Case Report

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

Terence Verla, MD MPHa,*, Venita Simpson, MDb, Alexander E. Ropper, MDa

a Department of Neurosurgery, Baylor College of Medicine, 7200 Cambridge St, Suite 9A, Houston, TX 77030 USA
b Naval Medical Center Portsmouth. 620 John Paul Jones Circle, Portsmouth VA 23707

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.

© 2020 The Authors. Published by Elsevier Inc. on behalf of University of Washington.

This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/)

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...
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 × 12 mm 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. 1 – Initial pre-operative MRI, T2 weighted sequence: MRI revealed an 8 × 10 × 12 mm 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 measuring 10 × 10 × 13 mm. 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.

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].
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 homogeneous 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 ineffectiveness 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.

References

[1] Klemperer P, Coleman BR. Primary neoplasms of the pleura. A report of five cases. Am J Ind Med 1992;22(1):1–31.
[2] Wushou A, Jiang YZ, Liu YR, Shao ZM. The demographic features, clinicopathologic characteristics, treatment outcome and disease-specific prognostic factors of solitary fibrous tumor: a population-based analysis. Oncotarget Dec 8 2015;6(39):41875–83.
[3] Albert GW, Golken M. Solitary fibrous tumors of the spine: a pediatric case report with a comprehensive review of the literature. J Neurosurg Pediatr Mar 2017;19(3):339–48.
[4] Lang N, Zhang E, Xing X, Yuan H. Solitary fibrous tumour of the spine: imaging features of a commonly misdiagnosed entity. Eur Radiol Sep 2018;28(9):3986–95.
[5] Piana S, Putrino I, Cavazza A, Nigrioli E. Solitary fibrous tumor of the spinal nerve rootlet: report of a case mimicking schwannoma. Arch Pathol Lab Med Mar 2004;128(3):335–7.
[6] Nagano A, Ohno T, Nishimoto Y, Oshima K, Shimizu K. Malignant solitary fibrous tumor of the lumbar spinal root mimicking schwannoma: a case report. Spine J Jan 2014;14(1):e17–20.
[7] Sebaaly A, Raffoul L, Moussa R. Solitary fibrous tumor of the lumbar spine: the great mimicker-report of the fifth case. Case Rep Orthop 2014;2014:852830.
[8] Oike N, Kawashima H, Ogose A, et al. A malignant solitary fibrous tumour arising from the first lumbar vertebral and mimicking an osteosarcoma: a case report. World J Surg Oncol 2017;15(1):100.
[9] Farooq Z, Badar Z, Zaccarini D, Tavernier FB, Mohamed A, Mangla R. Recurrent solitary fibrous tumor of lumbar spine with vertebral body involvement: imaging features and differential diagnosis with report of a case. Radiol Case Rep Dec 2016;11(4):450–5.
[10] Yi X, Xiao D, He Y, et al. Spinal Solitary Fibrous Tumor/Hemangiopericytoma: A Clinicopathologic and Radiologic Analysis of Eleven Cases. World Neurosurg Aug 2017;104:318–29.
[11] Lee KS, Im JG, Choe KO, Kim CJ, Lee BH. CT findings in benign fibrous mesothelioma of the pleura: pathologic correlation in nine patients. AJR Am J Roentgenol 1992;158(5):983–6.
[12] Mariniello G, Napoli M, Russo C, et al. MRI features of spinal solitary fibrous tumors. A report of two cases and literature review. Neuroradiol J Nov 2012;25(5):610–16.
[13] Ginat DT, Bokhari A, Bhatt S, Dogra V. Imaging features of solitary fibrous tumors. AJR Am J Roentgenol Mar 2011;196(3):487–95.
[14] Walker CT, Amene CS, Pannell JS, et al. Hemorrhagic intramedullary solitary fibrous tumor of the conus medullaris: case report. Journal of neurosurgery. Spine. Oct 2015;23(4):438–43.
[15] Jia Q, Zhou Z, Zhang D, et al. Surgical management of spinal solitary fibrous tumor/hemangiopericytoma: a case series of 20 patients. European spine journal: official publication of the European Spine Society, the European Spinal Deformity Society, and the European Section of the Cervical Spine Research Society Apr 2018;27(4):891–901.
[16] Galanis E, Buckner JC, Scheithauer BW, Kimmel DW, Schomberg PJ, Piepgrass DG. Management of recurrent meningeal hemangiopericytoma. Cancer May 15 1998;82(10):1915–20.
[17] Ecker RD, Marsh WR, Pollock BE, et al. Hemangiopericytoma in the central nervous system: treatment, pathological features, and long-term follow up in 38 patients. J Neurosurg Jun 2003;98(6):1182–7.
[18] Munoz E, Prat A, Adamo B, Peralta S, Ramon y Cajal S, Valverde C. A rare case of malignant solitary fibrous tumor of the spinal cord. Spine May 20 2008;33(12):E397–9.
[19] Gengler C, Guillou L. Solitary fibrous tumour and haemangiopericytoma: evolution of a concept. Histopathology Jan 2006;48(1):63–74.
[20] Beaman FD, Kransdorf MJ, Menke DM. Schwannoma: radiologic-pathologic correlation. Radiographics Sep-Oct 2004;24(5):1477–81.