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*Case Report*

# Successful excision of intradural extramedullary plexiform T1–T4 schwannoma

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## ABSTRACT

**Background:** Plexiform schwannomas (PSs) are rare. Here, we describe the clinical features, diagnosis, treatment, and outcome of a 17-year-old male presenting with a T1–T4 intradural extramedullary (IDEM).

**Case Description:** A 17-year-old male presented with back pain and pain radiating down both legs. The thoracic magnetic resonance revealed a left-sided T1–T4 IDEM mass. The patient underwent a T2–T4 laminectomy for gross total tumor excision, followed by posterior fusion. Immunohistochemical examination revealed S100 positivity, supporting the diagnosis of PS.

**Conclusion:** IDEM PSs are rare, may be readily diagnosed with MR, and can be successfully resected.

**Keywords:** Case report, Magnetic resonance imaging, Plexiform schwannoma, Spinal cord, Thoracic vertebrae

## INTRODUCTION

Plexiform schwannomas (PSs) are rare, with approximately 5% of schwannomas progressing into this histological subtype [Table 1].[5,6] Here, we describe the clinical features, diagnosis, treatment, and outcome following the resection of a T1–T4 intradural extramedullary (IDEM) PS in a 17-year-old male.

## CASE REPORTS

A 17-year-old male presented with back pain and bilateral leg pain of 1 month's duration. The magnetic resonance (MR) revealed a left-sided T1–T4 IDEM PS (6.5 cm × 1.8 cm × 6.3 cm) mass consistent with a schwannoma. On MR, the lesion was slightly hypointense on T1-weighted images, slightly hyperintense on T2-weighted/short-tau inversion recovery sequences, and homogeneously enhanced with contrast [Figure 1]. It caused severe spinal cord compression and extended into or blocked the left neural foramina at both the T2–T3 and T3–T4 levels, further extending into the left T3–T4 paraspinal region. The IDEM was completely removed during a T1–T4 laminectomy.

**Table 1:** Cases of plexiform schwannoma based on literature.

| Study | Age (years)/Sex | Site | MRI Result | Surgery | Follow-up (Months) | Outcome |
|---|---|---|---|---|---|---|
| Sakaura *et al.* (2007)[5] | 16/M | C3–C4: Multinod DB T C cord comp left for C3–C4 | T1WI contrast: inhomog-enhanced IDEM T. T2WI: multinod inhomog-enhanced | HL C3–C4, Left Fac, unilat LMS, SubT Resect | 12 | 100% Imp of pain and weakness in LUE, NTR |
| Capone *et al.* (2012)[1] | 28/F | C5–Th1: Mult IDEM T | T2WI: multinod inhomog signal, trachea displace, For extend, cord Comp | Lam C5–T1, ID Total Removal ED partial-Enuc | - | Imp Resp Fx |
| Mori *et al.* (2015)[4] | 61/F | L2–L4: IDEM T | T1WI+T2WI: hypointense. Gd-MRI: Homog IDEM T | Lam L2–L4, GTR | 12 | 100% Imp pain, NTR |
| Cheung *et al.* (2016)[2] | 65/M | T12–L1: IM T | T1WI: hypointense T2WI: hyperintense and HEC | Lam T12 Partial, Lam T11–L1, GTR | 3 | Imp leg motor Fx, Residual UR, NTR in 15 months follow up. |
| Yu *et al.* (2020)[6] | 66/M | L1–S3: IDEM T Large Multilob+Cyst | T1WI: Iso- to slightly low intense and HEC. T2WI: Isointense | Lam L1–S2, SubT Resect, Multifocal S2-S3 | 24 | 100% Imp pain and motor Fx, NTR |
| Current study | 17/M | Th1–Th4: IDEM T+Cyst | TIWI contrast: iso/ hypointense. T2WI/T2FS: Hyperintense | Lam T1–T4, GTR | - | 100% Imp pain |

C: Cervical, Comp: Compression, DB: Dumbbell, ED: Extradural, Enuc: Enucleated, Fac: Facetectomy, For Foraminal, Fx: Function, Gd-MRI: Gadolinium-enhanced magnetic resonance imaging, GTR: Gross total resection, HEC: Heterogeneous enhancement with contrast, HL: Hemilaminectomy, Homog: Homogeneous, ID: Intradural, IDEM: Intradural extramedullary, Imp: Improved, inhomog: Inhomogeneous, L: Lumbar, Lam: Laminectomy, LMS: Lateral mass screw, LUE: Left upper extremity, Mult: Multiple, MultiNod: Multinodular, Multilob: Multi-Lobulated, NTR: No tumor recurrence, Resp: Respiratory, S: Sacrum, SubT: Subtotal resection, T: Tumor, TIWI: T1-weighted imaging, T2WI: T2-weighted imaging, Th: Thoracic, UR: Urinary retention, Unilat: Unilateral

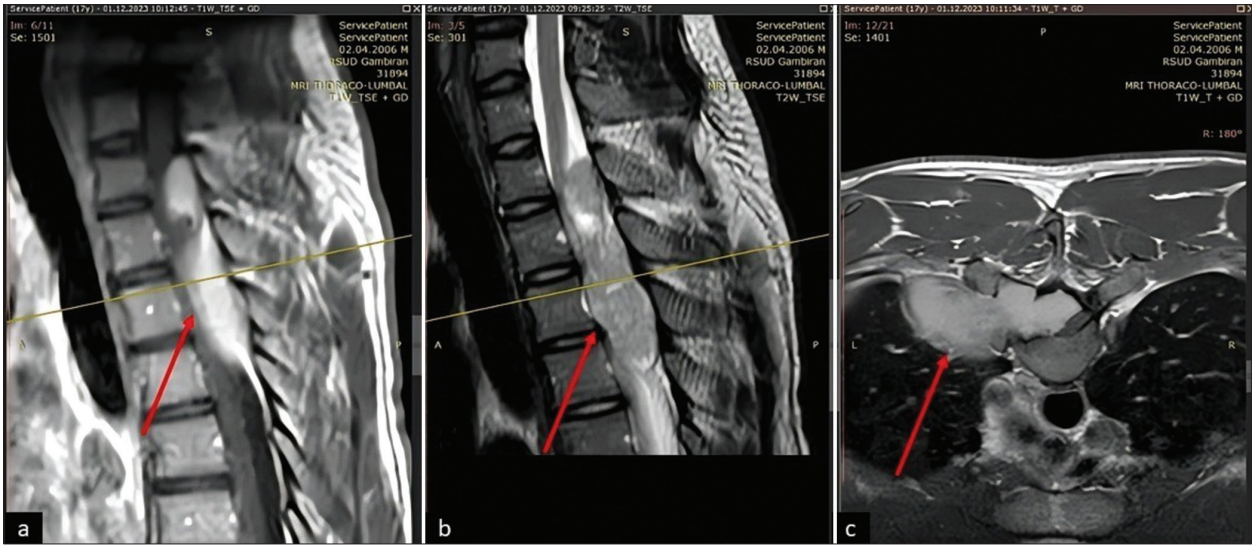


**Figure 1:** Thoracolumbar magnetic resonance imaging demonstrating a solid intradural extramedullary mass extending from T1 to T4. Red arrows indicate the mass in all images. a. T1-weighted sagittal with contrast. b. T2-weighted sagittal. c. T1-weighted axial with contrast.

### Histology

Histological examination confirmed the diagnosis of PS, revealing a growth pattern with whorling, varying cellularity, spindle-shaped cells, and Verocay bodies. Immunohistochemistry: S-100 positive in both nuclei and cytoplasm, consistent with PS.

## DISCUSSION

IDEM PS are very rare clinical entities, with a few documented cases in the medical literature [Table 1].[1,2,4-6] Spinal PS can occur at various spinal locations and typically present symptoms or signs ranging from pain alone to paraplegia.[3] PS

is characterized by three distinctive growth patterns: multifocal, intraneural, and multinodular.[2] On MR scans, these lesions are uniformly hyperintense on T2-weighted image (T2WI); among five documented cases, four (including ours) showed hyperintense T2WI lesions, while the other two showed lower signal intensity. Histologically, PS and schwannoma both exhibit a dominant Antoni A pattern. However, PS can exhibit more complex growth involving multiple fascicles compared to the single fascicle involvement of schwannoma.[2,6] It is important to distinguish PS from plexiform neurofibroma; plexiform neurofibroma is often associated with neurofibromatosis type I, a genetic condition with the potential for malignant transformation, whereas PS is not.[2,6]

### Surgery for IDEM PS

Surgical management is the primary treatment for PS, typically resulting in a cure with complete tumor removal. Of the five cases documented in the literature, two underwent total extirpation, while three had subtotal extirpation. Total excision can be challenging due to tumor size and adhesion to neural structures; in some cases, the relatively benign prognosis of PS may justify subtotal excision. All reported cases were safely excised without complications, leading to a significant improvement in neurological symptoms. (i.e., no tumor progression over 1- and 2-year follow-up periods).

## CONCLUSION

IDEM PS of the thoracic spine is exceedingly rare. In this case, the tumor was successfully resected after being diagnosed using MR imaging.

### Ethical approval

The Institutional Review Board approval is not required.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

### Conflicts of interest

There are no conflicts of interest.

### Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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