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CASE REPORT

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Giant spinal thoracic dumbbell Schwannoma in pediatric

Schwannoma gigante en mancuerna de columna vertebral torácica en pediatría

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ABSTRACT

Introduction: Giant spinal thoracic dumbbell schwannoma is a benign tumor extremely rare in pediatric age, which may be associated with neurofibromatosis. **Case description:** A 14-year-old girl who presented paresthesia in the lower extremities and back pain of 6 months onset, with 2 weeks of neurological deficit progression, associated with clinical data of neurofibromatosis. Posterior total excision was performed in a single surgery with transpedicular instrumentation without complications. **Conclusion:** This is the second case of type IVb pediatric thoracic giant spinal schwannoma reported, and the largest extracted by single posterior route in a single surgical time without complications.

Keywords: Schwannomas, pediatric spine, surgical approach, neurofibromatosis, spinal tumor.

RESUMEN

Introducción: *El schwannoma torácico espinal gigante con mancuerna es un tumor benigno extremadamente raro en la edad pediátrica, que puede estar asociado a neurofibromatosis.* ***Descripción del caso:*** *Niña de 14 años que presentó parestesias en miembros inferiores y dolor lumbar de seis meses de evolución, con dos semanas de evolución del déficit neurológico, asociado a datos clínicos de neurofibromatosis. Se realizó escisión total posterior en una sola cirugía con instrumentación transpedicular sin complicaciones.* ***Conclusión:*** *Este es el segundo caso reportado de schwannoma espinal gigante torácico pediátrico tipo IVb, y el mayor extraído por vía posterior única en un solo tiempo quirúrgico sin complicaciones.*

Palabras clave: *Schwannomas, columna vertebral pediátrica, enfoque quirúrgico, neurofibromatosis, tumor espinal.*

Introduction

Spinal schwannoma is a slow-growing encapsulated benign tumor that originates in a myelinated nerve sheath with a diagnostic peak between 4 and 5 decades of life in a male: female ratio of 1:1, constituting up to 30% of all spinal tumors and are usually located in the intradural extramedullary region (72%), they can also be located extradurally (13%), intradural and extradural (13%) and intramedullary (1%).[1] Giant Schwannomas can be dumbbell shaped tumors that invade other body cavities or vertebral bodies as K. Sridhar mentioned in 2001 when developed the current morphological classification.[2]

Case presentation

A 14-year-old girl who presented paresthesia in the lower extremities and back pain of six months onset,

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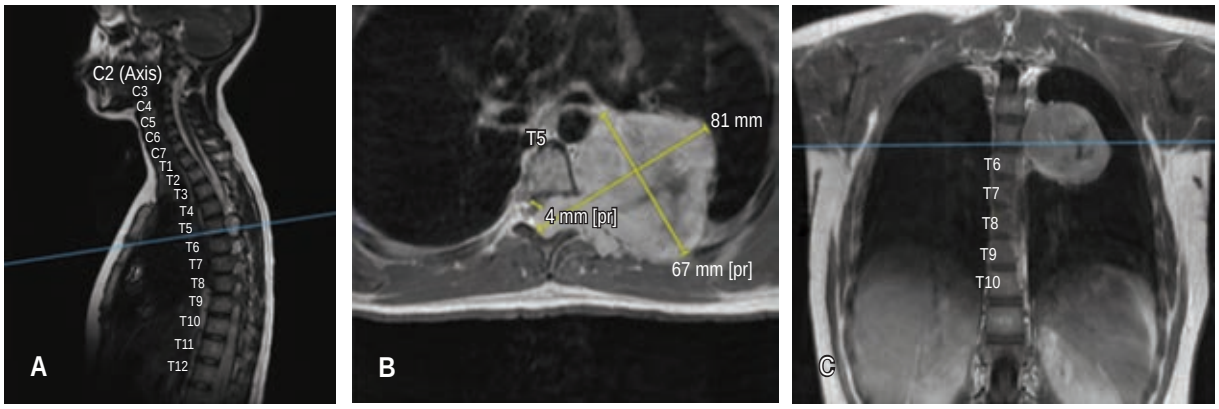

Figure 1: Magnetic resonance study. **A)** Sagittal view T2 sequence that shows the foraminal trayect of T5-T6 level of the Schwannoma. **B)** Axial view T1 sequence where the dumbbell tumor can be visualized with the measures of 81 × 67 mm. **C)** Coronal view T1 sequence left intrathoracic expansion with delimited borders.

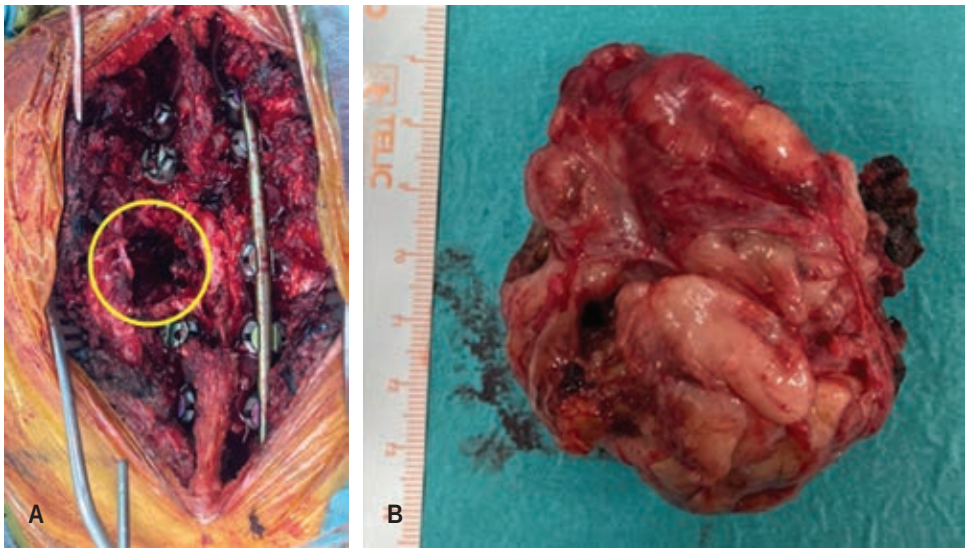

Figure 2:

Intraoperative images. **A)** Posterior approach with transpedicular instrumentation, yellow circle shows T5-T6 costotransversectomy for tumoral resection. **B)** Macroscopic capsulated tumor specimen.

with two weeks of neurological deficit progression, strength 3/5 and sensitivity 1/2 in the lumbosacral plexus with adequate sphincter control. Cafe-au-lait spots on back, abdomen and legs, rest of neurological examination normal. Magnetic resonance study was taken where the tumor is visualized at T5-T6 level that compromises 80% of the medullary canal and extends to the mediastinum and thorax, in the shape of a giant dumbbell with defined edges with a size of 81 × 67 × 55 mm *(Figure 1)*. Emergency surgery was performed due to progressive deficit with a posterior approach, placing transpedicular instrumentation of bilateral T2-T4, right T6, bilateral T7-T8 plus left costotransversectomy at left T5-T6 level that improved the entire intracanal and thoracic tumor without present complications *(Figure 2)*. The tumor was sent to pathology and the genetic study was carried out; the result was positive for Schwannoma with association to Neurofibromatosis type 2. Currently with 9 months of neurologically complete follow-up and without local recurrences of the tumor *(Figure 3)*.

Discussion

Spinal tumors are a relatively rare diagnosis, accounting for 1 to 10% of all pediatric central nervous system tumors. Pediatric spinal schwannomas constitute 2.5 to 4% of all pediatric spinal tumors, with a female: male ratio of 2:1, data that contrast with the

adult population.[3] Additionally, giant spinal schwannoma is an even rarer presentation, which may or may not be associated with neurofibromatosis.[4] Within the Sridhar classification, there is only one 14 years old male with a schwannoma type IVb (dumbbell) reported at T7-T8 level that invaded the thorax requiring a single posterior approach by costotransversectomy for its complete macroscopic excision, complementing with transpedicular instrumentation, which had complete clinical improvement at six months.[5] Case like ours that was treated with a single posterior approach plus transpedicular instrumentation to avoid risk of instability,[6] despite being larger, we obtained excellent postoperative results, complete neurological recovery without recurrence at nine-month follow-up *(Table 1)*.

Conclusions

Giant thoracic type IVb spinal schwannoma is extremely rare in pediatric age. At the present time,

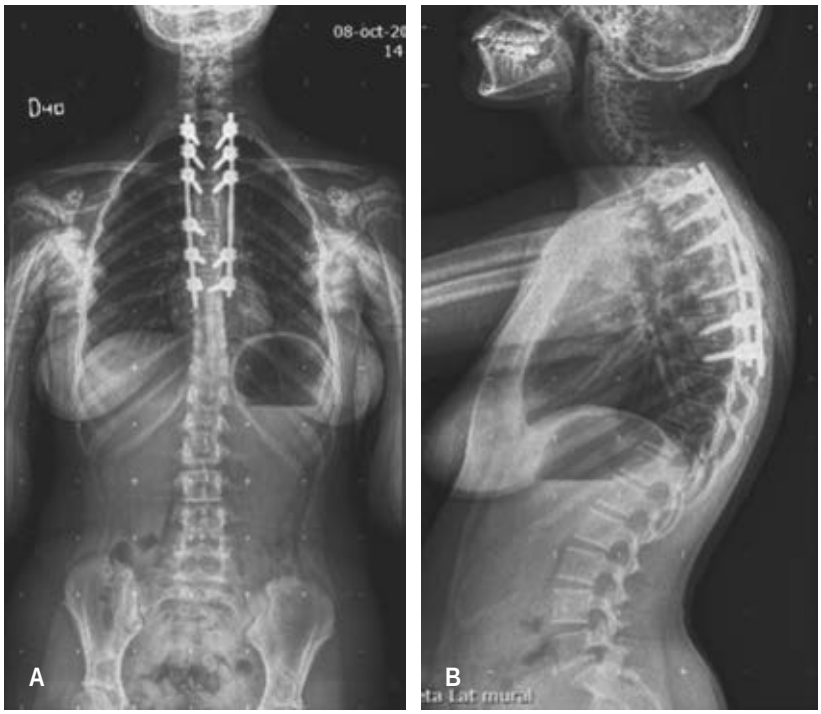

Figure 3:

A) Radiography anterior posterior view that shows transpedicular instrumentation from T2 to T8. **B)** Lateral radiography with adequate sagittal alignment. There is no evidence of tumoral recidive.

Table 1: Cases of pediatric giant spinal thoracic dumbbell Schwannoma.

Case	Author	Age	Sex	Localization sites of tumor	Size of the tumor	Symptoms	Treatment	Outcome
1	Vadivelu S, et al (2013)	14	Male	T7-T8	40 × 50 × 64 mm	Dorsal pain, bilateral pain and weakness of legs, unsteady gait	Lateral extracavitary approach with posterior transpedicular arthrodesis of T6-T8	Without neurologic alterations at 6 months of surgical procedure
2	Marroquin-Herrera, et al (2021)	14	Female	T5-T6	81 × 67 × 55 mm	Dorsal pain, and paresthesias of bilateral legs, after 2 weeks the patient presents weakness 3/5 and alterations in sensitivity in both legs. Without alterations of sphincters	T2-T4 bilateral transpedicular instrumentation, T6 right transpedicular instrumentation, T7-T8 bilateral transpedicular instrumentation, T5-T6 left posterior costotransversectomy	Without neurologic alterations and no evidence of tumoral recidive after 9 months of the surgical procedure

only 1 case has been reported in the world literature, this is the second report. Both cases were treated by a single posterior approach, with total macroscopic resection plus transpedicular instrumentation with good postoperative outcomes.

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Conflict of interest

The authors declare no conflict of interest.