



## Case Report

# A rare case of intramedullary schwannoma at conus medullaris: A case report with review of literature

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## ABSTRACT

**Background:** Intramedullary spinal schwannomas constitute only 0.3% of primary spine tumors. We could identify only 13 such cases involving the conus that were not associated with neurofibromatosis (NF). Here, we report a 70-year-old male without NF who was found to have a paraparesis due to a schwannoma of the thoracolumbar junction/conus (D11-L2).

**Case Description:** A 70-year-old male presented with an L1-level paraparesis with urinary incontinence. The magnetic resonance showed an intramedullary mass of 85 × 10 mm extending from D11 to L2; it demonstrated significant patchy enhancement. The patient underwent a D12 and L1 laminectomy with gross total excision of the mass that proved to be a schwannoma. Three months postoperatively, he was able to ambulate with support, and regained sphincter function.

**Conclusion:** Intramedullary schwannomas involving the conus/thoracolumbar junction are rare, and can be successfully excised resulting in good outcomes.

**Keywords:** Conus medullaris, Intramedullary, Schwannomatosis

## INTRODUCTION

Schwannomas constitute approximately 30% of all spinal tumors. Most of the cases are sporadic and solitary; if multiple, they are usually associated with neurofibromatosis (NF) Type 2 or schwannomatosis. They are most commonly intradural and extramedullary in location; intramedullary schwannomas are very rare. We could identify only 13 cases of solitary intramedullary schwannomas not associated with NF involving the conus.<sup>[5]</sup> Here, we present a 70-year-old male with a schwannoma involving the conus medullaris that was successfully excised.

## CASE PRESENTATION

A 70-year-old male presented with 2.5 years of progressive paraparesis and sphincter dysfunction (power 1/5 right and 3/5 left leg, and sensory loss below L1). He had no features of NF.

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**Figure 1:** T1-weighted images showing hypo to isointense lesion from D11 to L2.



**Figure 2:** T2-weighted images with hyperintense lesion from D11 to D12.

### Magnetic resonance (MR) study

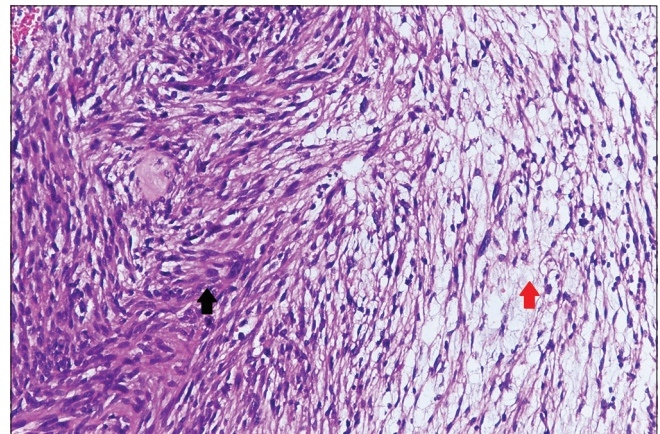
The thoracic MR showed an 85 × 10 mm intramedullary lesion extending from D11 to L2; it was hypointense to isointense on T1-weighted [Figure 1] and hyperintense on T2-weighted [Figure 2] images. The post contrast scan showed significant patchy enhancement with few central non-enhancing areas [Figure 3] (likely to be cystic or foci of necrosis). Other findings included; internal fibrinous reticulations, septations, and debris consistent with foci of hemorrhages. The provisional diagnoses included ependymoma versus astrocytoma; there were no other lesions involving the neuraxis.

### Surgery

With intraoperative neuromonitoring, D12-L1 laminectomy was performed. On exposure, there was a firm greyish-white mass exophytic from the conus that was adherent to not only



**Figure 3:** Post contrast image showing patchy enhancement with few non enhancing areas likely hemorrhage or necrosis.

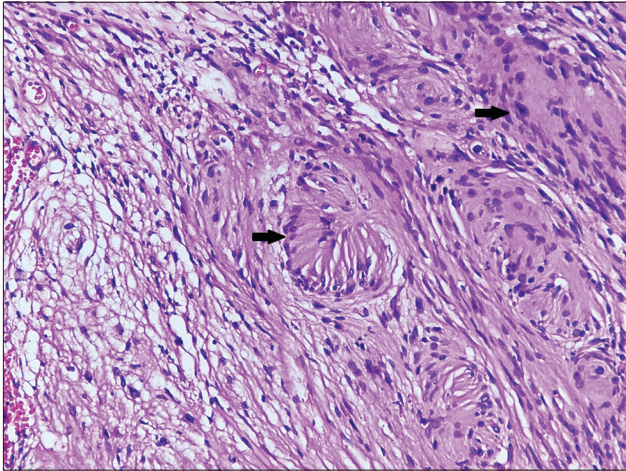


**Figure 4:** Spindle cells arranged in alternating compact hypercellular (Antoni A, black arrow) and loose myxoid hypocellular (Antoni B, red arrow) areas (H and E ×200).

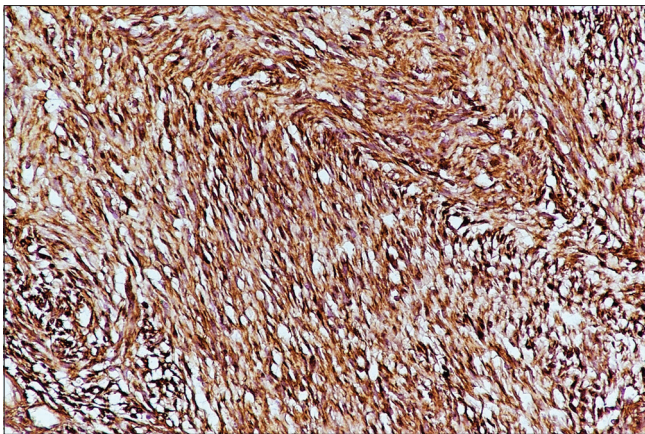
the conus but also multiple roots of the cauda equina. Under the microscope, a gross total resection was accomplished.

### Pathology

The histopathological examination revealed tumor tissue arranged in alternating hypercellular and hypocellular areas (Antoni A and B) [Figure 4]. Tumor cells were haphazardly arranged in the loose myxoid stroma and were oval to spindle-shaped with areas of palisading. The cells showed indistinct cytoplasmic outlines, hyperchromatic nuclei, and mild anisonucleosis with some nuclei showing buckling and tapered ends. No cytologic atypia or mitotic activity was noted. Many hyalinized blood vessels and Verocay bodies [Figure 5] were interspersed. Immunohistochemistry showed strong diffuse immunostaining with S-100 [Figure 6].



**Figure 5:** Verocay bodies showing palisaded nuclei arranged around acellular fibrillary zones (black arrows) (H and E  $\times 200$ ).



**Figure 6:** Strong diffuse cytoplasmic and nuclear immunostaining with S-100 (H and E  $\times 200$ ).

### Postoperative course

Immediately postoperatively, the patient's neurological function remained unchanged. However, 3 months later motor function was 4/5 in the left and 3/5 in the right lower extremity, and the patient regained bladder control.

### DISCUSSION

Schwannomas are most commonly intradural and extramedullary in location; <1% are intramedullary.<sup>[1]</sup> They can be present throughout all levels of the spinal cord, and typically are found in the cervical (58%), followed by the thoracic (32%) and lumbar regions (10%).<sup>[3]</sup> We could identify only 13 cases of solitary intramedullary schwannomas involving the conus without a history of NF.<sup>[5]</sup> The peak incidence for intramedullary schwannomas is between the 4<sup>th</sup> and 5<sup>th</sup> decades of life with neurological deficits reflecting their location. On MR, schwannomas are

typically hypointense or isointense on T1-weighted images and hyperintense on T2-weighted images. They usually heterogeneously or uniformly enhance with contrast. However, intramedullary schwannomas usually do not have distinctive radiological features, making it difficult to establish this diagnosis.

Surgery is the treatment of choice for symptomatic or growing intramedullary schwannomas. However, some cases of intramedullary schwannomas show infiltrative patterns making total resection impossible. Radiotherapy is a possible alternative in these cases with incomplete resections.<sup>[4,6]</sup> The recurrence rate for intramedullary schwannomas is approximately 5% at 2 years postoperatively. Risk factors for recurrence are subtotal resection, the higher number of spanned levels, increasing tumor size in the cranial-caudal direction, and the tumor location in the cervical or sacral regions.<sup>[2]</sup>

### CONCLUSION

Schwannomas are rarely seen as intramedullary tumors involving the conus. Here, we presented a 70-year-old male with such a lesion without a history of NF who underwent surgical resection of the lesion with postoperative neurological improvement.

### Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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### Conflicts of interest

There are no conflicts of interest.

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