Progressive ascending telangiectasias

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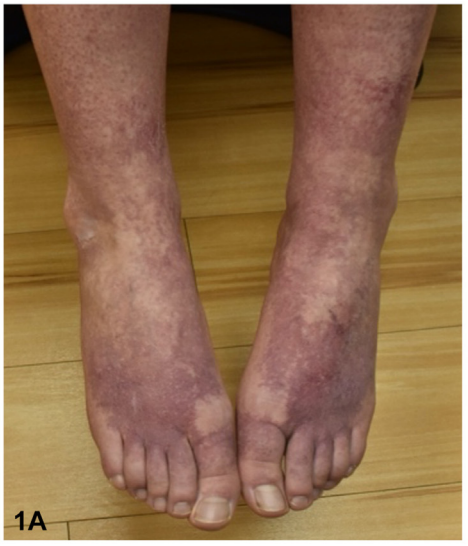

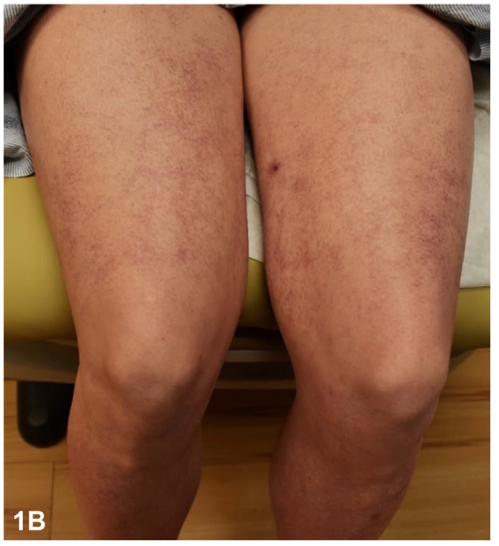

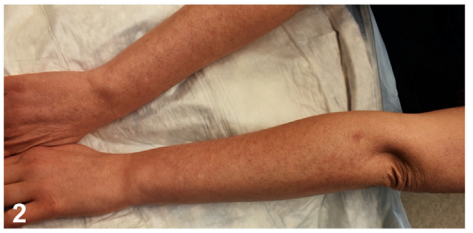

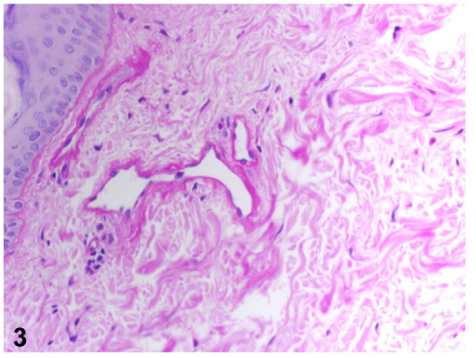

INTRODUCTION

A 39-year-old woman presented with a 2-year history of progressive ascending telangiectasias on the extremities associated with intermittent pain and sensitivity. The use of triamcinolone cream failed to alleviate symptoms. Her medical history included basal cell carcinoma and rosacea, but no abnormal bleeding. Family history was negative for similar lesions or abnormal bleeding. A review of her system was unremarkable. Physical examination revealed generalized nontender, nonurticating erythema and telangiectatic macules and patches distributed symmetrically on both lower extremities (Figs 1 and 2) with no involvement of the nails or mucosal surfaces. Biopsy revealed telangiectatic vessels within the papillary dermis with variably thickened periodic acid—Schiff (PAS)-positive walls and a sparse perivascular lymphocytic infiltrate (Fig 3).

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Question 1: What is the most likely diagnosis?

A. Cutaneous collagenous vasculopathy (CCV)

B. Generalized essential telangiectasias

C. Hereditary hemorrhagic telangiectasia

D. Pigmented purpuric dermatosis, Schamberg type

E. Telangiectasia macularis eruptiva perstans (TMEP)

Answers:

A. CCV — Correct. CCV is a rare form of superficial microangiopathy, affecting a wide demographic population often with medical comorbidities such as diabetes mellitus and hypertension and intake of medications at the time of diagnosis.[1] Despite this, no causal link has been established with regard to the onset of CCV. Clinically, CCV characteristically presents with diffuse, blanching telangiectatic macules that begin on the lower extremities and progressively spread to the trunk and upper extremities, usually sparing the head and neck.[1] Histopathology reveals dilated, thick blood vessels within the papillary dermis with PAS/PAS plus diastase-positive hyaline deposits, without significant inflammation or hemorrhage (Fig 3).[1] Electron microscopy studies show marked deposition of abnormal collagen referred to as "Luse bodies" within the walls of postcapillary venules (PCV).[1]

B. Generalized essential telangiectasias — Incorrect. The histopathology of generalized essential telangiectasias classically reveals mildly ectatic, thin-walled vessels in the dermis. Despite largely overlapping clinical features, the key distinguishing factor in this case presentation are the histopathologic finding of PAS-positive, thickened vessel walls and the sparing of mucosal surfaces and nails that are unique to CCV.[1]

C. Hereditary hemorrhagic telangiectasia — Incorrect. Hereditary hemorrhagic telangiectasia is clinically indistinguishable from CCV. However, unlike CCV, it is an autosomal dominant disorder characterized by a history of bleeding diathesis, a family history of similar telangiectasias, and frequently exhibits mucosal and/or nail involvement.[1]

D. Pigmented purpuric dermatosis, Schamberg type — Incorrect. Pigmented purpuric dermatosis, manifests as (nonblanching) petechiae and purpura with characteristic cayenne pepper-like discoloration. Similar to CCV, pigmented purpuric dermatosis also routinely manifests in the lower limbs and does not typically involve the upper extremities. Histopathologic findings often include superficial lymphocytic infiltration and marked hemosiderin deposition with erythrocyte extravasation.[2]

E. TMEP — Incorrect. TMEP is a rare form of mastocytosis classically affecting young adults, presenting with telangiectatic macules on the chest and the extremities. Although TMEP often manifests cutaneously, symptoms such as flushing, dyspnea, and gastrointestinal disorders are not uncommon. Histology reveals small mononuclear infiltrates with increased mast cells around superficial venous plexus capillaries.[3]

Question 2: Which of the following treatments is most likely to result in successful treatment in this patient?

A. Pulsed-dye laser

B. Sclerotherapy

C. Narrowband UV-B

D. Topical clobetasol

E. Oral methylprednisone

Answers:

A. Pulsed-dye laser — Correct. Pulsed-dye laser produces pulsed light at wavelengths of 585 or 595 nm, specifically targeting intravascular oxyhemoglobin by selective thermolysis, resulting in destruction of blood vessels within the dermis. It has previously been used successfully to treat CCV.[4] Our patient was treated with 12 sessions of pulsed-dye laser with marked improvement in appearance and symptoms.

B. Sclerotherapy — Incorrect. Sclerotherapy has been used previously to treat vascular disorders successfully but has not been found effective in the treatment of CCV.

C. Narrowband UV-B — Incorrect. Narrowband UV-B delivers energy at a 311 to 312—nm wavelength for effective penetration of the epidermis and epidermodermal junction. It has been used for effective treatment of skin diseases like psoriasis, vitiligo, and atopic dermatitis.

D. Topical clobetasol — Incorrect. Topical corticosteroids that classically act through antiinflammatory and immunosuppressive mechanisms will not treat CCV, where characteristic findings include

dilation and thickening of blood vessels from hyaline and collagen deposition. In our case, the patient used triamcinolone cream that did not alleviate symptoms.

E. Oral methylprednisone — Incorrect. Oral steroids, which are typically used to treat systemic inflammatory conditions including vasculitis, do not have a role in the treatment of CCV.

Question 3: Direct immunofluorescence staining for superficial blood vessels and PCVs will be positive for which immunoreactant in this patient?

A. Fibrinogen

B. C3

C. IgA

D. Immunoglobulin M (IgM)

E. Collagen type IV

Answers:

A. Fibrinogen — Incorrect. Fibrinogen deposits can be seen in the papillary dermis, specifically within the walls of PCV in cutaneous small-vessel vasculitis.[5]

B. C3 — Incorrect. C3 deposits within the walls of PCV is commonly seen in cutaneous small-vessel vasculitis. Homogenous deposits of C3 on the walls of superficial blood vessels are also seen in porphyrias, more commonly erythropoietic protoporphyria, but also infrequently in porphyria cutanea tarda and pseudo-porphyria cutanea tarda.[5]

C. IgA — Incorrect. IgA deposition on the walls of superficial blood vessels, often in a granular pattern, is frequently seen with Henoch-Schönlein purpura.[5]

D. IgM — Incorrect. IgM deposits on the walls of superficial blood vessels in the dermis has been observed in cutaneous small-vessel vasculitis.[5]

E. Collagen type IV — Correct. CCV is characterized by thickened walls of superficial blood vessels, including PCVs, because of the deposition of PAS-positive hyaline material.[1] These deposits are also positive for collagen based on Masson trichome staining and are reactive for collagen type IV based on immunohistochemical studies.[1]

Abbreviations used:

CCV: cutaneous collagenous vasculopathy
PAS: periodic acid—Schiff
PCV: postcapillary venules
TMEP: telangiectasia macularis eruptiva perstans

Conflicts of interest
None disclosed.

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