

Eosinophilic Fasciitis – Clinical Features and Therapeutic Management

Eosinophilic fasciitis is a rare disease from the group of scleroderma-like connective tissue diseases with unclear etiopathogenesis. It may be occasionally accompanied with other eosinophilic or autoimmune dysfunctions (1,2). Lack of international diagnostic criteria and treatment consensus may lead to diagnostic and therapeutic difficulties.

The 61-year-old man with no significant personal or family pathological history was admitted to the Dermatology Department presenting persistent induration for several months as well as erythema and pain of the shins that gradually extended to thighs and forearms, with limited mobility of peripheral

joints. (Figure 1) Additional tests showed leukocytosis with 16% eosinophilia, elevated CRP, and hypergammaglobulinemia. *Borrelia burgdorferi* antibodies (classes IgM and IgG) were negative twice. A biopsy that included deep fascia was taken for histopathological examination. Antinuclear antibody screening was negative, but the direct immunofluorescence showed complexes in the dermo-epidermal junction and around the vessels. The diagnostics conducted toward malignant process showed no disturbing abnormalities (i.e. tumor markers in serum, chest, and abdomen computed tomography imaging, panendoscopy). The treatment was carried out with cephalosporin and nonsteroidal anti-inflammatory drugs (NSAIDs). The condition did not improve much but was stable. Histopathological results were indicative of eosinophilic fasciitis with fibrous thickening of deep fascia and perivascular infiltrations of plasma cells and lymphocytes; oral prednisone was initiated



Figure 1. Erythema and induration on both shins, the thighs, and the forearms.



Figure 2. Orange peel skin with linear furrows over the superficial venous vessels – the “groove” sign.

and the condition begin to improve. After 12 weeks, we observed disease progression with fever and very hard and cyanic skin lesions, which presented as an orange peel with linear furrows over the superficial venous vessels (Figure 2). The lesions extended to the trunk and caused troubles in moving. A complex rehabilitative intervention was started to minimize the inflammatory fascial restrictions. The prednisolone dose was increased, and oral methotrexate was added. After two weeks, the patient suffered from abdominal pain and periodic bleeding diarrhea. Methotrexate was suspected of inducing gastrointestinal adverse effects, and antipyretic NSAIDs were completely withdrawn. Colonoscopy showed features of mucosal edema with erythema, and histopathological examination revealed eosinophilic colitis. The patient was referred to a gastroenterologist, and methotrexate was ceased and switched to azathioprine. In summary, the consensus therapy of the rheumatologist, dermatologist, and gastroenterologist consisted of prednisolone and azathioprine. As of this writing, the patient's condition is gradually improving.

The most characteristic symptoms of eosinophilic fasciitis is sudden onset with induration, sclerosis, and pain of the skin, with subcutaneous tissue and fascia usually appearing on the upper and lower limbs (3,4). The skin surface forms a characteristic orange peel appearance. The "groove" sign refers to the linear furrows over the superficial vessels of the extremities (1). Typical abnormalities are eosinophilia, elevated CRP, and hypergammaglobulinemia. The presence of eosinophilia is the most characteristic feature, occurring in 60-93% cases, but it is not necessary for diagnosis (1,5). Antinuclear antibodies are commonly absent with positive lesional direct immunofluorescence (6). If antinuclear antibodies are positive, it is recommended to broaden the diagnostic process to include other connective tissue diseases. Eosinophilia must be differentiated from hematological disorders and paraneoplastic syndrome. (4,6).

Eosinophilic colitis is an eosinophilic gastrointestinal disease (EGID). It is the least frequent manifestation of EGID. It may be associated with connective tissue diseases, mostly systemic sclerosis – to our knowledge, there is no information in the literature about coexisting eosinophilic fasciitis. (7,8). The case described herein demonstrated that such a connection may occur.

In treatment, it is important to prevent the patient from contractures and to maintain joint mobility by appropriate physiotherapy (2,9). The fascia forms a functional integral and continuous structure. Inflammation of one part of it changes the elasticity of the

whole and produces fascial restrictions with movement limitation and pain. The fascia is profusely innervated, which favors constriction as a result of inflammation, and is also poorly vascularized which disrupts its regeneration (9,10). Myofascial techniques improve fascia elasticity by breaking up the tissue adhesions caused by inflammation (11).

Eosinophilic fasciitis is a rare clinical entity, but knowing the possible clinical symptoms and laboratory abnormalities should help in taking the appropriate diagnostic path. It is important to treat the patient with attention to all concomitant diseases in consultation with different specialists.

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