Cutaneous Collagenous Vasculopathy

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PRACTICE **POINTS**

- Cutaneous collagenous vasculopathy (CCV) should be in the differential diagnosis of widespread telangiectases.
- Biopsy is needed to differentiate between CCV and generalized essential telangiectasia because of their similar clinical features.
- There may be underlying comorbidities associated with CCV, but the exact cause of the condition has yet to be found.

To the Editor:

Cutaneous collagenous vasculopathy (CCV) is a rare idiopathic microangiopathy characterized by diffuse blanchable telangiectases that usually develop in late adulthood. It appears morphologically identical to generalized essential telangiectasia (GET), but skin biopsy characteristically shows dilated superficial blood vessels in the papillary dermis that are surrounded by a thickened layer of type IV collagen.[1] We report a case of CCV occurring in an elderly white man.

A 72-year-old man presented with an asymptomatic rash on the arms, legs, and abdomen of 3 years' duration. His medical history was remarkable for hypothyroidism, hypertension, reflex sympathetic dystrophy syndrome, coronary artery disease, and nonmelanoma skin cancer. He denied any changes in medications or illnesses prior to onset of the rash. Physical examination revealed diffuse, erythematous, blanchable telangiectases on the arms, legs, and trunk (Figure 1). No petechiae, atrophy, or epidermal changes were appreciated. Darier sign was negative.

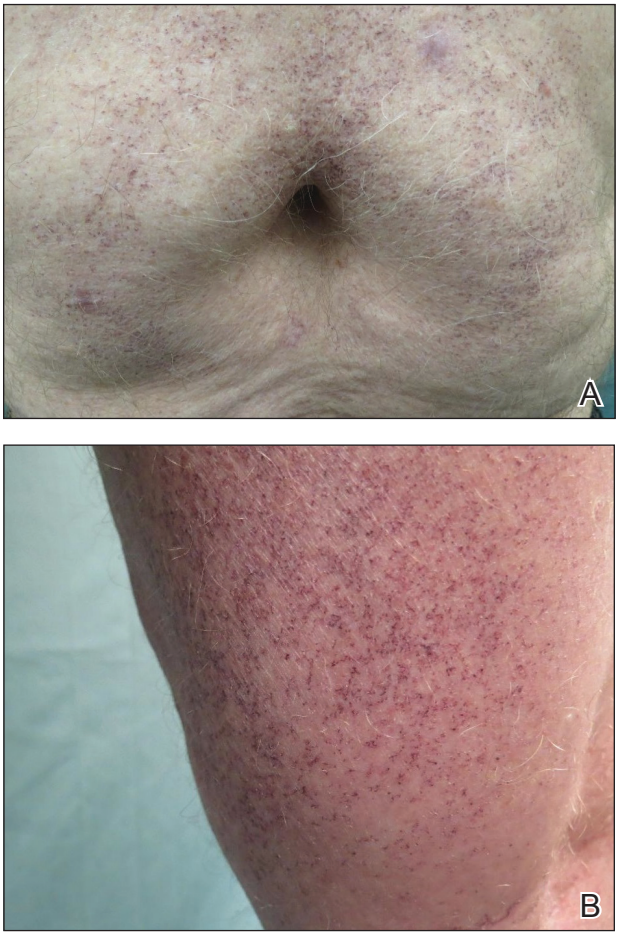

FIGURE 1. A and B, Diffuse, erythematous, blanchable telangiectases on the abdomen and left leg in a patient with cutaneous collagenous vasculopathy.

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Hematoxylin and eosin–stained sections of skin from the abdomen showed an unremarkable epidermis overlying a superficial dermis with dilated blood vessels with thickened walls that contained eosinophilic amorphous hyaline material (Figure 2A). This material stained positive with Masson trichrome (Figure 2B), a finding that was consistent with increased collagen fiber deposition within the vessel walls. Phosphotungstic acid–hematoxylin and Congo red stains were negative. No histologic features of a vaso-occlusive disorder or vasculitis were identified. These histologic findings were consistent with the rare diagnosis of CCV.

Cutaneous collagenous vasculopathy is a rare idiopathic microangiopathy that was first reported by Salama and Rosenthal[1] in 2000. They reported the case of a 54-year-old man with spreading, asymptomatic, generalized cutaneous telangiectases of 5 years' duration. Similar to our patient, skin biopsy showed dilated superficial dermal vasculature with deposition of eosinophilic hyaline material, which stained positive with periodic acid–Schiff with diastase and exhibited immunoreactivity to type IV collagen.[1]

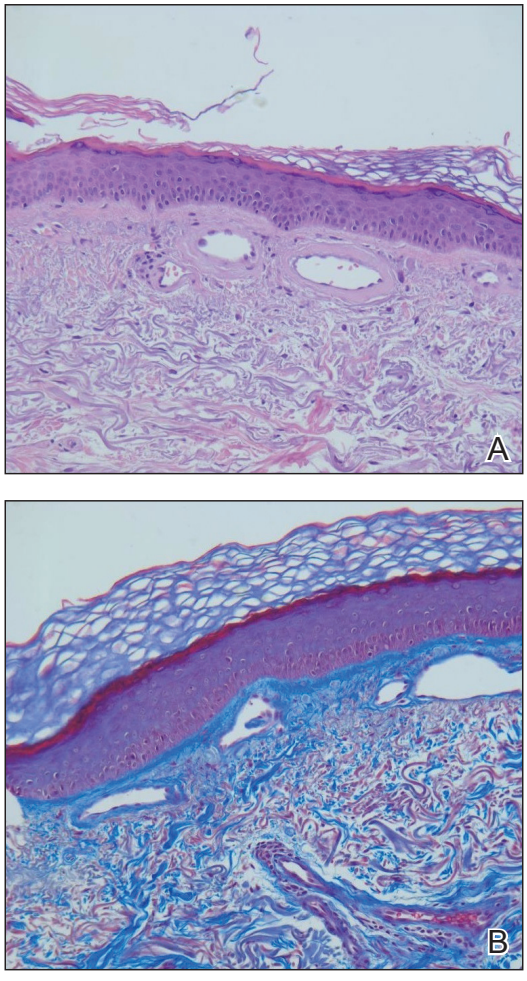

FIGURE 2. A, Biopsy from the abdomen revealed telangiectatic blood vessels with hyalinized thickened walls within the superficial dermis (H&E). B, Blood vessel walls stained positive with Masson trichrome, which was consistent with increased collagen fiber deposition within the vessel walls.

A PubMed search of articles indexed for MEDLINE using the search term *cutaneous collagenous vasculopathy* yielded 19 additional patients with biopsy-proven CCV.[2-6] The condition has shown no gender prevalence but generally is seen in middle-aged or elderly white individuals, with the exception of a white pediatric patient.[4] Cutaneous collagenous vasculopathy usually presents as telangiectases on the legs that progress to involve the trunk and arms while sparing the head and neck, nail beds, and mucous membranes.[5] However, it also has been described as first presenting on the bilateral breasts[2] as well as a nonprogressive localization on the thigh.[6]

Skin biopsy is essential to differentiate CCV from GET, which appears morphologically identical. Cutaneous collagenous vasculopathy may be underreported as a result of clinician choice not to biopsy due to a presumptive diagnosis of GET.[3] Successful treatment with a pulsed dye laser has been reported,[7] though the extent of disease may make complete destruction of the lesions difficult to accomplish. Although it is theorized that CCV may be a marker for underlying systemic disease or even a genetic defect causing abnormal collagen deposition, its cause has yet to be ascertained.[5] Previously reported patients have had a variety of comorbidities, including several cases of type 2 diabetes mellitus.[6] Another patient was reported to have recently started treatment with an angiotensin receptor blocker prior to onset of CCV.[5]

Our case contributes to the small series of reported patients with this rare diagnosis and further suggests that it may be underreported at this time. Similar to previously reported cases, our patient was an elderly white individual. Although our patient had long-standing iatrogenic hypothyroidism, no recent medication changes or underlying comorbidities could be tied to the development of CCV. Further studies are needed to determine if this disease process is associated with any underlying systemic illnesses, medications, or family history.

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