CASE REPORT

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Necrotizing Myositis as an Extraintestinal Manifestation of Crohn's Disease

Dong Hyun Kim, Kyu Hyun Paik, Hee Jun Shim, Su Yeon Rhie, Lae Ik Jung, Beom Jin Kim, Jae Gyu Kim
Department of Internal Medicine, Chung-Ang University Hospital, Chung-Ang University College of Medicine, Seoul, Korea

Inflammatory bowel disease is associated with extraintestinal manifestations involving almost every organ system in the body. Crohn's disease (CD) appears to be more commonly associated with an inflammatory myopathy than ulcerative colitis. However, myopathy of the thigh in patients with CD is rare. We report an unusual site of necrotizing myositis in a patient with CD. A 23-year-old woman presented with swelling and pain at the left popliteal area that had lasted for 1 week. Twenty-two months before admission, she had presented with pyoderma gangrenosum on the left upper chest and was diagnosed with CD. A magnetic resonance imaging scan of her leg revealed diffuse swelling in the left semimembranous muscle and biceps femoris muscle that was compatible with myositis, and a cystic lesion in the distal portion of the semimembranous muscle. The findings from semimembranous muscle biopsy were also consistent with necrotizing myositis. In conclusion, myositis, although rare, can be an extraintestinal manifestation of CD. **(Intest Res 2013;11:303-305)**

Key Words: Inflammatory bowel diseases; Crohn disease; Myositis

INTRODUCTION

Inflammatory bowel disease (IBD) is associated with extraintestinal manifestations (EIMs) involving almost every organ system in the body.[1] Common sites for these EIMs include the joints, skin, and eyes.[2] Crohn's disease (CD) appears to be more commonly associated with an inflammatory myopathy than does ulcerative colitis (UC).[3] Inflammatory myopathy in skeletal muscles may develop in a number of pathological conditions such as polymyositis, dermatomyositis, and inclusion-body myositis. However, other inflammatory changes can be present in CD or UC,[4] although myopathy, especially of the thigh, in patients with CD is rare.[5-7] EIMs contribute significantly to morbidity and mortality and respond well to immunosuppressive therapy; hence, careful attention to muscular symptoms and a high index of suspicion are important. We report an unusual site of necrotizing myositis in a patient with CD.

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Correspondence to Jae Gyu Kim, Department of Internal Medicine, Chung-Ang University Hospital, Chung-Ang University College of Medicine, 102 Heukseok-ro, Dongjak-gu, Seoul 156-755, Korea. Tel: +82-2-6299-1355, Fax: +82-2-6299-1137, E-mail: jgkimd@cau.ac.kr

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CASE REPORT

A 23-year-old woman presented with swelling and pain of the left popliteal area that had begun 1 week earlier. Twenty-two months before admission, she had presented with pyoderma gangrenosum on the left upper chest and was diagnosed with CD. She was treated with steroid and mesalazine for 3 months, which brought about improvement in her condition. Diarrhea continued intermittently, and she decided to discontinue the maintenance therapy 3 months before admission. The patient had never smoked and did not have a remarkable familial history. Blood pressure was 120/70 mmHg, body temperature was 36.2°C, and heart rate was 84 beats/min. The left popliteal area was reddish, swollen, and tender (Fig. 1). Examination of the other systems was unremarkable. Laboratory findings were as follows at admission: white blood cell count, 9,000/μL; hemoglobin, 12.4 g/dL; platelet count, 254,000/μL; AST, 13 IU/L; ALT level, 8 IU/L; serum albumin, 4.0 g/dL; BUN, 12 mg/dL; and serum creatinine, 0.7 mg/dL. ESR and CRP level were slightly elevated (26 mm/h and 32.5 mg/L, respectively), but CK (35 IU/L), LDH (126 IU/L), and antineutrophil cytoplasmic antibodies of the

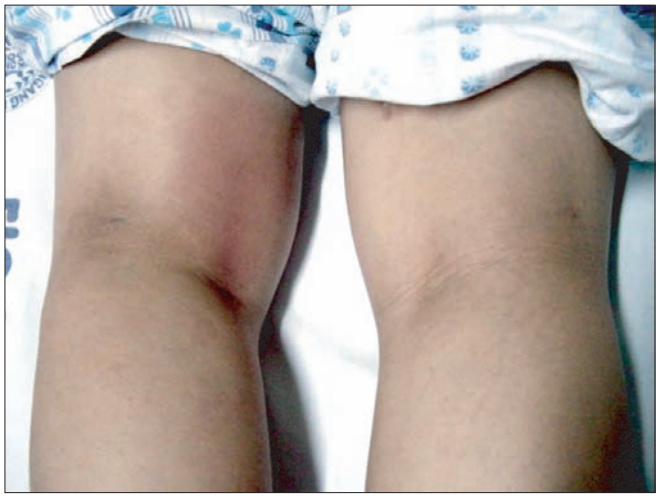

Fig. 1. Photograph of the patient's left popliteal area. The left popliteal area was reddish, swollen, and tender.

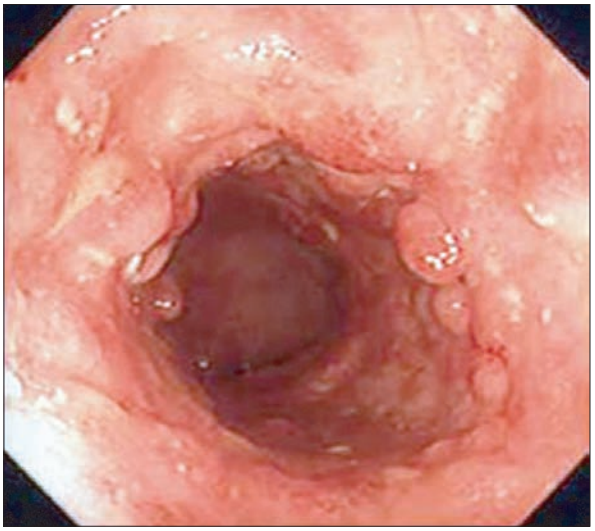

Fig. 2. Colonoscopy findings. Colonoscopy shows erythema, ulcerations, and pseudopolyps.

cytoplasmic type (c-ANCAs) levels were unremarkable. Colonoscopy showed skipped edematous mucosa with multiple aphthous ulcerations with pseudopolyps and focal stenosis in the cecum and sigmoid colon that were highly suggestive of CD (Fig. 2). The biopsies taken from the sigmoid colon showed chronic lymphocyte infiltration with crypt abscess rather than noncaseous granuloma. MRI scan (Magnetom Avanto 1.5; Siemens, Berlin, Germany) of the leg simultaneously revealed diffuse swelling in the left semimembranous muscle and biceps femoris muscle that was compatible with myositis, and a cystic lesion in the distal portion of the semimembranous muscle (Fig. 3). Ultrasonography-guided aspiration biopsy was performed to confirm an abscess in the cystic lesion. The aspirated material did not appear to be pus.

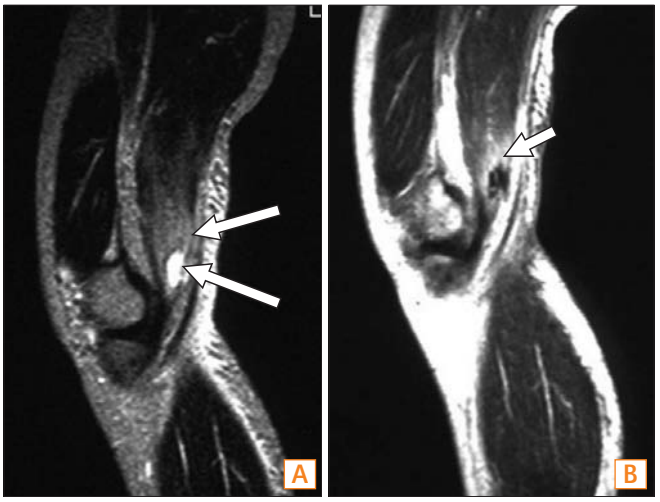

Fig. 3. MRI findings in the patient's left lower leg. (A) Sagittal T2 weighted image with fat saturation shows a well-defined high signal intensity lesion (arrows) in the sartorius muscle with increased signal intensity of surrounding muscle. (B) Sagittal contrast enhanced T1 weighted image shows enhancement in surrounding muscle (arrow) without enhancement in the central lesion or with peripheral enhancement in the central lesion.

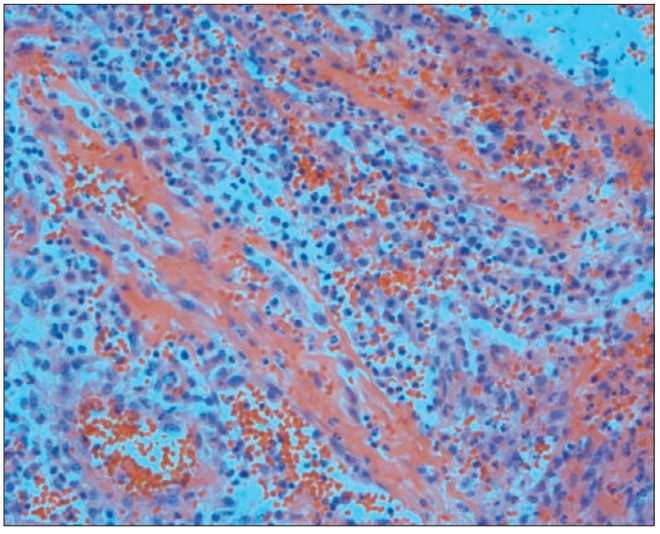

Fig. 4. Microscopy findings. Microscopic image shows perimysial and endomysial inflammation, and muscle fiber necrosis (H&E stain, ×400).

There was no growth on bacterial and *Mycobacterium tuberculosis* culture of the aspirated material.

The results of electromyography and nerve conduction velocity were unremarkable. On the fifth day of hospital stay, fever developed and swelling of the left popliteal area was aggravated. Surgical incision and drainage were performed. Acid fast bacilli stain was negative, and there was no growth on bacterial and *M. tuberculosis* culture of the aspirated ma-

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terial. The findings from semimembranous muscle biopsy were also consistent with necrotizing myositis (Fig. 4). The patient received mesalazine 3 g treatment daily and antibiotics intravenously for 4 weeks. Diarrhea and myositis improved without complications.

DISCUSSION

The association between IBD and muscle involvement was first reported in 1970 by Spiro, who illustrated granuloma formation in striated muscle in a patient with CD, but did not give clinical details.[8] To date, several cases of inflammatory myositis associated with CD have been reported. Muscular manifestations of CD usually affect the gastrocnemius muscles and present as muscle weakness, local swelling, or myalgia.[2,5,6,9-14] In contrast, our case showed unusual asymmetric necrotizing myositis in the left semimembranous muscle and biceps femoris muscle, without evidence of vasculitis or granuloma. In the previous seven cases, myositis preceded the bowel manifestation of CD,[2,5,10-14] in contrast to our case. Myositis usually occurred during an acute attack of CD, but the severity of CD was variable.[8,15] Our patient also showed myositis and mild active Crohn's colitis. The reason for vasculitis or myositis to have a predilection for the calves in patients with CD compared with the proximal muscles of the extremities and girdle in patients with polymyositis and dermatomyositis remains unclear.[13]

Infection with measles virus, paramyxovirus, and other infectious agents such as *M. paratuberculosis*, *Listeria*, *Escherichia coli*, and herpes virus can probably initiate antigen-driven production of autoantibodies (anti-Jo-1), resulting in the development of myositis.[4] IBD therapy using corticosteroids, 5-aminosalicylic acid preparations, or azathioprine could also result in myopathy development. However, this does not explain myositis that develops before IBD.[4] Autoantibodies are usually negative, and C3 and C4 complement levels are normal or minimally elevated in inflammatory myositis related to CD.[5,13] Most previous cases responded well to immunosuppressive treatment, usually in the form of monotherapy with prednisolone 0.5-1.0 mg/kg.[13,14] This is consistent with an immune pathogenesis that could involve perimysial and endomysial homing of migrant activated lymphocytes that have been antigenically stimulated in the intestinal wall.[14] However, because we were not able to exclude the possibility of infection and the Crohn's disease activity was below the reference value of 150, we did not use corticosteroid or immunosuppressive treatment. This patient responded well to mesalazine treatment without either corticosteroid or other immunosuppressive treatment. A few cases showed improvement in muscle symptoms with 5-aminosalicylic acid preparations only, similarly to our patient.[4,5,8] The lesion shown as part of an abscess in the scan might be a secondary change from necrotizing myositis because there was no growth on the culture of the aspirated material.

In conclusion, myositis, although rare, can be an EIM of CD. If proximal muscle weakness, myalgia, and swelling are present in a patient with CD, myositis should be considered as one of the differential diagnoses, along with arthritis, which is the most commonly encountered EIM in patients with CD.

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