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NEUTROPHILIC MYOSITIS ASSOCIATED WITH *PYODERMA GANGRENOSUM* IN A BREAK-DANCER

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Abstract
Neutrophilic myositis is an extremely rare condition, cases of which have been reported in association with neutrophilic dermatosis, inflammatory bowel disease and malignant hematological disease. The disorder is histologically characterized by a sterile infiltration of neutrophils throughout muscle, with necrosis of muscle fibres. We here report the case of a young male who also had associated pyoderma gangrenosum, and who presented with necrotizing fasciitis-like manifestations. In this case, although there were no other underlying disorders, compulsive exertional stress due to break-dancing was thought to be a precipitant. Debridement of the necrotic tissues combined with oral corticosteroid treatment was effective.

Key words: neutrophilic myositis; pyoderma gangrenosum; hard exercise

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Introduction

Neutrophilic myositis, which is extremely rare, is characterized by a histologically sterile infiltration of neutrophils throughout the muscle accompanied by disruption of the muscle architecture and numerous areas of muscle fiber necrosis [1]. This condition has been reported as an extracutaneous manifestation of neutrophilic dermatosis, and also been associated with inflammatory bowel disease and with hematological malignancies [1-6]. Here, we report a young male with neutrophilic myositis of the lateral aspect of the knee, accompanied by pyoderma gangrenosum. A clinical characteristic of our case was that exercise-induced stress on the knee brought on by compulsive break-dancing was thought to be a trigger of the onset.

Case Report

A 27-year-old man came to our hospital with a complaint of a one-month history of intractable pain and swelling of his right knee. He had experienced prior, similar ‚phlegmonous' swelling of his right knee, with three episodes over the past three years. Previously, oral or intravenous antibiotics relieved his symptoms, but this time, his eruption failed to respond to treatment with several courses of oral antibiotics that included cephems, new-generation quinolones, and tetracycline. When asked about his habits, he disclosed that he had started to break-dance at the age of nineteen, and that in the past the symptoms that affected his right knee consistently appeared after straining his knees during vigorous dancing. At his visit, his right knee was remarkably swollen with marked black cutaneous necrosis and skin ulcers (Fig. 1a). Body temperature was elevated to 38.8oC. Laboratory examination showed the following: white blood cell count, 16,800/μl (neutrophils: 77.6%, lymphocytes: 14.5%, eosinophils: 1.6%); C-reactive protein, 10.49 mg/dl; creatine phosphokinase, 560 IU/L; procalcitonin negative. Other tests, such as liver and renal function, quantitative serum immunoglobulins, blood cultures, urinalysis, and viral serology (cytomegalovirus, Epstein-Barr virus, human immunodeficiency virus), rheumatoid factor, antinuclear antibodies, anticardiolipin and antiphospholipid antibodies, and antineutrophil cytoplasmic antibodies were all normal or negative. Magnetic resonance imaging showed that inflammation involved not only the subcutaneous tissues but also the right musculus quadriceps femoris. We considered this condition as necrotizing fasciitis. We therefore performed emergency debridement of the necrotic tissue including the skin and muscles. Histological study of the debrided tissues revealed marked inflammatory infiltrates, composed predominantly of neutrophils that extended throughout all layers of the skin and involved muscle fibers, accompanied by disruption of the muscle architecture and numerous areas of muscle fiber necrosis (Fig. 2a, b).

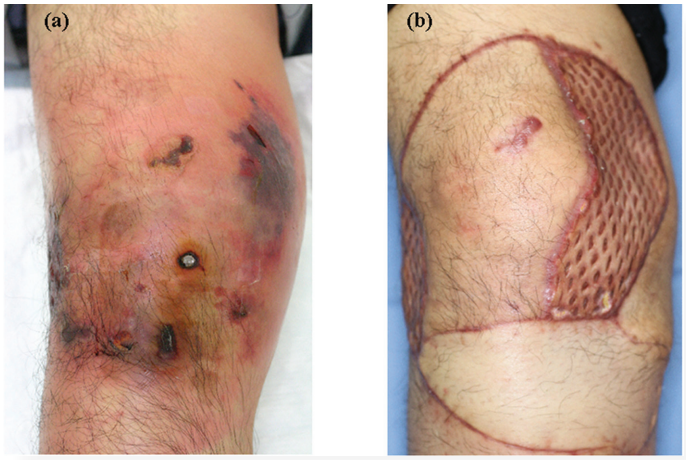

Figure 1. Clinical appearance (a) before, and (b) six months after treatment with debridement and oral steroid. (a) The right knee demonstrates remarkable swelling with marked, black cutaneous necrosis and skin ulcers. (b) After debridement and administration of oral steroids, the ulcerative lesion on the right knee healed with good granulation tissue, and could then be managed successfully with a skin graft and pedicle skin flap, without reactivation.

However, tissue cultures for bacteria, fungi, acid-fast microorganisms and parasitic amoebae were all negative. On the basis of these findings we made a final diagnosis of neutrophilic myositis associated with pyoderma gangrenosum. Investigation for an underlying illness, such as inflammatory bowel disease or a myeloproliferative disorder, (respectively by fibreoptic colonoscopy and bone marrow aspiration) revealed no abnormal findings. After debridement, we started treatment with oral methylprednisolone at a dosage of 40mg per day. His postoperative wound subsequently healed with good granulation tissue and, after one month, we performed a two-stage operation to address the residual ulcer successfully with the combination of a skin graft and pedicle-based skin flap. The dose of oral steroid could be tapered to 10mg per day for about six months, and there has been no exacerbation (Fig. 1b).

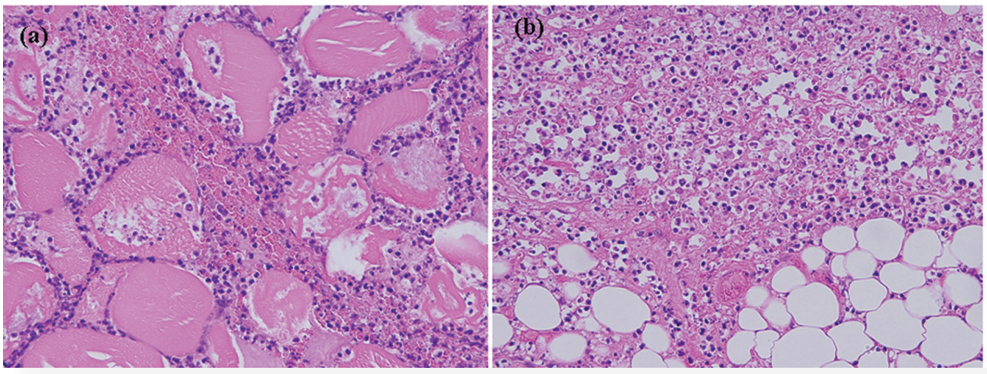

Figure 2. Histological study of the debrided tissues demonstrates severe inflammatory infiltrates, composed predominantly of neutrophils that extend throughout both the right musculus quadriceps femoris (a) and the skin of the same lesion (b). The muscle lesion was accompanied by disruption of the muscle architecture and numerous areas of muscle fiber necrosis. (Haematoxylin and Eosin stain, original magnification x400).

Discussion

Neutrophilic myositis is an extremely rare condition. To the best of our knowledge, only six cases have been reported, apart from ours [1-6]. The cases of neutrophilic myositis are summarized in Table I. The ratio of males to females is 5: 2, and the favorite involved sites were extremities. Three cases were associated with a neutrophilic dermatosis such as pyoderma gangrenosum [1] or Sweet's syndrome [2,3], and all of these cases coexisted with underlying leukaemia. The other cases had no cutaneous involvement, but were accompanied by underlying diseases, such as inflammatory bowel disease [4,5] or a myeloproliferative disorder [6]. Although coexistent pyoderma gangrenosum was seen in our case, there was no other underlying illness, in contrast to the reported cases [1-3]. We should however be vigilant for the later occurrence of such a disorder, particularly inflammatory bowel disease or a myeloproliferative disorder.

The diagnosis of neutrophilic myositis must be based on both histological demonstrations of intense neutrophilic infiltration throughout the affected muscles, with necrosis of muscle fibers, and exclusion of other muscle diseases, for example, polymyositis, dermatomyositis, inclusion body myositis, and pyomyositis [1].

Authors(s) [Ref.]	Sex	Age (years)	Coexisting disease	Involved site of myositis	Treatment	Prognosis
Marie I et al. [1]	Male	65	pyoderma gangrenosum acute myelogeneous leukemia	bilateral lower, upper limbs	Methylpredniso1one chemotherapy	death
Melinkeri SR et al. [2]	Female	6	acute myeloid leukemia	left thigh	Methylpredniso1one chemotherapy	death
Attias D et al. [3]	Female	60	Sweet's syndrome myeloblastic leukemia	bilateral thighs, left shoulder	Methylpredniso1one chemotherapy	death
Alawneh K et al. [4]	Male	42	celiac disease	right thigh, left lowe-re limb	Methylpredniso1one	remission
Qureshi JA et al. [5]	Male	36	ulcerative colitis	right shoulder, lower extremities	Methylpredniso1one	remission
Kim MK at al. [6]	Male	35	acute myeloid leukemia	upper arm	Methylpredniso1one	remission
our case	Male	27	pyoderma gangrenosum	right thigh	Methylpredniso1one debridement	remission

Table I. Summary of past cases of neutrophilic myositis.

Our case could finally be diagnosed as neutrophilic myositis related to pyoderma gangrenosum for the following reasons: the simultaneous onset of both neutrophilic myositis and pyoderma gangrenosum; negative stains and bacterial cultures of muscle and skin biopsy specimens; extensive, negative investigation for other causes of myositis such as infectious and connective tissue disorders; the efficacy of systemic steroids; and the ineffectiveness of broad-spectrum antibiotic therapy.

The etiology of neutrophilic myositis remains unclear. Heterogeneous factors, such as allergy to pathogenic organisms, traumatic injury and immunological abnormality due to co-existing disorders may be associated, as well as pyoderma gangrenosum [7]. The present case is the first in which compulsive exercise-induced stress on the knee due to break-dancing was a potentially causative trigger. This observation suggests that damage to or degeneration of muscle tissue induced by compulsive exercise may induce an abnormal, neutrophilic inflammatory reaction without any underlying immunological imbalance.

Finally, therapy for neutrophilic myositis has not been established. Based on the cases described, including our case, systemic treatment with moderate-to-high dose steroids seems effective [1-6]. Furthermore, a coexisting myeloproliferative disorder seems to indicate a poor prognosis [1-3, 6].

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