

A Case of Neutrophilic Myositis

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Neutrophilic myositis is a very rare disease histologically characterized by neutrophil infiltration of muscle tissues. We report a case of a 47-year-old man who presented with acute onset of severe swelling and pain on his left shoulder with high fever. He was initially suspected of having cellulitis, but intravenous antibiotics did not improve his symptoms. Similar swelling and pain then developed on both calves. Investigations with magnetic resonance imaging of the lower legs and muscle biopsy led to a diagnosis of neutrophilic myositis. High dose glucocorticoid dramatically improved his symptoms within days. Clinicians need to be aware of this rare disease as a cause of acute febrile myositis mimicking infection. (*J Rheum Dis* 2017;24:161-164)

Key Words. Myositis, Sweet syndrome, Neutrophil infiltration

INTRODUCTION

Neutrophilic myositis (NM) is a very rare disorder histologically characterized by dense neutrophil infiltration in skeletal muscles [1-7]. The etiology and pathogenesis of this rare disease is unclear, but it is suspected as in the spectrum of neutrophilic disease such as acute febrile neutrophilic dermatosis (AFND) [1,2]. Similar to AFND, NM was associated with several underlying hematologic and inflammatory diseases such as acute myeloid leukemia (AML) [1-4], ulcerative colitis (UC) [5] and celiac disease [6]. However, isolated NM developing in a previously healthy individual without any concurrent disease development has not been reported yet. We report a case of NM in a previously healthy man with a literature review.

CASE REPORT

A 47 year-old man was admitted due to painful swelling on his left shoulder that developed 3 days before admission. He had been in good health until that time when he suddenly developed swelling and pain on his left shoulder with a febrile sense. At the time of admission, he was fe-

brile with the body temperature of 38.5°C and diffuse edema, tenderness and heating sense were noted around his left shoulder. No specific skin lesions were found on the affected site. It was difficult to ascertain the presence of joint tenderness or muscle weakness due to severe pain. Initial laboratory findings were as follows: white blood cell count 7,400/ μ L, hemoglobin 13.8 g/dL, platelet 159×10^3 / μ L, C-reactive protein (CRP) 25.4 mg/dL (normal range, <0.5 mg/dL), creatinine kinase (CK) 467 IU/L (normal range, 39 ~ 308 IU/L), myoglobin 65.1 ng/mL (normal range, 28 ~ 72 ng/mL) and procalcitonin 4.31 ng/mL. He was initially treated with intravenous (IV) cefazolin 3 g per day in suspicion of infection such as cellulitis involving the upper arm. Fever and pain around his shoulder seemed to improve slightly during the first few days of IV antibiotic use. His body temperature fell to 37°C and the pain on his left shoulder began to improve on hospital day (HD) 3, which is the 3rd day of antibiotic use. However, on HD 6, the body temperature rose up to 39°C and the pain and swelling on his left shoulder became worse again. In addition, severe pain and diffuse edema developed on his both calves despite the use of antibiotic. On the follow-up laboratory test performed on HD 6, CK level rose from 467 to 3,442 IU/L, myoglobin

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rose from 65.1 to 1,006 ng/mL and the level of CRP was still high (28.35 mg/dL). No specific organism was isolated from blood cultures taken on HD 1. On the gadolinium enhanced magnetic resonance imaging (MRI) of both lower leg performed to evaluate the swollen calves, diffuse, T2-high signal intensity lesions was noted on the bilateral gastrocnemius muscles with a heterogenous enhancement pattern (Figure 1). Ultrasonography guided gun biopsy of the left gastrocnemius muscle revealed dense and confluent neutrophil infiltration into endomysium with resultant necrotic muscle fibers (Figure 2). He was finally diagnosed as NM and started to take high dose glucocorticoid (prednisolone 60 mg per day). The intense inflammation on his left upper arm and both lower leg dramatically improved after the administration of high dose glucocorticoid. Fever subsided on the day after glucocorticoid use. Pain and swelling on his arm and legs began to improve in 2 days and completely disappeared in a

week. The levels of CK, myoglobin and CRP were normalized in 10 days. He was discharged and started to taper glucocorticoid with methotrexate 10 mg weekly in out-patient clinic.

DISCUSSION

In this patient, we confirmed the diagnosis of NM based on the followings: (1) neutrophil infiltration was demonstrated in muscle tissues; (2) culture studies for bacteria and fungi were negative; (3) Other myopathies such as polymyositis (PM), dermatomyositis (DM), inclusion body myositis were excluded based on clinical and histologic findings; (4) the patient responded dramatically to glucocorticoids.

NM is quite a rare disease characterized histologically by infiltration of neutrophils in muscle tissues. To the best of our knowledge, only 7 cases [1-7], including a report of a

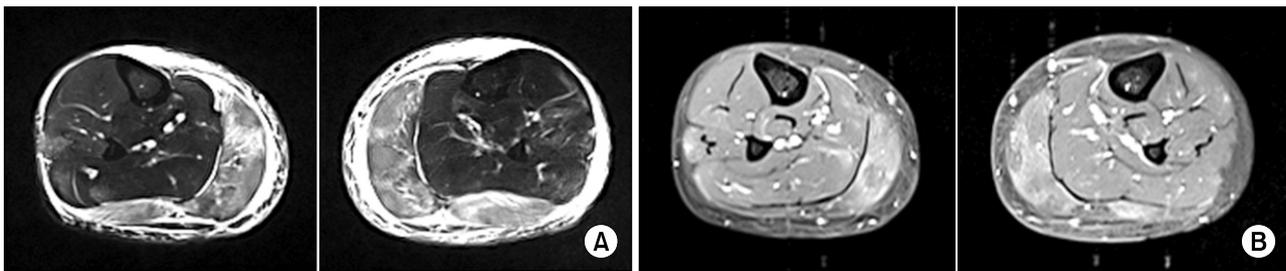


Figure 1. (A) T2-weighted magnetic resonance imaging (MRI) of both calves reveals diffuse T2 high signal intensities on both gastrocnemius muscles. (B) T1-weighted MRI with gadolinium enhancement shows heterogeneously enhanced lesions on both gastrocnemius muscles.

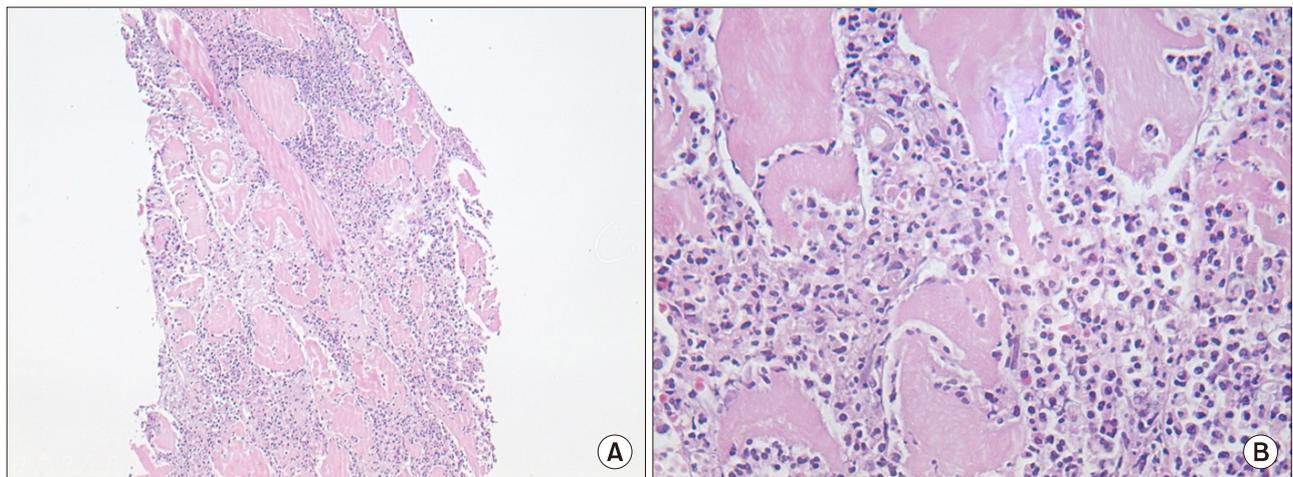


Figure 2. (A) Destructive inflammation leading to saw-toothed architecture of the muscle bundles (muscle fiber necrosis). (H&E, original magnification $\times 100$). (B) Inflammatory cells composed predominantly of neutrophils admixed with some lymphocytes and histiocytes, with occasional eosinophils. (H&E, original magnification $\times 400$).

Korean patient [2], have been reported so far. All the reported cases were associated with several inflammatory and hematologic disorders: 4 with AML [1-4], 1 with myelodysplastic syndrome (MDS) [7], 1 with celiac disease [6] and 1 with UC [5]. In the cases associated with AML, NM developed concurrently with AML [1,2] or after treatment with all-*trans* retinoic acid for AML [3,4]. In the other reports, NM developed in conditions where a certain disease (celiac disease, UC and MDS) was established as a background [5-7]. However, in the present case, NM developed in a previously healthy man and no other specific disorders were identified during the course of treatment, which is a unique feature of this case.

Based on the previous reports [1-7], the following clinical features were commonly found in NM. First, the presentation of disease in the present report was abrupt onset of diffuse edema and pain on the shoulder and both calves in succession, which was accompanied by high fever. The pattern of acute or subacute development of pain and swelling around distal lower leg and upper arm is commonly seen in the previous reports, which is different from other inflammatory myopathies. In PM or DM, the onset of disease is usually insidious with gradual worsening of proximal muscle weakness. Myalgia and muscle tenderness may also be seen, but they are mild in degree [8]. Second, NM is associated with several inflammatory and hematologic diseases. Lastly, NM showed a dramatic response to high dose glucocorticoid. In the present case, the profound inflammation in the muscles resolved within a week with normalization of inflammatory markers within 10 days. Similar dramatic responses were commonly seen in the previous reports.

The clinical features described above are also commonly seen in AFND, also known as Sweet syndrome, which is characterized by skin infiltration with neutrophils. Sweet syndrome is frequently associated with inflammatory bowel disease [9], AML [10], and the use of granulocyte colony stimulating factor [11], the association of which has also been reported in NM. Sweet syndrome also shows dramatic responses to glucocorticoids [12]. Moreover, NM has been reported in association with Sweet syndrome [1,3]. These findings suggest that NM may be regarded as in the spectrum of neutrophilic disorders including Sweet syndrome.

Severe swelling on the extremities accompanied by high fever may initially be suspected as infection such as cellulitis and be treated with antibiotics. This may be a reasonable practice to rule out infection, which, if untreated,

may result in disastrous results. However, if the patient does not respond to initial antibiotic, NM, though extremely rare, should also be suspected and checking the levels of muscle enzymes and imaging study such as MRI should be considered before considering to change the initial antibiotic regimen.

SUMMARY

We presented a rare, clinically distinct case with NM, characterized clinically by acute onset of diffuse edema and pain on the affected muscles with fever and histologically by neutrophil infiltration in muscle tissues. Our patient responded dramatically to glucocorticoids like the other previously reported cases. Clinicians need to be aware of this rare disease as a cause of acute, febrile myositis mimicking bacterial infection.

CONFLICT OF INTEREST

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

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