Poikiloderma of Civatte

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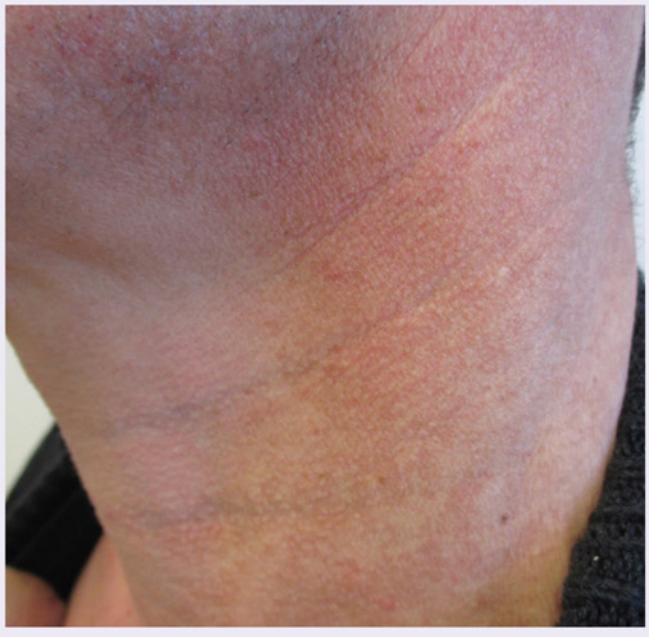
A 49-year-old white man presented with gradual progression of reddish brown pigmentation on the sides of the neck and lateral cheeks for 3 years. The man's lesions were asymptomatic.

The patient worked mainly outdoors. His past health was unremarkable. He was not aware of any family history of similar skin disease. He had never experienced an allergic reaction to medications, foods, metals, or perfumes.

Physical examination revealed skin atrophy, telangiectasia, and mottled, reddish brown, reticulate pigmentation that was symmetrically distributed on the sides of the neck and lateral cheeks. The shaded area under the chin was spared. The rest of the physical examination findings were normal.



Based on the history and clinical findings, a diagnosis of poikiloderma of Civatte was made. The patient was treated with 3 treatments of intense pulsed light (IPL), which evened out his skin tone to a satisfactory degree.

Discussion. Poikiloderma of Civatte is a rather common, benign skin condition characterized by erythematous to brownish patches consisting of superficial atrophy, telangiectasia, and mottled hyperpigmentation seen mainly on the sides of the neck and peripheral face.[1] The condition was first described by Achille Civatte in 1923 and now bears his name.[2]

Epidemiology and etiology. The exact prevalence is not known, given that many patients do not seek medical attention. It is estimated, for example, that the condition accounts for approximately 1.4% of dermatologic consults in Greece.[1] The condition is most common in fair-skinned individuals in the fourth to seventh decade.[3] There is a female predominance.[1,3] The occurrence is usually sporadic, but familial occurrence has been described.

Cumulative exposure to solar radiation appears to be a primary etiologic factor, given that the condition is most commonly seen in sun-exposed areas.[3] Other predisposing factors include photosensitizing chemicals in cosmetics and perfumes, hormonal changes related to menopause or a low estrogen state, and genetic susceptibility.[3,4] An autosomal dominant mode of inheritance with variable penetrance has been described.[5]

Histopathology. Histologic examination typically reveals an atrophic epidermis with parakeratosis, irregular distribution of melanin in the lower epidermis, melanophages in the upper dermis, solar elastosis associated with vasodilation in the papillary dermis, and mild perivascular lymphohistiocytic infiltrate.[3,6,7]

Clinical manifestations. Poikiloderma of Civatte presents with a combination of linear telangiectasia, erythema, mottled hyperpigmentation, and superficial atrophy in a reticular pattern, symmetrically affecting the sun-exposed areas such as the sides of the neck, lateral cheeks, and upper chest.[1,7,8] The shaded area under the chin is characteristically spared.[4] Based on the predominating clinical features, poikiloderma of Civatte can be

classified into 3 clinical types, namely, the predominant erythematotelangiectatic type, the pigmented type, and the mixed type.[1,3,8] The lesions usually are asymptomatic, although some patients report mild itching, burning, stinging, flushing, and increased sensitivity of the affected skin.

Diagnosis and differential diagnosis. The diagnosis is mainly clinical, based on clinical features and characteristic symmetric distribution of the lesion with sparing of the submental area.[1,3]

Differential diagnosis includes rosacea, erythromelanosis follicularis faciei et colli, Riehl melanosis, Berloque dermatitis, eczema, acquired brachial cutaneous dyschromatosis, cutaneous T-cell lymphoma, Bloom syndrome, acute cutaneous lupus erythematosus, dyskeratosis congenita, and parapsoriasis.[1]

Referral to a dermatologist is advised if the diagnosis is in doubt.

Prognosis and management. While the condition can be cosmetically unsightly and socially embarrassing,[1] poikiloderma of Civatte poses no health hazards. Although the disease is benign, it runs a slowly progressive and irreversible course.[7]

Apart from reassurance about the benign nature of the condition, treatment usually is not necessary. For those who desire treatment for cosmetic reasons, modalities include IPL, pulsed dye or other vascular laser therapy, and ablative fractional photothermolysis.[8,9]

Avoiding sun exposure especially during hours of peak UV intensity (11 AM to 4 PM), regular use of broad-spectrum sunscreens, and wearing protective hats and clothes when outdoors should be emphasized for prevention. ■

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Progressive Cutaneous Scleroderma

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A 60-year-old woman presented with painful and sclerotic hands and fingers. She had been an airline attendant but had to quit her job as a result of the pain and her inability to use her hands over the past 7 years.

History. The patient recently had been seen by a pain physician, had been started on opioids, and had received stellate ganglion blocks that had yielded very little relief. A rheumatologist had prescribed methotrexate and corticosteroids in an attempt to stop excessive overproduction of connective tissue and to help control her disease state, which also did not help. She had opted out of sacroiliitis therapy, which can soften the fibrosis but also is associated with an increased risk of malignancy.

Physical examination. The patient had no lung or heart problems or complaints. No vascular abnormalities or telangiectasia were seen on examination. Her fingers were unable to be opened or flexed and were wooden-like on palpation; some digits were missing from self-amputation with chronic pain (**Figures 1 and 2**).

The woman received a clinical diagnosis of progressive cutaneous scleroderma.

Figure 1. The patient's hands at presentation.

Discussion. Localized scleroderma has an estimated prevalence of 50 per 100,000 before age 18 years and 129 per 100,000 by age 80.[1] Scleroderma is a chronic autoimmune disorder that is manifested by excess synthesis and deposition of collagen in skin and connective tissue.[2]

Vascular abnormalities occur as Raynaud phenomenon and swelling of the acral portions of the extremities, with thickening of the skin of the fingers as the most common complaints in CREST syndrome (calcinosis, Raynaud phenomenon, esophageal dysfunction, sclerodactyly, and telangiectasia). Cutaneous manifestations were the only findings present in our