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Case Report

Inflammatory Pseudotumor in the Epidural Space of the Thoracic Spine: A Case Report and Literature Review of MR Imaging Findings

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Summary: We present a rare case of pathologically proven inflammatory pseudotumor in the thoracic spine. The lesion showed an isointense signal on T1-weighted images, a heterogeneous iso- and hyperintense signal on T2-weighted images, and strong homogeneous enhancement. There was no evidence of abnormalities in the adjacent bone. Whereas the exact pathogenesis of this lesion is unknown, it has been regarded as an unusual response to insults such as trauma or acute infection, a postinflammatory reparative process, or low-grade malignancy.

Inflammatory pseudotumor (inflammatory myofibroblastic tumor) is a benign tumorlike lesion of unknown cause, which has been described in very small numbers at various locations throughout the body (1). It is believed that inflammatory pseudotumor is an inflammatory process that includes a diverse group of lesions characterized by inflammatory cell infiltration and variable fibrotic responses (2, 3). Inflammatory pseudotumor is a lesion characterized by proliferation of myofibroblastic spindle cells with mixed inflammatory infiltrates of plasma cells, lymphocytes, eosinophils, and histiocytes. Myofibroblast is a modified fibroblast, differentiating into smooth muscle. It is usually located in granulation tissue, fibrous tissue, inflammatory reactive tissue, and some normal mesenchymal tissue. It is believed to perform a certain crucial role in the wound-repairing processes (4). Inflammatory pseudotumor is found in the lung and occasionally in the mouth, gastrointestinal tract, thyroid gland, kidney, lymph nodes, and skin (3). It has been treated with surgical removal, radiation therapy to the residual mass, and steroid therapy, with good results (4). This pseudotumor is important because of the difficulty in differentiating it from true neoplasms clinically and radiologically.

We present an extremely rare case of inflammatory pseudotumor originating in the spine. To our knowledge, only 11 cases originating in the spinal canal have been reported (1, 3–12).

Case Report

A 44-year-old man was admitted because of paraplegia and urinary incontinence. He had experienced steadily worsening thoracic pain for 10 months. For 4 weeks before admission, he had noticed difficulty in walking but no sphincter disturbance. For 1 week, he had experienced sphincter disturbance. On examination, he was paraplegic with numbness and sensory disturbance at and below the T4 dermatomes. Bilaterally, his knee and ankle reflexes were accelerated with positive ankle clonus. Hemoglobin level, white blood cell count, erythrocyte sedimentation rate, C-reactive protein level, and results of liver function tests were normal, as were the findings on chest radiography.

Thoracic spine CT showed a posterior epidural mass compressing the spinal cord without bony destruction (Fig 1). MR imaging demonstrated an expansile epidural mass from T1 to T7, compressing the thecal sac (Fig 2). The lesion also involved the left paraspinal space. There was no evidence of bone marrow abnormality in the vertebrae, adjacent bony destruction, or bony sclerosis on CT and MR images. Preoperatively, the initial diagnosis was a spinal epidural malignancy, such as a spinal lymphoma or metastatic tumor. We performed a T1–T7 laminoplastic laminectomy and subtotal resection of the mass, which was located in the epidural space inside the ligamentum flavum. The mass was slightly hard, yellowish, easily separated from the adjacent bone, and not hypervascular. It was firmly attached to the dura at its midlevel. After removing the mass,

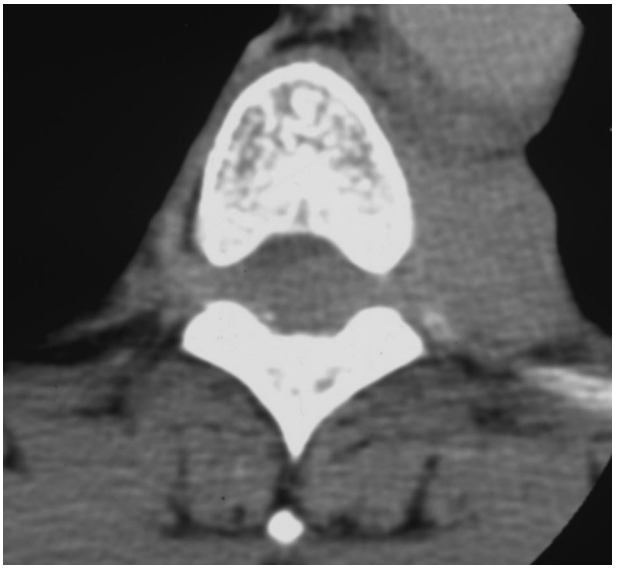

FIG 1. Thoracic spine CT scan shows an epidural mass compressing the spinal cord without bony destruction.

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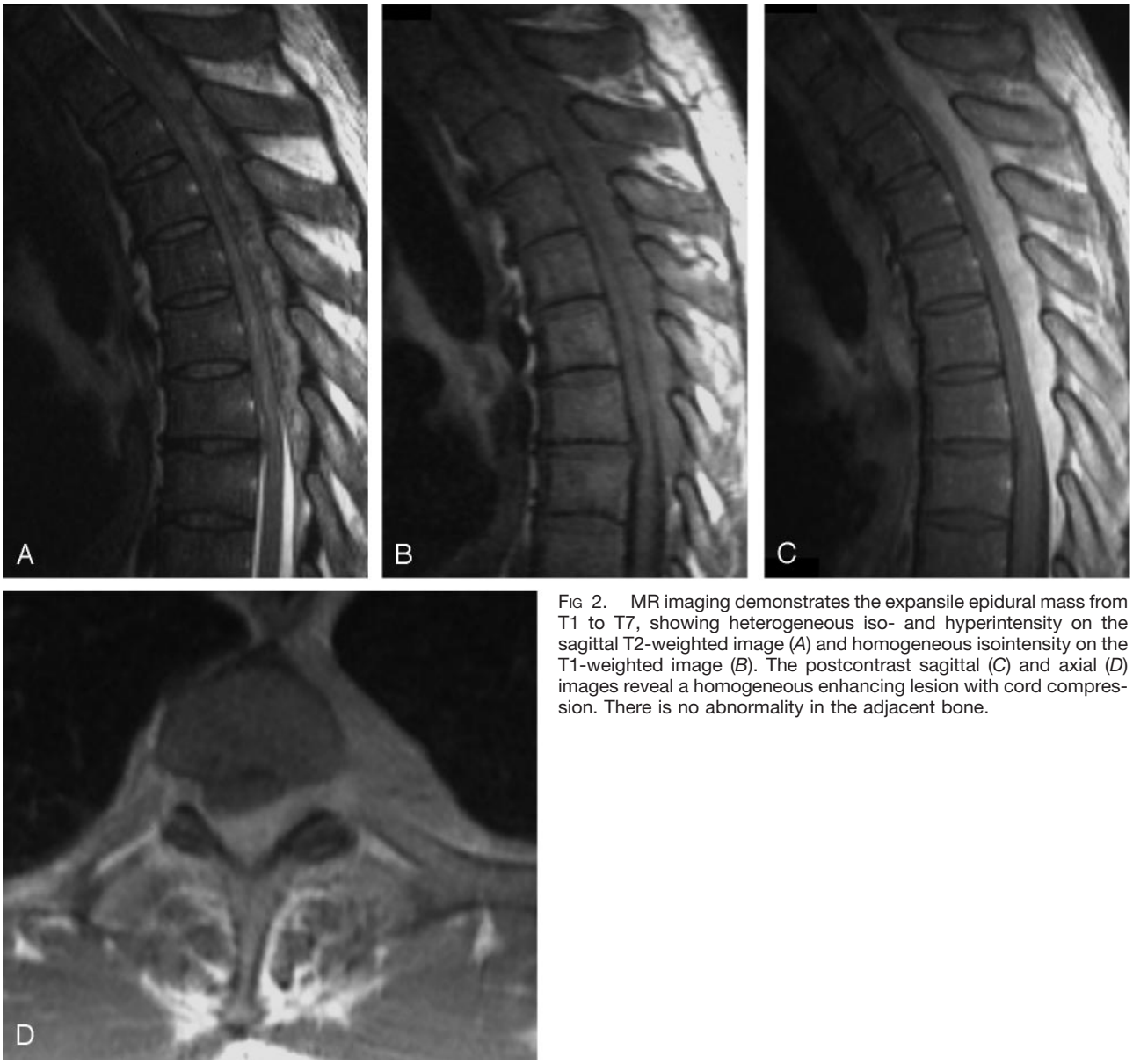

FIG 2. MR imaging demonstrates the expansile epidural mass from T1 to T7, showing heterogeneous iso- and hyperintensity on the sagittal T2-weighted image (*A*) and homogeneous isointensity on the T1-weighted image (*B*). The postcontrast sagittal (*C*) and axial (*D*) images reveal a homogeneous enhancing lesion with cord compression. There is no abnormality in the adjacent bone.

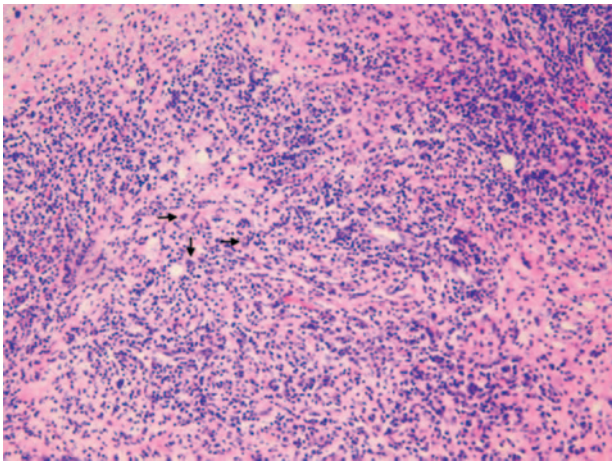

FIG 3. Microscopically, there is a polymorphic infiltrate composed of plump myofibroblasts (*arrows*) and lymphoplasma cells in loose fibromyxoid stroma, suggesting inflammatory myofibroblastic tumor (hematoxylin and eosin, original magnification ×100).

we sutured the dural defect by using a fascial flap. The results of staining and culture for bacteria and fungi were all negative.

Histopathologic examination revealed loosely arranged plump myofibroblasts in edematous stroma with extensive inflammatory infiltrates, characteristic of inflammatory myofibroblastic tumor (Fig 3). Some portions contained massive lymphoplasmacytic aggregates with vascular proliferation, resembling granulation tissue (Fig 4*A*). Nonetheless, others showed attenuated hyalinized collagenous stroma (Fig 4*B*). It was compatible with inflammatory pseudotumor. The possibility of multiple myeloma could be ruled out by immunohistochemical results revealing that kappa and lambda light chain expression was rarely present in different cells of the same morphologic type and that there was no evidence of clonality in the specimen. One year postoperatively, our patient's sensory disturbance and motor weakness had improved steadily, and he was ambulatory with a cane.

Discussion

Inflammatory pseudotumor is a chronic inflammatory tumefaction of unknown origin. The exis-

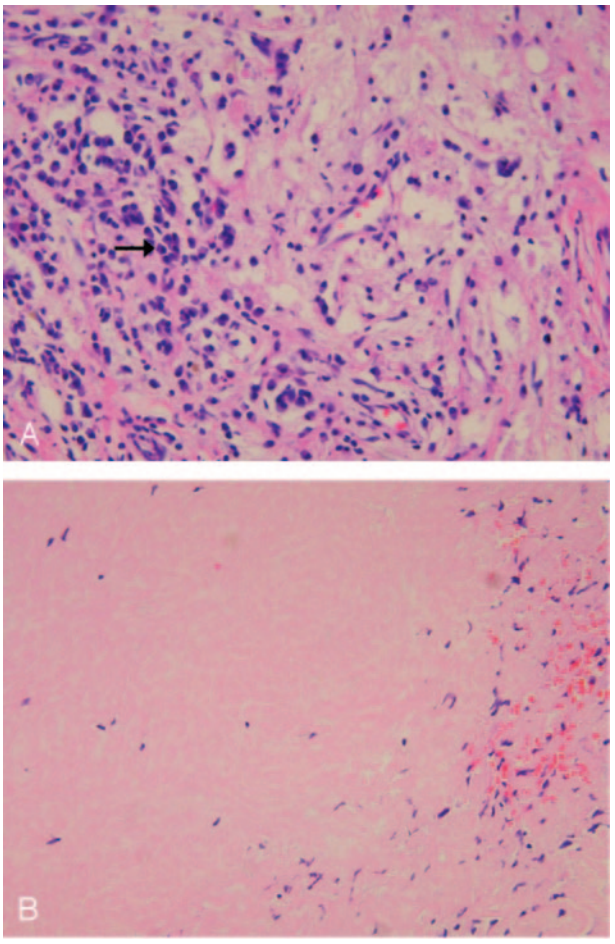

FIG 4. *A*, Abundant inflammatory cells such as plasma cells (*arrow*) in rich vascular stroma are seen in some areas (hematoxylin and eosin, original magnification ×200.)

B, Paucicellular area shows platelike collagen resembling scar tissue (hematoxylin and eosin, original magnification ×200).

tence of many synonyms for inflammatory pseudotumor and the varied histopathologic findings of this process suggest that it is not a single disease entity, but rather an umbrella term for any nonspecific chronic inflammatory mass lesion (13). The pathogenesis of inflammatory pseudotumor remains a matter of debate. Some cases have been associated with malignancy or tuberculosis as satellite lesions. The multiplicity of sites that can be involved suggests no particular route of entry or any specific agents. Prior surgery, trauma, or immune disturbances, in addition to infection are included for the possible etiology (11). Lesions can be located in the cervical, thoracic, or thoracolumbar spine. In relation to their locations in the spinal canal, of the 11 reported cases, 3 were in the extradural space, 4 were subdural, 1 was intra- and extradural, and 3 were intramedullary. Their important features are summarized in the Table.

Inflammatory pseudotumor has no distinguishing characteristics, either clinically or radiologically. Hence, the diagnosis of inflammatory pseudotumor can be made only after other specific disorders are ruled out. A few articles have reported that inflammatory pseudotumor shows low signal intensity on T1- and T2-weighted images and strong enhancement with gadopentetate dimeglumine (3, 5, 8). As the Table shows, low signal intensity on T2-weighted images appears radiologically suggestive of this disease entity, despite the limited number of cases. Han et al (14) suggested that T2 hypointensity of a soft-tissue lesion, which might be explained by a relative lack of both free water and mobile protons within fibrotic lesions, was characteristic of fibrosing inflammatory pseudotumor.

In our patient, the signal intensity showed heterogeneous iso- and hyperintensity on T2-weighted images, which differ from the signal intensity in reported cases. The histopathology can vary from polymorphous inflammatory cells and fibrosis with a matrix of granulation tissue, eosinophil, plasma cells, histiocytes, lymphoid follicles with germinal centers, and lymphocytes to a predominantly lymphocytic form embedded in a loose fibrous stroma (15). We speculate that the signal intensity on the T2-weighted images is dependent on the degree of reactive and fibrotic lesions within the inflammatory pseudotumor. Even though the areas of hyper- and isointensity on T2-weighted images could not correspond to the 2 different portions of the previously mentioned pathologic illustrations (Fig 4 *A*, *-B*), it is suggested that the area showing abundant inflammatory cells in vascular stroma could be hyperintensity on T2-weighted images. On the contrary, the paucicellular area in collagenous stroma might be isointensity or low signal intensity on the images.

Commonly, the differential diagnosis considered in spinal inflammatory pseudotumor cases includes spinal lymphoma, metastatic tumor, multiple myeloma, and meningioma. Without a biopsy, differentiating the diagnoses is very difficult. In general, typical findings of the lesions in spinal extradural lymphoma, metastasis, and myeloma include a low signal intensity on T1-weighted images and inhomogeneous hyperintensity on T2-weighted images compared with lesions in the spinal cord (16–18), and most lesions cause bone marrow abnormality, bone destruction, or hyperostosis. Therefore, it is impossible to distinguish these diseases from inflammatory pseudotumor on the basis of imaging findings, except for the relatively high frequency of intact adjacent bone in inflammatory pseudotumor.

Meningiomas are mostly isointense with the spinal cord on both T1- and T2-weighted images, and moderate relatively homogeneous enhancement is seen following contrast administration. Most spinal meningiomas have a broad-based dural attachment or a dural tail sign (19). Consequently, inflammatory pseudotumor cannot be distinguished from epidural lymphoma, metastasis, or myeloma preoperatively. When the preoperative MR imaging reveals no bony destruction and a normal fatty marrow, as in our patient, inflammatory pseudotumor should be included in the differential diagnosis.

Characteristics of cases of inflammatory pseudotumors reported in the literature that originated in the spinal canal

Source	Age (y)/Sex	Location	Relation with Meninges	Bony Destruction	Signal Intensity on MR Images Compared with Spinal Cord		
					T1-weighted	T2-weighted	Contrast-enhanced
Roberts et al, 1997 (1)	58/F	T9–T11	Epidural	Yes	Iso	Hypo	NR
Aizawa et al, 2002 (3)	46/M	C3–C7	Intramedullary	No	Iso	Hypo	Well
Jeon et al, 2005 (4)	60/F	L	Etramedullary intradural	NR	NR	NR	Well
Hsieh and Lin, 1995 (5)	37/M	T5, T12–L1	Extramedullary intradural	No	Low	NR	NR
Eimoto et al, 1978 (6)	37/M	C4–C5	Extramedullary intradural	No	NR	NR	NR
Gilliard et al, 2000 (7)	45/M	C3–T2	Epidural	Yes	Iso	NR	Well
Hsiang et al, 1994 (8)	57/M	T12–T13	Intra- and extradural	Yes	Low	NR	Well
Roberts et al, 2001 (9)	39/F	T5–T6	Epidural	No	Iso	Hypo	NR
Lee et al, 1998 (10)	3/F	C2–T10	Intramedullary	No	NR	NR	Well
Kilinc et al, 2002 (11)	34/M	T9–T12	Intramedullary	No	Low	Hyper	Well
Despeyroux-Ewers et al, 2003 (12)	22/F	T9	Extramedullary intradural	No	Iso	Hypo	Well

Note.—Iso indicates isointensity; Hypo, hypointensity; NR, not reported.

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