

Silent Spinal Cord Tumor

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ABSTRACT

Introduction: Intramedullary spinal cord tumors are rare, representing 4 to 10% of all central nervous system tumors. They account for 20% of all intraspinal tumors in adults and 35% of all intraspinal tumors in children.

Study design: Observational study.

Purpose: Understanding the natural progression of an intramedullary spinal cord tumor.

Materials and methods: To report a case of silent intradural intramedullary spinal cord tumor in a 38-year-old patient, on regular follow-up in the outpatient department since 6 years.

Investigations: Magnetic resonance imaging thoracic spine revealing intramedullary tumor which is localized, central, uniformly enhancing on contrast and is associated with syrinx formation from D1 to D7.

Management: Conservative.

Conclusion: The rare incidence of intradural tumors commonly results in misdiagnosis and improper diagnostic workup, resulting in delayed diagnosis and treatment. Dilemma exists in the management of clinically silent intramedullary tumors as no specific guidelines have been formulated.

Keywords: Asymptomatic, Conservative, Intradural, Spinal cord tumor, Syrinx.

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INTRODUCTION

Intramedullary tumors of the spine make up 4% of all central nervous system (CNS) tumors and are a rare cause of spinal cord dysfunction. They refer to a subgroup of intradural spinal tumors that arise from cells within the spinal cord, as opposed to adjacent structures, such as the nerve roots or meninges.¹

In 1887, Victor Horsley performed the first successful removal of an intradural tumor.²

The most common initial symptom is generalized back pain, which is very difficult to distinguish clinically from back pain from musculoskeletal conditions. Patients are often diagnosed only after the development of neurologic signs and symptoms that may occur later in the course of the disease.³ Early imaging is important for diagnosis. Neurological deficits resulting from intramedullary spinal cord tumors are seldom reversible.^{4,5}

The subject presented to us early in the course of disease with minimal neurological deficits and has been followed up with serial magnetic resonance imaging (MRI) scans for a period of 6 years providing an insight to the natural history of the disease.

CASE REPORT

This patient was first seen in the outpatient department 6 years back when he was 32 years old. He was in good general health condition. He works in an office (sedentary job) and is well built.

He presented with some vague pain in the dorsal region which has subsequently disappeared. There was no neurological deficit. No motor involvement, autonomic disturbance, or gait imbalance was present.

His MRI showed a spinal tumor at D3-D4 region which was intramedullary, localized, central, uniformly enhancing on contrast and associated with syrinx formation from D1 to D7 (Figs 1A to F).

In view of paucity of neurological symptoms and signs, we decided to observe him with serial imaging and no specific treatment.

Since that time, over the last 6 years he has come regularly for follow-up every 6 months and an MRI has been repeated every year. Currently, he is asymptomatic. There is no neurological or autonomic deficit. He is healthy and attends his work regularly. He does not suffer from diabetes, hypertension, or thyroid abnormalities.

DISCUSSION

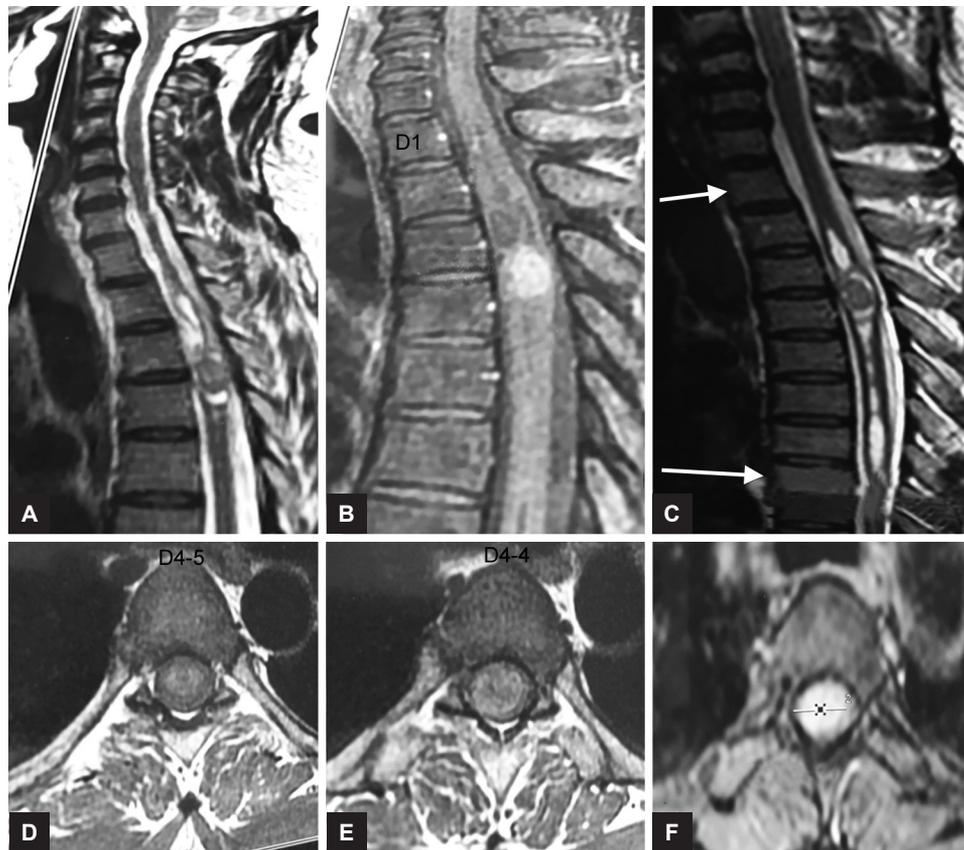
Intramedullary spinal cord tumors are rare, representing 4 to 10% of all CNS tumors. They are generally slow growing, histologically benign tumors and are often definitely treated with surgery. A long duration of symptoms prior to diagnosis is typical; 90 to 95% of all intramedullary neoplastic lesions are of glial origin, with spinal

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Figs 1A to F: Magnetic resonance imaging showed a spinal tumor at D3-D4 region, which was intramedullary, localized, central, uniformly enhancing on contrast, and associated with syrinx formation from D2 to D7

ependymoma comprising 60% and spinal astrocytoma accounting for 33%. Among the intramedullary nonglial neoplasms, spinal hemangioblastoma is the most common, being the third most commonly encountered entity. The lesions have characteristic radiographic appearance which helps in their differentiation (Table 1).

Our patient is 38 years of age. In adults, ependymomas are the most common tumor type, accounting for 40 to 60% of all intramedullary spinal tumors, with the mean age of presentation being 35 to 40 years. Intramedullary spinal tumors can arise anywhere in the spinal cord, from the cervicomedullary junction to the filum terminale, but they are found most frequently in the cervical cord. The etiology of intramedullary spinal tumors varies according to histology. Patients with the NF1 gene are predisposed

to spinal astrocytomas, whereas patients with the NF2 gene are predisposed to spinal ependymomas. Hemangioblastomas occur as a result of mutations in a tumor suppressor gene called *vhl*, which was found to be altered in patients with the neurocutaneous disorder von Hippel–Lindau disease (VHL).^{7,8}

The usual symptoms associated with these tumors are gait imbalance, upper or lower extremity weakness, upper or lower extremity numbness, urinary incontinence, nocturnal back pain, hand in-coordination, burning dysesthesias, and sudden paraparesis or quadriplegia. The signs are consistent with an upper motor neuron lesion consisting of spasticity, hyperreflexia, upgoing plantars, objective bilateral motor, and sensory weakness.⁹

Table 1: Radiographic differentiation of intramedullary tumors

Diagnosis/characteristics	Cord expansion	Contrast enhancement	Well circumscribed	Heterogeneous signal	Cord edema
Ependymoma	+	+	+	-	±
Astrocytoma	+	+	±	-	±
Hemangioblastoma	+	+	+	-	+
Cavernoma	+	+	+	+	±
Multiple sclerosis	±	±	±	-	+
Transverse myelitis	±	±	-	-	+
Spinal cord infarct	±	-	+	-	+
Arteriovenous fistula	-	-	-	-	+

The most important prognostic factor is the histopathology of the tumor. This is the most important predictor of patient outcome because it predicts if the tumor will be resectable and if it will recur. The recurrence rate for low-grade tumors is less than 5%.¹⁰ Malignant astrocytomas of the spinal cord have an overall recurrence rate of greater than 95%, with outcome unaffected by extent of surgery.¹¹

The identification of mitotically active neural stem cells and neural progenitor cells throughout the CNS has altered current thinking about how all intrinsic CNS tumors arise. Many lines of evidence point toward neural stem cells as the cells of origin of these tumors.¹²

In our patient, in spite of development of syrinx and apparently significant displacement of the spinal cord, he has remained asymptomatic over a follow-up period of 6 years. Such cases further help in our understanding of the natural progression of the disease.

OPTIONS FOR MANAGEMENT

Conservative

Medical management has been generally restricted to patients who have minimal neurological signs, presuming it to be a low-grade tumor. Our patient had hardly any neurological symptom except back pain and investigations showed cord expansion, contrast enhancement, and well circumscribed mass, and we preferred to observe him. We neither gave radiation therapy nor high-dose corticosteroids but observed him with serial imaging studies.¹³

Surgical management is the first-line treatment for intradural tumors.¹⁴⁻¹⁶ Traditional approach using standard microsurgical techniques has been the most popular in the past.¹⁷ Minimally invasive surgical (MIS) approach and stereotactic spinal radiosurgery are now gaining popularity over the traditional approach. Recent advancements like intraoperative neuromonitoring in the form of somatosensory evoked potentials, motor evoked potentials, and D-wave monitoring¹⁸⁻²⁰ are now in vogue. "Mini-open" procedure, a variant of MIS, and the use of minimally invasive retractor system to prevent intraoperative cord damage have recently been popularized.

One study has reviewed the natural history of such tumors with comparative analysis of those operated surgically and those conserved. Among the 10 patients conserved, the mean follow-up was 80 months, and all patients had same or better McCormick grade at the last follow-up. The four patients operated had 42 months follow-up and last follow-up suggested same (two patients) or worsening (two patients) McCormick grade. Authors

concluded that conservative approach of observation did not see neurological decline over long follow-up just as we have observed in our case.²¹

CONCLUSION

Described here is a case of clinically silent intramedullary tumor observed with serial MRI imaging over 6 years with patient still remaining clinically asymptomatic.

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