A Case of Polymyositis Presenting as Bent Spine Syndrome

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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...
Iliac crest and paraspinal muscles edema with enhancement, especially worse on the right iliocostalis muscle (arrow) and longissimus (arrow head) at the level of L2 and L3.

**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.

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/mm³. Blood chemistry showed serum alanine aminotransferase (ALT), serum aspartate aminotransferase (AST), and lactate dehydrogenase (LDH) levels were normal. The patient exhibited no 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.

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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, endomyssial 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-
Table 1. Patient cases of polymyositis with bent spine syndrome

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


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 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 prior to perform laboratory tests and imaging studies.

**CONFLICT OF INTEREST**

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