

# Epidural inflammatory pseudotumor in the thoracic spine in a patient with polymyalgia rheumatica

メタデータ	言語: eng 出版者: 公開日: 2017-10-05 キーワード (Ja): キーワード (En): 作成者: メールアドレス: 所属:
URL	<a href="http://hdl.handle.net/2297/31995">http://hdl.handle.net/2297/31995</a>

**Title**

Epidural inflammatory pseudotumor in the thoracic spine in a patient with polymyalgia rheumatica

**Authors**

Satoshi Kato, MD, Hideki Murakami, MD, Satoru Demura, MD,  
Katsuhito Yoshioka, MD, Okamoto Yoshiyuki, MD, Hiroyuki Hayashi, MD,  
Hiroyuki Tsuchiya, MD

**Affiliations**

Department of Orthopaedic Surgery, Kanazawa University School of Medicine,  
Kanazawa, Japan

**Corresponding author**

Satoshi Kato, MD

Department of Orthopaedic Surgery, Kanazawa University School of Medicine,  
13-1 Takara-machi, Kanazawa, 920-8641, Japan

Phone: +81-76-265-2374

Fax: +81-76-234-4261

E-mail: skato323@gmail.com

1 **Title**

2 Epidural inflammatory pseudotumor in the thoracic spine in a patient with  
3 polymyalgia rheumatica

4

5

6

7

8

9

10

11

12

13

14

15

16

17

18

19

20

21

22

23

24

25

26

27

28

29

30

31

32

33

34

35

1 **Introduction**

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

11  
12  
13  
14  
15  
16  
17  
18  
19  
20  
21  
22  
23  
24

1 **Case report**

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

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

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

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

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

19  
20  
21  
22  
23  
24

1 **Discussion**

2 Inflammatory pseudotumor is a chronic inflammatory tumefaction of unknown  
3 origin. It is found most often in the lung with extrapulmonary occurrence at sites  
4 including orbit, nasal sinuses, liver, spleen, pancreas, bowel, kidney, urinary  
5 bladder, testis, heart and lymphatic system (2). We only found the six previous  
6 cases of epidural inflammatory pseudotumor in the spine published in the  
7 literature (2-6) (Table 1). Inflammatory pseudotumor has no distinguishing  
8 characteristic, either clinically or radiologically. Some articles have reported that  
9 inflammatory pseudotumor shows low signal intensity on T1- and T2- weight  
10 images and strong enhancement with gadolinium (7-9). As the Table 1 shows,  
11 low signal intensity on T2-weighted images appears radiologically suggestive of  
12 this disease entity. Han et al. (10) suggested that T2 hypointensity of a  
13 soft-tissue lesion, which might be explained by a relative lack of both free water  
14 and mobile protons within fibrotic lesions, was characteristic of fibrosing  
15 inflammatory pseudotumor.

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

1 protein (CRP) are usually elevated. Bursitis or tenosynovitis in the proximal limb  
2 and joint areas is usually identified using scintigraphy, MRI, and ultrasonography  
3 (1, 12). Some articles have reported cervical interspinous bursitis in PMR  
4 identified using MRI (13, 14). Although active interspinous bursitis was not  
5 observed in the patient at that time, it might have the potential to lead to epidural  
6 inflammatory pseudotumors. PMR may occur as an isolated disease or it may be  
7 observed in the setting of giant cell arteritis (GCA). GCA is a chronic vasculitis of  
8 large and medium-sized vessels. Temporal-artery abnormality on physical  
9 examination characterized as tenderness or decreased pulsation and vasculitis  
10 proven by biopsy of the artery are very important for the diagnosis of GCA (15).  
11 Sailer et al. described two patients with epidural inflammatory pseudotumors in  
12 the cervicothoracic spine with biopsy-proven GCA. Inflammatory pseudotumors  
13 are also exceptional in the course of GCA (6). They can involve the genital tract,  
14 the breast, retro-orbital tissue, the aorta, and the small bowel. Pachymeningitis  
15 rarely has been reported (16), and not as a cause of spinal cord compression.  
16 The diagnosis in the patient was established as PMR without any findings in his  
17 temporal-arteries. However PMR and GCA are closely related conditions and  
18 some authorities consider them to be different phases of the same disease (1).

19 Surgical excision is usually mandatory in inflammatory pseudotumor  
20 compressing the spinal cord because of the emergent need of relieving the mass  
21 effect; it is generally curative when total excision is performed (2-8, 11). Systemic  
22 steroid and immunosuppressive drugs or radiotherapy are also given in  
23 inflammatory pseudotumor and lead to a decrease in volume of the mass (3, 7-8,  
24 11).

1 **Conclusion**

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

5

6

7

8

9

10

11

12

13

14

15

16

17

18

19

20

21

22

23

24

## 1   **References**

2  
3  
4  
5  
6  
7  
8  
9  
10  
11  
12  
13  
14  
15  
16  
17  
18  
19  
20  
21  
22  
23  
24  
25  
26  
27  
28  
29  
30  
31

1. Carlo S, Fabrizio C, Luigi B, Gene GH. Polymyalgia rheumatica and giant-cell arteritis. *N Engl J Med* 2002; 347: 261-71.
2. Roberts GA, Eldridge PR, Mackenzie JM. Case report: inflammatory pseudotumour of the spine, with literature review. *Br J Neurosurg* 1997; 11: 570-2.
3. Gilliard C, De Coene B, Lahdou JB, et al. Cervical epidural pseudotumor and multifocal fibrosclerosis: case report and review of the literature. *J Neurosurg Spine* 2000; 93: 152-6.
4. Roberts G, Farrell M, Allcutt D. Spinal inflammatory pseudotumours. *Br J Neurosurg* 2001; 15: 197-8.
5. Seol JH, Kim SS, Kim JE, et al. Inflammatory pseudotumor in the epidural space of the thoracic spine: a case report and literature review of MR imaging findings. *Am J Neuroradiol* 2005; 26: 2667-70.
6. Sailer LJ, Porte L, Ollier SM, et al. Giant cell arteritis and spinal cord compression; an overlap syndrome? *Mayo Clin Proc* 2006; 81: 89-91.
7. Aizawa T, Sato T, Tanaka Y, et al. Intramedullary plasma cell granuloma in the cervicothoracic spine: case report. *J Neurosurg Spine* 2002; 92: 235-8.
8. Hsieh PC, Lin CN. Multicentric plasma cell granuloma of spinal cord meninges. *Clin Orthop* 1995; 317: 188-92.
9. Hsiang J, Moorhouse D, Barba D. Multiple plasma cell granulomas of the central nervous system: case report. *Neurosurgery* 1994; 35: 744-7.
10. Han MH, Chi JG, Kim MS, et al. Fibrosing inflammatory pseudotumor involving the skull base: MR and CT manifestations with histopathologic comparison. *AJNR Am J Neuroradiol* 1996; 17: 515-21.
11. Boutarbouch M, Arkha Y, Rifi L, et al. Intradural cervical inflammatory pseudotumor mimicking epidural hematoma in a pregnant woman: case report and review of the literature. *Surg Neurol* 2008; 69: 302-5.
12. Chuang TY, Hunder GG, Ilstrup DM, Kurland LT. Polymyalgia rheumatica: a 10-year epidemiologic and clinical study. *Ann Intern Med* 1982; 97: 672-80.

1 13. Gonzalez-Gay MA. The clinical implication of cervical interspinous bursitis  
2 in the diagnosis of polymyalgia rheumatica. *Ann Rheum Dis* 2008; 67:  
3 733-4.

4 14. Salvarani C, Barozzi L, Cantini F, et al. Cervical interspinous bursitis in  
5 active polymyalgia rheumatica. *Ann Rheum Dis* 2008; 67: 758-61.

6 15. Hunder GG, Bloch DA, Michel BA, et al. The American College of  
7 Rheumatology 1990 criteria for the classification of giant-cell arteritis.  
8 *Arthritis Rheum* 1990; 33: 1122-8.

9 16. Marano E, D'Armiento FP, Scarano V, et al. Focal hypertrophic cranial  
10 pachymeningitis associated with temporal arteritis: a new case report. *J*  
11 *Neurol.* 2003; 250: 98-100.

12  
13  
14  
15  
16  
17  
18  
19  
20  
21  
22  
23  
24  
25  
26  
27  
28  
29  
30  
31  
32  
33  
34  
35

1 **Figure Captions**

2

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

6

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

9

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

15

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
Sailer 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.



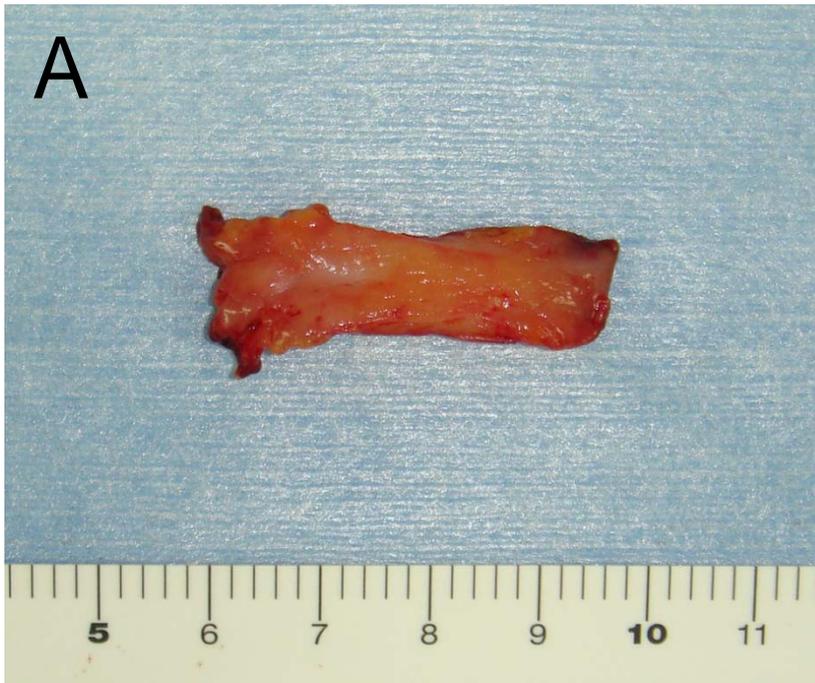
Fig. 1(A)



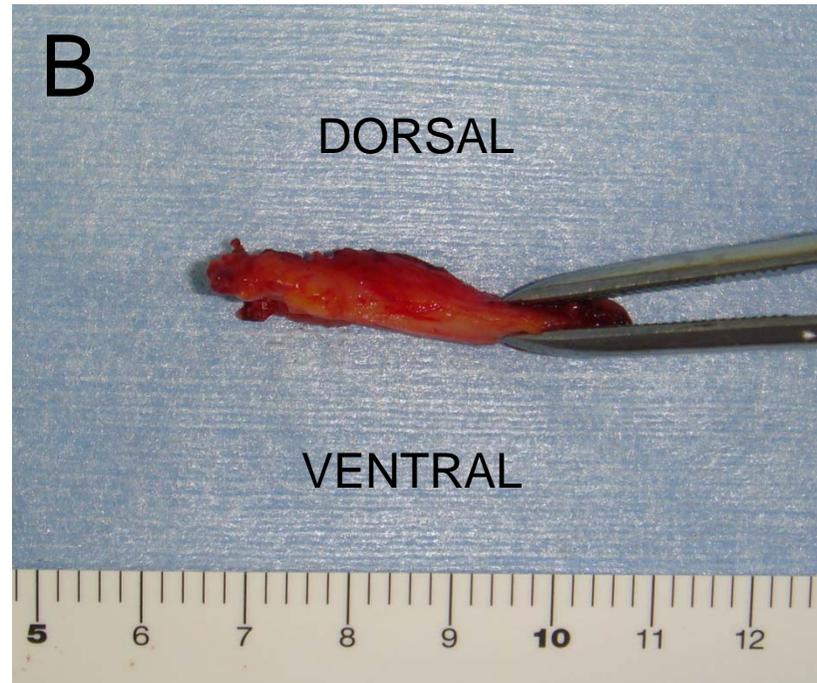
Fig. 1(B)



Fig. 1(C)



**Fig. 2(A)**



**Fig. 2(B)**

Fig. 3

