The Spine Journal 14 (2014) e1–e7

Case Report

Intradural extramedullary cavernoma of a lumbar nerve root mimicking neurofibroma. A report of a rare case and the differential diagnosis

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Received 20 January 2014; revised 16 June 2014; accepted 18 August 2014

Abstract

BACKGROUND CONTEXT: Intradural extramedullary (IDEM) cavernomas are rare vascular malformations. They are well-circumscribed dark berry-like lesions with a histologic appearance of sinusoidal vascular channels. Neurofibromas are the most common IDEM tumors, originating from all nerve elements and leading to firm enlargement of the affected nerve root. These lesions are completely different; however, they may involve the spinal nerve roots or the major nerve trunks. Any similarities in clinical findings are based on different pathophysiology.

PURPOSE: To present a rare resemblance of an IDEM cavernoma to a neurofibroma.

STUDY DESIGN: This is a case report with review of the literature focused on the differential diagnosis.

METHODS: A 79-year-old patient presented with acute sensorimotor disturbance from L2–S1 levels. The investigations showed an L2–L3 lesion occupying the canal. Findings resembled a neurofibroma and a surgical resection was decided.

RESULTS: The complete surgical resection revealed a vascular lesion originating from a nerve root. The histology confirmed an IDEM cavernoma. This is a unique case as such a clinical resemblance and a macroscopical appearance has not been reported for an IDEM cavernoma as yet. The patient showed full postoperative recovery from his initial symptoms.

CONCLUSIONS: Intradural extramedullary cavernoma is a rare cause of compression to spinal cord or nerve roots. Its manifestation characteristics are well defined and should always be part of the differential diagnosis. Intraoperative findings aid the diagnosis in nontypical cases before the final histology. The nontraumatic and nerve tissue sparing surgical resection warrants optimal postoperative results and excellent prognosis.

Keywords: Intradural extramedullary cavernous angioma; Cavernoma; Intraspinal tumours; Vascular malformations; Neurofibroma; Neurofibromatosis

Introduction

Spinal cavernomas are vascular malformations with a reported incidence of 5% to 12% of all spinal vascular abnormalities [1–5]. Only 3% of spinal cavernomas are intradural, usually with an intramedullary location [1,6]. Intradural extramedullary (IDEM) cavernomas are even more rare entities [6,7]. They are well-demarcated vascular lesions causing symptoms related to their space-occupying effect, the obstruction of the cerebrospinal fluid (CSF) flow, and the potential intradural hemorrhage [8]. On the contrary, neurofibromas are

FDA device/drug status: Not applicable.

Author disclosures: ***GM:*** Nothing to disclose. ***SP:*** Nothing to disclose. ***SN:*** Nothing to disclose. ***MM:*** Nothing to disclose. ***NT:*** Nothing to disclose.

The resources and funding for the realization of this study came out from the regular budget of our department.

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http://dx.doi.org/10.1016/j.spinee.2014.08.447

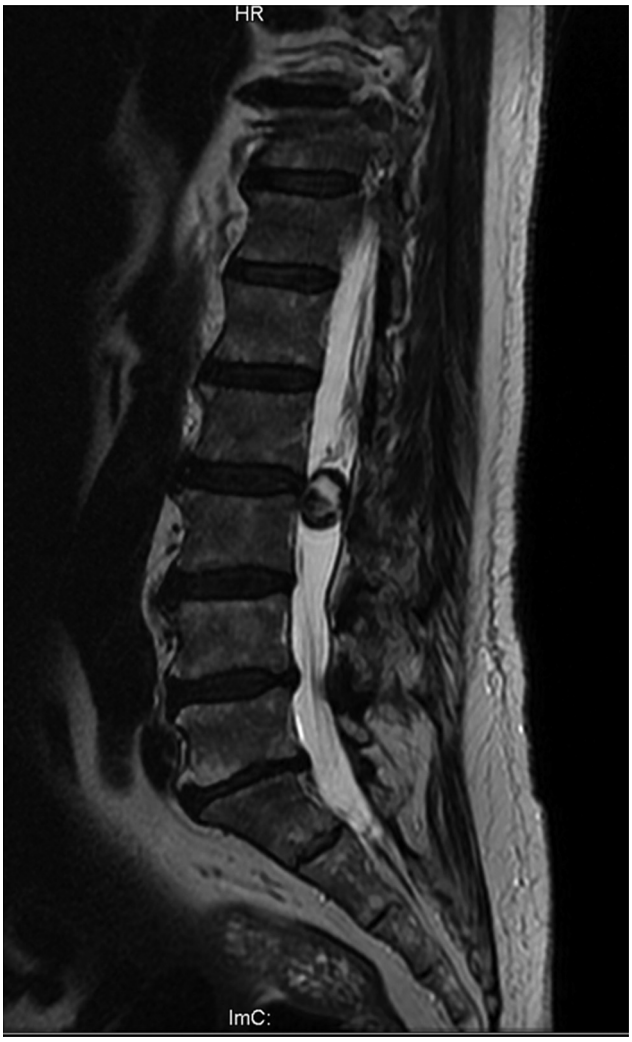

Fig. 1. The midsagittal T2-weighted preoperative magnetic resonance imaging. A lesion at L2–L3 level is shown obliterating the spinal canal. The lesion has a hyperintense central part with a heterogeneous signal that is slightly less intense than that of the cerebrospinal fluid.

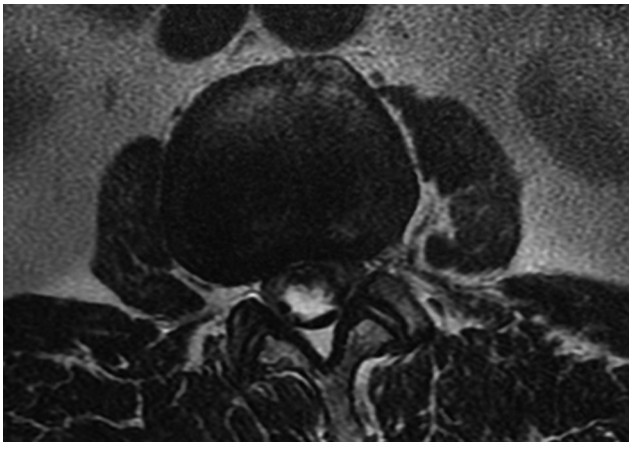

Fig. 2. An axial T2-weighted preoperative magnetic resonance imaging at the level of L2–L3. The obliteration of the canal is demonstrated. The signal intensity of the lesion has the same characteristics as in Fig. 1.

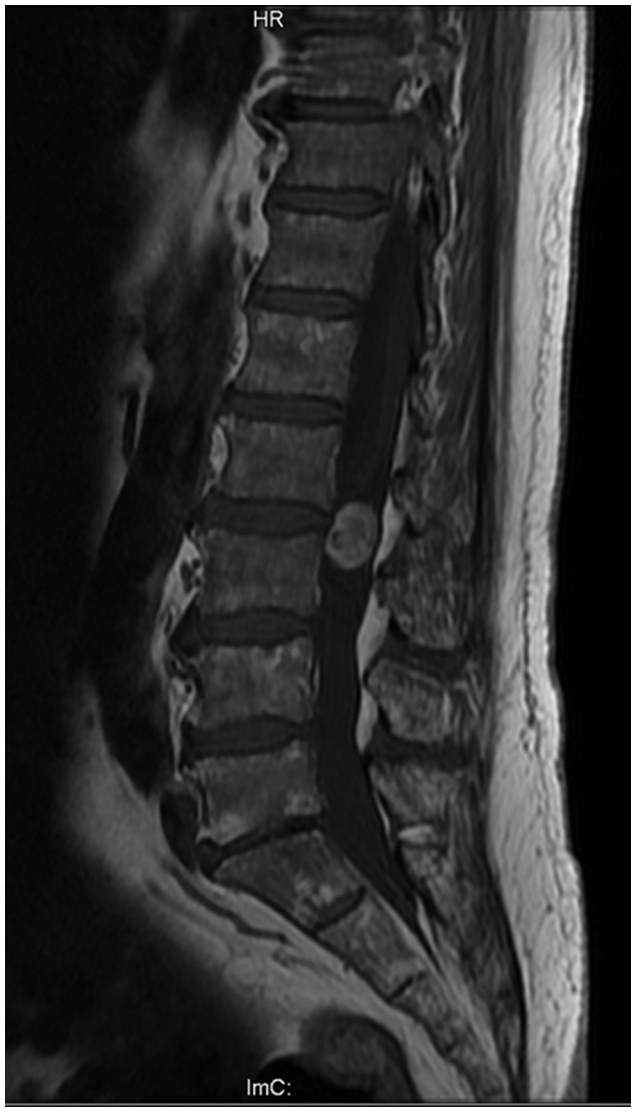

Fig. 3. The midsaggital T1-weighted preoperative magnetic resonance imaging. The lesion obliterating the canal is shown. The signal of the lesion is iso- or slightly hypointense to that of the muscle.

the most common IDEM tumors [9]. They most commonly involve the spinal nerve roots and the major nerve trunks [10], leading to enlargement of the affected nerve.

This report highlights an IDEM cavernoma case regarding its clinical and imaging resemblance to a neurofibroma, which influenced the surgical treatment before the final histology report. This is the first report in the literature and all the up to date available information for this entity is discussed.

Case report

A 79-year-old man was referred to our spinal service with a 2-week history of back pain, cramping feelings in his right leg, numbness in his toes, and weakness in both