EOSINOPHILIC FASCIITIS

A Scleroderma-Like Condition

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Eosinophilic fasciitis is a hypereosinophilic disorder characterized by a thickening of the fascia as well as skin changes similar to those seen in scleroderma.

What is Eosinophilic Fasciitis?

Eosinophilic fasciitis, which was first described in 1975, is a rare scleroderma-like hypereosinophilic (increased levels of blood eosinophils) disease without the characteristic Raynaud’s phenomenon or systemic involvement seen in systemic sclerosis. Although its cause isn’t known, eosinophilic fasciitis is suspected of having an autoimmune origin. In addition, eosinophilic fasciitis is known to occur in persons with other autoimmune disorders, including autoimmune thyroiditis, Sjogren’s syndrome, alopecia areata, vitiligo, and autoimmune thrombocytopenia (platelet deficiency).

Symptoms and Signs

Symptoms of eosinophilic fasciitis include rapid weight gain, swelling, inflammation of the skin followed by induration (causing an orange-peel appearance) over the anterior surface of the extremities, and muscle pain. Although rapid weight gain is common, weight loss, fever, and fatigue can also occur. Unlike the skin involvement of scleroderma, the fingers and toes are not affected in eosinophilic fasciitis. However, the face and trunk of the body may be affected with changes to the skin resembling those of scleroderma including morphea lesions, redness, thickening, hyperpigmentation, blisters, and a claw-like deformity of the hands similar to that which often occurs in scleroderma.

Symptoms usually appear gradually and cause morning stiffness, edema (swelling), inflammatory arthritis, and a gradual restriction of arm and leg movement. Restriction occurs because the fascia (thin tissue sheath covering muscle) is affected to a greater degree than the skin. Although the fascia is primarily affected, the tendons, synovial membrane, and muscles may also be affected. Contractures (locked joints; chronic loss of joint motion due to structural changes and muscle shortening in non-bony tissue) often evolve affecting the elbows, wrists, ankles, knees, hands and shoulders, and accompany the fascial changes. Peripheral nerve involvement may occur causing symptoms of carpal tunnel syndrome. Rarely, spleen and liver enlargement have also been reported.

The acute disease state is characterized by increased blood eosinophil levels, an elevated erythrocyte sedimentation rate, and elevated IgG immunoglobulin levels (hypergammaglobulinemia). Less than 40 percent of patients show evidence of antinuclear antibodies (ANA). In severe cases, changes to blood cells (hematological changes) may occur including aplastic anemia.
Who is Affected?

Women are more often affected than men, and whites are affected more often than blacks. Eosinophilic fasciitis has been reported in patients from ages 2-88 although it’s most likely to occur in people between 30 and 60 years.

Eosinophilic fasciitis often occurs after intense exercise in usually sedentary persons and it sometimes occurs after infection with Borrelia burgdorferi, the causative agent of Lyme disease.

Toxins are also suspected of triggering eosinophilic fasciitis. Other hypereosinophilic syndromes have been reported to occur after ingestion of L-tryptophan and other medications, particularly anti-hypertensive medications.

Diagnosis

Eosinophilic fasciitis is suspected in patients with characteristic scleroderma-like skin changes and the absence of Raynaud’s phenomenon. The diagnosis is confirmed by biopsy of affected skin and fascia deep enough to include muscle fibers. The subdermal fascia is markedly thickened with increased collagen production and cellular infiltration.

MRI studies are used to show fascial thickening, and electromyogram is used to show myositis (muscle inflammation) which may occur even in the absence of elevated muscle enzymes.

Treatment

Treatment consists of prednisone in doses ranging from 20-60 mg daily. Improvement is seen with an immediate improvement in eosinophilia and edema. Most patients are able to stop the use of prednisone when symptoms resolve without relapse occurring.

Resources:


Eosinophilic Fasciitis, Merck Manual of Diagnosis and Therapy, Musculoskeletal and Connective Tissue Disorders.


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