Cutaneous collagenous vasculopathy

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DESCRIPTION

A 47-year-old Caucasian woman presented with a 4-year history of macular skin lesions. They started as small, non-pruritic, erythematous macules on the lower limbs ([figure 1](#)). They gradually spread over several years, to involve the torso and upper limbs. She remained systemically well, with no hair, nail or mucosal membrane involvement. Her medical history was significant for ulcerative colitis, diagnosed shortly after the onset of the skin lesions and hypothyroidism. Examination revealed widespread 3–5 mm blanching hyperpigmented macules, sparing the face. Blood tests, including immunology and virology, were normal. Four punch biopsies were all initially reported as non-specific. However, on specialist review, histology revealed scattered ectatic capillaries with collagen deposition, cinching the diagnosis of cutaneous collagenous vasculopathy (CCV) ([figure 2](#)).

Figure 1 Clinical photograph demonstrating hyperpigmented macules on the anterior lower limbs.

Figure 2 Dilated superficial dermal vessels with thickening of the vessel wall associated with hyaline eosinophilic material on H&E staining at low (top left) and high power (top right), Masson's trichrome stain with positive material seen in the vessel wall (bottom left) and periodic acid–Schiff stain with positive material seen in blood vessel walls (bottom right).

CCV is a microangiopathy of dermal blood vessels originally described in the year 2000.¹ It is a rare disease with fewer than 44 cases in the literature to date; however, is likely to be under diagnosed. CCV is mainly seen in Caucasians with a wide age range and equal gender prevalence. Clinically, it presents as asymptomatic blanching, non-urticarial macules and diffuse telangiectasia starting on the lower limbs with cephalic spread, sparing mucosal surfaces and without systemic involvement. Other features include ecchymoses and petechiae. It has also been associated with a history of diabetes mellitus, autoimmune and cardiovascular disease.² Although benign, the widespread distribution of the disease alongside the diagnostic uncertainty it often creates can lead to distress to the patient.

The main differential of CCV and a common misdiagnosis is generalised essential telangiectasia. This is a benign disorder with widespread telangiectasia most often found on the lower limbs. Unlike in our case where the lesions were hyperpigmented macules, most cases of CCV present as diffuse telangiectasia and are therefore clinically indistinguishable from generalised essential telangiectasia. There are several other differentials to consider, including telangiectasia macularis eruptiva perstans a form of cutaneous mastocytosis.³ A normal serum tryptase made this less likely. These alternative diagnoses can also be differentiated from CCV by their histological appearance.

The pathogenesis remains unclear. A proposed mechanism is that following endothelial cell injury,

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Images in...

veil cells (immature dendritic cells) are activated and a reparative fibrosis ensues, involving deposition of abnormal and disorganised collagen in blood vessel walls.²

The diagnosis of CCV is reached by correlating the clinical presentation with characteristic histology. Typical histological features are dilated capillaries and postcapillary venules with collagen deposits. Superficial dermal vessel walls are thickened due to eosinophilic hyaline material deposits. Veil cells and Luse bodies (abnormal collagen) with focal endothelial damage can also be seen on electron microscopy.⁴

Treatment options are limited. Pulsed dye laser therapy has been suggested and was successful in one case, warranting further exploration.⁵

In summary, this is a disorder of dermal blood vessels, likely under diagnosed by clinicians but that can be confirmed by its characteristic histological features.

Learning points

- ▶ Cutaneous collagenous vasculopathy (CCV) is a disorder of dermal blood vessels with characteristic clinical and histological findings.
- ▶ CCV presents with asymptomatic blanching, non-urticarial macules and diffuse telangiectasia, often misdiagnosed as generalised essential telangiectasia.
- ▶ Histology typically demonstrates superficial dermal blood vessel wall thickening due to collagen deposits on H&E staining, and veil cells and Luse bodies on electron microscopy.

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