Radiology Case Reports 16 (2021) 472–475

Available online at www.sciencedirect.com

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journal homepage: www.elsevier.com/locate/radcr

Case Report

Solitary Fibrous tumor of the lumbar spine mimicking a sequestered disc fragment☆,☆☆,★

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ARTICLE INFO

Article history:
Received 22 August 2020
Revised 1 December 2020
Accepted 3 December 2020

Key Words:
Lumbar spine
Solitary fibrous tumor
disc herniation
sequestered disc
case report

ABSTRACT

Solitary fibrous tumor in the lumbar spine is a rare pathology with non-specific radiographic features, sometimes resulting in misdiagnosis. Our patient was a 41-year old female who presented with low back pain and bilateral leg pain. Initial MRI showed a lesion misdiagnosed for a sequestered disc at the mid L4-5 level, which was subsequently characterized appropriately and treated surgically, with resolution of symptoms. Pathologic diagnosis was most consistent with a solitary fibrous tumor due to STAT 6 and CD 34 reactivity. Long-term follow up is recommended in these patients to monitor tumor recurrence and evidence of metastasis.

Introduction

Solitary fibrous tumor (SFT) can present in the central nervous system in several ways, mimicking other pathologies resulting in misdiagnosis. This lesion was first described as a rare spindle cell tumor in the chest/abdomen/pelvis [1]. Only 24.5% of SFT are CNS based [2], with the lumbar spine accounting for about 13.2% [3,4]. Due to its nonspecific clinical and radiographic features, spinal SFT can mimic other pathologies such as schwannoma [5,6], meningioma [7], osteosarcoma [8], among others. Based on our search in PubMed, Embase and Cochrane Library, we could not find any other reported case describing SFT mimicking a lumbar extruded disc fragment.

Case report

Patient is a 41-year-old previously healthy female presented with low back and bilateral leg pain for 5 years that had worsened in the past year. Pain radiated across her low back and travelled down through her buttocks to posterior thighs and hips, with right leg pain worse than left. She had full strength

☆ **Disclosure:** Dr Ropper is a Consultant for Globus Medical and Stryker. However, they have no bearing on this manuscript.
☆☆ **Informed Consent Statement:**
★ Written informed consent was obtained from the patient for the spinal surgical procedure and for utilization of radiology and pathology data for research and academic purposes provided there is no patient identifying information and HIPAA policy is strictly followed.

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https://doi.org/10.1016/j.radcr.2020.12.009

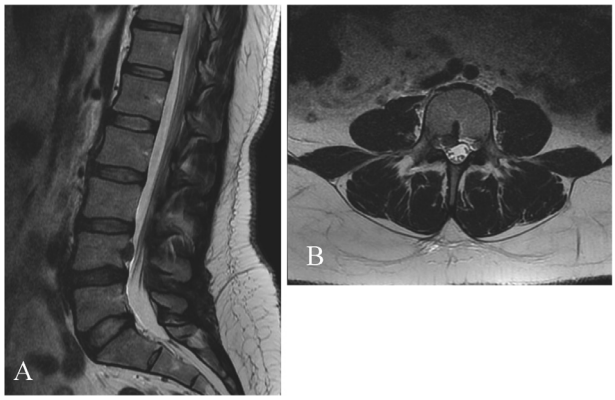

Fig. 1 – Initial pre-operative MRI, T2 weighted sequence: MRI revealed an 8 x 10 x 12mm right paracentral extradural lesion effacing the ventral theca sac at the L4 vertebral body level. This was initially thought to be a sequestered disc fragment and was managed conservatively.

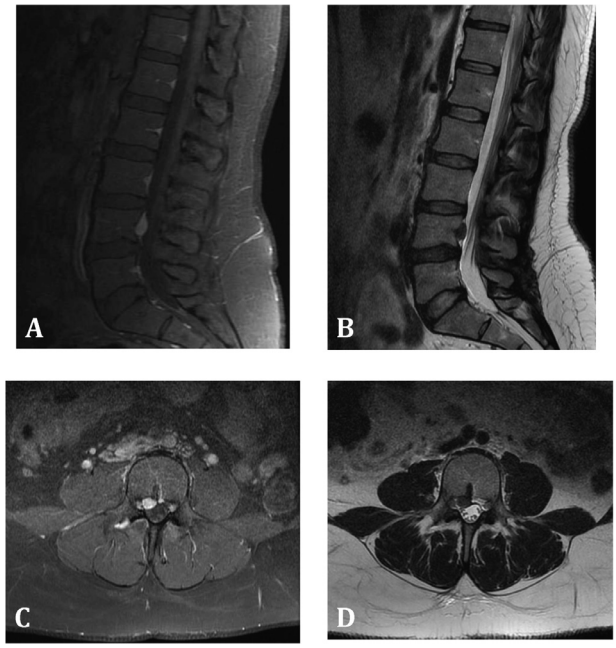

Fig. 2 – Follow-up MRI two years later demonstrated slight increase in size of the lesion, now measuring10 x 10 x 13mm. There is effacement of the ventral theca sac and the right L4 nerve root as it enters the right lateral recess with mild displacement. [A] **and** [C] **- Sagittal and axial post-contrast T1-MRI, with homogenous contrast enhancement.** [B] **and** [D] **– Sagittal and axial T2-MRI.**

in all extremity muscle groups, with normal sensation and reflexes. Pain was elicited with bilateral positive straight-leg-raise test at 30-degrees.

Initial MRI from outside facility revealed an 8 × 10 × 12mm right paracentral extradural soft tissue signal intensity at the level of L4 vertebral body effacing the ventral thecal sac and right L4 nerve root (Fig. 1). Pathology was thought to be a sequestered disc fragment and patient was managed conservatively. MRI 2 years later demonstrated slight increase in size of the "lesion," measuring 10 × 10 × 13 mm. Due to the increased size and abnormal location of the lesion, contrast was administered and the lesion demonstrated homogenous enhancement (Fig. 2). Due to the enhancement of the lesion, an extradural tumor was considered, with the differential considerations including schwannoma and meningioma. Surgical resection of the lesion was recommended.

Laminectomy was performed at the level of L4 and the canal was gently decompressed. A large bulge was felt under the theca at the level of the right L4 pedicle. The theca sac and nerve root were retracted medially and we encountered a large extradural mass on the floor of the spinal canal, not attached to the nerve root. There was effacement of the right L4 nerve root as it entered the right lateral recess with mild displacement. The L4 nerve was not tethered to the mass and thus easily mobilized. The mass was covered by a capsule and was rubbery, mobile and fibrous and did not have the typical appearance of a disc fragment. After coagulating and opening the capsule, there was no evidence of CSF egress. Gross total resection (GTR) was achieved without intraoperative complications. Pathology revealed a histologically bland cellular spindle-cell neoplasm in a background of collagen. S100 immunoperoxidase stains were negative, excluding a nerve sheath tumor. CD34 highlighted thin-walled blood vessels. STAT-6 showed reactivity for tumor cells, making the pathologic diagnosis most consistent with a solitary fibrous tumor, WHO Grade-I (Fig. 3).

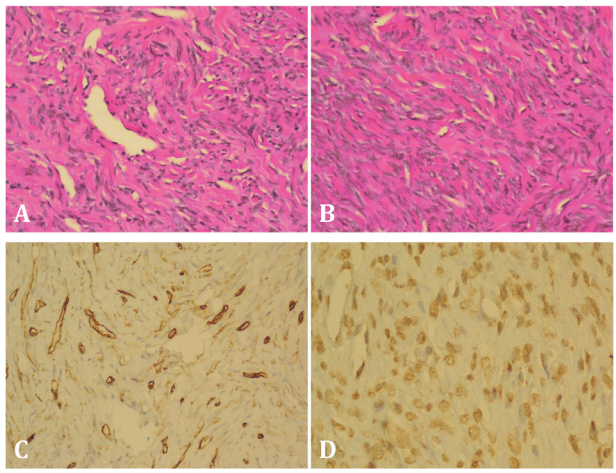

Fig. 3 – Pathologic Examination: [A] **and** [B] **are Hematoxylin-eosin staining that show bland cellular spindle cell neoplasm in a background of collagen without excessive mitotic activity, high cellularity or necrosis. Immunohistochemistry showed positive CD34 highlighted thin walled blood vessels** [C] **and STAT 6 showed reactivity** [D]**.**

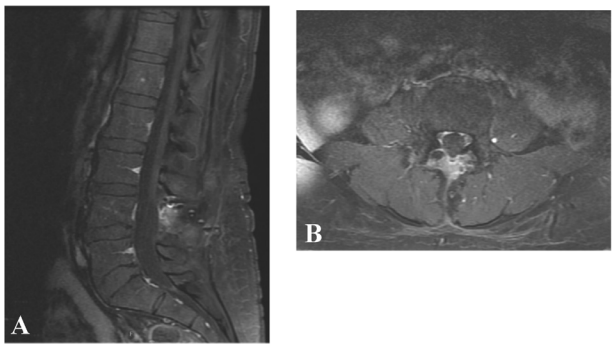

Fig. 4 – Post-operative MRI at 2 years showing gross total resection of lesion in the lumbar spine. No evidence of recurrence or residual disease.

The patient had significant improvement in pain and continued to retain full strength without any neurologic deficits. She was discharged home on postoperative day 1. One-year and 2-year follow-up MRIs (most recent: Fig. 4) revealed no recurrence of the tumor.

Discussion

Solitary fibrous tumor is a rare soft-tissue lesion, first described by Klemperer and Rabin in 1931 as a neoplastic lesion from the pleura [1]. Only a small percentage is present in the central nervous system with even a smaller subset in the lumbar spine [2-4]. Despite reported cases in the lumbar spine mimicking extradural tumors, this case report is the first describing SFT mimicking an extruded disc fragment.

Presenting symptoms of lumbar SFT are often variable and nonspecific, including low back pain and radiculopathy (as seen in our patient) [6,7,9], palpable mass with leg weakness [6], paresthesia and urinary dysfunction [10]. Lesions can be intradural (mimicking schwannoma and meningioma), or extradural (mimicking disc herniation, fibrosarcoma, and osteosarcoma). Nagano et al [6] presented a large extradural malignant SFT, in the retroperitoneal space, next to the L4-5 vertebra mimicking schwannoma. Yi et al [10] showed that SFT can also mimic tuberculosis, metastasis or bone tumors in patients with bony destructive lesions. There is no unique imaging diagnostic feature of SFT. Plain x-rays are usually nondiagnostic for soft tissue lesions where there is no bony invasion or destruction. CT sometimes shows soft tissue mass with areas of calcification and necrosis [11]. MRI remains the most sensitive imaging modality [3,9,12], with radiographic similarity to disc extrusion or sequestered disc fragment on T1 and T2 weighted imaging (low signal intensity). However, there is avid contrast enhancement, usually a well-circumscribed homogenous lesion [12,13]. A PET scan can provide some insight on aggressiveness and metastatic burden from avid FDG uptake in malignant cases [9]. These radiographic features present a diagnostic challenge as the lesion can be misdiagnosed for other spinal pathologies such as schwannoma, meningioma, chordoma, fibrosarcoma, neurofibroma, ependymoma, or cavernous malformation [7,9,12,14].

Tissue diagnosis is the only reliable method of confirming SFT since its clinical presentation in the spine can mimic a spectrum of other pathologies. Thus, biopsy ± resection is highly recommended in patients with symptomatic pathologies. Histologically, these lesions are different from meningioma and schwannoma due to the presence of CD34 and STAT-6, and the absence of S100 [4,6,7,9,12].

Surgery is the primary treatment for patients with symptomatic spinal SFT. GTR and tumor grade are factors that influence prognosis [3,15]. For benign pathologies, outcome is largely driven by extent of surgical resection, with approximately 100% five-year survival rate [3,4]. A study showed 89.1% of patients with no recurrence at 1.2 years follow-up after GTR [3]. Prognosis in patients with malignant features of SFT continues to be very poor regardless of extent of resection [3,15]. This is likely due to local tissue invasion and systemic metastasis. Jia et al [15] showed a recurrence rate of 40% and 60% for grades II and III respectively.

Chemotherapy and radiation therapy have traditionally been reserved for malignant disease, recurrent pathology or metastatic disease. Our patient did not require adjuvant therapy due to the benign nature of the pathology. Where there is concern for local recurrence, or in small inoperable tumors, radiation therapy could be beneficial [15,16]. However, several studies stipulate the ineffective long-term role of adjuvant chemo-radiation in the management of SFT [16-19]. Without effective adjuvant therapy, outcomes in malignant cases tend to be very poor.

Regardless of pathologic grading and treatment modality, long-term surveillance is recommended due to risk of delayed recurrence or metastasis [8,9,20]. There currently is no guideline regarding post-operative surveillance. Our patient will follow up with annual MRI for the first three years and then one last MRI at 10 years. For grade II and III, we recommend serial annual MRI for the first 5 years, then at 7th and 10th years. This will allow determination of late recurrence or metastasis.

Conclusion

To the best of our knowledge (based on extensive online search), we describe the first reported case of solitary fibrous tumor in the lumbar spine mimicking a herniated disc. Although a rare entity in the spinal column, SFT should be included in the differential diagnosis for spinal lesions, specifically when there is serial increased in size of the lesion and when there is contrast enhancement on T1-weighted MRI sequence. Regardless of clinical suspicion, tissue diagnosis is needed for pathologic confirmation. Long-term follow up is recommended to monitor tumor recurrence and evidence of metastasis.

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