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Giant Thoracic Cystic Intradural Extramedullary Schwannoma Causing Severe Abrupt Paraparesis: A Case Report and Review of the Literature

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Abstract

Background: Spinal schwannomas represent approximately 20% - 30% of all spinal tumors and are the most common type of intradural extramedullary tumors. These slow-growing tumors arise from spinal nerve root sheaths and can cause progressive spinal cord compression, which is usually seen in many patients. They are mainly solid or heterogeneous in nature and can increase in size, undergo degenerative changes in some rare cases leading to cyst formation, hemorrhage, calcification, and hyalinization. However, pure cystic intradural extramedullary schwannomas are very rare and uncommon. The severity of these symptoms depends on the location and the size of the tumor on the spinal cord. The aim of this study is to describe an unusual manifestation of this tumor, to discuss the sudden deterioration observed in this case, and to emphasize the differential diagnosis. **Case Presentation:** We report a case of a giant cystic schwannoma in a 17-year-old girl revealed by a severe abrupt paraparesis. Magnetic resonance imaging revealed a huge intradural extramedullary lesion that extended from T10-T11 and caused a remarkable spinal cord compression. This lesion had been totally removed surgically and the postoperative course was uneventful. **Conclusion:** Cystic schwannomas are rare spinal lesions responsible for cord compression in some rare cases. If symptomatic, complete surgical resection associated with physiotherapy might be the goal despite the clinical stage of presentation.

Keywords

Intradural, Extramedullary, Cystic Schwannoma, Spinal Cord Compression

1. Introduction

Spinal schwannomas are the most common intradural extramedullary spinal tumors that arise from spinal nerve root sheaths. These benign tumors represent approximately 20% - 30% of all spinal tumors and are mainly solid or heterogeneously in nature [1] [2]. They can increase in size, undergo degenerative changes in some rare cases leading to cyst formation, hemorrhage, calcification, and hyalinization. However, pure cystic intradural extramedullary schwannomas are very rare and uncommon. They can cause symptoms depending on their size and location. Herein, we report a case of a 17-year-old girl manifesting with a severe abrupt paraplegia.

2. Case Report

A 17-year-old girl with no previous medical history presented within our department with 3 weeks history of progressive thoracolumbar back pain exacerbated by her daily activities. These symptoms had been complicated rapidly by intermittent neurogenic claudication, progressive ascending bilateral numbness with weakness in the lower limbs just 4 days before admission. Neurological examination showed a well-nourished girl with severe paraparesis (strength 1/5 in both limbs), presented hyperreflexia in deep tendon reflexes with bilateral Babinski sign and hypoesthesia below T12. Bladder and bowel functions were normal and there was no stigmata of neurofibromatosis. There was no history of trauma or infection. Magnetic Resonance Imaging (MRI) revealed an intradural extramedullary cystic lesion that extended from T10-T11, hypointense on T1-Weighted Image (T1WI), hyperintense on T2-Weighted Image (T2WI) with rim-enhancement on contrast administration (**Figure 1(A)**, **Figure 1(B)**). The biological data were normal. In prone position and after general anesthesia and under fluoroscopy, a T10-T12 laminectomy was made through a posterior midline approach. The exploration revealed a huge heterogeneous distended cystic lesion measuring 5 cm × 2 cm × 1 cm in its largest diameters, located dorsally to the dural sac. This cyst was opened sagittally to diminish the pressure on the spinal cord. The contents of this cyst were yellowish. The cyst wall was progressively dissected from the spinal cord and completely removed from the adjacent nerve root sleeves. Histopathological study of excised tissues stained with hematoxylin and eosin confirmed the diagnosis of schwannoma with cystic changes (**Figure 2**). Low back pain rapidly resolved immediately and she was discharged 6 days after surgery and referred to intensive physiotherapy for rehabilitation. The postoperative course was uneventful and she recovered her mobility just 02 months after surgery. Postoperative MRI 6 months later showed no recurrence (**Figure 1(C)**).

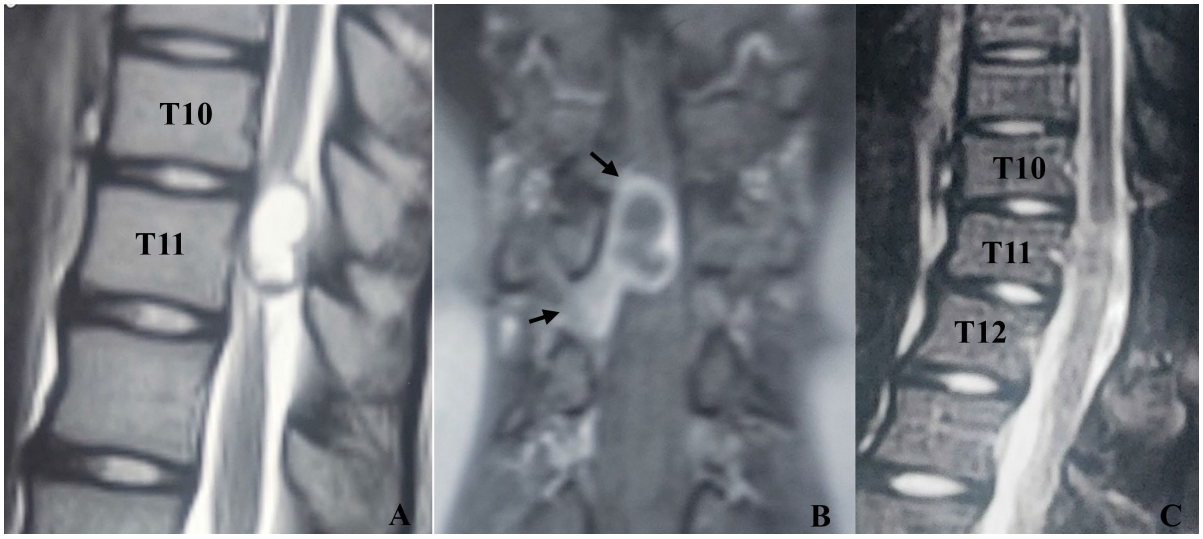

Figure 1. Sagittal T2 WI and coronal T1WI showing a hyperintense cystic intradural extramedullary lesion (A), with rim enhancement and prolongated adjacent nerve roots sleeves after gadolinium administration (black arrow) (B). Postoperative MRI showing complete resection of the lesion (C).

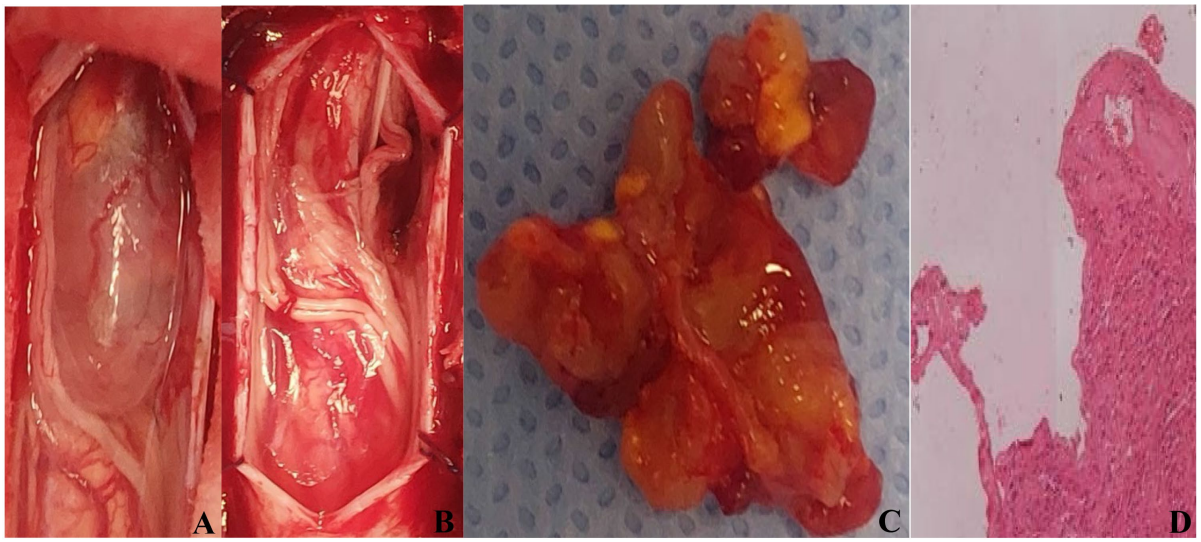

Figure 2. Operative view of intradural extramedullary cystic schwannoma before (A) and after complete resection (B) with cyst wall dissected from spinal cord (C) and sending for histopathological study confirming the diagnosis of schwannoma with cystic changes (D).

3. Discussion

Schwannomas represent the most frequent intradural extramedullary tumors accounting for 20% - 30% of all spinal tumors [1] [2]. They are mostly solid or heterogeneous in nature and are mainly seen in adults in the fourth or fifth decade of life [3] [4]. Pure cystic formation in schwannomas is a very uncommon phenomenon. The mechanism of this event might be resulting from Antoni B cells' degeneration or central ischemic necrosis within the tumor [5] [6]. The clinical manifestations ranged from progressive myelopathy to abrupt neurologic deterioration as seen in our case. The abruptness of this presentation led to a preliminary

diagnosis of tumor with hemorrhage or sudden expansion of the cyst. These data were supported by Jenkins *et al.* [7]. In this category, males are slightly more affected than females [8]. These tumors are more commonly seen in the lumbar and lumbosacral regions and less frequently in the thoracic and cervical regions [8] [9]. In the literature, about 57 cases of cystic intradural extramedullary schwannomas (including ours) have been described. The mean age of presentation was 47.7 years ± 13. Our patient was a 17-year-old and was the youngest published case reported in the recent literature. The most common clinical sign was back pain followed by muscle weakness and sensory deficit in 81.8%, 68.2% and 54.5% [8]. The real presentation of such lesions highly varies depending on the tumor location, size and the other related symptoms including gait difficulties, abnormal reflexes or sphincter dysfunctions and control disturbances [9] [10]. In all these patients, the neurological status ranged from abnormal reflexes to spastic paraparesis [8] [9] [11]. Our patient was 17 years old and was also admitted with severe paraparesis (strength 1/5), another historical event never related before. MRI with and without contrast strongly suggested the diagnosis of cystic schwannoma and was considered to be the best tool to investigate these tumors [12]. In the literature, on MRI cystic schwannomas seem generally to be well-defined, rounded lesions with or without adjacent bone remodelling or extension in the neural foramina. They are hypo or isointense on T1-Weighted (T1W) images, and variable degrees of hyperintensity on T2-Weighted (T2W) images with thick well-enhancing rim enhancement after gadolinium administration compared to other intraspinal cystic lesions such meningioma (cyst within the tumor with dural tail sign often associated to osseous erosion or soft-tissue calcification), epidermoid (slightly hyperintense to surrounding tissue on T1WI and show similar intensity to CSF on T2W images with no enhancement after contrast administration), dermoid (seen with dermal sinus tract and showed hypersignal intensity that is suggestive of fat tissue) or arachnoid cyst (isointense to Cerebrospinal Fluid (CSF) on all pulse sequences and do not show enhancement after gadolinium injection and are usually located posteriorly to the thecal sac). As for the neuroenteric cysts, the lesions are multi-lobulated, extended, and are usually located in the anterior intradural extramedullary region. Hydatid cysts have multiple well-circumscribed cystic lesions, internal echoes, and daughter cysts [5] [11] [13]-[15]. All these reported cystic schwannomas, excluding one underwent surgery with complete resection in most of them [8] [15]. Complete resection is safe and remains the treatment of choice in cystic schwannomas. Despite her advanced clinical signs, she had been operated successfully without delay and completely recovered in just 02 months postoperatively. In the literature, the best surgical outcome is strongly related to earlier diagnosis, meticulous investigation, and total resection [4] [11] [12]. Good to excellent outcomes are the rule in such conditions.

4. Conclusion

Cystic schwannomas are rare spinal lesions responsible for cord compression in

some rare cases. Contrast MRI is the investigation of choice and plays a major role in predicting these lesions preoperatively. Complete surgical resection associated to physiotherapy might be the goal despite the clinical stage of presentation.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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