

Intradural Extramedullary Schwannoma of the Thoracic Spine with Intratumoral Haemorrhage

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Abstract

Schwannomas are the commonest intradural extramedullary tumors of the spine, mostly present in the cervical and lumbar regions. Spinal schwannomas with intratumoral haemorrhage are very rare lesions. Authors here present a 40-year-old male with a 2 year history of pain in the thoracic region and acute onset of spastic paraparesis and cauda equina syndrome. Magnetic Resonance Imaging (MRI) of his thoracic spine demonstrated a heterogeneous intradural extramedullary lesion with no associated enhancement. After emergent removal of the mass, histopathology revealed schwannoma with intratumoral haemorrhage.

Keywords: Spinal tumors; Schwannoma; Haemorrhage; MRI

Introduction

Spinal schwannomas are schwannomas arising from nerves within the spinal canal. They are the most common intradural extramedullary spinal tumors representing 30% of such lesions followed by meningiomas (25% cases) [1]. They are most commonly located in cervical and lumbar regions because of the higher density of nerve roots at that regions [2,3]. Haemorrhagic spinal schwannomas are usually presented with motor symptoms as well as with pain, numbness and paresthesias. Until present, only 11 cases of haemorrhagic schwannomas have been described in the international literature.

We report a case of a thoracic intradural extramedullary schwannoma with acute intratumoral haemorrhage in an adult male patient who had sudden onset of monoplegia of the right lower extremity and paraparesis of the left.

Case Report

40-years old male patients with symptoms of thoracic spine ache since 2 years ago. He presented to the emergency department of our hospital with increasing pain and numbness of bilateral lower limbs and associated symptoms of cauda equina syndrome within the last 24 h. Neurologic examination revealed bilateral Babinski sign and very brisk tendon reflexes. Severe muscle weakness of the left rectus femoris, quadriceps and anterior tibial muscles as well as of the right foot muscles on the motor examination also noted. MRI examination of the thoracic spine demonstrated an intradural mass lesion in the right lateral aspect of the intradural compartment at the level T4-T5 displacing the spinal cord to the left side (Figure 1). The tumor showed high signal intensity on non-contrast T1WI consistent with blood degrading products (Figure 2). After intravenous contrast administration, no evidence of abnormal enhancement pattern presents (Figure 3).

The patient was operated on emergency. Under general anesthesia, posterior laminectomy from T4 to T5 performed. A "chocolate" colored vascularized mass with a surrounding capsule and associated displacement of the regional spinal cord to the left and ventrally was identified and en bloc removed (Figure 4). Consequently, it was sent to the pathologic department for histopathologic correlation. Wound closure was performed in layers and the patient was transferred to the intensive care unit for recovery. Post-operative period was uneventful. The patient presented promptly significant clinical improvement and managed to become ambulant with support in 5 days.

In histological examination, a spindle cell tumor was observed. The tumor cells had elongated normal nuclei without atypia or mitoses and fascicular or storiform arrangement. Extensive haemorrhagic infiltration was observed, but no necrosis (Figure 5A). Immunohistochemically the tumor cells were positive for S100 protein (Figure 5B). According to the previously described features, the diagnosis of a schwannoma was documented.

Discussion

Schwannomas are benign encapsulated neoplasms of Schwann cells (WHO grade I). They arise eccentrically from their parent nerve. They are composed of spindle cells of types: Antoni A and Antoni B [3,4]. In the Antoni A pattern of growth, elongated cells with cytoplasmic processes are arranged in fascicles in areas of moderate to high cytoplasmic ratio with little stromal matrix; acellular areas lie between opposing rows of parallel nuclei (Verocay bodies). In the Antoni B pattern of growth, the tumour is less densely cellular with meshwork of cells along with microcysts and myxoid changes [1].

Patients commonly present with segmental pain. As spinal schwannomas usually arise from the dorsal sensory roots, radicular sensory changes also occur. Weakness is less common. Myelopathy may occur if the lesion is large.

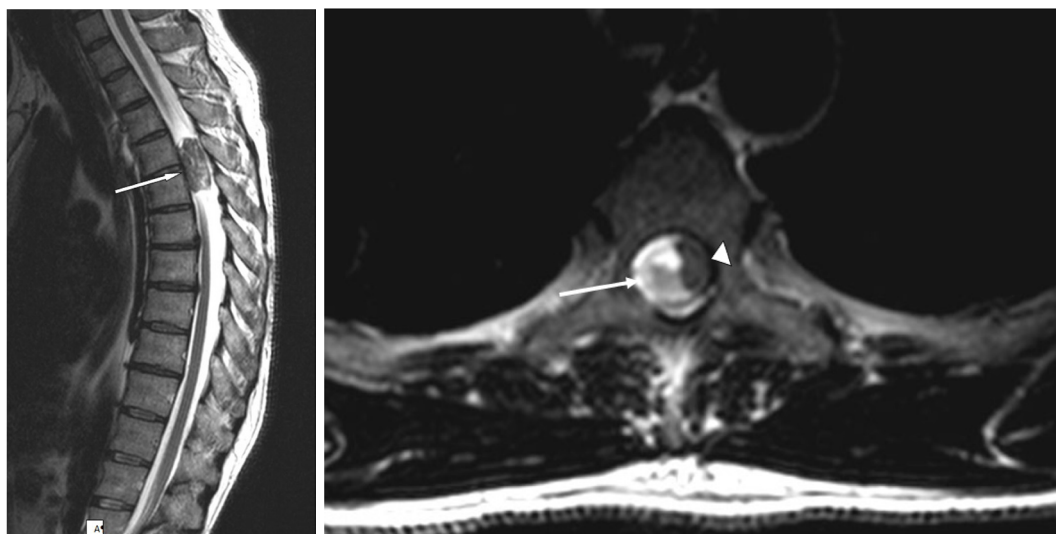
MRI is the examination of choice in the diagnosis of spinal tumors. Typical schwannomas are iso- or hypointense on T1-weighted image, markedly hyperintense on T2-weighted sequence and shows intense homogeneous enhancement in post contrast study [5]. Sometimes they are inhomogeneous on T2-weighted images with hyperintense and hypointense areas due to cyst formation, hemorrhage or dense collagen deposition [6].

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A

B

Figure 1: Sagittal (A) and axial (B) T2WI show an intradural mass at the level Th5-Th6 with low signal intensity (arrows) displacing the spinal cord to the left side (arrowhead).



Figure 2: On sagittal non-contrast T1WI the intradural tumor (arrow) shows high signal intensity due to the intratumoral blood products.

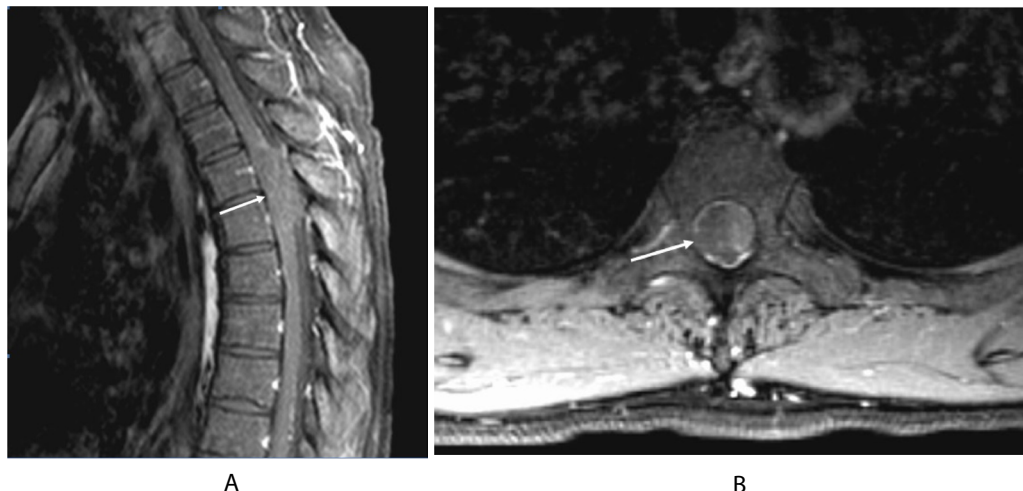


Figure 3: On sagittal (A) and axial (B) post-contrast fat-sat T1W images the intradural tumor (arrows) remains unenhanced.



Figure 4: Intraoperative view shows the hemorrhagic schwannoma (asterisk).

Haemorrhagic schwannomas show variable signal intensity on MRI depending on the ages of blood product. In acute haemorrhage the tumor is isointense on T1WI and hypointense on T2WI, in early subacute hemorrhage, as in our patient, hyperintense on T1WI and hypointense on T2WI. In late subacute hemorrhage appears hyperintense on T1/ T2WI and hypointense on T1/T2WI in chronic haemorrhage [7].

Two main theories have been proposed for the occurrence of tumoral haemorrhage. According to the vascular theory, there occurs spontaneous thrombosis of hyalinised tumor vessels with resultant distal tumor necrosis and secondary bleed or alternately, vascular tumors may get obliterated by endothelial proliferation with recanalization resulting in haemorrhage. The second one, mechanical theory, postulates that movements of spine induce traction on tumor vessels resulting in haemorrhage and this usually occur in thoraco-lumbar lesions [8-10]. Other postulates of haemorrhage (including subarachnoid space) include central ischemic necrosis due to tumor growth or malignant transformation with neovascularisation [9,11]. In

our case, because of the absence of prior clinical manifestations and occurrence of symptoms after a minor trauma, we speculate that the latter theory would be more suitable. This is due to its location in the cervico-thoracic region wherein, probably because of the increased mobility of this segment compared to the thoraco-lumbar ones noted in most other cases, haemorrhage would have occurred.

Conclusion

Intradural extramedullary schwannoma with intratumoral haemorrhage is a very rare occurrence. MRI represents the examination of choice in the diagnosis of an intradural extramedullary tumor with spontaneous haemorrhage. Patients with acute paraplegia from a haemorrhagic spinal tumor are considered as neurosurgical emergency. Urgent tumor resection represents the treatment of choice [12].

Conflict of Interest

All authors certify that they have no affiliations with or involvement in any organization or entity with any financial interest (such as

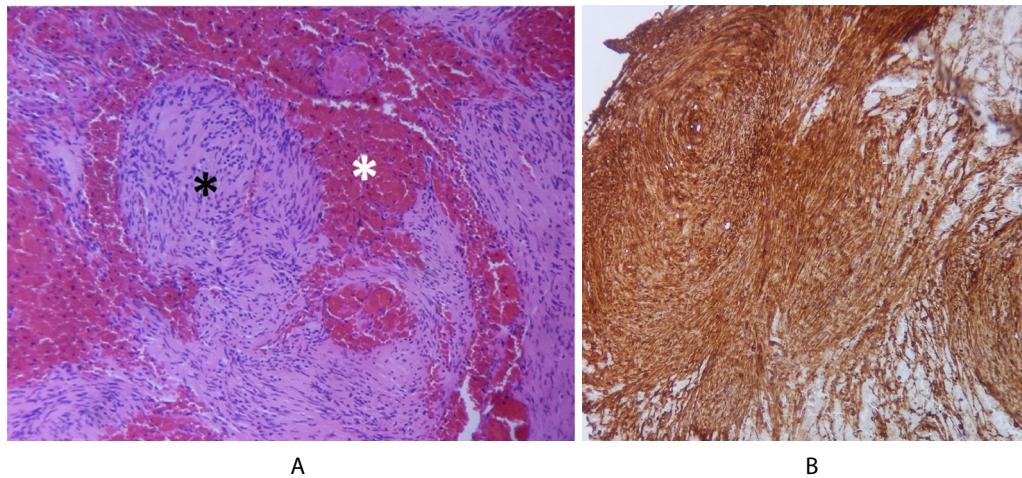


Figure 5: (A) Spindle cells without atypia or mitoses in fascicular-storiform arrangement (black asterisk) and extensive hemorrhagic infiltration (white asterisk) (H+E 200X). (B) The tumor cells are positive for S100 protein (Immunostain 200X).

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