A case of overlap between eosinophilic fasciitis and generalized morphea

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Dear Editor,

Eosinophilic fasciitis (EF) is a rare connective tissue disease that presents as a cutaneous edema and skin sclerosis of the distal extremities.[1-3] Morphea is also a rare connective tissue disease but presents as a sclerotic plaque, typically unilateral on the trunk or proximal extremities. Generalized morphea, a variant of morphea, sporadically manifests as well-circumscribed sclerotic lesions on the trunk and/or extremities. EF often accompanies morphea. However, concurrent EF and generalized morphea are extremely rare. To the best of our knowledge, only eight cases have been reported in the literature.[4,5]

A 65-year-old man was referred to our hospital with lower extremities edema and skin sclerosis on the trunk that appeared one month prior to the visit. He had a medical history of diabetes mellitus, hyperlipidemia, and spinal canal stenosis. Physical examination revealed skin sclerosis, irregularly shaped erythematous patches, and plaques with symmetrical gloss on the mid-front chest and bilateral thorax, lumbar region, and buttocks. The bilateral lumbar regions had a bottleneck shape due to the skin sclerosis. The region from the thighs to the foot dorsum on both legs presented with edema, partial skin sclerosis, and a rash. Moreover, the ankle has a limited range of motion. Mild skin sclerosis was observed in the brachium bilaterally. No significant findings were observed in the face, neck, forearms, hands, or fingers. He had no history of excessive exertion or physical trauma prior to the onset of symptoms. No Raynaud's symptoms, enlarged/giant capillaries, or capillary microhemorrhage in the eponychium were identified. Laboratory data revealed high levels of eosinophils (20.5%), erythrocyte sedimentation rate (47 mm/h), C-reactive protein

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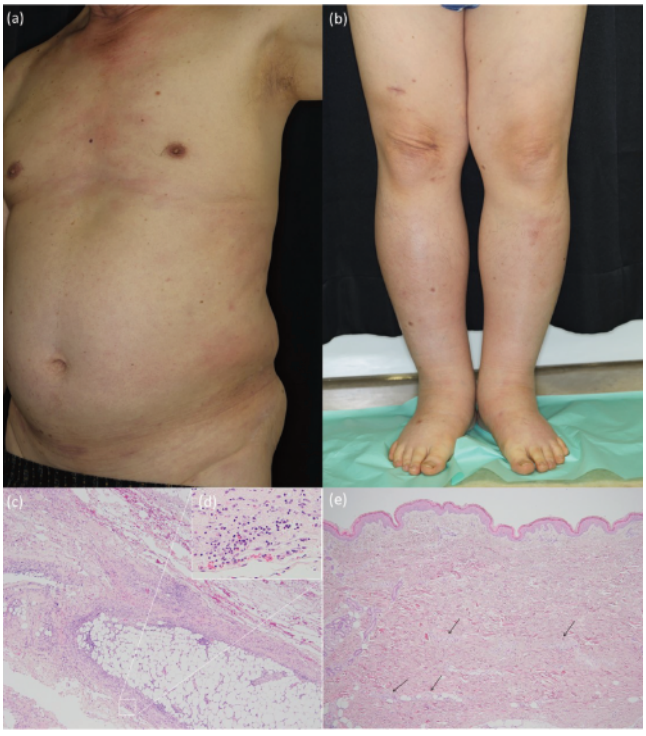

Figure 1. a) Skin sclerosis and irregularly shaped erythematous patches or plaques with gloss symmetrically observed on the mid-front chest, lateral thorax, and lumbar region. The lateral lumber region appeared bottleneck-shaped due to the skin sclerosis; **b**) the region from the thighs to foot dorsum of both legs presented edema with partial skin sclerosis and rash; **c, d**) a skin biopsy of the left thigh revealed widening of the septa in subcutis and thickened fascia with fibrosis, edema and infiltration of eosinophils, lymphocytes, and plasma cells; **e**) the skin biopsy of the erythematous plaque with skin sclerosis in the left lateral lumber revealed thickened collagen bundles. The arrows indicate atrophy of sweat glands in the reticular dermis.

(4.4 mg/dL), and soluble interleukin-2 receptor (5080 IU/mL). Anti-nuclear, anti-centromere, and anti-Scl-70 antibodies were all within the normal limits. Magnetic resonance imaging of the thigh and lower extremities revealed thickened fascia with hyperintensity on short-term inversion recovery sequences. Histopathological evaluation of a sclerotic lesion from the left thigh revealed subcutis septal widening and a thickened fascia with fibrosis, edema, and infiltration of eosinophils, lymphocytes, and plasma cells, consistent with EF (Figure 1a-b). Skin biopsy from an erythematous sclerotic plaque on the left lateral lumbar region revealed thickened collagen bundles and sweat gland atrophy in the reticular dermis, consistent with morphea (Figure 1c). The patient's symptoms gradually improved after oral treatment with 40 mg/day of prednisolone.

In EF, typical histopathological findings include thickening and fibrosis of the fascia, with infiltration of eosinophils, lymphocytes, and plasma cells. The inflammation and fibrosis extend to the subcutaneous tissue and dermis with EF progression. Fibrosis in morphea occurs mainly in the reticular dermis and, in advanced cases, extends to the subcutaneous tissues or fascia. Both EF and morphea present with fibrosis and can be differentiated by the depth of skin involvement. Therefore, they may represent the same spectrum of scleroderma-like diseases.[1-3] Moreover, 30% of EF patients present with morphea.[1-3] Patients with EF and associated morphea are more refractory to treatment than those without morphea.[6] Therefore, if EF is suspected, a detailed physical examination of the entire body and a histopathological examination are recommended for prognosis considering generalized morphea.

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