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Case Report

Giant lumbar spinal schwannoma: a case report and literature review ☆,☆☆

Thi Kieu Loan Nguyen, MD.^a, Nhu Quynh Vo, MD.^a, Dac Hong An Ngo, MD.^a,
Trong Binh Le, MD. PhD.^a, Thanh Minh Nguyen, MD.^b, Thao Nguyen Thanh, MD. PhD.^{a,*}

^a Department of Radiology, University of Medicine and Pharmacy, Hue University, Hue, 53000, Vietnam

^b Department of Surgery, University of Medicine and Pharmacy, Hue University, Hue, Vietnam

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ABSTRACT

We report a case of a 40-year-old female patient admitted to the hospital due to lumbar pain that spread to both legs and was associated with weakness of the lower extremities. Magnetic resonance imaging revealed an intradural - extramedullary tumor at the level of the T12 - L2 vertebra. The lesion was over 7 cm in greatest diameter and compressed the conus medullaris. The patient underwent surgery to remove the entire tumor. Postoperative pathology confirmed the diagnosis of schwannoma. The symptoms resolved almost completely without significant complications.

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Introduction

Nerve sheath tumors can be found in different regions of the spinal cord, with the cervical cord being the most common site, followed by the thoracic and lumbosacral regions [1, 2]. Spinal schwannomas, along with meningiomas, are the most common spinal nerve sheath tumors [3, 4]. Schwannomas are often intradural- extramedullary masses and are more commonly seen in adults. Most schwannomas are solitary (90%), benign, and sporadic [5]. Multiple schwannomas often associated with neurofibromatosis type 2 [6]. Most patients present with mild sensory symptoms consisting of sudden pain or paraesthesia with nerve palpation; weakness can occur, but is

less common [7]. Typical characteristics of schwannomas on magnetic resonance imaging (MRI) are a well-defined mass, isointensity on T1W, heterogeneously hyperintensity on T2W, and intense heterogeneous enhancement on T1W after intravenous contrast administration [8]. Herein, we report a case of a 40-year-old female patient diagnosed with giant spinal schwannoma.

Case report

A 40-year-old female patient presented with a history of low back pain for two years that had spread down both legs. Her

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* Corresponding author.

E-mail address: ntthao@hueuni.edu.vn (T. Nguyen Thanh).

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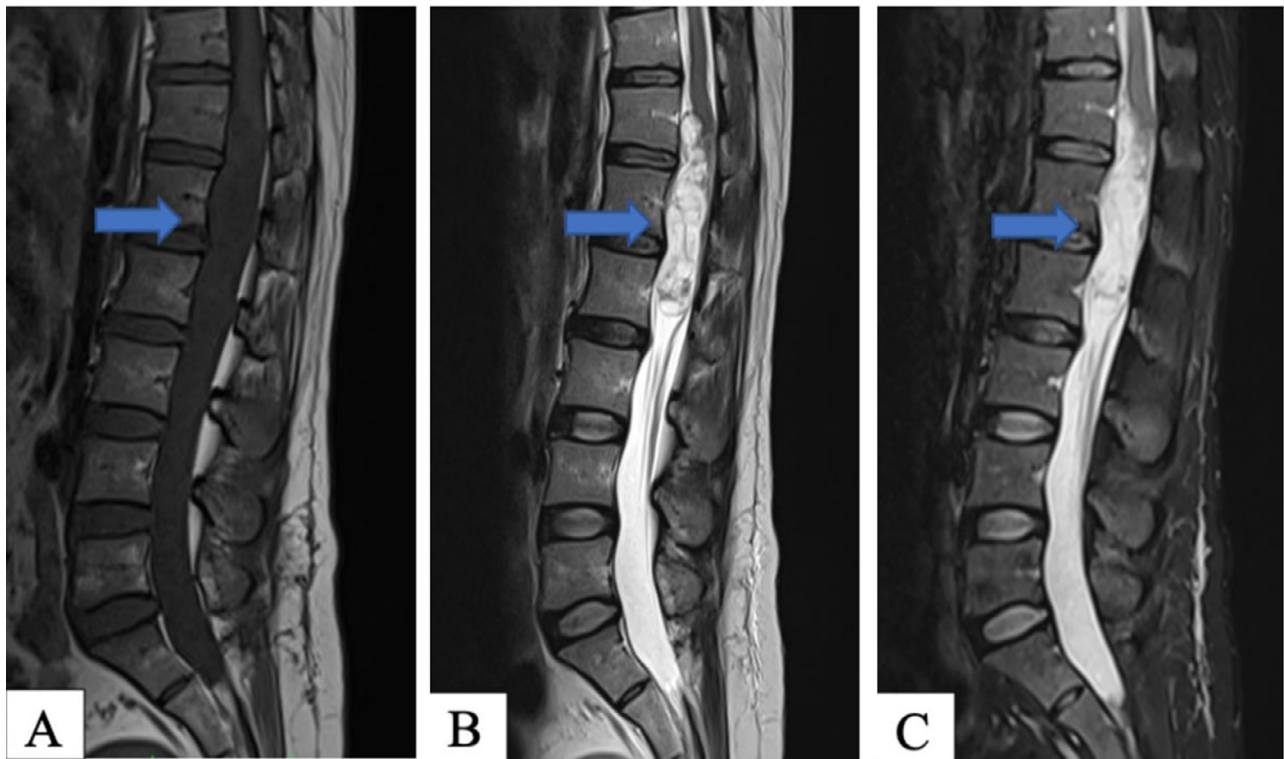


Fig. 1 – Median sagittal plane on T1W (1A), T2W (1B), and STIR (1C) of the lumbar spine showing an extramedullary – intradural lesion at the level of vertebra T12-L2 (arrows). The mass is hypointense on T1W, heterogeneous hyperintense on T2W and shows no signal suppressed on STIR.

symptoms worsened upon bending forward or walking and improved when sitting. She could not lie on her back because of the pain and had to sleep in a sitting position. On physical examination, there was evidence of weakness of the lower extremities and mild urination difficulty. The Lasque sign was positive at 50° on the right leg and 70° on the left leg. Schober test was 12/10 cm. Neri's sign was positive. The neurological reflexes were limited. The patient did not show any sign of paresthesia or sensory disorder in her legs.

Lumbar spinal magnetic resonance imaging (MRI) (Fig. 1) revealed an intradural - extramedullary mass of 72 × 21 × 18 mm in dimension, with left deviation, compressing the cauda equina at the level of the T12 - L2 vertebra. It had a hypointense signal on T1W sequence and a hyperintense signal on the T2W sequence, which was enhanced heterogeneously after injection of intravenous gadolinium (Fig. 2). The mass was partially hyperintense on DWI and hypointense on ADC, demonstrating diffusion restriction (Fig. 3). The mass had a well-defined border and occupied most of the posterior and left spinal canal. The patient was diagnosed with schwannoma of the conus medullaris and underwent surgery to remove the tumor. During surgery, the tumor was partially adhered to the dura but had not yet invaded the spinal cord. It was gently separated from the cord without damaging the conus medullaris. Macroscopically, the tumor had a smooth capsule with Dural attachment (Fig. 4). Histopathology results confirmed a schwannoma tumor. Microscopically, the mass showed hyperplastic tissue of spindle cells arranged in mul-

tidirectional intersecting layers to form bundles (Fig. 5). No other abnormal cell type was detected. Institutional Review Board approval was waived due to the nature of the case report. The patient gave consent to publish the case details and any accompanying images.

Discussion

Schwannoma, along with meningioma are the most common intradural - extramedullary tumors. Both entities can be associated with neurofibromatosis [6]. Schwannomas are typically benign tumors arising from the spinal nerve root sheaths and are the most common intradural extramedullary spinal tumors [6]. Schwannomas are related to Schwann cells that produce the myelin sheath of peripheral nerves. The incidence of spinal schwannomas varies with age. The incidence of spinal schwannomas varies with age. The peak incidence is in the fourth to fifth decade of life [9, 10]. There was no reported correlation between the prevalence of this tumor and sex. Symptom vary depending on the involved regions. Early-stage root pain is due to disruption of nerve transmission (direct or indirect neuritis or compression of the tumor) [11]. Later, when spinal cord compression is more severe, weakness of the lower extremities or myelopathy may present. Symptoms of motor paralysis rarely occur initially [9].

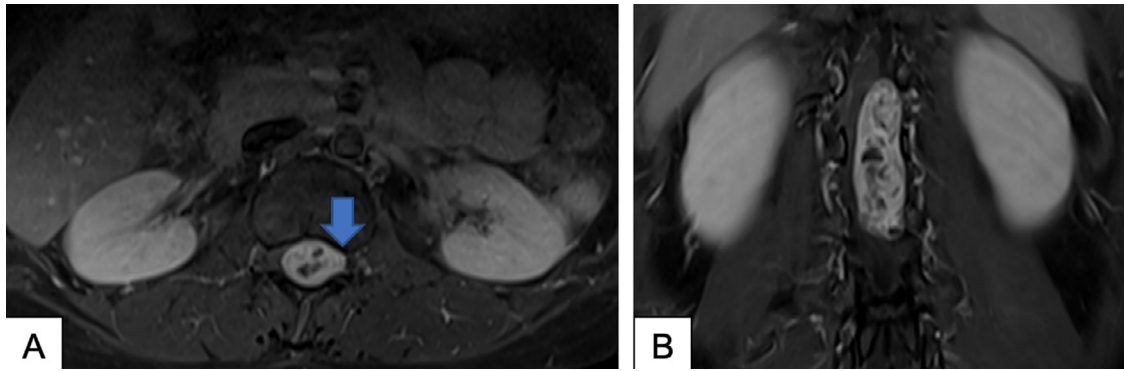


Fig. 2 – A, B. Axial (A) and coronal (B) T1W FATSAT sequence after injection of gadolinium showed the intradural extramedullary mass (arrow) with heterogeneous enhancement.

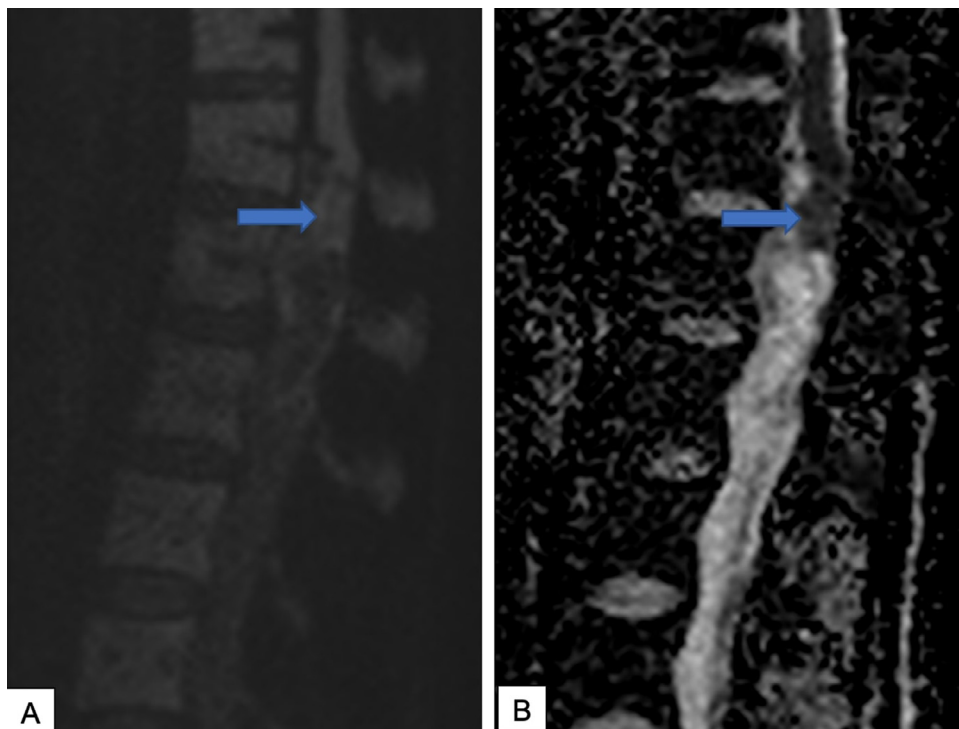


Fig. 3 – On sagittal DWI (A) and ADC map (B), the mass (arrows) shows partial diffusion restriction (arrows).



Fig. 4 – Macroscopic image of the tumor. The tumor was 72 × 21 × 18 mm in dimension with a smooth capsule and dural attachment (black arrow).

On MRI, schwannomas are commonly isointense on T1W images and hyperintense on T2W images with heterogeneous enhancement after intravenous administration of gadolinium [6]. Its heterogeneity is not an indicator of malignant changes [12]. Large lesions may show extension into neural foramina and erosion of the posterior aspect of the vertebral body, creating the “dumbbell” sign [6]. The differential diagnosis of schwannomas is primarily based on the location, clinical presentation, age, and sex of the patient [13]. Schwannomas are typically round, encase nerve roots and often present with hemorrhage or cystic changes, while neurofibromas are usually fusiform, homogeneous and displace nerve roots due to asymmetric growth [5]. On MRI, the “dumbbell” sign is suggestive of schwannomas, as it is present in 69% of schwannomas, and is less common in neurofibromas (12%)

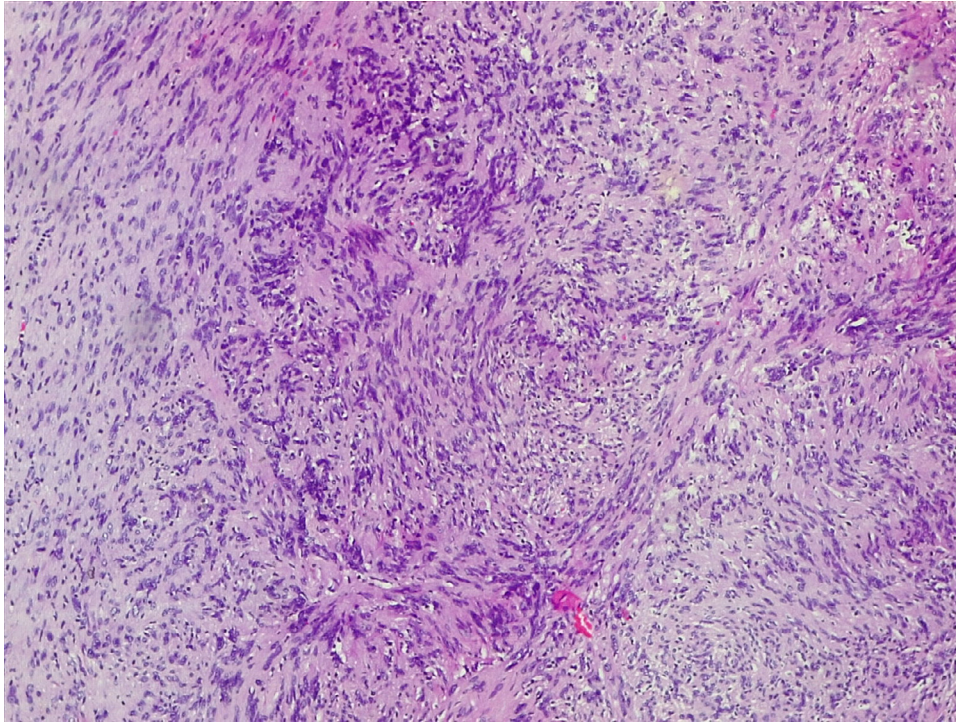


Fig. 5 – Histopathology showed hyperplastic spindle cells and tumor cells layered in multiple directions to form bundles and intersect, consistent with a schwannoma. No other abnormal cell detected. HE, X100.

and meningiomas (5%) [14]. One of the differential diagnoses for intradural-extramedullary spinal lesions in middle-aged females was meningioma. Meningiomas are commonly located at the upper and mid thoracic levels [14]. Iso-/hypo intensity on T1W and slight hyperintensity of the spinal cord on T2W with intense enhancement and a “Dural tail sign” are the most common characteristics of meningiomas. In fact, spinal intradural-extramedullary tumors with hyperintensity on T2W or intense enhancement without a “Dural tail sign” should be considered schwannomas [15]. A large schwannoma usually shows heterogeneity because of cystic degeneration or hemorrhage, while calcification is rare [16]. Other differential diagnoses should include chordoma, paraganglioma, ependymoma, and astrocytoma. In a case series report, 1/5 of spinal schwannomas were associated with syringomyelia, and in 20% of cases perilesional edema was observed [17].

Sridhar was the first to suggest a classification system of benign spinal schwannomas [18]. This classification was later modified by Kotil in 2014 [19]. Giant spinal schwannomas are defined as lesions that extend over more than two vertebral levels (type II), have an extraspinal extension of more than 2.5 cm (giant dumbbell, type IVb), or erode the vertebral bodies and extend posteriorly and laterally into the myofascial planes (giant invasive tumors, type V) [18, 19].

Surgical resection of the tumor is the first choice if feasible to improve the patient’s symptoms with a low rate of complications [3]. MRI characterization is essential in evaluation and treatment planning. In our case, the mass showed extension into the neural foramina and erosion of the posterior aspect of

the left vertebral lamina without signs of aggressive invasion. Therefore, surgical resection of the tumor was chosen for complete removal. The dura was opened from below. The tumor was covered by nerve clusters of the cauda equina that were dissected and lifted to sides to reveal the tumor and to observe the internal organization. The lower pole of the tumor was exposed and separated from the dura. The tumor was lifted toward the contralateral side of the spinal cord and separated from the conus medullaris. The nerve roots and blood vessels feeding the tumor were identified, burned, and cut to separate the tumor from the upper pole and remove it. Microdissection was performed to remove the entire tumor, delivering the best treatment results and a lower risk of recurrence. In our case, to limit complications during surgery that can result in weakness, curvature of the spine and possibly compression of the spinal cord or nerve roots, we performed posterior lumbar interbody fusion (PLIF) fixation of the spine to help immobilize the patient’s spine. Pathological exams contributed to accurately determining the nature of the tumor as well as guiding treatment for the patient.

Conclusions

Schwannomas are common nerve sheath tumors of the spinal cord. Our paper reports a giant schwannoma in the thoracolumbar spine that was symptomatic due to nerve compression. Clinical examination and MR imaging must be well coor-

dinated to achieve accurate diagnosis and contribute to surgical planning, mostly by microsurgery for complete tumor removal.

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