

Cutaneous Collagenous Vasculopathy

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PRACTICE POINTS

- In cutaneous collagenous vasculopathy (CCV), skin biopsy may demonstrate eosinophilic hyaline thickening of superficial dermal blood vessels with mild perivascular lymphocytic inflammation and rare extravasated erythrocytes.
- Lack of mucous membrane and nail involvement differentiates CCV from hereditary hemorrhagic telangiectasia.

the patient had no history of hepatic or renal dysfunction, cancer, or heart disease. Her family history was negative for clotting disorders or bleeding diatheses.

On physical examination, telangiectatic matting was present on the bilateral ankles and dorsal feet with an associated blanchable erythema (Figure 1). The matting extended into a fine, mottled, pretibial telangiectasia associated with Schamberg purpura. She had no pitting edema, and both dorsalis pedis and posterior popliteal pulses were intact and symmetric bilaterally. No popliteal lymphadenopathy or palpable cords were present.

To the Editor:

Cutaneous microangiopathy describes pathology of the small blood vessels within the dermis. Cutaneous collagenous vasculopathy (CCV) is a rare manifestation of cutaneous microangiopathy with only 38 reported cases in the literature, according to a PubMed search of articles indexed for MEDLINE using the term *cutaneous collagenous vasculopathy*. This disorder must be distinguished from generalized essential telangiectasia, which generally does not warrant a biopsy. Prior case reports have demonstrated predominance of CCV in middle-aged men; however, women and adolescents also may be affected.¹⁻⁵

We report a case of CCV in a 41-year-old woman who presented for evaluation of a rash on the bilateral lower extremities of 7 to 8 months' duration. The eruption had started on the left ankle and spread over several weeks to the bilateral dorsal feet followed by the ankles and shins. The patient noted associated swelling and a pressure-like dysesthesia of the lower legs. She was otherwise in good health, though she had started an oral contraceptive 1 year prior for heavy menstrual bleeding. A review of systems was negative for deep vein thrombosis, pulmonary embolus, and other thromboembolic phenomena, and



FIGURE 1. Cutaneous collagenous vasculopathy presenting as telangiectatic matting with blanchable erythema on the bilateral ankles and dorsal feet.

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The authors report no conflict of interest.

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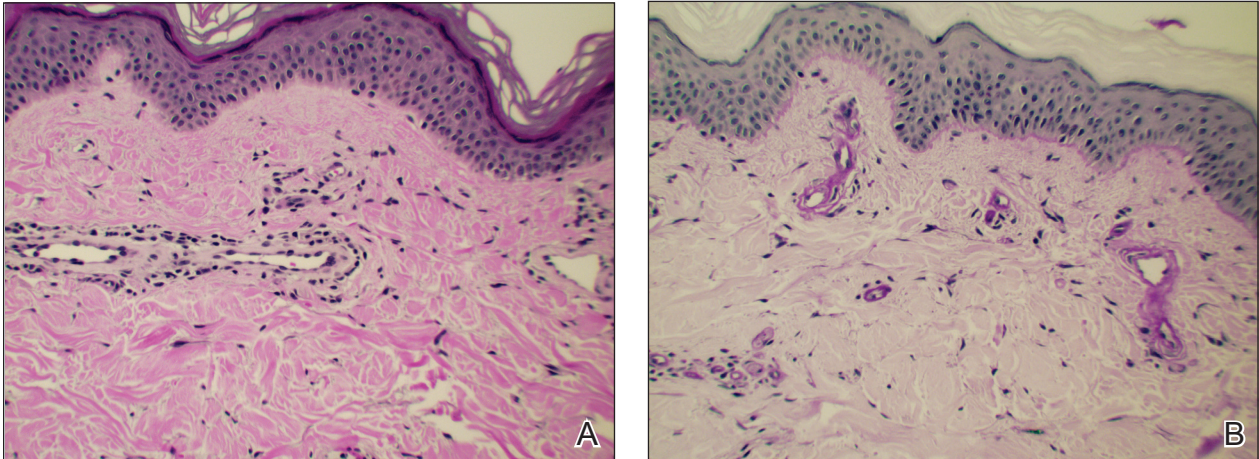


FIGURE 2. A, Punch biopsy revealed mild vascular dilatation with thickened eosinophilic walls and mild perivascular lymphocytes (H&E, original magnification $\times 20$). B, Periodic acid–Schiff staining demonstrated vessel walls thickened with positive eosinophilic hyaline material (original magnification $\times 20$).

Two punch biopsies taken from the erythematous telangiectatic area on the left foot and metatarsal region demonstrated an unremarkable epidermis without interface change, thickening of the epidermal basement membrane, or single-cell dyskeratosis. There was mild dilatation of blood vessels within the superficial dermis with mild perivascular lymphocytic inflammation and rare extravasated erythrocytes. Leukocytoclastic debris, fibrinoid necrosis of vessel walls, and endothelial cell necrosis were not seen. As is classic in CCV, the vessel walls appeared thickened by eosinophilic hyaline material, which was periodic acid–Schiff positive and diastase resistant (Figure 2). Sclerotic thickening of collagen bundles or absence of periadnexal adipose tissue was not seen. CD34 immunohistochemical staining demonstrated normal retained CD34 interstitial dermal positivity, which excluded morphea. Additionally, direct immunofluorescence testing was negative for IgG, IgA, IgM, C3, fibrin, and C1q. Nodular reduplication of vessels or other changes of stasis were not seen. Fibrin thrombi or neoplastic cells were not identified. The clinical and histopathologic findings were suggestive of CCV.

Prior case reports of CCV have described a similar clinical manifestation with blanching macules that occur symmetrically on the lower extremities and spread

cephalically.^{1–6} A distinction from hereditary hemorrhagic telangiectasia is the noninvolvement of mucous membranes and nails. The etiology of this rare microangiopathy has not been elucidated, though disease concurrence with local trauma, stressful events such as childbirth, and diabetes mellitus has been documented.⁶ As the body of literature continues to grow, more research regarding the etiology, mechanism, prognosis, and treatment options will enhance our understanding of CCV.

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