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Case Report

Recurrent focal myofasciitis of Behçet syndrome mimics infectious myofasciitis: a case report

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Behçet syndrome (BS) is a chronic inflammatory disease with multiorgan manifestations. However, muscular involvement in BS has rarely been reported. Herein, we report the case of a 30-year-old male with BS who had recurring pain and swelling of the lower legs. The patient was administered antibiotics on several occasions as the condition was misinterpreted to be infectious myositis. Magnetic resonance imaging revealed myofascial involvement with focal necrotic lesions, and muscle biopsy revealed acute suppurative myositis with perivascular infiltration of polymorphonuclear leukocytes. His symptoms improved after treatment with corticosteroids. Azathioprine and colchicine therapy was beneficial for preventing further relapse after short-term corticosteroid treatment. Therefore, BS should be considered in the differential diagnosis of focal suppurative myofasciitis.

Keywords: Case reports, Behcet syndrome, Myositis, Myalgia, Vasculitis

INTRODUCTION

Behçet syndrome (BS) is a rare, relapsing, multisystem disease that is often classified under vasculitis, involving small, medium, and large vessels. The prevalence of BS varies widely according to geographical location. According to a recent meta-analysis, it is approximately 10.3 per 100,000 inhabitants [1]. The clinical manifestations of BS are highly variable. Recurrent orogenital ulcers are the most common manifestations, and other manifestations include the involvement of the skin, eyes, joints, nervous system, and gastrointestinal tract [2]. But, muscular involvement in BS is a rare manifestation; Taarit et al. [3] reported only two cases of myositis in 450 patients with BS. To date, there have been only few cases reported in the Korean literature [4,5].

Herein, we report a BS case who presented with recurrent pain and swelling in the lower legs that was repeatedly mistaken for cellulitis and infectious myositis. He was finally diagnosed with acute suppurative myositis associated with BS based on clinical features and biopsy pathology.

The study was approved by the Institutional Review Board of the Seoul National University Bundang Hospital (IRB no. SNUBH IRB NO 2111-721-702). Informed consent was obtained from the patient included in the study.

CASE REPORT

A 30-year-old male had pain and swelling in both lateral lower legs for three days. He had a recurrent aphthous ulcer in his early twenties and a single episode of a painful scrotal ulcer. Additionally, he had intermittent papulopustular lesions on the neck,

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upper back, and lower extremities. Five years prior, he suddenly developed pain and erythematous swelling in the right calf. He underwent a local incision with drainage and intravenous antibiotic therapy under the impression of cellulitis and myositis at a referral hospital. Magnetic resonance imaging (MRI) of the leg revealed a 1-cm lesion with T1-low/T2-high intensity in the right gastrocnemius and edematous changes in the muscle and subcutaneous fat layer (Figure 1A). However, tissue culture studies were negative. Subsequently, unilateral or bilateral myalgia in the lower leg recurred with a maximum frequency of once every 2 months. Myalgia was occasionally associated with fever and erythema of the overlying skin and sometimes resolved over 4 to 7 days with non-steroidal anti-inflammatory drugs. One month prior, he was admitted to another hospital because of fever and pain in the left calf. MRI also revealed an abscess-like lesion in the left gastrocnemius and edematous changes in the muscle, fascia, and overlying subcutaneous skin layers (Figure 1B). Under the suspicion of cellulitis with myositis and BS, he received a broad-spectrum antibiotic and prednisolone at a dosage of 10 mg/day. Sixteen days after discontinuation of medication, the patient experienced severe pain with swelling in both the distal lateral legs. Physical examination did not reveal much, except for tenderness in the lower extremities and a few areas with folliculitis. Laboratory examination revealed the following: white blood cell count, 9.44×10^9/L (neutrophils 72.6%); erythrocyte sedimentation rate, 47 mm/h; C-reactive protein, 3.31 mg/dL; and HLA-B51 (+). Liver and renal function test results were normal. His serum creatinine kinase and electrolyte levels were within the normal ranges. Ultrasonography showed diffuse swelling with heterogeneous echogenicity in the peroneus longus. Ultrasonography-guided muscle biopsy revealed acute suppurative myositis with interstitial and perivascular infiltration of inflammatory cells, predominantly polymorphonuclear leukocytes, and abscess formation (Figure 2). He was administered a single dose of intramuscular methylprednisolone (60 mg), and methylprednisolone (20 mg/day) was tapered over 7 days. To prevent further attacks, colchicine at a dose of 1.2 mg/day and azathioprine (maximum 100 mg/day) were added. Although they were helpful in reducing the attack frequency and intensity, azathioprine was discontinued 4 months later because of intolerable dyspepsia. Even with colchicine at 1.2 mg/day alone, he had an uneventful course during the next 20 months.

DISCUSSION

The present report describes the clinical features and course of unique muscular involvement in a patient with BS presenting with recurrent focal suppurative myofasciitis. The patient satis-

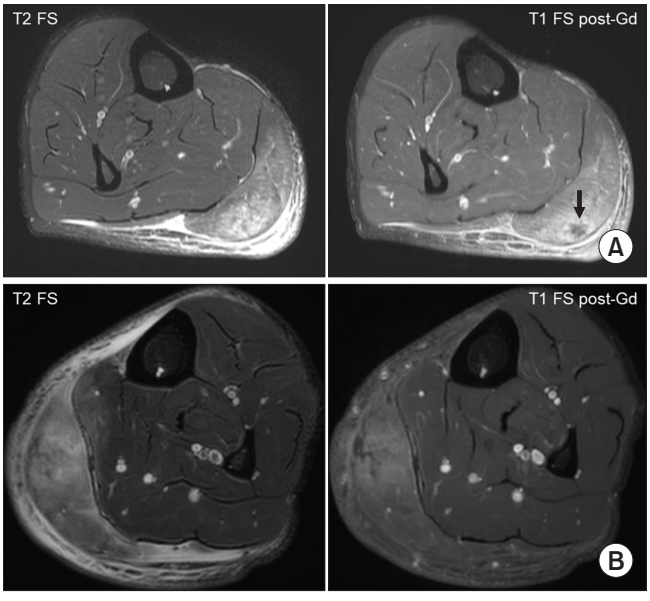

Figure 1. Axial magnetic resonance imaging scans of the legs revealed gastrocnemius myofasciitis with a focal centrally necrotic lesion (arrow) and edematous change of the dermis and subcutaneous tissues 5 years ago (A) and 1 month ago (B). FS: fat suppressed, post-Gd: post-gadolinium enhancement.

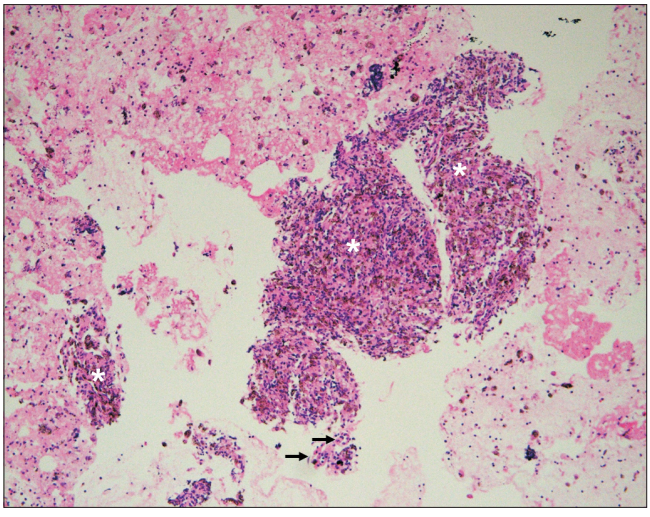

Figure 2. Muscle pathology of the peroneus longus. Acute suppurative myositis showing interstitial and perivascular infiltration (arrows) of inflammatory cells and abscess formation (asterisks). Main infiltrating cells were polymorphonuclear leukocytes (H&E, x200).

fied the 1990 criteria of the International Study Group for Behçet's disease [6] and the other two criteria. Similar to most BS manifestations recurring with varying frequencies and intensities, the current case repeatedly developed uni- or oligo-locular pain with swelling in the lower extremities. When he had severe attacks with systemic symptoms, such as fever, muscle imaging showed acute focal myofasciitis with necrotic lesions. BS is pathologically characterized by neutrophilic vascular and/or perivascular inflammatory reactions, and a biopsy reveals nonsterile neutrophil-dominant perivascular inflammation in the muscle tissue. Because muscular involvement is rarely associated with BS and is unfamiliar to physicians, these features could lead to confusion in physicians, especially if BS is not considered for differential diagnosis [7].

To the best of our knowledge, a single male patient was presented in a poster in Korean literature [4]. This patient was suspected to have BS based on recurrent oral ulcer, clinical pathergy history, and deep vein thrombosis, according to the 2014 revised International Criteria for Behçet's Disease (ICBD) [8]. In the first episode of pain in the right upper thigh, edema in the muscular and subcutaneous layers was found on MRI scans, and nonspecific inflammation with type 2 muscle atrophy was observed in muscle biopsy specimens. The above case improved over two weeks with colchicine treatment alone. While we prepared the manuscript, Kim et al. [9] recently reported 10 Korean patients presenting with focal vasculitic myositis who were diagnosed with BS at a rehabilitation medicine clinic. Unfortunately, the authors did not describe the clinical features and course in detail. Additionally, approximately half of the patients were classified as having possible (but unlikely) or probable BS based on the 2014 revised ICBD [8] or did not satisfy the complete or incomplete type of the 1988 revised Japanese criteria [10]. Moreover, muscle biopsy was performed in only 3 cases of which one showed myositis with lymphocytic vasculitis and perivasculitis. Furthermore, the follow-up period described in article was limited to 3 weeks in 9 cases, although oral prednisolone (10~60 mg) or colchicine for 1 to 3 weeks resolved the acute muscular symptoms in all patients.

BS myopathy has been proposed to be classified as generalized or localized condition. The generalized type was defined as one affecting ≥2 parts of the body and showing graver prognosis [11]. According to a literature review of some journals, localized or focal myositis is a predominant manifestation of BS myopathy. The most commonly affected parts were the lower extremities, followed by upper extremities and orbital muscles [9]. MRI most commonly revealed increased heterogenous signal intensity in the intermuscular or subcutaneous fascia on T2-weighted images. Concurrent thrombophlebitis and myositis were reported in one case [9]. These features are in line with the Korean cases in previous and our studies.

Yilmaz et al. [12] summarized MRI findings in BS myositis into five categories, one of which was focal centrally necrotic lesions with a rim peripheral hyperintense appearance or rim enhancement. Such MRI findings commonly suggest the likelihood of necrotizing infectious conditions or severely ischemic portions, and pyomyositis or diabetic myopathy can be at the top of the differential diagnoses. Therefore, radiological differentiation between BS and infectious myositis is difficult.

Although BS has been viewed as a kind of systemic vasculitis and vascular inflammatory injury is the main pathologic finding of BS, all BS manifestations do not result from true vasculitis [13]. Additionally, neutrophils are the crucial effector cells of an acute stage of BS, but mononuclear cell infiltrates are predominant in the late stage. Therefore, the pathology of BS myositis has been heterogeneously reported and may be dependent on the time and site of muscle biopsy. Some studies have described pathologically definite vasculitis, including leukocytoclastic and necrotizing vasculitis with multifocal interstitial infiltration of lymphocytes, eosinophils, and plasma cells [7,9]. However, others have revealed only muscular degeneration/atrophy. When a muscle biopsy shows neutrophilic inflammation in the muscle tissue, pyomyositis is considered the first presumptive diagnosis. However, physicians have to consider BS in the differential diagnosis of patients in a clinical context suggestive of the disease, as in our case.

The prognosis of BS-associated myositis was variable, with most cases responding well to steroids, some were treated with additional immunosuppressive drugs, and one patient was treated with infliximab [14]. The initial dose of steroids ranged from 10mg prednisolone to 1 g methylprednisolone, and the time taken for recovery varied from one week to fourteen months. One patient died because of uncontrolled myositis [15]. Considering the previous Korean and our cases together, short-term, low-to-moderate doses of corticosteroids can be beneficial for most cases of BS focal myositis. Additionally, colchicine maintenance therapy may have a favorable effect in reducing the risk of frequent relapses.

Owing to the low prevalence of focal myositis in BS and the

absence of "BS myositis" in the clinical diagnostic criteria, physicians tend to under-diagnose myositis if patients present with muscular symptoms before the diagnosis of BS. In patients with recurrent focal myositis, especially in the lower extremities, a thorough consultation of history and review are necessary to diagnose BS.

SUMMARY

Here, we report a case of myositis associated BS that mimicked infectious myofasciitis. His symptoms improved after treatment with corticosteroids. This is the first reported full-length case report of BS-associated myopathy in Korea.

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CONFLICT OF INTEREST

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

AUTHOR CONTRIBUTIONS

All authors contributed to case design, data collection, and interpretation. All authors have approved the final manuscript as submitted and agree to be accountable for all aspects of the work.

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