

Inflammatory myofibroblastic tumor of the lumbar spinal canal

A Case Report With Literature Review

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Abstract

Rationale: Inflammatory myofibroblastic tumor (IMT) is a rare type of mesenchymal tumor. IMT can arise in multiple anatomic locations. IMT of the lumbar spinal canal is exceptionally rare.

Patient concerns: Here, we report the case of a 56-year-old male patient with an IMT who was in good health until 1 year prior to admission, when he began experiencing pain in both lower extremities and the lower back.

Interventions: A space-occupying lesion in the lumbar canal was identified by magnetic resonance imaging and then surgically resected.

Diagnoses: Histopathological analysis of the lesion revealed a composition of mucous edema, inflammatory cells, collagenous fibers, and spindle cells that were diffuse and positive for smooth muscle actin and CD68; focal positive for vimentin and desmin; and negative for CD34 (marker of vascular endothelial cells), CD21, CD23, CD35, S-100, Epstein–Barr virus infection, Ki-67, and anaplastic lymphoma kinase. Thus, the diagnosis was an IMT of the lumbar canal.

Outcomes: In the spinal canal, IMT should be considered in the evaluation of tumors although it is a very rare diagnosis. It is a benign lesion, but it has potential for invasion and recurrence.

Lessons: There are no characteristic imaging features of these tumors, but they can be addressed by complete surgical excision. Patients with these lesions should undergo frequent long-term follow-up to detect and address recurrence.

Abbreviations: ALK = anaplastic lymphoma kinase, CT = computed tomography, EBV = Epstein–Barr virus, IMT = inflammatory myofibroblastic tumor, MRI = magnetic resonance imaging, SFT = solitary fibrous tumor, SMA = smooth muscle actin.

Keywords: inflammatory myofibroblastic tumor, lumbar canal, magnetic resonance imaging

1. Introduction

Inflammatory myofibroblastic tumor (IMT) is a rare type of mesenchymal tumor^[1] of unknown pathogenesis that is composed of neoplastic myofibroblasts, which are about an inflammatory stroma that is also well vascularized.^[2] Most patients with an IMT are younger than 16 years of age.^[3] An IMT can arise in multiple anatomic locations, with the most common sites being the lungs and omentum.^[4] IMTs have also been found

in the head and neck region,^[5] gastrointestinal tract,^[6] spleen,^[7] liver,^[8] genitourinary tract,^[9] thyroid,^[10] and larynx.^[11] Herein, we report a case of a 56-year-old male patient with an IMT in the lumbar spinal canal, and to the best of our knowledge, this is the first case of an IMT in that location to be reported.

2. Case presentation

Informed consent was obtained from the individual participant included in the study.

A 56-year-old male patient who reported experiencing pain in the lower back as well as headache for 1 year and pain in both lower limbs for 7 days was admitted to the Binzhou Medical University hospital (Shandong, China).

The patient's low back and bilateral lower limb pain and numbness had no obvious cause and was reported to be intermittent, dull pain. He experienced some relief from the pain with bed rest, and his pain was greatest after walking and when he was tired. Physical examination revealed the following findings. His lumbar curvature was straighter than normal, without palpable deformity, and the interspinous paraspinal tenderness was level L4/S5/S1 without sciatica.

He reported restricted movement at the joint. His lower limb muscle tone was normal. Acupuncture performed previously to both lower limbs did not help to diminish the pain. Computed tomography (CT) scanning of the lumbar region revealed the following findings. The lumbar curvature was less natural, and the posterior margin of the vertebral body was continuous. The edges of the vertebrae and the lumbar facet joints showed varying

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Figure 1. Plain and enhanced MRI revealed a space-occupying lesion at the L4/5 level located in the epidural space of the spinal cord. (A–C) Magnetic resonance imaging (MRI) scans (A: T1WI SAG; B: T2WI SAG; C: T2WI-FS SAG) showing a nonhomogeneous soft tissue mass arising from the lumbar spinal canal. MRI enhanced scans (D: T1WI SAG) showing a mass in the lumbar spinal canal with inhomogeneous enhancement. The modified area was showing with white arrows inserted on the picture. MRI = magnetic resonance imaging.

degrees of hyperosteoegeny. The L2/3/4/5 intervertebral discs were prominent, whereas the dural sacs at the corresponding levels were compressed. Plain and enhanced magnetic resonance imaging (MRI) revealed a space-occupying lesion at the L4/5 level located in the epidural space of the spinal cord (Fig. 1A–D). The MRI findings suggested a benign tumor and provided a clear indication for surgical resection in this patient who exhibited obvious nerve damage, which had a serious impact on his daily life. Therefore, surgical treatment involving lumbar spinal tumor resection was required to achieve decompression of the dura.

Peri-operatively, the tumor was located on the right side of the L4/5 level and had a diameter of approximately 1 cm. Although the tumor was well circumscribed, it involved the dura and the right L5 nerve root. Pathological analysis revealed a gray piece of tissue with a size of $0.9 \times 0.7 \times 0.3$ cm. The histological examination showed mucous edema and spindle cells with no nuclear division and no necrosis as well as inflammatory cells in dense areas and obvious degeneration of the collagen fibers (Fig. 2A and B). Immunohistochemistry showed that the tumor cells were diffusely positive for smooth muscle actin (SMA) and CD68; focal positive for vimentin, and desmin; and negative for CD34 (marker of vascular endothelial cells) (Fig. 3A–E), CD21, CD23, CD35, S-100, Epstein–Barr virus infection, Ki-67, and anaplastic lymphoma kinase (ALK). According to this expression profile, the tumor was considered to be an IMT.

3. Discussion and review of literature

The first case of an IMT, or an inflammatory pseudotumor as it was previously named, was described in the lungs in 1939 by Brunn.^[12] The World Health Organization defines IMTs as lesions composed of myofibroblastic spindle cells accompanied by an inflammatory infiltrate of lymphocytes, plasma cells, and eosinophils. Cases of IMTs have been documented across a wide age range,^[13] but predominantly occur in the soft tissue and viscera of children and young adults.^[14] The various anatomic locations in which IMTs have been observed include the lung, retroperitoneum, orbit, and abdomino-pelvic region.^[15] Lung IMTs are more frequently observed in the lower lobe of the right lung.^[16] To our knowledge, the present case is the first reported case of an IMT in the lumbar spinal canal, making this an extremely rare pathologic diagnosis.

The radiological features of IMTs are variable, and no characteristic findings on computed tomography or MRI have been established despite recent improvements in imaging technology.^[17] IMTs have a similar appearance to other typical spinal epidural lesions, such as meningioma, lymphoma, metastasis, and myeloma, except that other lesions can lead to changes in bone. Thus, biopsy or immunochemical examination is needed to determine the specific tumor type. In our case, MRI showed a hypointense lesion on T1- and T2-weighted images (Fig. 1A–C), and enhanced MRI showed a mass in the epidural space of the spinal cord with nonhomogeneous enhancement (Fig. 1D). Preoperative diagnosis of IMTs remains clinically challenging, and final diagnosis is currently based on histopathologic evaluation of the involved tissue. In the present case,

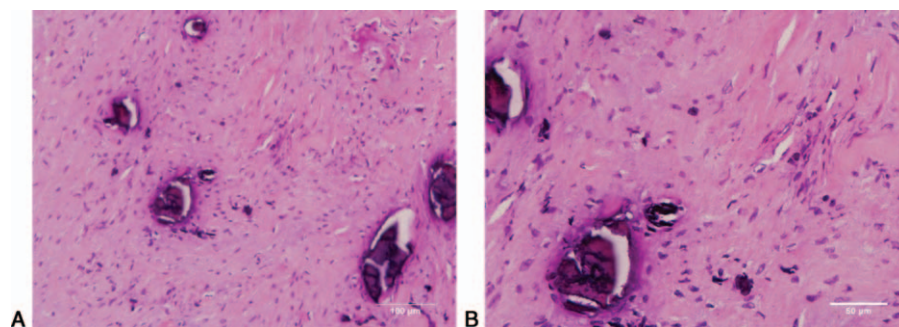


Figure 2. Histological examination showed an area of mucous edema, spindle cells showing no nuclear division and no necrosis, and inflammatory cells in dense areas, with obvious degeneration of the collagen fibers. (A) Hematoxylin and eosin staining (original magnification $\times 200$). (B) Hematoxylin and eosin staining (original magnification $\times 400$).

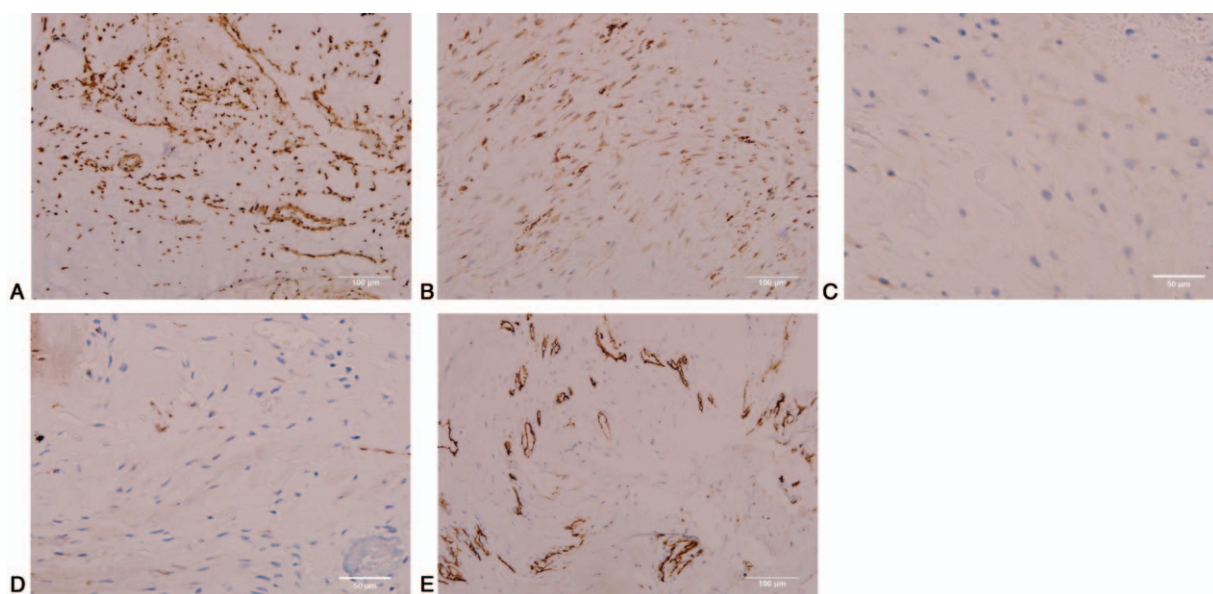


Figure 3. The tumor cells stained diffusely positive for (A) SMA, (B) CD68 (original magnification, $\times 200$), and focal expression of (C) vimentin and (D) desmin (original magnification, $\times 400$), and (E) negative for CD34 (marker of vascular endothelial cells, original magnification, $\times 200$), suggestive of an inflammatory myofibroblastic tumor. SMA = smooth muscle actin.

histopathological evaluation suggested an IMT and tumor cell expression of SMA and CD68 along with focal expression of vimentin and desmin and the absence of CD34 expression. We did not find CD68 expression as reported previously for IMTs,^[18] whereas the tumor cells were CD68 positive in our case. We tend to believe that these cells were modified histiocytes.

The etiopathogenesis of IMTs remain unresolved. IMTs are defined as a proliferating neoplasm due to their invasive potential and the frequency of recurrence and metastasis.^[19] Arber et al^[20] suggested that IMTs are associated with viral infection such as Epstein-Barr virus (EBV). The patients usually have nonspecific clinical symptoms such as fever and pain. In our case, the patient did not have features of chronic inflammation, and the test for EBV infection was negative. IMTs are classified as minimally aggressive mesenchymal malignancies^[2] based on their typically aggressive clinical course, and when it is feasible, the first-line therapeutic option is surgical resection. Researchers have suggested that studying ALK gene expression may be correlated with a higher recurrence rate.^[21] Another study reported a recurrence rate of 45% for ALK-positive IMTs compared with a recurrence rate of only 20% for ALK-negative IMTs.^[4] Staining for ALK expression was negative in our case.

IMTs involve tumor dedifferentiation of CD34-positive bone marrow-derived mesenchymal stem cells.^[22] Tumors such as Kaposi sarcoma or myofibroblastoma maintain CD34 positivity,^[23] whereas it can be lost in other tumors during histogenesis.

On histological and immunohistochemical evaluation, an IMT needs to be differentiated from myofibroblastoma and solitary fibrous tumor. Myofibroblastoma is a rare solitary benign tumor characterized by fascicles of spindle cells having features of myofibroblasts, with intervening large hyalinized collagenous stroma, and a surrounding pseudocapsule composed of compressed breast tissue. It has characteristic expression of vimentin, CD34, and desmin. Solitary fibrous tumor (SFT) cells are enriched with the alternating distribution of different shapes of collagen fiber bundles and sparse area composition, which can appear as mucinous degeneration. Inflammatory cells infiltrating

SFTs include mast cells that expression CD34, CD99, and bc1-2 but do not express SMA or desmin.

Anticancer therapies including chemotherapy,^[24] radiotherapy,^[25,26] nonsteroidal anti-inflammatory drugs,^[27] and steroids^[28] have been used to treat patients with disease recurrence, with variable outcomes. The patient in the present case was treated by complete surgical resection. After surgery, the patient was given mannitol and was instructed to strengthen the lower limbs through exercise. MRI examination after 3 months showed no recurrence of the lesion. Follow-up at 6 months was recommended based on the recurrence rate for this tumor type. Accordingly, after surgical resection, long-term follow-up is needed for early identification of recurrence in patients with IMTs.

In conclusion, although IMT in the spinal canal is a very rare diagnosis, it should be considered in the evaluation of tumors in this space. It is a benign lesion, but the associated pathological changes have potential for invasion and recurrence. There are no characteristic imaging features of these tumors, but they can be treated by complete surgical excision. Overall, patients with these lesions should undergo complete surgical resection with frequent long-term follow-up to detect and address recurrence.

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