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Intradural Extramedullary Spinal Tumors – A Review of Modern Diagnostic and Treatment Options and a Report of a Series of 40 Cases

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Abstract

Spinal tumors are uncommon lesions and affect only a minority of the population. Spinal tumors comprise 15 % of all CNS tumors, with an incidence of 2-10 per 1,00,000. Location of the tumor is the most important aspect in the diagnosis of spinal tumor with regard to treatment and prognosis. Extradural lesions are more common (60% of all spinal tumors), with majority of lesions originating from the vertebrae. The clinical symptoms are often nonspecific and include back pain, radicular symptoms, and slowly progressive neurological deficits. Primary and most important diagnostic modality for intradural spinal tumors is MRI scan. Contrast material (defining the extent of intramedullary neoplasms and identifying areas of blood brain barrier break-down) and multiplanar imaging have broadened the use of MRI. Gross total tumor resection while preserving and improving the neurological function is the usual goal of surgery. If the complete removal of the tumor demands removal of more than one column of the spine, instrumented fusion of the adjacent levels may be deemed necessary. Here we discuss the literature review of intradural extramedullary tumors in a glance and clinical presentation, symptomatology, and prognosis of 40 extramedullary intradural tumor cases that we have operated on in a span of 15 years. Out of these 40 cases, majority were schwannomas. The incidence of schwannoma was found to be higher in the 30-50 age group. 5 cases of meningiomas, 5 cases of cystic schwannomas and 1 each of intradural metastasis, paraganglinoma, AV malformation, ependymoma, cellular schwannoma, intradural hematoma and intradural lipoma with tethering. All cases had postoperative improvement in neurological status to normalcy.

Keywords- Spinal tumors, Intradural extramedullary tumors, schwannomas, meningiomas, cystic schwannomas, intradural metastasis, paraganglinoma, AV malformation, ependymoma, cellular schwannoma, intradural hematoma, intradural lipoma , symptomatology, MRI, prognosis

Introduction

Spinal tumors, comprising 15% of all central nervous system (CNS) tumors, with an incidence of 2–10/ per 1,00,000 are uncommon lesions affecting only a minority of the population. More than 90% of these are found in patients above 20 years of age [1]. Location of the tumor is the most important aspect in the diagnosis of the spinal tumor with regard to treatment and prognosis. Extradural lesions are more common (60% of all spinal tumors), with majority of lesions originating from the vertebrae. The most common extradural tumor is metastasis. Intradural tumors are rare, and among them, majority are extramedullary (30% of all spinal tumors), with meningiomas, nerve sheath tumors (schwannomas and neurofibromas), and drop metastasis being the most frequent [1]. Tumors of the spine, though uncommon, might cause severe morbidity ranging from pain syndromes, through weakness and bladder involvement [2]. Also Furthermore, Brown brown-sequard syndrome, signs of long tract involvement such as clonus, hyperreflexia, and babinski sign are commonly found, but not pathognomonic. With current advancements in imaging and diagnostics, magnetic resonance imaging (MRI) has become the single most important tool in diagnosing the level, location, and extent of spinal tumors[3]. So Hence, when there is suspicion of tumor based on clinical history and symptomatology, MRI scan should be done, unless contraindicated. Neither the presence nor the absence of abnormal findings on plain film imaging, computed tomography (CT) or CT myelography can exclude or sufficiently delineate and characterise characterize an intradural tumor. In this paper, we attempt to

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discuss various aspects of treatment, complications, and outcome of surgical treatment of intraduralextramedullary tumors. In addition, we include our own retrospective experience of 40 patients treated over a time interval of 15 years.

Presenting Signs and Symptoms

The clinical symptoms are often nonspecific and include back pain, radicular symptoms, and slowly progressive neurological deficits such as weakness, paresthesia, gait problems, impotence, and bowel and bladder dysfunctions. In children, intradural tumors may be associated with skeletal deformities such as kyphoscoliosis and scalloping of vertebral bodies. Since the symptoms are mostly nonspecific, the diagnosis is often delayed. Meantime to diagnose is 12 months.

Investigations

Primary The primary and most important diagnostic modality for intradural spinal tumors is an MRI scan. Contrast material (defining the extent of intramedullary neoplasms and identifying areas of blood blood-brain barrier break-down) and multiplanar imaging have broadened the use of MRI with respect to imaging capabilities and pathophysiological characterization. Pre-operative imaging protocol should also be inclusive of plain radiographs and CT scan with thin cuts and reconstructions to evaluate bony anatomy. In patients whom whose MRI is contraindicated, CT Myelography myelographyis an option.

Intradural Extrame dullary Lesions

The most common intradural extrame dullary lesions are schwannomas, meningiomas, and neurofibromas. Less common lesions include paragangliomas, metastases, lipomas, spinal nerve sheath myxomas, sarcomas, and vascular tumors.

Nerve Sheath Tumors

Nerve sheath tumors are the most common lesions in this category. Most common nerve sheath tumors are neurofibromas and schwannomas. Ganglioneuromas and malignant nerve sheath tumors make up the rare varieties of nerve sheath tumors.

Neurofibromasare the most common type of nerve sheath tumors. Classically seen in patients with neurofibromatosis Type 1, but can also be seen sporadically. . This tumor arises from the dorsal sensory nerve roots and invests the adjacent nerve fibers.

Schwannomas, the second most common type of non stress test NST are is composed of schwann Schwann cells with fibrous tissue. MRI imaging gives the characteristic target lesion appearance with the decreased signal intensity in the center representing the fibrous Antoni A tissue, while the increased signal intensity in the periphery represents the myxomatousAntoni B tissue. Schwannomas are more commonly seen in adults and in association with Neurofibromatosis neurofibromatosis Type 2. The tumor most commonly are is eccentrically located in relation to the nerve fibers and usually does not involve the nerve fiber themselves.

Though Although malignant changes in schwannomas are extremely rare, cystic degenerations in schwannomas have been reported. Them being a benign, extremely slow slow-growing tumor, the symptoms are usually vague and often leads to a diagnostic dilemma [4,5].

Meningiomas

They are the second most common intradural extramedullary tumors. Classically described as a benign, slow-growing tumor that arises from the arachnoid cap cells. Thoracic spine is the most common location for spinal meningiomas (80%). The attachment to the dura is broad based (dural tail). Malignant transformation is rare and mostly they can be resected out safely.

Filum Terminale Ependymomas

About 50% of all ependymomas are spinal, and of these, quite few of them are located in the filumterminale. Although technically intramedullary, from the anatomical and surgical perspective they are considered intraduralextramedullary tumors. They occur most commonly in patients in the third to fifth decades of life. Recurrence is rare after radical resection, but subarachnoid seeding is a possibility [1].

Other Primary Tumors

The remaining intraduralextramedullary tumors include epidermoid tumors, paraganglinomas, lipomas, plasmacytomas, and chloromas.

Metastatic Disease

Disseminated CNS tumors make up the majority of spinal metastasis. CNS sources include medulloblastoma, high-grade astrocytoma, ependymoma, oligodendroglioma, retinoblastoma, pineal tumors, and choroid plexus papilloma. Systemic neoplastic sources for intradural metastasis include breast, lung, melanoma, lymphoma, gastrointestinal carcinoma, genitourinary tumors, neuro endocrine tumors, and head-and-neck cancers [1,2].

Tumor Like Lesions

These include inflammatory disorders such as sarcoidosis and arachnoiditis, vascular anomalies such as arteriovenous (AV) malformations and intradural hematomas and other cysts associated with neural tube defects[2].

Pre-operative Planning and Treatment

Gross total tumor resection while preserving and improving the neurological function is the usual goal of

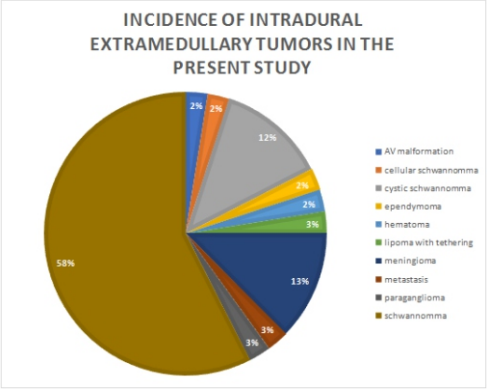

Chart 1: Incidence of intradural extramedullary tumors in our study (sample size of 40 cases).

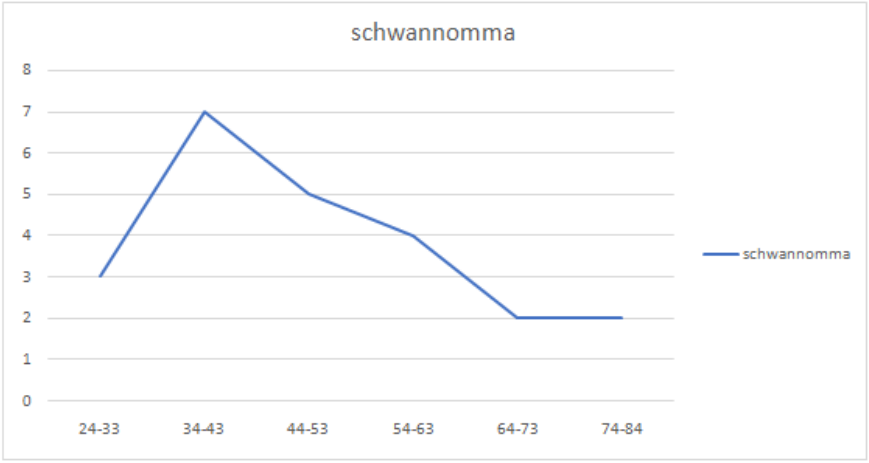

Chart 2: Incidence of schwannoma in different age groups.

surgery. Intraoperative somatosensory evoked potentials and motor evoked potentials are both great tools to assess neurological damage with the patient in general anesthesia. After a detailed clinical and radiological pre-operative evaluation, the appropriate operative approach is planned. The extent of incision and exposure may be limited with the judicious use of intraoperative C arm navigation to determine the exact level of pathology. Intraoperative ultrasound scanwas usedto determine the depth of invasion of tumor tissues. If the complete removal of the tumor demands removal of more than one column of the spine, instrumented fusion of the adjacent levels may be deemed necessary. Most common complications include Cerebrospinal fluid (CSF) leak, pseudomeningoceleformation, and wound infections. Post-operative spinal instabilities and residual neurological deficits are also not uncommon [1,3].

Discussion

Here, we discuss the clinical presentation, symptomatology, and prognosis of 40 extramedullary intradural tumor cases that we have operated on in a span of 15 years. Out of the 40 cases that were operated, majority were schwannomas (Chart. 1). In our study, schwannomas showed a predilection towards thoracolumbar spine. The incidence of schwannoma was found to be higher in the 30-–50 years of age groups, this being comparable to the incidence in other published studies (Chart. 2). Five cases of meningioma were operated, and they were clearly more common in the upper thoracic spine and mostly presented as paraplegias. Five cases of cystic schwannomas were reported.,And and 1 one each of intradural metastasis, paraganglinoma, AV malformation, ependymoma, cellular schwannoma, intradural hematoma, and intradural lipoma with tethering. All cases had post-operative improvement in neurological status to normalcy.

Among them, one case presented with paraplegia with urinary incontinence and bilateral Grade IV gluteal ulcers. He had a history of coronary artery disease, and was on the permanent pacemaker. Patient The patient was sitting on bed for the past 8 months, unable to walk due to paraparesis. He was evaluated in another outside hospital with CT scan of lumbosacral spine and was treated for intervertebral disc prolapse. At our hospital, we insisted on a CT myelogram, which revealed a complete obstruction at D12/L1 region. He was operated and the specimen on histopathological evaluation, proved to be an intradural schwannoma. Patient started neurological recovery soon after surgery and was mobilized with walker on the 14th post-operative day. The gluteal ulcers started granulating and healing, once the patient was operated. The wound was fully granulated by the 14th post-operative day. Evaluation by an MRI of the spine is a must to diagnose or to rule out the diagnosis of a spinal tumor. In cases where MRI is contraindicated, like as the case in question, a CT myelogram should be considered.

One case presented with bilateral lower limb weakness and bladder involvement. He was evaluated from an outside hospital with an MRI of the lumbar spine, which showed no obvious pathology. He was treated symptomatically and was referred to our hospital for further evaluation. On admission and further evaluation with MRI of the whole spine, an intradural tumor at the thoracolumbar junction compressing the conus medullaris was detected. He was operated for the same and the tumor on histopathological evaluation, was proved to be an intradural schwannoma. The patient completely recovered clinically after the excision. The patient presented to us 4 years later with another intradural extramedullary tumor at the L2 level. He was operated again, and the tumor was once again proved histologically to be an intradural schwannoma. He has been on yearly follow follow up ever since, and there is no sign of further recurrence yet. This case has been an eye eye-opener in the sense that a tumor was missed on initial evaluation despite an MRI of the

lumbosacral spine. Lumbosacral spine MRI evaluation often tends to miss lesions at the thoracolumbar junction. Hence, a screening of the whole spine should be done mandatorily along with detailed MRI of the region under evaluation.

There was a case of multiple intradural tumors, including a dumbbell tumor in the cervical region (Fig. 1) of a patient presented with quadriparesis. Patient The patient was a known case of neurofibromatosis. The dumbbell tumor and the tumors causing cord compression were excised with a warning for possible recurrence, immunohistochemistry evaluation showed Ki proliferation index to be zero, which indicated low chance of recurrence.

One patient presented with abdominal pain and was evaluated, the initial evaluation by the Gastroenterology gastroenterology Department department with investigations ranging from simple blood tests to an endoscopic evaluation, which failed to reveal the cause of the pain. He was then evaluated with an MRI spine which showed an intradural tumor in the D11/D12 region. Radicular pain can often mimic other system involvement. The patient recovered clinically once the tumor was removed.

The intradural extramedullary metastasis in a 75 year -year-old female who presented with paraparesis, on excision and histological examination was proved to have a neuroendocrine tumor as the primary lesion. Neuroendocrine The neuroendocrine tumor which metastasizes to spine is an extremely rare entity (Only one such case report was found in the literature search). She was treated with excision of tumor, circumferential decompression, and stabilization of spinal architecture according to the NOMS guideline [6,7]. The patient improved clinically following the surgery. The spinal metastases are often very vascular and may cause uncontrollable bleeding which makes visualization of the lesion difficult and may even lead to hemodynamic instability. In such situations, if the tumor is suspected to be highly vascular, a pre-operative angioembolization of the tumor might help [8].

One case presented to us with paraplegia and skin ulceration in the lower limb which was not healing. A biopsy from the ulcer revealed it to be a case of Systemic systemic Lupus lupus Erythematosiserythematosus. MRI of the spine revealed an intradural tumor at the D12/L1 level. She was advised to undergo surgical excision, but despite the advice, she underwent ayurvedic massage for 5–6 months. She became paraplegic during the massage and reported back to our hospital willing for surgery. The tumor was excised and histopathological examination revealed it to be paraganglioma of the spine. Paragangliomas are tumors arising in the paraganglia and rarely occur in the spinal canal. These tumors are usually regarded as benign, but malignancy is found in 6.5% of all extra extra-adrenal paragangliomas, and local tumor recurrence has been described in 12% [9]. Paragangliomas of the spinal canal are more common than previously thought and can be located anywhere along the spine, although the lumbosacral level is the most common. Their appearance on MRI cannot make them distinguish them from other tumors in the spinal canal. Even though paragangliomas in general, in general, are benign and slowly growing their growth pattern can vary and be more aggressive, to the point of metastatic spread [10].

5 Five cases of cystic schwannomas were operated (Fig. 2). These represent benign slowly growing schwannomas which undergo cystic degeneration. These variants are not as uncommon as was thought earlier before and should most definitely be considered as a differential diagnosis when the MRI

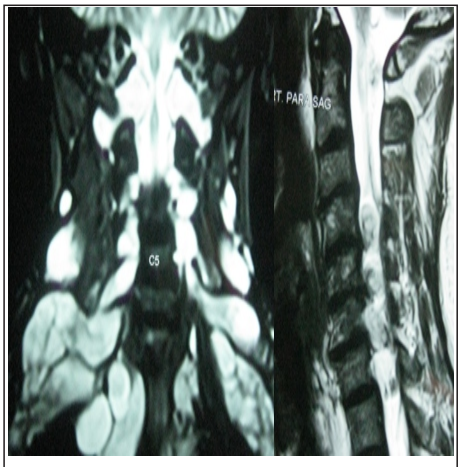

Figure 1: multiple Multiple neural tumors in the cervical region.

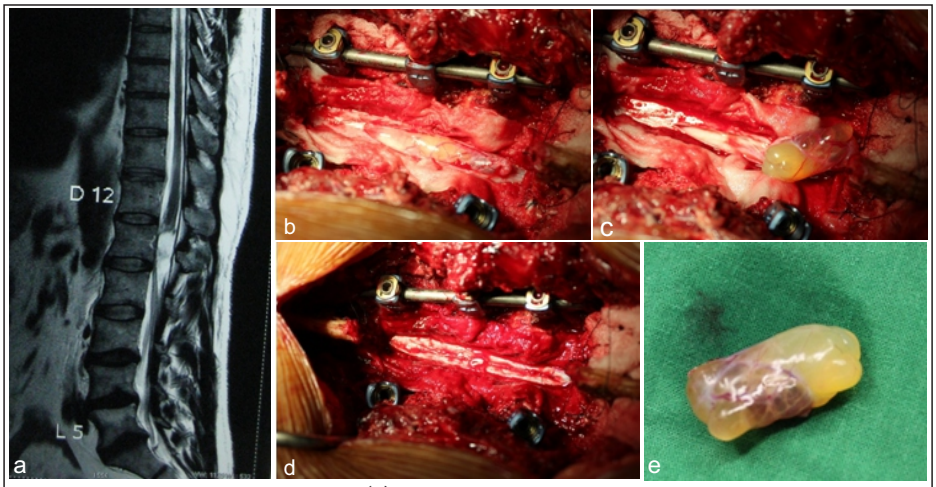

Figure 2: Cystic schwannoma (a)-magnetic resonance imagingMRI sagittal view, (b–)intraoperative, (c-) removal of cystic lesion, (d) –cystic schwannomaafter excision).

spine reveals an intradural cystic lesion [4,5].

Summary/Conclusion

The present study puts forward a few eye eye-openers and pitfalls which the surgeons should be careful about while dealing with intradural spinal tumors. A detailed clinical examination which includes the physical examination and neurological status of the patient should be done, which should be invariably followed by a detailed MRI of the concerned spinal region. The above mentioned detailed that MRI should be entailed with a mandatory screening of the whole spine. If this is not done, junctional tumors can be easily missed. In patients whom whose MRI is contraindicated, a CT myelogram should be performed to detect spinal canal stenosis and obstruction of CSF flow.

While dealing with cystic intradural tumors, cystic schwannomas, though rare, should be considered as a differential diagnosis.

Gluteal pressure sores should be treated with the utmost care and a detailed evaluation of neurology of the patient and an MRI of the spine should be considered.

Intradural metastases, though rare, are not uncommon and would often need resection of the lesion with circumferential decompression of the cord and stabilization of the spinal column. The resection of vascular tumors may cause torrential bleeding, and a pre-operative embolization of the lesion may be considered.

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