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%). 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.

Case presentation

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

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.
adult population. Additionally, giant spinal schwannoma is an even rarer presentation, which may or may not be associated with neurofibromatosis. 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. Case like ours that was treated with a single posterior approach plus transpedicular instrumentation to avoid risk of instability, 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, the few cases that have been described in the literature indicate that this pathology should be considered as a rare entity. Our case is the first reported in the literature that demonstrates the feasibility of using the lateral extracavitary approach to excise a dumbbell-type IVC schwannoma, which invaded the thoracic cavity, making the thoracic approach unnecessary.

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

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

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.
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.

References


Conflict of interest

The authors declare no conflict of interest.