CASE REPORT

Cutaneous collagenous vasculopathy: a rare cause of generalised cutaneous telangiectasia

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SUMMARY

Cutaneous collagenous vasculopathy is a rare cutaneous microangiopathy of unknown aetiology with only 27 cases reported to date. It is characterised clinically by generalised cutaneous telangiectasias and microscopically by dilation and marked thickening of the walls of superficial dermal blood vessels. Differential diagnosis should be performed with other causes of disseminated telangiectasias, including generalised essential telangiectasia, from which it is clinically indistinguishable. We report a new case of cutaneous collagenous vasculopathy in a 61-year-old woman presenting with a 5-year history of asymptomatic telangiectasias distributed symmetrically on her upper and lower limbs and highlight the importance of clinicopathological correlation for the diagnosis of this disease.

BACKGROUND

Cutaneous collagenous vasculopathy (CCV) is an idiopathic cutaneous microangiopathy first described by Salama and Rosenthal in 2000.¹ Since the initial report, only 26 cases have been reported which, together with our case, gives a total of 28 patients.^{2–5} CCV is characterised by progressive development of generalised cutaneous telangiectasias with unique but subtle histological features, consisting of dilated blood vessels of the superficial dermal plexus with marked wall thickening due to collagen deposition.^{6–7} Because it is clinically indistinguishable from generalised essential telangiectasia (GET), differing in its histology, it is possible that some cases clinically suspected to be GET may actually represent CCV.⁸ We describe a new case of CCV to increase awareness of this still poorly known condition and emphasise the importance of clinicopathological correlation in the differential diagnosis of generalised telangiectasias.

CASE PRESENTATION

A 61-year-old woman presented to our department with a 5-year history of asymptomatic telangiectasias that had initially appeared on the distal legs and forearms, and gradually spread reaching her thighs and upper arms. She did not report any bleeding events or photosensitivity. Her medical history included hypertension, coronary heart disease, obesity, vesicular lithiasis, hyperlipidaemia and chronic venous insufficiency. Treatment included perindopril, bisoprolol, clopidogrel and atorvastatin. Family history was negative for telangiectasias and bleeding diathesis. Physical examination revealed symmetrical, telangiectatic, reticulated,

reddish macules that blanched with pressure, distributed on her upper and lower extremities (figures 1 and 2). Darier's sign was negative and there was no nail or mucosal involvement.

INVESTIGATIONS

Routine blood analyses were within normal limits and autoantibodies screen revealed positive anti-nuclear antibodies with a 1/320 titre and a speckled pattern, without further alterations.

Histopathological examination of a skin biopsy taken from the left arm showed multiple dilated blood vessels with thickened walls within the superficial dermis (figure 3). The vessel walls contained amorphous, hyaline, eosinophilic material (figure 4) that was highlighted with periodic acid-Schiff (PAS) stain and was resistant to diastase (figure 5A). This material was also emphasised by immunohistochemical staining for collagen type IV, which showed a duplication of the basal membrane (figure 5B). Epidermis and adnexal structures were normal and no significant haemorrhage or inflammation was present.

DIFFERENTIAL DIAGNOSIS

The clinical, histopathological and immunohistochemical findings were consistent with CCV.

TREATMENT

Given the absence of symptoms, it was decided not to treat the lesions.

OUTCOME AND FOLLOW-UP

The patient has been followed up at our department for 1 year, and is in good general condition without specific treatment.

DISCUSSION

CCV is an uncommon primary cause of generalised cutaneous telangiectasia involving the postcapillary venules of the horizontal dermal plexus.¹ It presents in both sexes without predilection and in various age groups, although mainly in the middle-aged and elderly.²

Figure 1 Erythaematous, mottled, telangiectatic macules symmetrically involving the forearms and upper arms.

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Figure 2 Detail of telangiectatic macules affecting the lower limb.

Clinically, it is characterised by the progressive development of symmetric telangiectatic macules that typically first arise in the lower extremities and gradually spread to reach the trunk and upper limbs, sparing the mucosal surface and nail bed.^{2 9} The head and neck are usually not involved.² Lesions are typically asymptomatic but can rarely be pruritic.^{2 9} The individual telangiectatic macules characteristically remain stable but may gradually darken.² Family history is negative for telangiectasias or bleeding disorders and there is no consistent association with a specific medical disorder.² Although most cases of CCV described had some associated disease and received various treatments, these associations are probably fortuitous and related to the age of the patients.^{2 8}

There are currently no recommended laboratory studies to aid in the diagnosis of CCV. Reported laboratory studies have usually been unremarkable, including routine laboratory studies, autoantibodies screen and viral serology. The primary role for such studies would be to help exclude other entities in the clinical differential diagnosis.⁹

Histopathological findings are essential for the diagnosis of CCV,⁸ but can be subtle.⁹ The characteristic finding is dilation and marked thickening of the walls of superficial dermal blood vessels, due to deposition of eosinophilic hyaline material around the endothelial basal lamina.⁹ This material stains with PAS, is diastase resistant,⁹ and is also highlighted with iron colloidal stain⁷ and by immunohistochemistry for type IV collagen,

Figure 3 Dilated thickened small blood vessels within the superficial dermis (H&E, ×100).

Figure 4 On higher magnification, the presence of amorphous pink hyaline material can be seen in the vessel walls (H&E, ×400).

fibronectin and laminin antibodies.^{1 6} It stains blue with Masson trichrome and often contains mucinous material.⁸ Elastin stains, Congo red and crystal violet stains are negative,⁹ and there is decreased or absent actin staining.² A reduplication and splitting of the basement membrane can be seen.^{2 10} There are no inflammatory cells present within vessel walls,⁹ but a few lymphoid cells can be seen around occasional vessels.² Little to no haemorrhage or haemosiderin is present.¹

Electron microscope analysis shows that the affected vessels correspond to postcapillary venules.² Ultrastructurally, characteristic features include reduplication of the basal lamina, paucity of pericytes lacking contractile fibrils, and marked deposition of both normal and abnormal collagen within the vessel walls.² Luse bodies, or abnormal long spacing collagen, are non-specific and may be absent in some cases of CCV.^{9 10} Their origin is as yet unknown but it is believed to be related to abnormal collagen synthesis and organisation.^{1 2}

CCV may be under-recognised because it can be mistaken both clinically and histopathologically for other entities.⁹ The differential diagnosis includes other primary and secondary causes of disseminated telangiectasias: GET, hereditary haemorrhagic telangiectasia, hereditary benign telangiectasia, telangiectasia macularis eruptiva perstans, angioma serpiginosum, lymphomas, drug-induced telangiectasias, radiodermatitis, collagen vascular diseases and amyloidosis.^{4 8} Distribution of telangiectasias, age of onset, associated systemic features and histopathology aid in the diagnosis.³ Clinically, CCV is indistinguishable from GET and differs in its histology. It is possible that some non-biopsied cases clinically suspected to be GET may actually represent CCV.⁸

In the absence of a good clinical history, other entities with amorphous, PAS-positive hyaline material could be considered in the histopathological differential diagnosis, including porphyria, pseudoporphyria, lipoid proteinosis, atrophie blanche and amyloidosis. In entities in which the deposits are not composed of basement membrane material, immunohistochemistry with type IV collagen antibody may help in the differential diagnosis.⁹

The aetiopathogenesis of CCV is uncertain, probably because of the rarity or scarcity of reported cases.^{2 9} Ultimately, CCV seems to be clearly related to damage to the superficial dermal microvasculature, and recent reports suggest that, at least in some cases, intravascular occlusive microthrombosis secondary to repeated endothelial cell injury may play a role.^{2 10} However, the initial triggering factor causing the endothelial cell damage remains unknown.²

Figure 5 The hyaline material within the vessel walls is (A) periodic acid-Schiff (PAS)-positive resistant to diastase (PAS with diastase, $\times 400$) and (B) emphasised by immunohistochemical staining for collagen type IV (collagen type IV antibody stain, $\times 400$).

There is no cure for this chronic and progressive condition,⁷ and in most reported cases, treatment was not performed or this was not specified.⁸ Laser therapy with 585 nm pulsed dye laser was reported in one patient, with important aesthetical improvement.⁷

Learning points

- Cutaneous collagenous vasculopathy (CCV) is a rare microangiopathy of superficial dermal vessels that is possibly underdiagnosed; clinicians may interpret it as generalised essential telangiectasia and not proceed to biopsy.
- Histopathological findings are essential for the diagnosis of CCV but can be subtle.
- Increased awareness of this condition may lead to more biopsies from patients with generalised telangiectasia and further improve its recognition and understanding of its aetiopathogenesis.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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