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Introduction

Polymyalgia rheumatica (PMR) is an inflammatory condition of unknown cause. It is characterized by aching and morning stiffness in the cervical region, the shoulder and pelvic girdles. It usually responds rapidly to low doses of corticosteroids and has a favorable prognosis (1). Inflammatory pseudotumor (inflammatory myofibroblastic tumor) is a benign tumor-like lesion of unknown cause. It occurs at various location in the body and shows up in only a small number of people (2). We present a rare case of epidural inflammatory pseudotumor mimicking epidural hematoma in the thoracic spine in a patient with polymyalgia rheumatica.

Case report

A 63-year-old male had a 6-year history of PMR. At the time of diagnosis of PMR, his symptoms were myalgias in cervical region and bilateral proximal regions of the arms. Laboratory testing, which included complete blood count, serum protein electrophoresis, biochemical survey and assays for antinuclear antibody and rheumatoid factor, was negative except for a C-reactive protein (CRP) of 9.4 mg/dl and a Westergren erythrocyte sedimentation rate (ESR) of 70 mm/h. Biopsy of the temporal artery did not show vasculitis related to giant cell arteritis. Treatment with prednisone and cyclosporine was associated with resolution of symptoms and lowering of his ESR to 25 mm/h. Over the next six years he received adequate relief of symptoms using low dose prednisone at the average dose of 5 mg/day and cyclosporine at the average dose of 50 mg/day.

The patient presented with a history of back pain for two weeks and numbness of the lower extremities for one week. He complained of rapid worsening of gait disturbance for the last 3 days. There was no history of trauma or anticoagulation therapy. At his physical examination upon admission, he had incomplete paraplegia and increased deep tendon reflexes of the lower extremities. He was unable to stand without a walker and had a mild bladder dysfunction. The results of chest and thoracolumbar spine x-rays and laboratory examinations were normal. Magnetic resonance imaging (MRI) on the 12th day after the onset of the symptoms showed spinal cord compression at the T5-T6 level; this was caused by a posterior epidural mass. It was isotense to the spinal cord in T1 sequence and hypointense in the anterior side and hyperintense in the posterior side in T2 sequence (Figure 1).

Based on the patient's MRI findings, it was believed that he had an epidural hematoma in the thoracic spine, -hence he was scheduled for surgery. The patient underwent a T5-T6 laminectomy and a total excision of the mass, which was located in the epidural space and involved the ligament flavum and epidural adipose tissue (Figure 2). The mass was slightly hard, yellowish, easily separated from the adjacent bone, and not hypervascular. It was firmly attached to the dura mater. The results of staining and culture for bacteria and fungi were all negative.

Histopathologic examination revealed severe lymphoplasmacytic infiltration with fibrosis in the mass; this consisted of the ligament flavum and the epidural adipose tissue. The inflammatory infiltration existed in the entire specimen and there were no evidence of hematomas or tumorous lesions (Figure 3). Immunohistochemical studies showed the infiltrating population to consist of both T and B lymphocytes, The B cells were polyclonal as assessed by light chain expression.

After surgery, the patient's pain and neurological symptoms disappeared immediately. Two years after surgery, the patient is now neurologically normal and has not had a recurrence on the follow-up MRIs.

Discussion

Inflammatory pseudotumor is a chronic inflammatory tumefaction of unknown origin. It is found most often in the lung with extrapulmonary occurrence at sites including orbit, nasal sinuses, liver, spleen, pancreas, bowel, kidney, urinary bladder, testis, heart and lymphatic system (2). We only found the six previous cases of epidural inflammatory pseudotumor in the spine published in the literature (2-6) (Table 1). Inflammatory pseudotumor has no distinguishing characteristic, either clinically or radiologically. Some articles have reported that inflammatory pseudotumor shows low signal intensity on T1- and T2- weight images and strong enhancement with gadolinium (7-9). As the Table 1 shows, low signal intensity on T2-weighted images appears radiologically suggestive of this disease entity. Han et al. (10) 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.

The pathogenesis of inflammatory pseudotumor is unknown. However it is considered an immunologic host response to infectious agents, microorganisms, neighboring necrotic tissue or chronic inflammation, neoplasms, or foreign bodies (11). The patient had an inflammatory pseudotumor in the course of PMR. Polymyalgia rheumatica (PMR) is a relatively common inflammatory condition that generally occurred in patients older than 50 years. It is characterized by aching and morning stiffness in the cervical region, the shoulder and pelvic girdles. The prevalence of PMR has been estimated to be 0.5% of the population (12). In these patients the erythrocyte sedimentation rate (ESR) and C-reactive

protein (CRP) are usually elevated. Bursitis or tenosynovitis in the proximal limb and joint areas is usually identified using scintigraphy, MRI, and ultrasonography (1, 12). Some articles have reported cervical interspinous bursitis in PMR identified using MRI (13, 14). Although active interspinous bursitis was not observed in the patient at that time, it might have the potential to lead to epidural inflammatory pseudotumors. PMR may occur as an isolated disease or it may be observed in the setting of giant cell arteritis (GCA). GCA is a chronic vasculitis of large and medium-sized vessels. Temporal-artery abnormality on physical examination characterized as tenderness or decreased pulsation and vasculitis proven by biopsy of the artery are very important for the diagnosis of GCA (15). Sailler et al. described two patients with epidural inflammatory pseudotumors in the cervicothoracic spine with biopsy-proven GCA. Inflammatory pseudotumors are also exceptional in the course of GCA (6). They can involve the genital tract, the breast, retro-obital tissue, the aorta, and the small bowel. Pachymeningitis rarely has been reported (16), and not as a cause of spinal cord compression. The diagnosis in the patient was established as PMR without any findings in his temporal-arteries. However PMR and GCA are closely related conditions and some authorities consider them to be different phases of the same disease (1).

Surgical excision is usually mandatory in inflammatory pseudotumor compressing the spinal cord because of the emergent need of relieving the mass effect; it is generally curative when total excision is performed (2-8, 11). Systemic steroid and immunosuppressive drugs or radiotherapy are also given in inflammatory pseudotumor and lead to a decrease in volume of the mass (3, 7-8, 11).

Conclusion

We present a rare case of epidural inflammatory pseudotumor in thoracic spine in a patient with polymyalgia rheumatica. Total excision confirmed the diagnosis and resulted in complete relief of the symptom.

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Figure Captions

Figure 1. MRI showing a dorsal epidural mass at T5-6 level causing spinal cord compression. (A) T1-weighted sagittal image (B) T2-weighted sagittal image (C) T2-weighted axial image at T6 pedicle

Figure 2. Excised specimen. The mass was firmly attached to the dura matter. (A) the ventral side (B) the lateral side.

Figure 3. Histopathologic examination revealed severe lymphoplasmacytic infiltration with fibrosis in the mass which involved the ligament flavum and epidural adipose tissue. The inflammatory infiltration existed in the entire specimen and there were no hematomas or tumorous lesions (stained with hematoxylin-eosin, magnification $\times$10 and 200).

Table 1. Characteristics of cases of epidural inflammatory pseudotumor in the spine reported in the literature

Source	Age(y) /Sex	Location	Comorbidity	Bony Involvement	Signal Intensity on MR images Compared with Spinal Cord		
					T1-weighted	T2-weighted	Contrast-enhanced
Roberts et al, 1997 (2)	58/F	T9-T11	Hypertension	Yes	Iso	Hypo	NR
Gilliard et al, 2000 (3)	45/M	C3-T2	Multifocal Fibrosclerosis	Yes	Iso	NR	Well
Roberts et al, 2001 (4)	39/F	T5-T6	None	No	Iso	Hypo	NR
Seol et al, 2005 (5)	44/M	T1-T7	NR	No	Iso	Iso-Hyper	Well
Sailler et al, 2006 (6)	78/M	C6-T3	Giant Cell Arteritis	NR	NR	Hypo	Well
	73/F	T5-T7	Giant Cell Arteritis	NR	NR	Hypo	Well
Our case	63/M	T5-T6	Polymyalgia rheumatica	No	Iso	Hypo-Hyper	NR

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

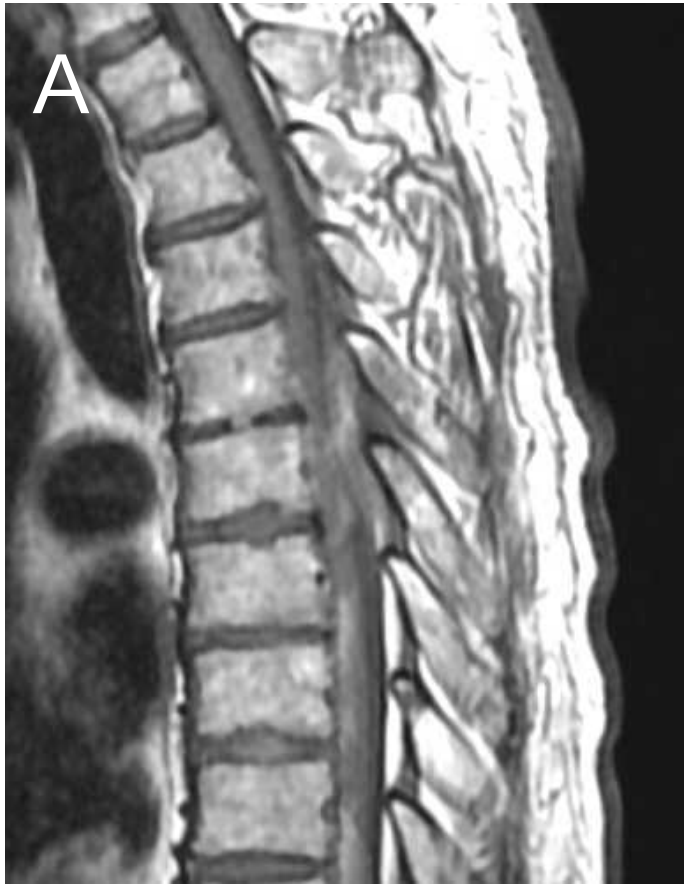
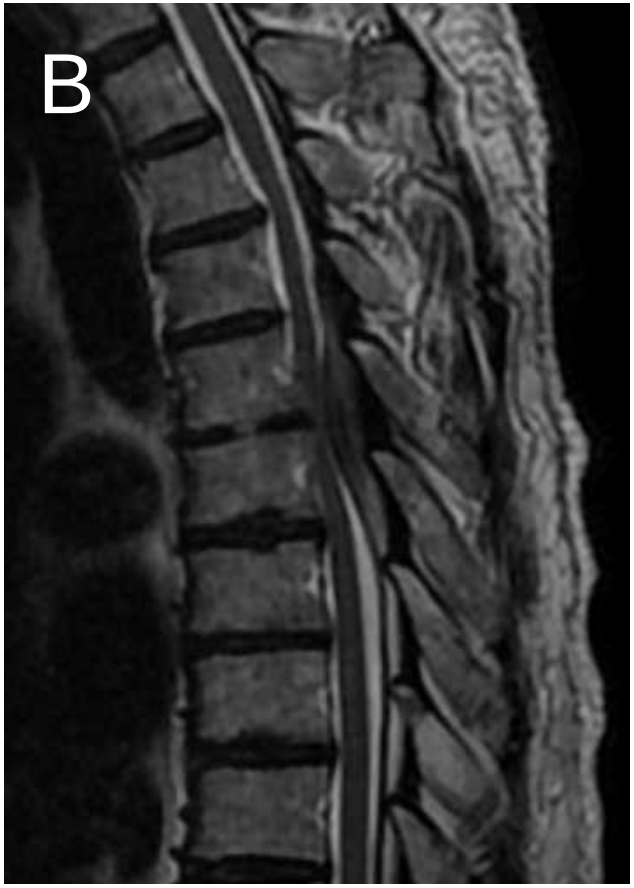


Fig. 1(A)



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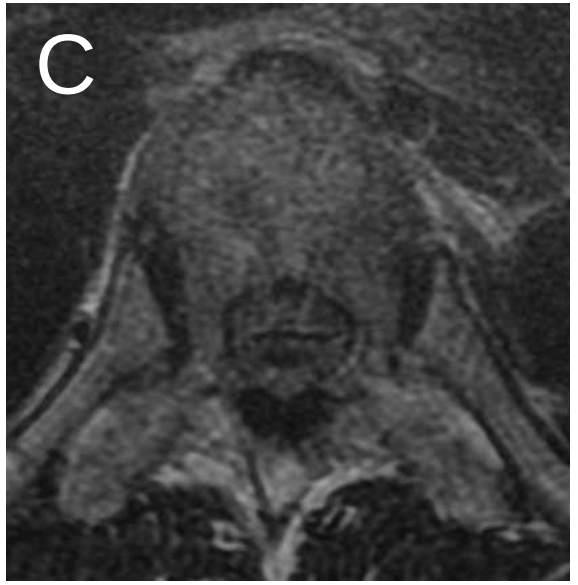

Fig. 1(C)

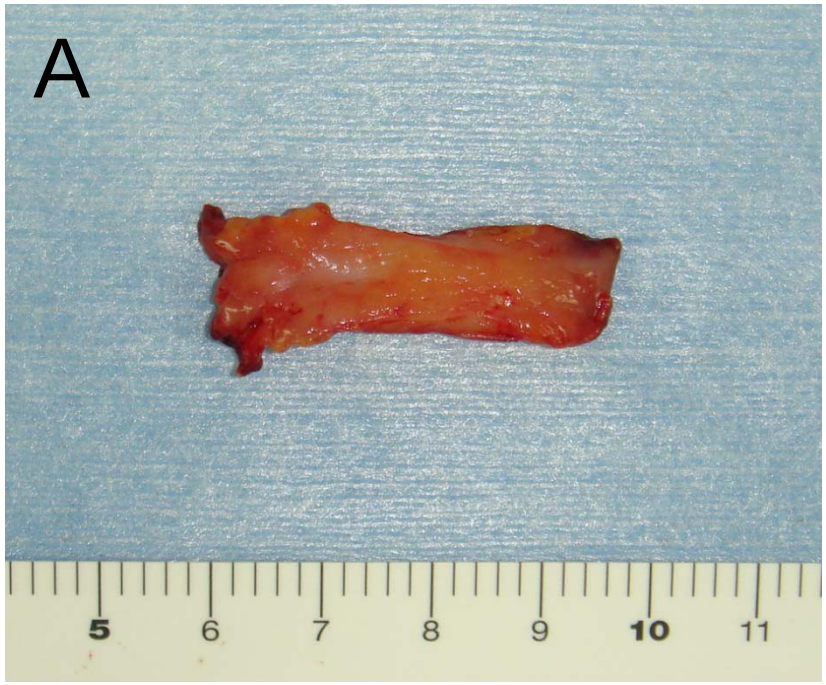

Fig. 2(A)

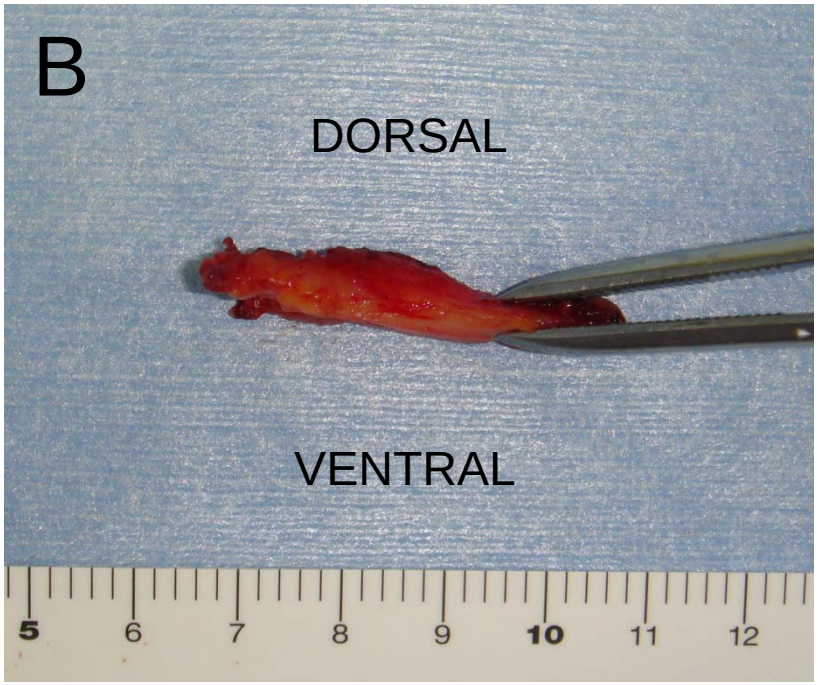

Fig. 2(B)

Fig. 3