



Case Report

Spinal Schwannoma presenting due to torsion and hemorrhage: case report and review of literature

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Abstract

BACKGROUND CONTEXT: The presentation of a tumor due to torsion, with hemorrhage from presumed reperfusion injury as a result of infarction of the lesion, is extremely rare and may be different than typical tumor presentation.

PURPOSE: The aim was to describe a patient with a rare case of twisted intradural nerve sheath myxoid Schwannoma.

STUDY DESIGN: This was a case report and a review of literature.

METHODS: A patient presented with acute onset of severe pain was found to have minimally enhanced intradural extramedullary cystic lesion. The patient underwent bilateral L2 and L3 laminectomy and microsurgically assisted intradural exploration.

RESULTS: At laminectomy and intradural exploration, it was found to be a Schwannoma, which had rotated above and below, with obvious color change consistent with either infarction or hemorrhage. Because the color change ceased abruptly at the site of the torsion, we presumed that the mechanism of the hemorrhage in and around the Schwannoma found at pathologic evaluation was due to the torsion. The torsion caused vascular insufficiency (likely venous) and produced subsequent reperfusion-related hemorrhage, because of the compression of the vascular supply coming from the proximal and distal ends of the root of origin. The patient did well with complete resolution of his symptoms and 11 years of pain relief.

CONCLUSIONS: This acute infarction of the tumor and the associated nerve caused the acute pain syndrome that is not commonly associated with lumbar Schwannomas. Patients with acute onset of severe radiating pain may have torsion of a benign tumor arising from the nerve in question. © 2015 Elsevier Inc. All rights reserved.

Keywords:

Schwannoma; Torsion; Pain; Spinal; Intradural tumor; Acute infarction of tumor

Objective

Schwannomas, a type of nerve sheath tumor, make up about one-third of primary spinal tumors [1–3]. Schwannomas develop from Schwann cells and are usually benign. Local pain and radiculopathy from compression of local structures is the

most common presenting syndrome; however, when the tumors are large enough, motor weakness or cauda equina syndrome may occur. The presentation of a tumor due to torsion of the lesion and its attached nerve is extremely rare [4], and hemorrhage within spinal tumors is also rare [5].

The following is a rare case of an intradural nerve sheath myxoid Schwannoma, which had twisted, resulting in torsion and hemorrhage within the lesion causing pain and weakness in the nerve distribution in question.

Clinical presentation

A 62-year-old otherwise healthy man presented to the emergency room because of extensive pain, and numbness

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in the right L4 distribution, and was unable to ambulate without assistance because of the pain. On examination, he was found to have mild (4/5) weakness of right dorsiflexion. A magnetic resonance imaging revealed an intradural lesion with cystic changes in the regions of L2 and L3 and minimal enhancement with gadolinium administration (Fig. 1, Left and Right). The abruptness of the presentation led to a preliminary diagnosis of tumor (benign, such as Schwannoma or metastatic) with hemorrhage, parasitic infection, or intradural focal abscess.

Operative findings

The patient underwent bilateral L2 and L3 laminectomy and microsurgically assisted intradural exploration. On exposure of the cauda equina, we were able to identify the lesion as a reddish round purplish mass arising from one nerve fascicle, approximately 1.5 cm in diameter. On further separating the rest of the cauda equina from the lesion, we found that the proximal and distal ends of the nerve on either side of the lesion had the appearance of more than one full turn of rotation of the nerve in a very focal manner, with associated compression of the concomitant vascular supply. This was associated with an abrupt border where

the root appearance changed within the coils of the torsion of the root, becoming profoundly darker, without spread above or below this point (Figs. 2 and 3). Provocative stimulation of the nerve failed to demonstrate any activation of either leg muscle electromyography, bladder manometry, or anal sphincter electromyography.

After bipolar cautery was applied proximally and distally to the rotated sections of the root on either side of the lesion, the root was cut with microscissors, and the lesion was sent in its entirety to pathology.

Histology

Received in formalin, the specimen consists of a tan-brown soft-tissue nodule measuring 1.8×1.2×1 cm. Microscopic sections using hematoxylin and eosin stains show neoplasm characterized by predominantly loose stroma with evenly distributed round to oval regular nuclei against a clear myxoid background, interspersed with thick-walled blood vessels and occasional more cellular foci with architecture occasionally reminiscent of palisading or Verocay bodies (Fig. 4). There is extensive recent hemorrhage throughout the tissue but no evidence of necrosis, atypia, or mitotic activity. The tumor is diffusely positive for

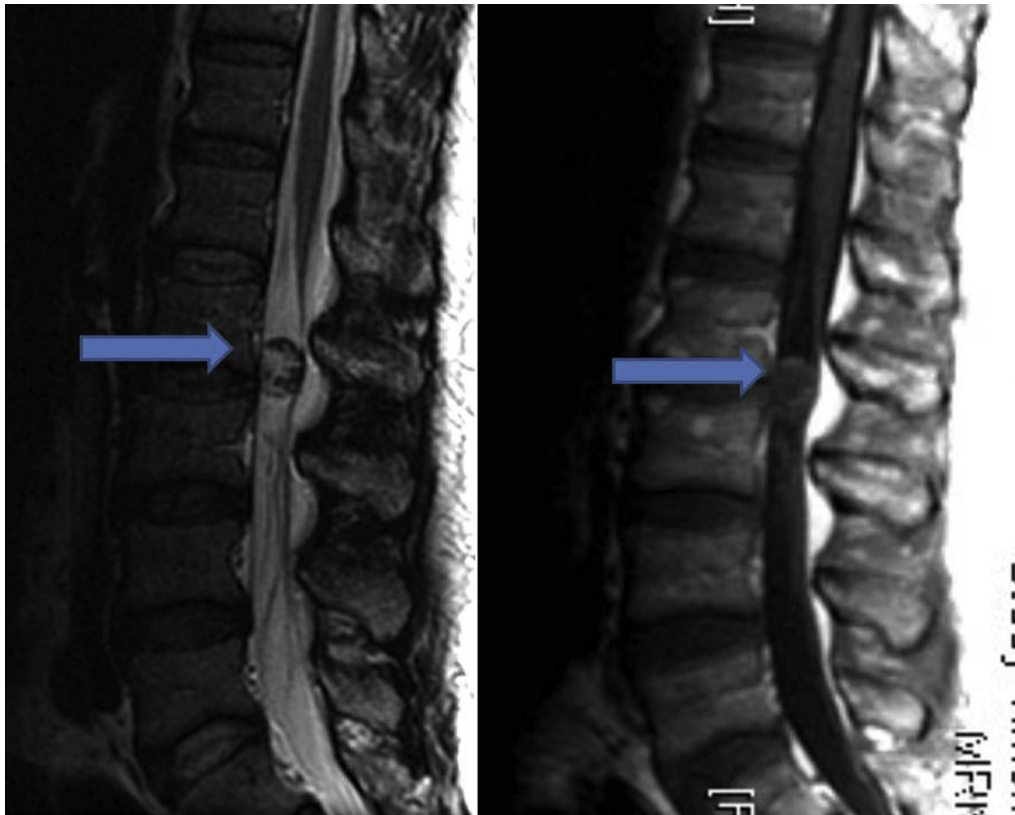


Fig. 1. (Left) Magnetic resonance imaging lumbar spine T2-weighted sagittal image showing the lesion, with heterogeneous internal architecture, in the intradural extramedullary space. The arrow points to the lesion. (Right) Magnetic resonance imaging lumbar spine T1-weighted sagittal image after gadolinium contrast infusion, with low homogenous uptake of contrast. The arrow points to the lesion.

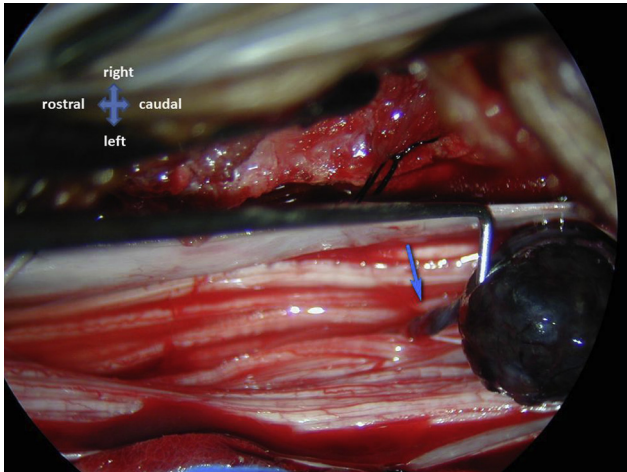


Fig. 2. Intraoperative photomicrograph, with the arrow demonstrating the proximal (rostral) rotation of the nerve root with the dusky appearance of the lesion distal to the point of the inferior torsion site.

S100 and shows weak isolated immunoreactivity with glial fibrillary acid-fast protein. Reticulin stain shows variably intense reticulin network surrounding individual cells of the neoplasm, whereas trichrome highlights the thick vessel walls and the stroma of both the myxoid and more cellular regions. Alcian blue is focally positive within the myxoid matrix. Additionally, there are scattered patchy zones of lymphohistiocytic infiltrate, evident by staining with leukocyte common antigen, and neurofilament is focally positive only adjacent to the tumor, in an adherent peripheral nerve but not in the tumor itself. Epithelial membrane antigen, chromogranin, and pancytokeratin (cam 5.2) are negative. Elastic Van Gieson stain used routinely to identify the internal elastic lamina in biopsy specimens is noncontributory. Taken together, the aforementioned findings are most consistent with a myxoid Schwannoma.

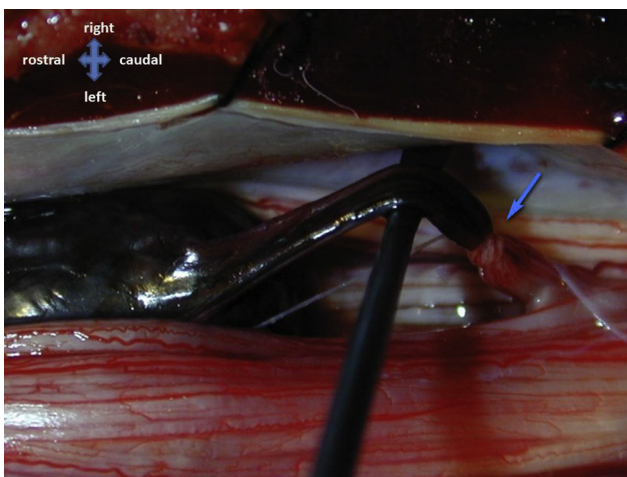


Fig. 3. Intraoperative photomicrograph of the caudal area below the same lesion, with the arrow demonstrating the rotation of the nerve root with the dusky appearance of the lesion proximal to (rostral to) the point of the superior torsion site.

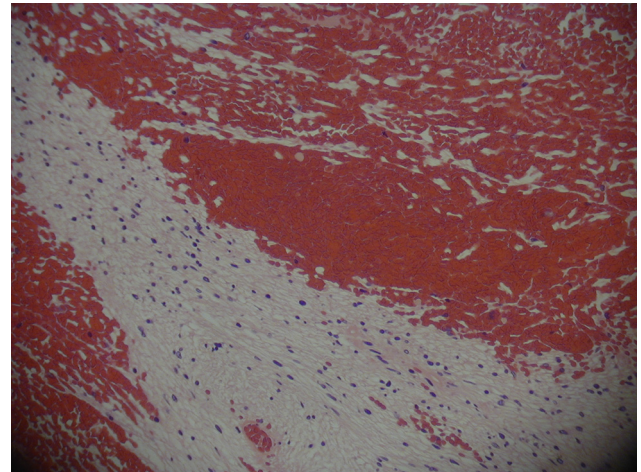


Fig. 4. Photomicrograph of the histologic staining with hematoxylin and eosin revealed a myxoid Schwannoma with recent hemorrhage.

Postoperative outcomes

After the operation, the patient was found to be moving all four extremities without any difficulty, and his preoperative pain was completely alleviated. A spine and brain magnetic resonance imaging survey with and without contrast done on postoperative Day 1 did not demonstrate any other lesions. Two months after the operation, he had no back pain, nor leg pain. A small amount of preexisting pain in the patient's left knee was present. On further follow-up 11 years after surgery, the patient reports that he has fully recovered, with no weakness, pain, or numbness. He is able to jog 6 miles at a time with no signs of neurologic compromise and is very satisfied with the outcome of his surgery.

Discussion

This is a very rare case of a nerve root Schwannoma, which caused excruciating pain due to torsion and presumed subsequent hemorrhage on the participating nerve. There is only one prior report of a nerve sheath tumor presenting with torsion [4] and one with hemorrhage [5]. We present the second documented case of torsion, presumed infarction, and subsequent hemorrhage within a cauda equina nerve root tumor resulting in acute symptoms and pain.

The blood supply to the nerves (and therefore to any nerve sheath tumors arising from nerves) within the cauda equina runs in both directions between the exiting nerve sleeve and the cord itself [6]. There is no bridging arterial or venous supply to the nerve, but typically a compression at one point will still allow circulation from either side, so the nerve does not become ischemic “downstream,” as there is no absolute “downstream.” Any factor that results in compression of the blood supply at two places, however, would result in ischemia in between the two obstructions. In such a case, a venous obstruction (which typically occurs at a lower pressure than arterial) may cause regional venous

engorgement and hemorrhage, or complete vascular obstruction followed by a partial reperfusion, which may result in a reperfusion injury and hemorrhage.

Because both ends of the nerve are fixed in relative position, rotation of a nerve sheath tumor in between the two ends of the nerve within the canal results in rotation of the nerve on either side of the tumor. This results in compression of the tissue at the point of rotation, resulting in kinking of the blood vessels. Kinking of the blood vessels would cause venous insufficiency initially rather than arterial because of the higher arterial pressures. The lack of enhancement of the tumor with gadolinium may be due to the continued decreased vascular supply from the persistent torsion, and this finding has been described previously in a patient with acute infarction of a Schwannoma [7].

There are known pulsations within the spinal canal, and this may contribute to focal movement of the nerve sheath tumor [8]. Within the region of the cauda equina, there is enough room for a lesion smaller than the diameter of the canal for such movement to occur. But the mechanism by which the tumor might become rotated and trapped in that rotated position is unclear. A ratcheting mechanism, possibly caused by subtle inhomogeneity on the surface, or adhesions of the arachnoid on the surface of the tumor, may result in rotation in one direction and impair potential for the lesion to rotate back. The observation that the subpial hemorrhage that seemed to propagate right up to the point of maximum torsion suggests that the subpial potential space was occluded by the tension of the torsion at that point, preventing further progression of the hemorrhage along the nerve.

The reason for the severity of the patient's pain is unclear, given that the sensory nerve ganglia are distal to the point of the tumor in the dorsal root ganglia under the pedicle of the body at the point of exit (more caudally located). This pain may represent either a localized subpial vascular irritation, tension on the nerve similar to a rubber band being twisted in model airplane, or some local pain fibers within the nerve itself. The immediate reduction in the pain after section of the nerve does not completely answer the question because the treatment in question

(sectioning of the nerve and resection of the tumor and associated hemorrhage) addressed all possible etiologies at the same time.

Immediately after the surgery, the patient's pain was reduced; and postoperatively, only a small amount of pain remained. The patient has done well with complete resolution of symptoms, and has maintained that status for more than 11 years and is satisfied with his outcome from the procedure.

Conclusions

Patients may present due to torsion-induced hemorrhage within a tumor on a nerve root. It is unclear whether the torsion or the hemorrhage was the inciting event of the painful presentation, but removal of the lesion in question was immediately beneficial to the patient. This presentation should be kept in the differential diagnosis of any patient with an acute presentation for an intradural tumor, especially one arising from a nerve root within the cauda equina.

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