

What Is Your Diagnosis?



A 43-year-old woman presented with pain and paresthesia of the bilateral legs of 3 months' duration with skin tightness and discoloration, which she attributed to a car accident that occurred 7 months prior. She also reported abdominal pain, shortness of breath, fever, double vision, dysphagia, voice changes, temperature sensitivity, and hair loss. The patient underwent outpatient steroid injections with limited symptomatic relief. She denied any antecedent exposure to vinyl chloride, rapeseed oil, or L-tryptophan. Physical examination revealed thickened skin on the bilateral legs (top), reddish discoloration of the feet, decreased sensation to light touch, and edema of the ankles and wrists, as well as a peau d'orange appearance of the skin on the arms (bottom), legs, and abdomen.

PLEASE TURN TO PAGE 43 FOR DISCUSSION

Lana X. Tong, MD, MPH; Julie Jackson, MD; Scott Binder, MD; Scott Worswick, MD

From the Division of Dermatology, Department of Medicine, David Geffen School of Medicine at the University of California, Los Angeles.

The authors report no conflict of interest.

Correspondence: Scott Worswick, MD, Division of Dermatology, Department of Medicine, David Geffen School of Medicine at UCLA, 200 Medical Plaza, Ste 450, Los Angeles, CA 90095 (sworswick@mednet.ucla.edu).

The Diagnosis: Eosinophilic Fasciitis

Eosinophilic fasciitis is a rare autoimmune disease of uncertain etiology first described by Shulman¹ in 1974. It is similar in presentation and is perhaps related to scleroderma. Classic differentiating features include a peculiar peau d'orange appearance, peripheral eosinophilia, and lack of Raynaud phenomenon, thus it is regarded as a unique disease.² Despite the name of the disease, eosinophilia has been known to be absent in later stages of eosinophilic fasciitis.¹

On physical examination, “prayer and groove signs” can sometimes be evident.³ Although it was not initially observed in our case, a groove sign was noted on the left forearm on a second inspection (Figure 1). In contrast with systemic sclerosis, visceral involvement rarely is seen with eosinophilic fasciitis. There are, however, 3 major exceptions to this rule. First, there can be widespread nerve deficits, esophageal dysmotility, and nonspecific electromyography findings (ie, denervation, reinnervation, fasciculations).⁴ There also can be a concomitant hematologic disorder or Hashimoto thyroiditis.⁵ Because eosinophilic fasciitis has been associated with monoclonal gammopathy, which our patient also demonstrated, it is important to conduct a workup with serum or urine protein electrophoresis. If the test is negative, it should be followed up with an immunofixation assay or serum light chain assays.^{6,7}

Some proposed risk factors for eosinophilic fasciitis include trauma, extensive exercise, and *Borrelia burgdorferi* infection, but many cases have none of these associations.⁸ Although not firmly proven in the literature, there have been reports of eosinophilic fasciitis after isolated trauma.⁵ A causal link could not be established between our patient's car accident and eosinophilic fasciitis, but the coincidence was notable.

The treatment of eosinophilic fasciitis is similar to scleroderma. Corticosteroids are effective in most cases and recovery often occurs with monotherapy.⁵ Case series have demonstrated efficacy in adding methotrexate, azathioprine, colchicine, and hydroxychloroquine in refractory patients.^{2,9} Our patient demonstrated a good response with a combination of prednisone and methotrexate. Relapses have been known to occur.²

A punch biopsy obtained from the right arm showed thickened acellular collagen bundles throughout the dermis and extending into the underlying subcutis. There also was obliteration of adnexal structures, loss of perieccrine fat, and sparse perivascular and

interstitial lymphoplasmacytic infiltrate (Figure 2), consistent with a sclerosing disorder such as scleroderma or eosinophilic fasciitis.

A complete blood cell count revealed eosinophil levels of 12.5% (reference range, 2.7%). Rheumatologic workup was negative for antinuclear antibody, double-stranded DNA, thyroid-stimulating hormone, anticentromere antibodies, and Scl-70 autoantibodies. Computed tomography of the chest and pelvis revealed a thickened patulous esophagus. Endoscopy showed dysmotility of the lower esophagus. At this point the differential diagnosis included scleredema versus eosinophilic fasciitis, and the patient was started on oral prednisone 60 mg daily. She showed rapid improvement in sclerosis, joint mobility, and ability to swallow. Magnetic resonance imaging was then performed and revealed thickening and contrast enhancement of the forearm fascia, particularly along the distal aspect, confirming a diagnosis



Figure 1. Groove sign on the left forearm characteristic of eosinophilic fasciitis.

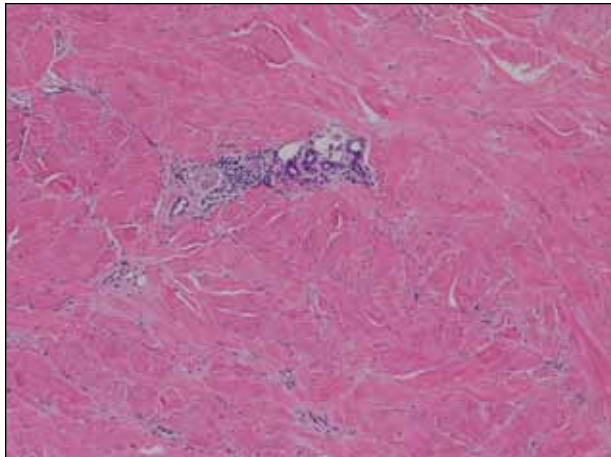


Figure 2. A punch biopsy from the right arm showed thickened acellular collagen bundles throughout the dermis and extending into the underlying subcutis. There also was obliteration of adnexal structures, loss of perieccrine fat, and sparse perivascular and interstitial lymphoplasmacytic infiltrate (H&E, original magnification $\times 10$).

of eosinophilic fasciitis. Further workup including immunofixation assay and serum light chain assays were performed, revealing IgG λ hypergammaglobulinemia. She was then additionally treated with oral methotrexate 15 mg weekly. Due to the rapid improvement of symptoms on oral prednisone over 2 weeks, the peripheral eosinophilia, the magnetic

resonance imaging findings, and the results of skin biopsy, a diagnosis of eosinophilic fasciitis was heavily favored over scleroderma and scleredema.

REFERENCES

1. Shulman L. Diffuse fasciitis with hypergammaglobulinemia and eosinophilia in a new syndrome. *J Rheumatol*. 1974;1(suppl):46.
2. Lakhanpal S, Ginsburg WW, Michet CJ, et al. Eosinophilic fasciitis: clinical spectrum and therapeutic response in 52 cases. *Semin Arthritis Rheum*. 1988;17:221-231.
3. Servy A, Clerici T, Malines C, et al. Eosinophilic fasciitis: a rare skin sclerosis. *Pathol Res Int*. 2010;2011:716935.
4. Satsangi J, Donaghy M. Multifocal peripheral neuropathy in eosinophilic fasciitis. *J Neurol*. 1992;239:91-92.
5. Antic M, Lautenschlager S, Itin PH. Eosinophilic fasciitis 30 years after—what do we really know? *Dermatology*. 2006;213:93-101.
6. Doyle JA, Ginsburg WW. Eosinophilic fasciitis. *Med Clin North Am*. 1989;73:1157-1166.
7. Naschitz JE, Yeshurun D, Miselevich I, et al. Colitis and pericarditis in a patient with eosinophilic fasciitis—a contribution to the multisystem nature of eosinophilic fasciitis. *J Rheumatol*. 1989;16:688-692.
8. Hausteil UF. Scleroderma and pseudoscleroderma: uncommon presentations. *Clin Dermatol*. 2005;23:480-490.
9. Lebeaux D, Francès C, Barete S, et al. Eosinophilic fasciitis (Shulman disease): new insights into the therapeutic management from a series of 34 patients. *Rheumatology*. 2012;51:557-561.