoma with neurofibromatosis is present. Early symptoms of these tumours are non-specific. They are slow progressing in nature and the duration of symptoms usually ranges from 3 to 4 years. Low back pain may be the only apparent symptom during this time (11). They are usually benign, but can be locally aggressive and cause severe neurological compromise. We present a case of intramedullary schwannoma of conus medullaris with related review of literature.

CASE REPORT

A 40-year-old female presented with complaint of diminished sensation, tingling & numbness in both lower limbs since one & half years duration. No history of any trauma or previous disorders such as neurofibromatosis or specific lesion on her skin were present. Neurological examination revealed diminished sensations of all modality below L1 dermatomes and weakness in B/L ankle & EHL dorsiflexion (power grade 3/5). Patient had no involvement of bladder or bowel.

Magnetic resonance imaging (MRI) demonstrated a well-defined expansile intramedullary lesion involving the conus medullaris, iso to hypointense on T1W image (Figure 1A,B).

The operation was done with standard posterior midline approach. Following skin incision and muscle dissection D10 to L1 laminectomy were performed. Dura & arachnoid were opened and D11 to D12 intramedullary lesion were dissected off the spinal cord tissue and excised (Figure 2,3). Frozen section findings were defined as schwannoma. After the operation patient was discharged on postoperative day 6th having 5/5 power in all four limb joints except bilateral ankle & EHL dorsiflexion having power grade 3/5 and improved sensory modality below L1. A month after operation in follow-up patient had improvement in sensation and motor weakness at B/L ankle and EHL. Histological analysis confirmed the diagnosis of schwannoma (Figure ABSTRACT

Intramedullary schwannoma unrelated with neurofibromatosis are uncommon. The conus medullaris level is unusual for schwannomas. Till date only eight cases of intramedullary schwannoma of conus medullaris without neurofibromatosis have been reported in the available English literature. Here, we are reporting a case of intramedullary tumor that turned out as schwannoma on histopathology. Total resection was done and histological analysis confirmed as a schwannoma. The patient recovered after surgery and shows improvement in follow up. This case report of a very rare tumour is presented with review of the literature.

KEY WORDS: Conus medullaris, intramedullary tumour, non neurofibromatosis, schwannoma
Intramedullary Schwannoma of Conus Medullaris: Case Report and Review of the Literature

Intramedullary lesion in 1932 (19). These tumors correspond to 30% of spinal tumors. According to the new WHO classification of tumors there are three types of schwannomas: cellular, plexiform and melanotic. Intramedullary (IM) schwannomas are rare tumors, accounting for 0.3-1% of intraspinal neoplasms and 1.1% of spinal schwannomas (3,12). Intramedullary spinal schwannomas most commonly occur in the cervical spine (61%) and are less common in the thoracic (29%) and lumbar (10%) spine (2). Till date only eight cases of intramedullary schwannoma of conus medullaris without neurofibromatosis have been reported in available English literature (Table 1). Their rarity may

Figure 1: Magnetic resonance image (A) sagittal and (B) axial image showing intramedullary mass like lesion on D11-D12 level.

Figure 2: Intraoperative picture showing gross enlargement of the conus; note the filum origin from distal end of enlargement (arrow) and other nerve roots around it.

4A,B). Immunocytochemical staining for S100 protein was strongly positive.

DISCUSSION

Spinal schwannomas are tumors originating from the Schwann cells thus, their location is usually intradural extramedullary and/or extradural (18). Penfield was the first to describe characteristics of schwannomas in an intramedullary lesion in 1932 (19). These tumors correspond to 30% of spinal tumors. According to the new WHO classification of tumors there are three types of schwannomas: cellular, plexiform and melanotic. Intramedullary (IM) schwannomas are rare tumors, accounting for 0.3-1% of intraspinal neoplasms and 1.1% of spinal schwannomas (3,12). Intramedullary spinal schwannomas most commonly occur in the cervical spine (61%) and are less common in the thoracic (29%) and lumbar (10%) spine (2).
be explained by the lack of Schwann cells in the spinal cord (6,17).

Presence of intramedullary schwannomas is a paradox in itself as there are no Schwann cells in the central nervous system hence the controversy about its pathogenesis. Several theories have been suggested. Kernohan et al proposed that the origin of such lesions could be derived from schwann cells in nerve fibers of spinal arteries. Ramamurthi et al (20) suggested that a few ectopic Schwann cells of the embryonal neural tube could be the source of origin. Focal intra-medullary proliferation of Schwann cells in reaction to chronic disease or trauma was also suggested (21). Rusell et al (21) suggested that these tumors emerge from the transformation of neuroectodermal pial cells into Schwann cells. A tumor arising from Schwann cells in this “critical area,” where the nerve root loses its sheath, could enter the subpial area in the spinal cord and appear as an intramedullary mass.

An extensive literature review by Conti et al, (5) found only about 50 reported cases of intramedullary schwannomas unrelated to neurofibromatosis. Most of these lesions had been reported as single intramedullary lesions, and more rarely as dumbbell intra- and extramedullary ones. The melanotic schwannomas are even rare with only 39 cases reported of which 5 were intramedullary lesion.

The male female ratio for intramedullary schwannomas is 3:1 with a mean age of 40 years. Intramedullary schwannomas are mostly located in the cervical spinal cord (51%), followed by the thoracic region (22%) (3,12). They have a slow growth pattern and because of this, the average interval between first symptoms and diagnosis ranges from six months to 20 years (23).

The most common initial symptom is pain which may proceed to motor or sensory loss and even loss of genitourinary function (17).

The X-ray findings are typical of intramedullary tumours. Magnetic resonance imaging (MRI) is the gold standard to study intramedullary tumours. On MRI, sagittal and axial images demonstrate a widening of the spinal cord. These tumors are well delineated with moderate perilesional edema (5) and no syringomyelia. These tumors are hypointense or isointense on T1-weighted sequences and generally hyperintense on T2-weighted sequences. When gadolinium is injected there is a heterogeneous enhancement. It also enhances the thickened spinal nerve root in continuity with the tumor which helps to differentiate these tumors from other intramedullary lesions (3) and also allows better differentiation from associated edema (3,12).

It has been debated whether or not MRI can or cannot allow preoperative differentiation of histological differentiation of intramedullary tumours. Takemoto et al (23) suggested that MRI allows pre-operative diagnosis of schwannomas, neurofibromas, meningiomas and hemangioblastomas whereas Nicoletti et al (16) claimed neither the MRI nor CT scan can differentiate the intramedullary tumour histological type. The Antoni A-type is characterized by the presence of compact wave-shaped cells surrounded by a reticular net. The Antoni B-type has large and loose cells surrounded by a collagenous web. Demachi et al found no correlation between Antoni A and B fibres and the MRI findings (7).
Intramedullary Schwannoma of Conus Medullaris: Case Report and Review of the Literature

Complete surgical excision is the treatment of choice for spinal schwannomas, including intramedullary ones. Gross total resection is the goal because of the benign nature of this tumour but sometimes this cannot be accomplished due to the infiltrative character of the tumor (6,17). The relative rarity of schwannomas in intramedullary location and dominance of gliomas in this location binds the surgeons not to attempt complete surgical removal. Therein lies the importance of preoperative and intraoperative diagnosis (5,12). The infiltrative pattern of some intra-medullary schwannomas make total gross resection impossible and some authors suggest in these cases the use of radiotherapy for residual lesions.

**CONCLUSION**

Intramedullary schwannoma of conus medullaris is extremely rare without neurofibromatosis. They are slow growing and usually benign in nature. Complete surgical excision is considered in noninfiltrative tumor. An early diagnosis and gross total resection of tumor accomplishes better prognosis and outcome for the patients.

**REFERENCES**


**Table 1:** Summary of intramedulary schwanomma in conus medullarlis in non neurofibromatosis patients in literature

<table>
<thead>
<tr>
<th>Author / year</th>
<th>Age/ Sex</th>
<th>Involved segment</th>
<th>Symptoms</th>
<th>Duration</th>
<th>Treatment</th>
<th>Results</th>
</tr>
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<tbody>
<tr>
<td>Guidetti, 1969(8)</td>
<td>NA</td>
<td>Conus</td>
<td>NA</td>
<td>NA</td>
<td>Total resection</td>
<td>Tr</td>
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<td>Mc Cormik, 1969(14)</td>
<td>M</td>
<td>L2</td>
<td>NA</td>
<td>6 weeks</td>
<td>Autopsy finding</td>
<td></td>
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<tr>
<td>Schmitt, 1975(12)</td>
<td>M</td>
<td>Conus</td>
<td>NA</td>
<td>6 weeks</td>
<td>Autopsy finding</td>
<td></td>
</tr>
<tr>
<td>Lesoin et al, 1983(13)</td>
<td>M</td>
<td>Conus</td>
<td>NA</td>
<td>5 years</td>
<td>Total resection</td>
<td>Tr</td>
</tr>
<tr>
<td>Gokmen Kahilogulleri et al, 2005(10)</td>
<td>38y/F</td>
<td>Conus</td>
<td>Pain &amp; numbness</td>
<td>1 year</td>
<td>Subtotal resection</td>
<td>NA</td>
</tr>
<tr>
<td>H Mouchaty et al, 2008(15)</td>
<td>56y/F</td>
<td>T12-L1 (conus)</td>
<td>NA</td>
<td>6 weeks</td>
<td>Total resection</td>
<td>Tr</td>
</tr>
<tr>
<td>T.Ohttonari et al, 2009(18)</td>
<td>29y/M</td>
<td>Conus</td>
<td>NA</td>
<td>8 months</td>
<td>Subtotal resection</td>
<td>Tr</td>
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<td>Suat Canbay et al, 2011(4)</td>
<td>F</td>
<td>Conus</td>
<td>Diminished sensation, tingling &amp; numbness</td>
<td>1.5-2 years</td>
<td>Total resection</td>
<td>Tr</td>
</tr>
<tr>
<td>Current case</td>
<td>F</td>
<td>Conus</td>
<td>Diminished sensation, tingling &amp; numbness</td>
<td>1.5-2 years</td>
<td>Total resection</td>
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