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ORIGINAL ARTICLE

# Surgical Outcome of Intradural Extramedullary Tumors: Single Institutional Experience—Assessment using Frankel Grading

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## ABSTRACT

**Background:** To report the surgical outcome of intradural extramedullary (IDEM) tumors in 52 patients. Clinical effect was evaluated based on Frankel grade.

**Purpose:** The results of a single neurosurgery institution are reported to highlight the peculiarities and pitfalls of the management of this disease. Tumors are analyzed from the point of view of their localization, histology, surgical technique and outcome was evaluated in term of Frankle classification.

**Methods:** Fifty-two cases of histopathologically confirmed IDEM were treated laminectomy with complete resection of tumors between January 2011 and February 2014. There were 34 males and 18 females with an average age of 41.53 years. The mean postoperative follow-up period was 22.44 months. The histopathological findings, locations of the tumors, and clinical results were analyzed. The neurological findings obtained during the preoperative stage and the postoperative follow-up was evaluated according to the Frankel grading.

**Results:** The histopathological results are as follows: 14 cases of a meningioma, 30 cases of nerve sheath tumors (28 cases schwannoma and 2 cases neurofibroma), 4 cases of an arachnoid cyst, 3 cases of Tarlov cyst, and one case of benign cystic teratoma. The locations of the tumors were as follows: 40 cases in the thoracic region, eight cases in the cervical region, and two cases in the lumbar and two cases of sacral region. The most common diagnosis was nerve sheath tumors (57.69%), followed by meningioma (26.92%). At the final follow-up, a 3-grade, 2-grade and 1-grade improvement was observed in 6, 22 and 24 cases, respectively. There were changes in the Frankel grade in every case. The preoperative neurological deficit improved within 6 postoperative weeks in most cases. Postoperatively, there were one case of cerebrospinal fluid leakage.

**Conclusion:** Intradural extramedullary tumors detected by magnetic resonance imaging (MRI) are mostly benign and good clinical results can be obtained when treated surgically. Therefore, more dynamic surgical approaches by neurosurgeons are suggested to decline morbidity.


**Keywords:** Extramedullary, Frankel grading, Intradural, Tumor.

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## INTRODUCTION

Intradural extramedullary (IDEM) tumors account for two-thirds of all primary intraspinal neoplasm and these include schwannomas, meningiomas and ependymomas.[1] While the clinical symptoms and pathology of IDEM tumors have not changed, there have been dramatic improvements in the diagnosis and treatment with the advances of the radiological and surgical techniques. Particularly, mylography, computed tomography (CT) and magnetic resonance imaging (MRI) allow making an easy diagnosis and accurate identification of the location of a tumor in the dura mater and its dural attachment before surgery, and a significantly improved surgical outcome can be expected with the use of an operating microscope[1-3] and new microsurgical techniques. However, given the high sensitivity of the radiological techniques, such as MRI, care should be taken not to overlook a tumor in the upper spine when observing radiographic evidence of nerve root compression in the lower lumbar region. In this study, we assessed the IDEM tumors that we surgically managed with regard to the pathological diagnosis, the preoperative medical history and the clinical symptoms, the surgical treatment outcome and recurrence, the prognostic factors and the reasons for a misdiagnosis. More aggressive treatment approaches aimed at preserving and further enhancing the neurological function have been developed over the past 30 years. In addition, great strides have been made recently in the prognosis of surgery due to the development of diagnostic tools, such as CT and MRI, an understanding of the precise anatomical structures, and the advancement of surgical instrumentation and techniques. Laminectomy is the traditional approach used to excise IDEM tumors; however, the stability of the spine is often affected, due to impairment of the posterior column. This study examined 52 patients who were followed for more than 1 year after the diagnosis and underwent a

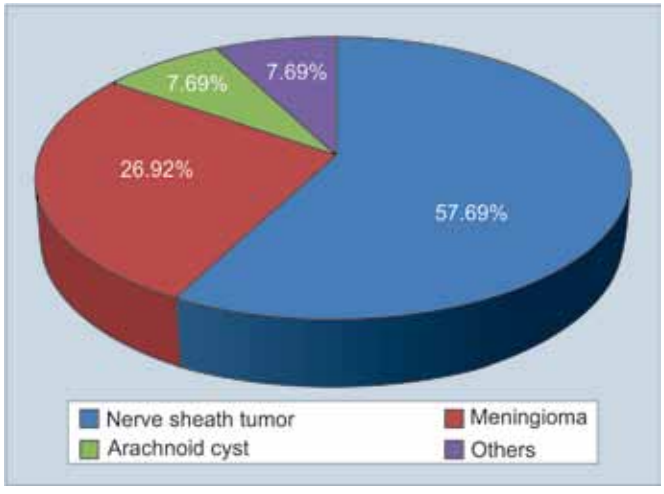
**Table 1:** Frankel grading system

| *Grade* | *Description* |
|---|---|
| A | Complete loss of motor and sensory function below the level of lesion |
| B | Complete motor paralysis with some sensory preservation (e.g. Sacral sparing) |
| C | Retain motor function but useless |
| D | Useful motor function |
| E | Free from neurological symptoms |

laminectomy of the IDEM tumor and clinical outcome evaluated based on Frankel grading (Table 1).

## MATERIALS AND METHODS

Patients treated surgically for IDEM tumors between January 2011 and February 2014 at our hospital, 52 patients who showed histopathological evidence and were available for a more than a 1-year follow-up were enrolled in this study. The mean age of the patients was 41.53 years (12–60 years). There were 34 males and 18 females. The mean postoperative follow-up period was 22.44 months (12–36 months). The histopathological findings, locations of the tumor, clinical symptoms, durations of symptoms, and radiological findings were analyzed and MRI was performed in all cases. All patients received general anesthesia and were placed in the prone position. Syringe needle markers were inserted into spinal processes under the fluoroscope of the C-arm X-ray machine to assure accurate positioning. A posterior midline longitudinal incision was made and the subcutaneous tissues and lumbodorsal fascia was divided. The supraspinal ligaments, interspinal ligaments and tendinous insertions of muscles exposed bilaterally, regardless of position of tumor and complete laminectomy was performed. Ligamentum flavum was removed and the dural sac was exposed. The oblique tilting of the operating table to the contralateral side ensured an adequate surgical field for the procedure. A microscope was positioned to begin microscopic complete excision of the IDEM tumors. A complete excision was made, when a nerve fiber was attached to a tumor, a nerve stimulator was used to determine if it was a sensory or motor nerve branch. The sensory nerve branch was removed while the motor nerve branch was preserved through careful detachment and then finished the removal of the IDEM tumors between the surrounding normal structures and the tumor. Bipolar coagulation was used to maintain hemostasis, and the dura mater was tightly sutured with 5-0 lines using continuous mattress suture, covered with an absorbable gelatin sponge. The postoperative evaluation involved a comparison of the neurological findings were graded according the Frankel classification preoperatively and at the last follow-up.




**Graph 1:** Percentage of IDEM

## RESULTS

The pathological diagnoses included 30 cases of nerve sheath tumors (57.69%), 14 cases of meningioma (26.9%) and four cases of Arachnoid cyst (7.6%) and four cases of others (Graph 1). There was one case of recurrence of schwannomas (Tables 2 and 3) (Figs 1 to 4).

As observed on the sagittal plane images, eight tumor was situated in the cervical spine (15.38%), 40 in the thoracic spine (76.92%), two in the lumbar spine (3.8%), two in sacral spine (3.8%). With look upon to the relative location all were located dorsal to the spinal cord.

### Symptoms

The mean duration of symptoms was 13.8 months (1–120 months) and the mean postoperative follow-up period was 22.4 months (12–36 months). Lower limb pain and numbness, which were observed in 46 patients 88.46% cases. The symptoms tended to augmented during walking relatively than during bed rest and sitting. Motor weakness was also observed in 50 patients (96.15%), Sensory disturbance in 36 patients (69.23%), and sphincter involvement in 18 patients (34.61%) (Table 3). All the cases' symptoms better postoperatively.

The preoperative Frankel grade was B in 14 cases, C in 18 cases, D in 20 cases. During the postoperative follow-up period, six patients showed a 3-grade improvement (11.53%) and 22 patients showed 2-grade improvement (42.30%) and 24 patients showed 1-grade improvement (46.15%) (Table 3). The mean intraoperative bleeding volume of Laminectomy was 287 ml (145–450 ml) and the mean surgery time was 118 minutes (85–185 min).

### Complications

Duraplasty using a fat with fibrin glue was performed in one case with a pseudomeningocele as a complication at

**Table 2:** Demographical data of all patients

| | *All type of IDEM* | *Nerve sheath tumor* | *Meningioma* | *Arachnoid cyst* | *Others* |
|---|---|---|---|---|---|
| Total no. (%) | 52 | 30 (57.69%) | 14 (26.92%) | 4 (7.69%) | 4 (7.69%) |
| Male-female ratio | 34/18 (1.88) | 24/6 ( 4) | 4/10 (0.4) | 3/1 (3) | 3/1 (3) |
| Mean age | 41.53 | 43.06 | 44.57 | 27.50 | 33.50 |
| *Age groups (year)* | | | | | |
| Children( 0–18) | 6 (11.53%) | 4 (13.33%) | 0 (0%) | 2 (50%) | 2 (50%) |
| Adults ( 19–44) | 20 (40%) | 12 (40%) | 6 (42.85%) | 0 (0%) | 2 (50%) |
| Seniors (>44) | 26 (50%) | 14 (46.66%) | 8 (57.14%) | 2 (50%) | 0 (0%) |
| *Tumor location* | | | | | |
| Cervical | 8 (15.38%) | 7 (23.33%) | 0 (0%) | 0 (0%) | 1 (25%) |
| thoracic | | 18 (60%) | 14 (100%) | 2 (50%) | 0 (0%) |
| Lumbar | 40 (76.92%) | 2 (6.66%) | 0 (0%) | 2 (50%) | 2 (50%) |
| sacral | 2 (3.84%)<br>2 (3.84%) | 2 (6.66%) | 0 (0%) | 0 (0%) | 1 (25%) |
| Tumor situation | All situated to post. to spinal cord | Post. to spinal cord | Post. to spinal cord | Post. to spinal cord | Post. to spinal cord |
| *Symptoms* | | | | | |
| Radicular/Local pain | 46 (88.46%) | 26 (86.66%) | 13 (92.85%) | 4 (100%) | 3 (75%) |
| Muscle weakness | 50 (96.15%) | 30 (100%) | 12 (85.71%) | 4 (100%) | 4 (100%) |
| Sensory dysfunction | 36 (69.23%) | 24 (80%) | 8 (57.14%) | 3 (75%) | 1 (25%) |
| Sphincter dysfunction | 18 (34.61%) | 8 (26.66%) | 6 (42.85%) | 2 (50%) | 2 (50%) |
| Treatment | Laminectomy (LE) | LE | LE | LE | LE |

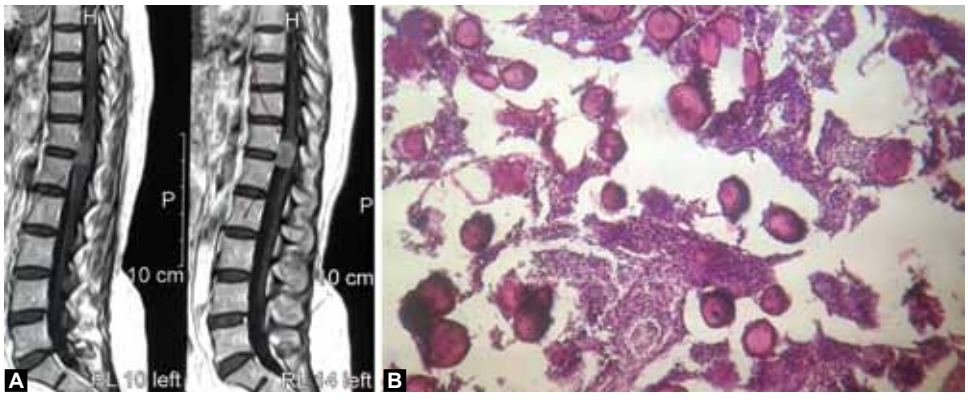
**Table 3:** Outcome assessment using pre- and postoperative Frankel grade

| | | *Postoperative Frankel grade* | | | | |
|---|---|---|---|---|---|---|
| *Preoperative Frankel grade* | | *A* | *B* | *C* | *D* | *E* |
| A | 0 | 0 | 0 | 0 | 0 | 0 |
| B | 14 | 0 | 0 | 0 | 8 | 6 |
| C | 18 | 0 | 0 | 0 | 4 | 14 |
| D | 20 | 0 | 0 | 0 | 0 | 20 |
| E | 0 | 0 | 0 | 0 | 0 | 0 |

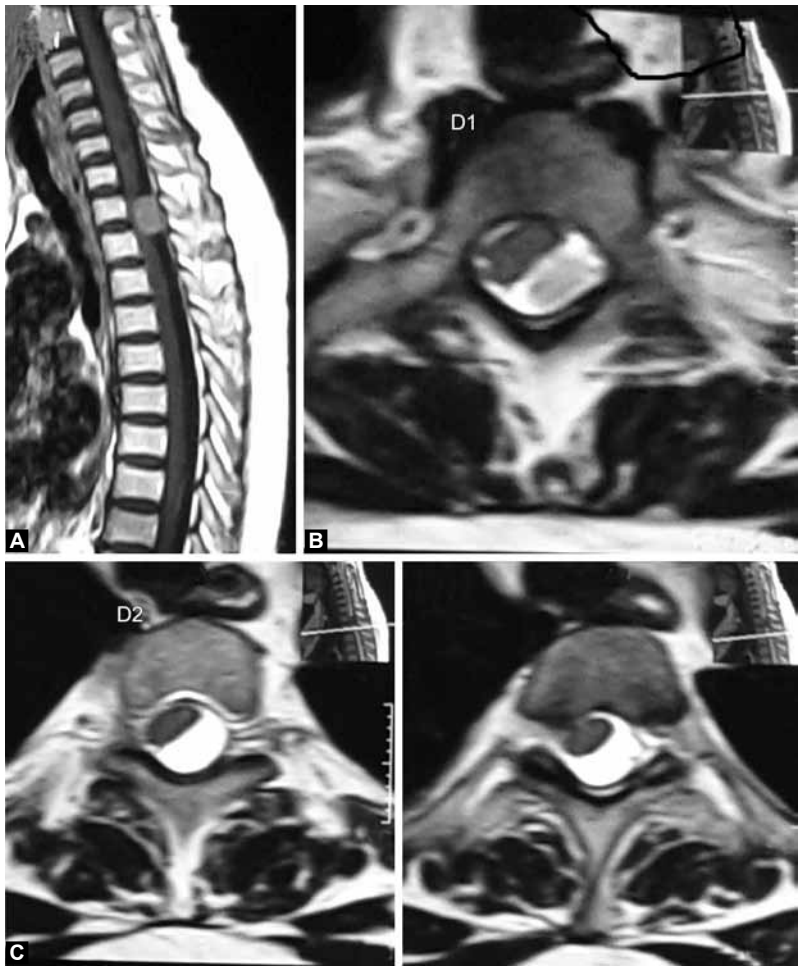
the 15th postoperative day. There was no deterioration of the neurological symptoms after surgery.

## DISCUSSION

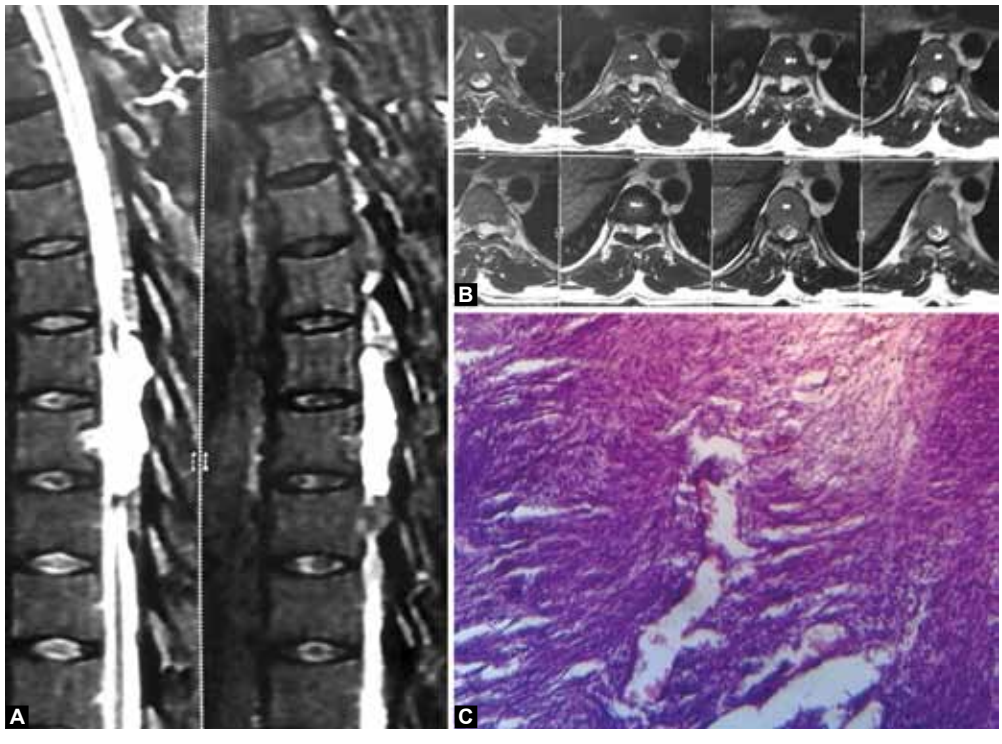
As a past, Sir Victor Horsley in 1888 succeeded for the first time in surgically excising an IDEM tumor situated in the thoracic region, and this was 44 years before the innovation of mylography.[2] Thereafter, the progression in radiological examination techniques and the use of surgical microscopes has brought about extraordinary improvements in the diagnosis and surgical treatment, but the basic surgical principles have not changed.[3] Meningiomas account for 25 to 46% of all primary intraspinal neoplasm's and spinal meningiomas are only 7.5 to 12.5% of all meningiomas because most meningiomas are found in the brain.[4] Spinal meningioma is mostly located in the thoracic vertebra and they are more common in females, which is presumably due to the influence of female hormones.[4] In our study, all meningiomas were also found in the thoracic vertebra of female patients. With regard to the treatment of the dural attachment of a meningioma, there are three common procedures:



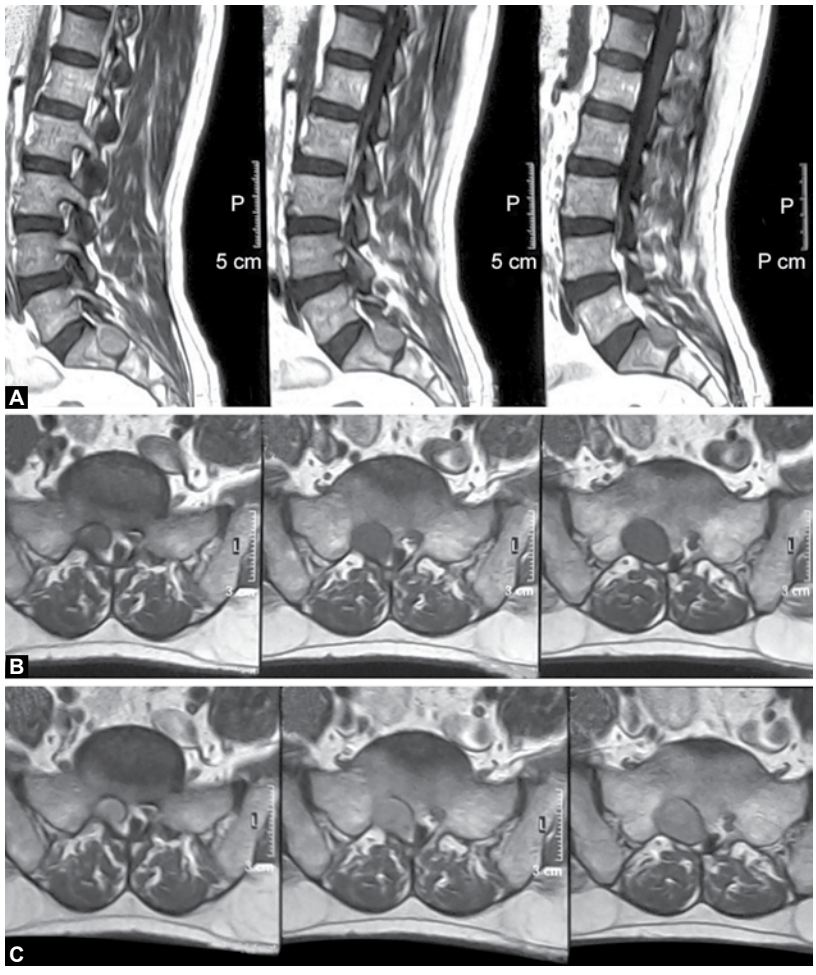

**Figs 1A and B:** A forty year-old female with intradural extramedullary meningioma with paraparesis: (A) T1-weighted sagittal MR image shows homogenous high signal intensity mass. The tumor is located at the D11-12 level, (B) Histopathology findings shows a psammomatous meningioma: The psammomatous variant is characterized by presence of numerous calcified psammoma bodie (original magnification, × 40)




**Figs 2A to C:** Meningioma (A) T1-weighted sagittal MR image shows homogenous high signal intensity mass: (A) The tumor is located at the D1-D2 level. (B and C) T1 and T2-weighted coronal MR image shows a space occupying mass compressing the spinal cord




**Figs 3A to C:** A forty-year-old male with intradural extramedullary nerve sheath tumor with paraparesis: (A) T2-weighted sagittal MR image shows homogenous high signal intensity mass. The tumor is located at the D7-8 level. (B) T1-weighted with post-contrast coronal MR image shows a space occupying mass compressing the spinal cord and (C) Histopathology was suggestive of neurofibroma; there are numerous area of myxoid degeneration (original magnification, ×40)




**Figs 4A to C:** A forty four year-old male with intradural extramedullary nerve sheath tumor with paraparesis without motor, sensory and sphincter involvement: (A) T1-weighted sagittal MR image shows isointense mass at L5-S1 level (B and C) T1-weighted pre- and post-contrast axial MR image shows a space occupying mass compressing the spinal cord at right paracentral aspect of bony spinal canal at L5-S1 level

- Some portion of the dura mater is resected with the tumor to remove any residual tumor cells and then duraplasty is performed
- Some of the internal dura mater is peeled off and the rest of it is sutured
- The dural attachment is cauterized.[5] In our study, the closures were performed without additional procedures because all the tumors were easily separated from the dura mater and only one recurrence was observed at the last follow-up. Schwannomas have been known to grow with displacing nerve fibers laterally rather than invading them. Even so, complete removal of a schwannoma is only possible when the nerve fibers are also excised in the case of their involvement within the tumor. Most authors have emphasized that the preservation of nerve roots compromises achieving complete tumor removal.[6-8] Yet according to Kim et al[9] only 23% of the complete excisions of schwannomas with functionally important nerve roots resulted in the development of neurological symptoms (not severe ones) because the nerve roots involved in the tumors had already become dysfunctional.

We used the Frankel grading for evaluation for outcome of IDEM. Our study similar the Song KW et al[10] studies, but drastic differences in results, sample size, and least complication rate and least recurrence.

According to El-Mahdy et al[2] the postoperative recurrence rate of IDEM tumors was 16%. According to Asazuma et al[6] the recurrence rate of intraspinal neoplasms was 7.2 and 46% of recurrenced masses were IDEM spinal tumors which recur more commonly than other intraspinal tumors. They also reported that the ventral location of a tumor, extradural invasion, neurogenic tumors and ependymomas were the risk factors for recurrence. According to the study on the treatment of ependymomas by Klekamp and Samii[11] the recurrence rate was 29.5% at 5 years after complete resection, and this rate was higher than that of other tumors. We believed incomplete removal of the dura mater, which is the origin of the tumor, caused the high rate of recurrence of meningioma. In our study, the recurrence rate was found to be 1.9%. However, it is our understanding that our obtained recurrence rate is not reliable because the mean follow-up period (22.44 months) was too short and various pathological

diagnoses were included. While some authors reported that pedicle erosion, vertebral body erosion, foraminal widening, neural foramen widening, and scoliosis were found on plain radiographs in approximately 38 to 56% of patients with an intradural extramedullary tumor[12] normal findings were observed in 46 cases except for six cases of neural foraminal widening. Recently, MRI was regarded as being a helpful tool with regard to tumors: in assessing the size, shape, and anatomical relations with the adjacent structures, particularly with the dura mater and spinal cord; and in determining the basic treatment guidelines and surgical approaches by allowing early detection of a metastatic tumor.[13]

The most prevalent spinal IDEM tumor was schwannoma. With nerve sheath tumors, two tumor populations need to be distinguished which are schwannoma and neurofibroma. Schwannomas are more common and are the largest category of nerve sheath tumors. Whereas schwannomas are encountered in patient with neurofibromatosis (NF-2) and in patient without NF, neurofibromas are found in patients with NF-1.[14] Most nerve sheath tumors arise from a dorsal nerve root. Neurofibromas represent a higher proportion of ventral nerve root tumor and often exhibit a dumbbell configuration.[15]

Histological appearance of neurofibromas consists of an abundance of fibrous tissue and conspicuous presence of nerve fibers within the tumor stroma. Grossly, the tumor produces fusiform (plexiform) enlargement of the involved nerve, which make it impossible to distinguish between tumor and nerve tissue.[16,11] Schwannomas appear grossly as smooth globoid mass that do not enlarge the nerve but are suspended eccentrically from it with a discrete attachment. Their histological appearance consists of elongated bipolar cells with fusiform, darkly staining nuclei arranged in compact interlacing fascicles with a tendency toward palisade formation ( Antoni-A). A loosely arranged pattern of stellate-shaped cells (Antoni-B) is less common.[17]

Traditionally, the surgical resection of a spinal cord tumor has been performed through total laminectomy.[18] This facilitates access and visualization. Seppälä MT et al[14] reported a series of 187 patients that underwent surgical resection for spinal schwannoma and reported satisfactory prognoses. In this series, 90% were completely resected, with a 10% surgical complication rate and 1.5% surgical fatality rate. Levy et al[19] also reported similar results for 66 patients with a 9% surgical complication rate and a 1.5% mortality rate. However, total laminectomy may cause spinal instability and kyphosis due to the damage to the musculoligamentous structures and posterior bony elements. And these complications may produce neurologic symptoms by compressing the spinal cord or nerve roots.[20] In order to prevent such complications, a total laminectomy with arthrodesis or a unilateral limited laminectomy, It may be assumed that the view is narrow in a unilateral laminectomy, so it is not easy to handle instruments and the exposure of the lesion is incomplete, and as a result, normal nerves may be damaged or it may be difficult to remove the lesion completely.

For making the diagnosis of IDEM tumors, taking a detailed history, a thorough physical examination and performing MRI scans on the proximal regions are suggested because the symptoms of IDEM tumors can be like with those of lumbar herniated nucleus pulposus and spinal stenosis.

## CONCLUSION

Intradural extramedullary tumors detected by MRI are mostly benign and good clinical results can be obtained when treated surgically. Therefore, more dynamic surgical approaches by neurosurgeons are suggested to decline morbidity.

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