JRD Journal of Rheumatic Diseases Vol. 23, No. 4, August, 2016
http://dx.doi.org/10.4078/jrd.2016.23.4.261

Case Report

A Case of Polymyositis Presenting as Bent Spine Syndrome

Jeongim Choi[1], Dam Kim[1], Jae-Bum Jun[1], Seunghun Lee[2], Seung Sam Paik[3], Yoon-Kyoung Sung[1]

[1]Department of Rheumatology, Hanyang University Hospital for Rheumatic Diseases, Departments of [2]Radiology and [3]Pathology, Hanyang University College of Medicine, Seoul, Korea

Polymyositis (PM) is a subset of idiopathic inflammatory myopathies. The muscles involved with PM are typically proximal and distal limb muscles, but paraspinal muscles are rarely affected. The primary PM clinical symptom is gradual proximal muscle weakness but unusually abnormal trunk posture. Bent spine syndrome (BSS), also referred to camptocormia, is defined as an abnormal flexion of the trunk, appearing in standing position. An idiopathic axial myopathy is the most common cause of primary BSS. A few cases of inflammatory myopathy, a secondary BSS, have been reported. We describe a 59-year-old polymyositis patient with normal finding on an magnetic resonance imaging femur scan who presented with BSS only, myopathic findings on electromyography and elevation of muscle enzymes. **(J Rheum Dis 2016;23:261-265)**

Key Words. Polymyositis, Bent spine syndrome, Camptocormia

INTRODUCTION

Idiopathic inflammatory myopathies are heterogeneous groups of muscle inflammation with unknown etiology [1,2]. Among them, polymyositis (PM) is generally manifested by progressive and symmetric proximal muscle weakness, myopathic finding on electromyography (EMG) and high elevation of muscle enzymes, and is confirmed by muscle biopsy [1,2]. The involved muscles in PM were typically proximal and distal limb muscles, neck flexor muscles, and oropharyngeal muscles but paraspinal muscles were rarely affected [3].

Bent spine syndrome (BSS), also called camptocormia, is defined as an abnormal flexion of the trunk, appearing in standing position, increasing during walking and disappearing in supine position [4-7]. It was initially considered as a psychogenic disorder, but now it has been established that apart from being a syndrome of psychiatric origin. Many cases of BSS originate from muscular diseases; idiopathic axial myopathy is the most common cause of primary BSS, while muscular dystrophy, mitochondrial myopathy, and endocrine disease such as hypothyroidism and osteomalacia might be secondary causes of BSS [4-8].

Though PM is one of causative diseases in secondary BSS, it is uncommon to see the patients with PM only manifested with BSS. Since we have experienced a patient with BSS due to PM without proximal muscle involvement, we would like to describe this case with literature review.

CASE REPORT

A 59-year-old woman came to our clinic with complaint of mild weakness in the lower extremities and flexion forward while walking that lasts 2 years. Laboratory testing revealed elevated lactate dehydrogenase (LDH) 699 IU/L (normal range, 60 to 200 IU/L), creatine kinase (CK) 2,780 IU/L (30 to 180 IU/L) and myoglobin 717 ng/mL (14.3 to 65.8 ng/mL). Serum aldolase level was 46.0 IU/mL (0.0 to 7.6 IU/mL). She had no pathognomic skin lesions and her magnetic resonance imaging (MRI) scan of thighs showed no increase of signal intensity. However, the EMG of both upper and lower extremities suggested

Received : September 27, 2015, **Revised :** November 2, 2015, **Accepted :** November 2, 2015

Corresponding to : Yoon-Kyoung Sung, Department of Rheumatology, Hanyang University Hospital for Rheumatic Diseases, 222 Wangsimni-ro, Seongdong-gu, Seoul 04763, Korea. E-mail : sungyk@hanyang.ac.kr

pISSN: 2093-940X, eISSN: 2233-4718

myopathy. We could not find out target muscle for muscle biopsy but she was compatible for possible PM. Therefore, we had started moderate dose of corticosteroid (prednisolone 20 mg daily) and immunosuppressant (azathioprine 50 mg daily), muscle weakness was improved and elevated muscle enzymes were lowered. After 6 months, she stopped visiting our clinic by herself and stayed without medication. Ten months after last visit, she came to our clinic by conspicuously abnormal flexion of trunk, during walking and standing without back pain which definitely differed from typical PM patients.

On admission, physical examination revealed proximal muscle weakness of both upper and lower extremities Grade as 4/5 Medical Research Council (MRC) for muscle strength scale [9] and the patient did not exhibit dermatomyositis-associated skin lesion: heliotrope rash, V sign, shawl sign or periungal change.

Laboratory studies revealed slight elevation of erythrocyte sedimentation rate 23 mm/h (0 to 20 mm/h) and C-reactive protein level 1.1 mg/dL (0 .0 to 0.8 mg/dL). A complete blood count revealed a white blood cell count of 8,000/ μL (66.4% neutrophil), hemoglobin 16 g/dL, platelet 269,000/mm^3. Blood chemistry showed serum alanine aminotransferase (ALT), serum aspartate amino-

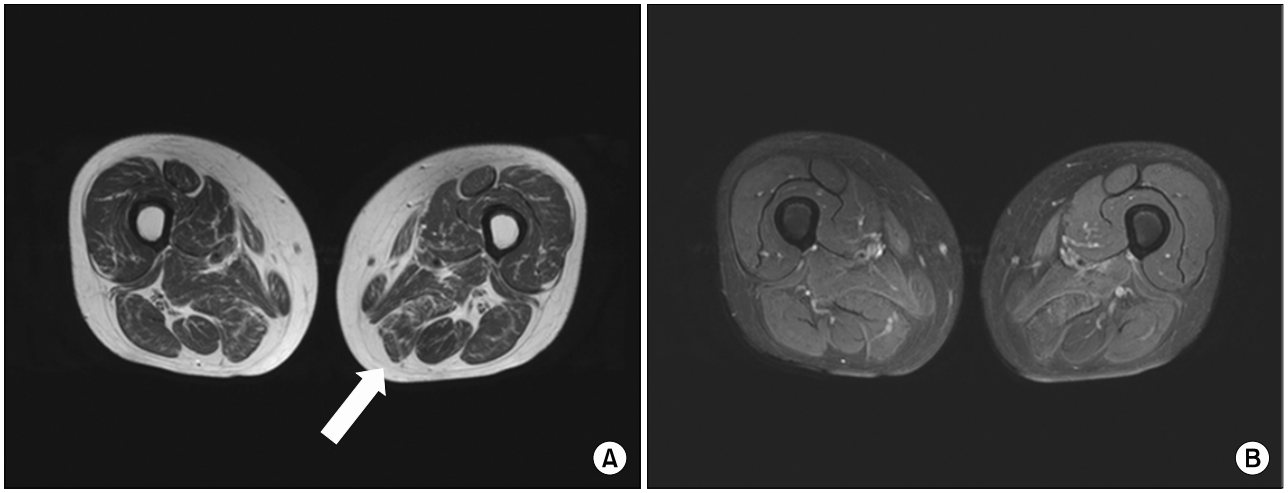

Figure 1. Magnetic resonance imaging scan for thighs at rehospitalization. Axial T2-weighted scan (A) and axial fat-saturated T2-weighted scan (B) revealed bilaterally fatty atrophy of semimembranosus muscle (arrow) without active inflammation.

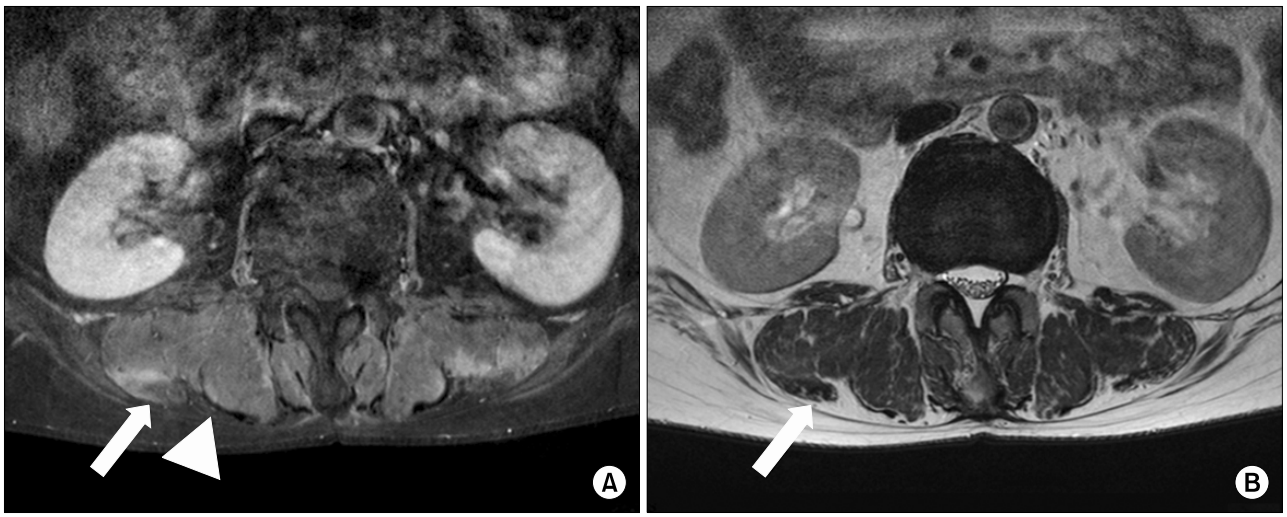

Figure 2. Axial lumbar spine magnetic resonance imaging. Fat saturated T1-weighted scan (A) and T2-weighted scan (B) showed paraspinous muscle edema with enhancement, especially worse on the right iliocostalis muscle (arrow) and longissimus (arrow head) at the level of L2 and L3.

transferase (AST) of 70/90 IU/L (5 to 40 IU/L/5 to 45 IU/L) elevated LDH 707 IU/L (60 to 200 IU/L), CK 2,590 IU/L (30 to 180 IU/L), myoglobin 801 ng/mL (14.3 to 65.8 ng/mL) and aldolase 47.2 IU/mL (0.0 to 7.6 IU/mL). Anti-nuclear antibody (ANA) testing was positive at high titer in a cytoplasmic pattern (1:2,560). Anti-Jo-1 antibody and other autoantibodies to specific antigens were all negative. Thyroid stimulating hormone (THS, thyrotropin) and free T4 levels were in normal range. A thorough screening for occult cancer yielded normal results.

Rechecked MRI scan for femur showed fatty atrophic change without active inflammation in thigh muscle (Figure 1). Therefore, additional gadolinium enhanced MRI scan for lumbar spine was performed to evaluate abnormal flexion of trunk, it showed diffuse edema of the paraspinal muscles and demonstrated intense enhancement of the muscles (Figure 2). We performed ultrasound-guided gun biopsy for the right paraspinal muscles and its pathologic findings revealed atrophy of muscular fibers, endomysial lymphocytic infiltration, fibrosis and infiltrations of CD8, CD68 positive T-cell (Figure 3).

PM with secondary BSS was diagnosed, and the patient was started on high dose of prednisolone (50 mg/day, 1 mg/kg). In the outpatient clinics, methotrexate 10 mg/week and azathioprine 50 mg/day were subsequently added to spare the dose of prednisolone.

During ten months after PM with secondary BSS diagnosis, the prednisolone was tapered to 5.0 mg/day, and bending spine was improved with decreasing muscle enzymes to their normal ranges.

DISCUSSION

BSS is a postural disorder characterized by abnormal flexions with walking and standing. It was first described by French neurologist during World War I that young soldier acutely bent a forward posture. BSS was regarded as conversion disorder but it was revealed abnormal fatty infiltration of paraspinal muscle in 1991 [4].

The majority of BSS of muscular origin was related to a primary idiopathic axial myopathy that described separate clinical entity. The primary idiopathic axial myopathy was characterized by a progressive weakness of paravertebral muscles in elderly patients, fatty infiltration of paraspinal muscles in computed tomography (CT)/MRI scan, and replacement of fat tissue and fibrosis in paraspinal muscle biopsy [4, 5,10]. Laboratory results was almost normal range, in some cases serum CK was elevated 2 to 5 times with upper normal limits [8] . Until now, there is still no cure and specific therapy for primary axial myopathy [4].

Secondary BSS was classified into various muscular disorders including inflammatory myositis, dystrophy and endocrine-metabolic causes and neurological disorders such as amyotrophic lateral sclerosis and Parkinson's disease [4-7,11,12]. The secondary BSS with idiopathic inflammatory myopathies were reported a few cases of PM [13-15] (Table 1). In 2009, the first case of BSS associated PM was reported in a 59-year old female [13]. Although they were compatible with secondary BSS due to inflammatory myopathy, they also have proximal muscle involvement. Hence, most of them were performed mus-

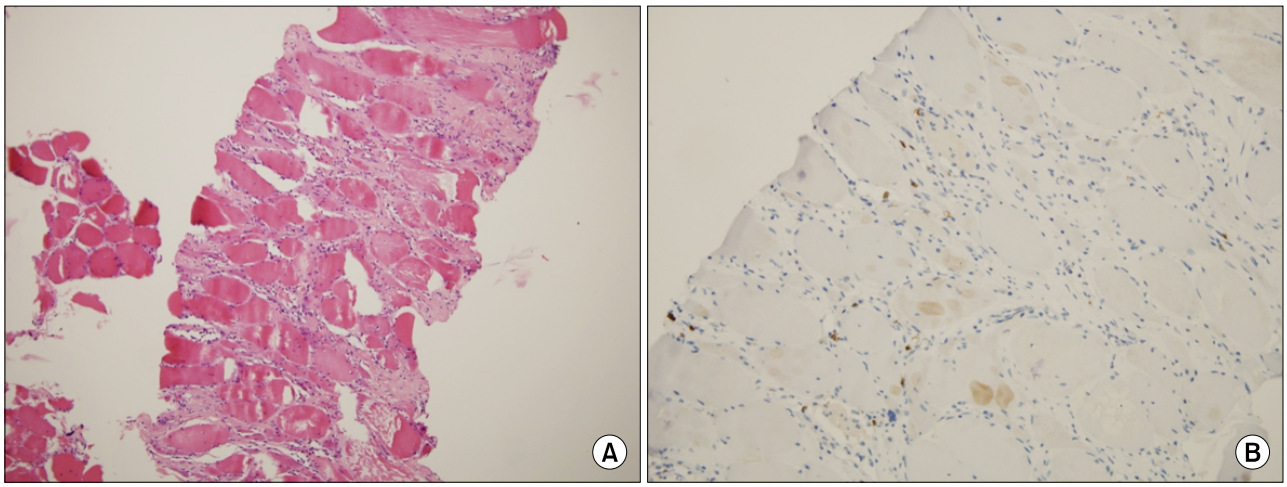

Figure 3. Pathologic finding of paraspinal muscle. (A) The right longissimus muscle demonstrates endomysial lymphocytic infiltration (H&E, ×100). (B) CD8 cytotoxic T-cell in the same tissue (immunohistochemical stain, ×200).

Table 1. Patient cases of polymyositis with bent spine syndrome

Case	Sex/ age (yr)	Presenting symptom	Serum CK	EMG	Biopsy site	Histology	Treatment	Reference
1	F/59	Difficulty holding back straight while standing or walking	526 (24～173 IU/L)	Neurogenic	Quadriceps	Lymphocytic infiltration of the endomysium	IVMP	[13]
2	M/67	Muscle weakness of thoracolumbar spine	1,103 (24～173 IU/L)	Proximal inflammatory myopathy	Biceps	Perimysial lymphomononuclear inflammatory infiltration	Corticosteroid, azathioprine, methotraxate	[14]
3	F/70	NA	248 (＜110 IU/L)	Myopathic	Quadriceps	Normal	Corticosteroid	[15]
4	F/66	NA	350 (＜110 IU/L)	Myopathic	Quadriceps	Normal	IVMP, IVIG	[15]
5	F/59	Flexion forward while walking	2,780 (30～180 IU/L)	Myopathic	Paraspinal muscle	Atrophy of muscular fibers, endomysial lymphocytic infiltration, fibrosis	IVMP, azathioprine, methotraxate	Present study

CK: creatine kinase, EMG: electromyography, F: female, IVMP: intravenous methyprednisolone, IVIG: intravenous immunoglobuline, M: male, NA: not available.

cle biopsy from thigh muscle. However, in our case, it was difficult to diagnose PM with secondary BSS because there was no definite inflammation in proximal muscle and only paraspinal muscles were involved. Although patient showed high elevation of muscle enzymes, compliant of mild proximal muscle weakness and myopathic finding on EMG, there was no evidence of active muscle inflammation on MRI scan of both thighs. Additionally checked L-spine MRI scan finally revealed paraspinal muscle inflammation. Hence, we performed muscle biopsy and it revealed inflamed muscle with CD 8-positive lymphocytic infiltration in endomysium. Finally, she was diagnosed as PM with paraspinal muscles involvement. We could make a definite diagnosis as PM with secondary BSS after taking a careful history and MRI for lumbar spine and biopsy.

Through this case, we have learned two lessons. One is the importance of careful history taking. She initially complained about the posture change but we had focused on the proximal muscle weakness and elevated enzymes. Since we could not point out the exact muscle involved with MRI scan, it might be connected to non-compliance of patient. The other one is the possibility of BSS in many patients with complaint of abnormal posture in our clinic. Among patients who have BSS, it is not easy to distinguish the etiology of BSS between idiopathic axial myopathy and inflammatory myopathy, since they are almost same symptom except more frequent accompanying with proximal muscle weakness in inflammatory myopathy. In addition, muscle enzyme is not a useful tool to identify the etiology of BSS in these patients. Therefore, it is important to take into account the possibility of BSS in patients with posture change and thoughtful history taking and physical examinations are essential prior to perform laboratory tests and imaging studies.

SUMMARY

We described a rare case of PM with normal finding on MRI scan of femurs and manifested as BSS. The physicians should take account BSS in patients with posture change regardless of elevation of muscle enzymes and thought history taking and physical examinations are essential to diagnose BSS.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

REFERENCES

1. Christopher-Stine L, Plotz PH. Adult inflammatory myopathies. Best Pract Res Clin Rheumatol 2004;18: 331-44.
2. Dalakas MC, Hohlfeld R. Polymyositis and dermatomyositis. Lancet 2003;362:971-82.
3. Dalakas MC. Inflammatory muscle diseases. N Engl J Med 2015;373:393-4.
4. Lenoir T, Guedj N, Boulu P, Guigui P, Benoist M. Camptocormia: the bent spine syndrome, an update. Eur Spine J 2010;19:1229-37.
5. Laroche M, Cintas P. Bent spine syndrome (camptocormia): a retrospective study of 63 patients. Joint Bone Spine 2010;77:593-6.
6. Serratrice G, Pouget J, Pellissier JF. Bent spine syndrome. J Neurol Neurosurg Psychiatry 1996;60:51-4.
7. Finsterer J, Strobl W. Presentation, etiology, diagnosis, and management of camptocormia. Eur Neurol 2010;64:1-8.
8. Kim JM, Song EJ, Seo JS, Nam EJ, Kang YM. Polymyositis-like syndrome caused by hypothyroidism, presenting as camptocormia. Rheumatol Int 2009;29:339-42.
9. Dyck PJ, Boes CJ, Mulder D, Millikan C, Windebank AJ, Dyck PJ, et al. History of standard scoring, notation, and summation of neuromuscular signs. A current survey and recommendation. J Peripher Nerv Syst 2005;10:158-73.
10. Mahjneh I, Marconi G, Paetau A, Saarinen A, Salmi T, Somer H. Axial myopathy--an unrecognised entity. J Neurol 2002;249:730-4.
11. Djaldetti R, Mosberg-Galili R, Sroka H, Merims D, Melamed E. Camptocormia (bent spine) in patients with Parkinson's disease--characterization and possible pathogenesis of an unusual phenomenon. Mov Disord 1999;14:443-7.
12. Gdynia HJ, Sperfeld AD, Unrath A, Ludolph AC, Sabolek M, Storch A, et al. Histopathological analysis of skeletal muscle in patients with Parkinson's disease and 'dropped head'/'bent spine' syndrome. Parkinsonism Relat Disord 2009;15:633-9.
13. Kuo SH, Vullaganti M, Jimenez-Shahed J, Kwan JY. Camptocormia as a presentation of generalized inflammatory myopathy. Muscle Nerve 2009;40:1059-63.
14. Mattar MA, Gordo JM, Halpern AS, Shinjo SK. Camptocormia secondary to polymyositis. Rev Bras Reumatol 2013;53:368-70.
15. Delcey V, Hachulla E, Michon-Pasturel U, Queyrel V, Hatron PY, Boutry N, et al. [Camptocormia: a sign of axial myopathy. Report of 7 cases]. Rev Med Interne 2002; 23:144-54. In French.