Solitary Dorsal Intramedullary Schwanomma – A Rare Lesion

Schwanomma intramedular dorsal solitário – Uma lesão rara

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Abstract

Intramedullary schwanommas are rare, and most cases are reported in cervical region. Less than 20 dorsal intramedullary schwanommas have been reported till date in literature. This is due to their cell of origin, the Schwann cell, which is not normally found within the parenchyma of the brain and spinal cord; therefore it is not surprising that these lesions are rare. We report a rare solitary dorsal intramedullary schwanomma in a young adult patient who presented with paraplegia.

Keywords

► dorsal
► intramedullary
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Palavras-chave

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Introduction

Schwanommas account for 30% of primary intraspinal tumors. Intra-spinal schwanommas are usually located in the intra-dural extramedullary (IDEM) space, and are rarely intrame- dullary. Intramedullary schwanommas (ISs) only account for 0.3% of intraspinal tumors, and for 1.1% of intraspinal schwanommas.1 Most ISs are found in the region of the cervical cord, and less than 20 cases of been reported in dorsal region to date. The cell of origin of the schwanomma is the Schwann cell, which is not normally found within the parenchyma of the brain and spinal cord; therefore, it is not surprising that these lesions are rare. Several theories have been postulated to explain the origin of these tumors, but none has gained universal acceptance. We report a rare case of solitary dorsal IS in a young patient who presented with paraplegia.2,3

Case

A 20-year-old female patient presented with back pain that had been felt for 1 year, with gradually progressive weakness in both lower limbs in the previous 2 months. Upon neurological examination, she had paraparesis in both lower limbs with a power of 3/5 on the myelomeningocele (MMC) scale. She also had bowel and bladder incontinence. Her sensory examination revealed loss of sensation below the level of D5.
She was submitted to a magnetic resonance imaging (MRI) scan of the dorsal spine with contrast, which revealed a well-defined heterogeneously enhancing intramedullary lesion with expansion of cord and perilesional edema at the level of D5 to D6 (Figs. 1–23). The patient operated with the differential diagnoses of intramedullary tuberculoma or glioma in mind. She underwent dorsal (D4 to D6) laminotomy with complete excision of the lesion. Intraoperatively, a greyish-white, well-defined, firm, non-suckable intramedullary lesion was found (Figs. 4–567). Postoperatively, there was minimal improvement in power in both lower limbs. The patient was discharged with an indication for physiotherapy and regular follow-up.

Fig. 1 Preoperative magnetic resonance imaging (MRI) scan of the dorsal spine with contrast showing the intramedullary contrast-enhancing lesion.

Fig. 2 Preoperative magnetic resonance imaging (MRI) scan of the dorsal spine with contrast showing the intramedullary contrast-enhancing lesion.

Fig. 3 Preoperative MRI of the dorsal spine: T2-weighted images showing the intramedullary lesion and cord expansion with syrinx formation.

Fig. 4 Preoperative MRI of the dorsal spine: T2-weighted images showing the intramedullary lesion and cord expansion with syrinx formation.
To our surprise, the histopathology was suggestive of spindle-cell tumor with pallisading architecture and intratumoral aggregates of pigments in the hemosiderin-laden macrophages with cystic changes that is, schwannoma (► Fig. 8).

Discussion

Spinal schwannomas are the most common primary spinal tumors, accounting for ~25% of primary intradural spinal cord tumors in adults. Males and females are equally affected, and the age of onset is usually between 25 and 50 years.

Hirano et al. reported an extended series of 678 spinal cord tumors: schwannomas were the most common histological type, with a slight prevalence of the male sex. The symptoms are related to tumor location and its proximity to the spinal...
cord. Most studies report pain as the first symptom, followed by sensory deficits. Motor deficits and sphincter impairment are observed relatively late. The gold standard for the preoperative diagnosis of spinal schwannoma is the MRI. Schwan-

noma is uncommon. Contrast-enhanced T1-weighted images better delineate the lesion and differentiate the solid from the cystic components and edema. After gadolinium administration, variable enhancement can be found, and heterogeneous enhancement is the most common, with a few cases showing homogeneous and circular enhancement. The preoperative diagnosis of an IS purely on radiological grounds is difficult, as it is also difficult to differentiate it from intramedullary gliomas. However, in ISs, the classic dumb-bell appearance is rarely observed.1,3

Histologically, schwannomas are composed of an Antoni-A cell areas comprising compact cells in a reticular framework, and Antoni-B cell areas comprising large cells in a loose collagenous background. Intramedullary schwannomas do not have any specific histological feature.6

As most of these lesions are well-demarcated, gross total excision of the lesion with minimal damage to the surrounding neural tissue remains the gold standard treatment. However, subtotal resection can be performed if it is adherent to surrounding neural tissue. The use of advanced neurosurgical techniques, surgical microscope and a cavitron ultrasonic suction aspirator (CUSA) have resulted in better removal of intramedullary tumors by reducing tumor volume, with minimal retraction of the spinal cord, yielding better results.3

Conclusion

Intramedullary schwannomas are histologically benign tumors, and complete functional recovery can be achieved after early total excision. They are difficult to diagnose preoperatively, as there are no pathognomonic signs that enable its differentiation from other intramedullary tumors. Therefore, IS should be considered in the differential diagnosis of an intramedullary lesion in the thoracic spine.

Conflict of Interests

The authors have no conflict of interests to declare.

References