IMAGING FEATURES AND TREATMENT OF AN INTRADURAL LUMBAR CYSTIC SCHWANNOMA

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ABSTRACT - Spinal schwannomas are frequently observed among patients treated in a reference neurosurgery center. Cystic spinal schwannomas, however, are very scantily found. Due to its indolent behavior and benign course, the diagnosis of schwannomas may pose a challenge to the care giver, and the imaging findings can be misleading. In this article, we illustrate an example of a pauci-symptomatic 55 year-old male patient whose complaint was solely a non specific lumbar pain. Investigation revealed a large cystic lesion comprising the lower lumbar intradural space. He was then treated with microneurosurgical technique involving complete removal of the tumor and reconstruction of the duramater. Histological and immunohistochemical diagnosis were consistent with cystic schwannoma. The patient presented with complete recovery of his symptom. In this article we aim to emphasize the clinical presentation and treatment of lumbar spine schwannomas, and to illustrate the imaging findings within this uncommon case.

KEY WORDS: schwannoma, cystic tumor, nerve sheath tumor, neurinoma.

Schwannomas are benign tumors of the peripheral nervous system, arising from Schwann cells¹. Schwannomas can occur virtually in any body location where peripheral or cranial nerves are present², and they correspond to 8% of primary intracranial and 29% of primary spinal tumors¹. Within the spinal region, Prevedello et al. have investigated the prevalence of different extramedullary intradural spinal tumors and observed that schwannomas comprised 66% of all cases³. The lumbar region is one of the most common sites for occurrence of spinal schwannomas, as demonstrated by Conti et al., who studied the distribution of spinal schwannomas and reported that 48% lie within the lumbar spine⁴. While schwannomas occurring within the lumbar spine are not rare, large and predominantly cystic schwannomas occurring in the lumbar spine have scantily been demonstrated, even though it is well defined that benign schwannomas can eventually display degenerative changes defined by cyst formation, calcification, hemorrhage and hyalinization¹. Interestingly, large schwannomas within the lumbar spine may go unnoticed for a large time span mainly due to the indolent growth of the tumor and the paucity of symptoms generated. With a more malleable structure, a cystic schwannoma may pose a challenge to the diagnosis.

We present a rare case of a patient with a large cystic schwannoma in the lumbar region. We aim to emphasize the differential diagnosis of cystic masses of the intraspinal region by illustrating the

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clinical picture, imaging findings and treatment of this unique case. This report was approved by the Committee of Ethics and Research of our Institution (UNICAMP).

CASE
A 55-year-old man without remarkable previous medical history was admitted in our Hospital due to a 12-month history of lumbar pain with a poorly defined irradiation to both lower limbs. His general physical examination was normal and did not exhibit signs compatible with neurofibromatosis. His neurological physical examination failed to disclose signs of radicular irritation, motor or sensory deficits. Magnetic resonance imaging (MRI) (Fig 1) depicted an extensive lesion (6.0x1.8x 1.8 cm) extending from the 4th to 5th lumbar vertebrae almost completely constituted by a large cyst. Gadolinium contrasted MRI depicted a ring-like enhancement of the cyst. Tumor was totally removed by microsurgical technique (Fig 2). The remaining thin duramater was not large enough for primary repair, and an artificial graft (DuraGen™) was successfully used to accomplish a water tight seal duramater closure. Histological examination of the excised specimen revealed typical schwannoma cell nuclei (Fig 3A). Immunohistochemical study included staining for S-100 protein and NGFR (nerve growth factor receptor), which were both positive diffusely across the tumor cells (Fig 3B and 3C). The immunohistochemical analysis alongside with the histological observations confirmed the diagnosis of a cystic schwannoma. After an unremarkable postoperative period, the patient experienced a complete remission of preoperative symptoms. At a follow-up visit performed 12 months after surgery, the patient was asymptomatic.

DISCUSSION
Schwannomas are slow growing benign tumors. They are usually encapsulated, and rarely undergo malignant transformation. Schwannomas arise from the Schwann cells of the nerve sheath, and they comprise the most common tumor type affecting the peripheral nerves. The most common location of schwannomas are around peripheral nerves in the extradural space. Intracranial schwannomas have also been observed, and they usually arise from the facial, trigeminal, or vestibular nerves. Conspicuously, schwannomas are more frequently observed in patients with neurofibromatosis type 2, schwannomas can be observed in rather

Fig 1. Pre-operative sagittal MRI scan of the lumbar spine: In A) T2-weighted image showing an intradural cystic lesion; In B) T1-weighted image (gadolinium injection) showing the enhanced thin ring-like cystic tumor capsule.

Fig 2. Intra-operative image showing the cystic lesion after separation from the roots by blunt microdissection. A rootlet remains adherent to the tumor disappearing inside its capsule (arrow).
Fig 3. A) Hematoxylin-eosin study of the histological section of the excised tumor reveals typical schwannoma cell nuclei. B) S-100 protein preparation of the histological section reveals intense staining. C) Histological preparation for investigation of NGFR (nerve growth factor receptor) antigen shows intense positive reaction.
rare locations. Women and men are equally affected by schwannomas, and the literature is prolific in showing cases of schwannomas affecting a large age range, albeit there is a predefined prediction for occurrence in between the fourth to sixth decades of life.

Benign schwannomas can occasionally display degenerative changes that are encompassed by cyst formation, calcifications, hemorrhage and hyalinization. When multiple degenerative changes are encountered, schwannomas fit into the category of “ancient schwannomas”, which are extremely benign in course, rarely demanding any form of treatment. Specifically, the cystic degeneration of schwannomas occurring in isolation, i.e., without additional features of ancient schwannomas, is scantily encountered. Cystic degenerations have been observed in the orbital region, in the olfactory groove, in the tentorial hiatus and posterior or cavernous sinus, in the presacral region, within the pancreas, in the maxillary sinus, within the spinal cord, and in the intraventricular region. Cystic schwannomas have also been observed surrounding cranial nerves such as the vestibular nerve, within the trigeminal nerve and within the jugular foramen.

Only seven cases of cystic lumbar nerve sheath tumours have been described in the literature pointing out its resonance magnetic imaging presentation.

As schwannomas are benign tumors with a slow growth rate, the diagnosis of extracranial schwannomas may pose a challenge to the care giver when few symptoms are observed. In the case we report, there is a striking contrast with the paucity of symptoms and the size of the tumor. This highlights the importance of suspecting of a cystic lumbar nerve sheath tumor when symptoms associated to lumbar spinal cord, or nerve roots compression are encountered. Moreover, schwannomas should be included in the differential diagnosis of a cystic mass in the spinal region.

Differently from cystic calcified schwannomas, i.e., “ancient schwannomas”, cystic schwannomas possibly behave in a similar fashion to solid schwannomas. Therefore the treatment of a cystic schwannoma in that setting should involve radical surgical excision of the tumor, which, as demonstrated in this case report, can be performed without inflicting any harm to the patient.

In conclusion, cystic schwannomas are rare benign tumors of the lumbar region. Clinical symptoms are usually due to the compression of lumbar spine or nerve roots structures, but given the slow growth of the tumor, few symptoms can be observed up until the tumor has reached a large mass. The treatment of cystic and non-calcified schwannomas involves safe radical excision of the lesion.

REFERENCES