

Case Report

Eosinophilic perimyositis manifesting as localized urticaria

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Case Report

A previously well 55-year-old Korean woman presented for evaluation after 3 months of recurrent, transient, urticarial plaques localized solely to the left thigh associated with intense pruritis and burning discomfort. The onset and amelioration of the eruptions occurred within hours, with over two episodes per week. Generalized painful swelling of the left thigh was initially associated with these cutaneous eruptions. She had traveled to Southeast Asia in the month preceding the commencement of cutaneous lesions. At the initial clinical onset, empirical treatment with oral albendazole 400 mg daily was administered due to the clinical suspicion of parasitic infection, with minimal improvement.

Cutaneous examination revealed multiple well-demarcated erythematous papules coalescing into irregularly shaped plaques on the left thigh (Fig. 1).

Pertinent laboratory findings included mild leukocytosis ($11.9 \times 10^9/l$; normal: $4.0-11.0 \times 10^9/l$) with 41.3% eosinophils ($4.92 \times 10^9/l$; normal: $0.02-0.50 \times 10^9/l$), elevated C-reactive protein (5 mg/l; normal: <3 mg/l), and raised erythrocyte sedimentation rate (21 mm/h; normal: 0-10 mm/h). Lactate dehydrogenase was normal. Magnetic resonance imaging of the left thigh revealed T2 hyperintensity of the rectus femoris

muscle. Histologic examination of a skin biopsy of the left thigh revealed nonspecific mild spongiosis with perivascular and interstitial eosinophils in the dermis (Fig. 2). Muscle biopsy specimens of the left thigh demonstrated intramuscular eosinophilic-rich inflammatory infiltrate with no parasitic parts or myofiber necrosis (Fig. 3). Extensive laboratory work-up revealed negative or normal results for parasitic infection, toxin exposure, connective tissue disorders, HIV, and hepatitis B and C. No previous medication or supplementation such as



Figure 1 Multiple erythematous, urticarial plaques on the left thigh

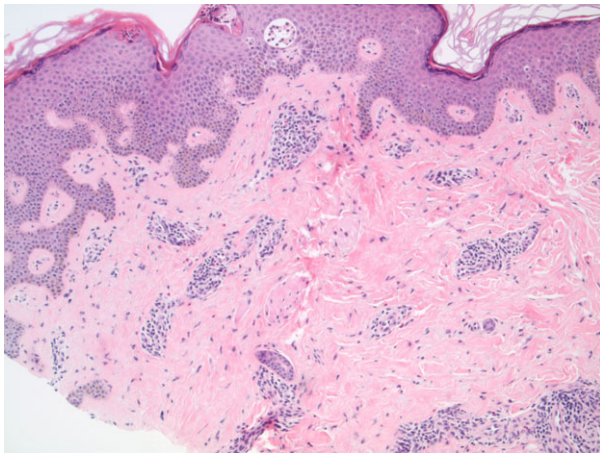


Figure 2 Histologic examination of skin biopsy demonstrates mild spongiosis with perivascular and interstitial eosinophils in the dermis (hematoxylin and eosin stain $\times 10$ magnification)

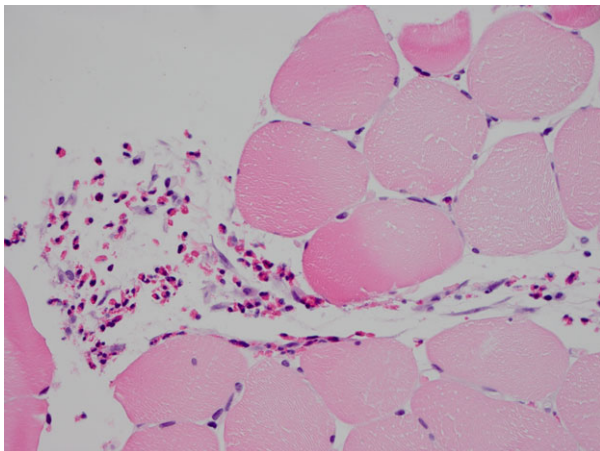


Figure 3 Histologic examination of muscle biopsy demonstrates eosinophilic-rich infiltrate surrounding normal muscle fibers (hematoxylin and eosin stain $\times 400$ magnification)

L-tryptophan was noted. Thus, a diagnosis was made of eosinophilic perimyositis presenting with myalgia and recurrent episodic urticarial plaques localized to the left thigh. Colchicine 0.5 mg daily and cetirizine 10 mg daily as required were commenced. However, colchicine was ceased after 1 week due to gastrointestinal side effects. Upon review, after 6 and 9 months, the episodes had reduced in frequency, and cetirizine was continued. Limited case studies have shown a favorable response of eosinophilic perimyositis to glucocorticoid treatment, which could be trialed should there be future relapses.¹

Discussion

The differential diagnosis of persistent eosinophilia includes hypereosinophilic syndrome and toxin exposure. Although hypereosinophilic syndrome can present with cutaneous manifestations, the syndrome is a multisystem disease that does not correlate with our case. Toxin exposure, such as the toxic oil syndrome in Spain in 1981 and the L-tryptophan associated eosinophilia–myalgia syndrome in Mexico in 1989, typically involves a history of toxin exposure and a systemic acute and chronic disease progression.^{1,2} Given the absence of systemic involvement and no history of toxin exposure, a diagnosis of eosinophilic perimyositis was reached.

Eosinophilic myositis is an idiopathic inflammatory skeletal muscle disorder characterized by eosinophilia and/or eosinophilic muscle infiltrate. The disorder is composed of three subtypes: focal eosinophilic myositis, eosinophilic polymyositis, and eosinophilic perimyositis.¹ The eosinophilic perimyositis subtype accounts for approximately 50% of cases of eosinophilic myositis.² Of the 15 cases of eosinophilic perimyositis published in the literature, the typical clinical presentation involves myalgia and an absence of systemic symptoms. Clinical consideration of eosinophilic perimyositis should be heightened by exclusion of parasitic infection, toxin exposure, and connective tissue disorders. On histology, eosinophilic perimyositis is distinguished by an eosinophilic rich perimysial inflammatory infiltrate without endomysial infiltration or myofiber necrosis.^{2–4}

Cutaneous manifestations have been described with eosinophilic perimyositis in five reported cases. Induration and erythema are the most common skin abnormalities observed.^{2–5} We describe an atypical presentation of eosinophilic perimyositis in which the histopathological findings were consistent with eosinophilic perimyositis, but the clinical presentation differed from those classically seen.

To our knowledge, this is the first report to describe episodic urticarial plaques localized to a region, accompanying eosinophilic perimyositis. The distribution of the cutaneous eruptions involving the proximal lower limb is also atypical.^{2–5} We present this case to expand the clinical spectrum of cutaneous manifestations of eosinophilic perimyositis to include urticarial plaques, increase awareness of atypical presentations of eosinophilic perimyositis, and facilitate earlier identification of the condition.

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