Cutaneous collagenous vasculopathy: papular form

Alberto Conde-Ferreirós[1], Mónica Roncero-Riesco[1], Javier Cañueto[1], Concepción Román-Curto[1], Ángel Santos-Briz[2]

Affiliations: [1]Department of Dermatology, University Hospital of Salamanca, Paseo de San Vicente, 58-182, 37007, Salamanca, Spain, [2]Department of Pathology, University Hospital of Salamanca, Paseo de San Vicente, 58-182, 37007, Salamanca, Spain

Corresponding Author: Dr. Alberto Conde Ferreirós, University Hospital of Salamanca, Department of Dermatology, Paseo San Vicente, 58-182, 37007, Salamanca (Spain), Email: alconfe25@yahoo.com albertoconde@saludcastillayleon.es, Tel: 34-923-29100/34-691-010273

Abstract

Cutaneous collagenous vasculopathy is a rare clinicopathological entity, first described in 2000. Cutaneous collagenous vasculopathy has been considered a form of microangiopathy of superficial dermal vessels and produce lesions that appear as telangiectasia. We present a patient with histopathologic features of cutaneous collagenous vasculopathy and scattered erythematous papules on the trunk with a striking dermatoscopic finding. We propose the term of "cutaneous papular collagenous vasculopathy" as a new clinical manifestation of this disease.

Keywords: cutaneous collagenous vasculopathy, collagen IV, microangiopathy, cutaneous telangiectasias

Introduction

Cutaneous collagenous vasculopathy is a rare clinicopathological entity; it was first described in 2000 by Salama and Rosenthal [1]. Cutaneous collagenous vasculopathy has been considered a form of microangiopathy, which affects superficial cutaneous vessels. It is characterized by the deposit of a homogeneous eosinophilic hyaline material in vascular walls. To date, all cases clinically consist in progressive asymptomatic cutaneous telangiectasias.

Case Synopsis

We report a 68-year-old man with a history of hyperuricemia, hypertension, atrial fibrillation, gout,

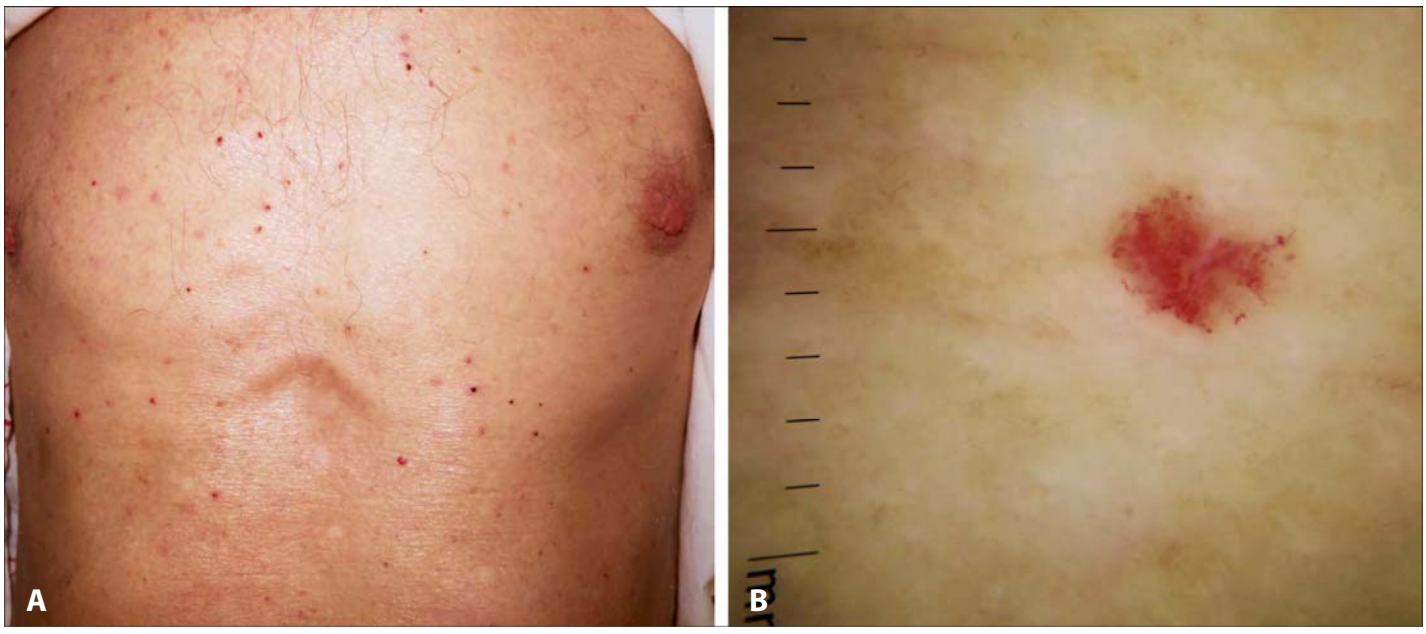

Figure 1. ***A)*** *Erythematous papules located to the trunk alternating with non-blanchable millimetre crusted lesions.* ***B)*** *Upon dermoscopy, vascular-like erythematous papules with tortuous branching telangiectasias that draw a pink crown. Each division = 1mm.*

peripheral arterial disease, internal carotid aneurysm, posterior carotid ischemic stroke, and chronic renal failure related to hypertensive nephrosclerosis. He was being treated with imidapril, bisoprolol, acetyl salicylic acid, warfarin, pentoxifyline, furosemide, atorvastatin, and omperazole. He was admitted to the hospital owing to pneumonia (*Streptococcus pneumoniae*).

Upon cutaneous examination, the patient exhibited blanchable millimetric eruptive erythematous papules located on his trunk alternating with non-blanchable minute macules (**Figure 1A)**. No arboriform networks of telangiectasias were observed. Some lesions clinically resembled cherry angiomas. However, upon dermoscopy they exhibited vascular-like erythematous papules with tortuous branching telangiectasias that showed a pink crown (**Figure 1B)**. The distinctive reddish lacunae pattern of cherry angiomas was not seen. Nail beds and the mucosal surfaces were both spared.

Skin biopsy showed dilated capillaries and post-capillary venules in the superficial dermis with perivascular deposits (**Figure 2A)**. Periodic acid-Schiff stain and periodic acid-Schiff stain with diastase digestion highlighted a deposit of a homogeneous eosinophilic hyaline material in vascular walls (**Figure 2B** and figure **2C)**.

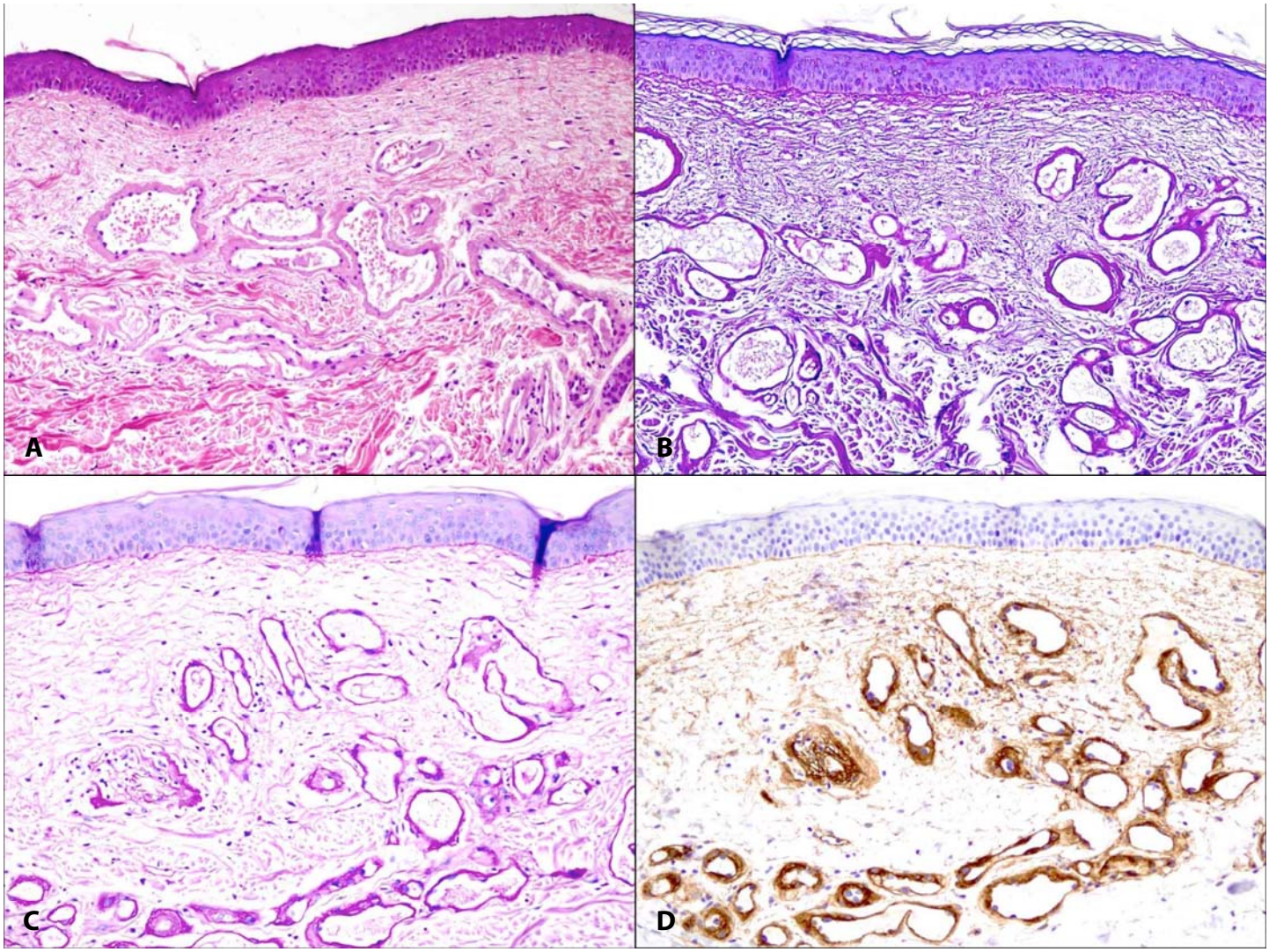

Figure 2. ***A)*** *Dilated capillaries and post-capillary venules in the superficial dermis, with perivascular deposits of a homogeneous acellular eosinophilic material. H&E, 100×).* ***B)*** *and* ***C)*** *Periodic acid-Schiff stain and Periodic acid-Schiff stain with diastase digestion highlight deposits of homogeneous eosinophilic hyaline material in vascular walls, 100×.* ***D)*** *Immunohistochemical staining for type IV collagen is positive, 100×.*

Immunohistochemical staining for type IV collagen was positive (**Figure 2D)**. Also, mild fibrosis throughout the capillary vessels and a mild perivascular lymphocytic infiltrate in the superficial dermis were observed. Congo red staining was negative, which confirmed that amyloid deposits were not present in the vessel wall.

Based on the histopathological findings, a diagnosis of cutaneous collagenous vasculopathy was made. Although treatment with 0.1% methylprednisolone aceponate was prescribed, the lesions did not improve significantly.

Case Discussion

Cutaneous collagenous vasculopathy has been described as a rare form of microangiopathy affecting superficial cutaneous vessels [1]. To date, 45 cases have been reported in the literature to our knowledge [1, 2]. It mainly affects adults but has also been described in children [3]. The etiopathogenesis of cutaneous collagenous vasculopathy is currently unknown but it might have a relationship with cardiovascular risk factors and extensive medication intake [4]. Despite this fact, there is no clear association between other diseases or medication usage and the development of cutaneous collagenous vasculopathy.

The present case is unique because it does not meet the typical clinical features of cutaneous collagenous vasculopathy. All cases described to date consist of progressive asymptomatic cutaneous telangiectasias that initially appear on the lower extremities and then gradually progress to the abdomen, trunk, and arms [5]. Dermatoscopic exploration of papular cutaneous collagenous vasculopathy exhibits some erythematous papules with a characteristic dermatoscopic visualization consisting of thick telangiectasias arranged in a crown. However, the isolated observation of just one case is not sufficient to consider this as a unique finding of papular cutaneous collagenous vasculopathy. Histopathological features in cutaneous collagenous vasculopathy are similar to classic telangiectatic cutaneous collagenous vasculopathy found in the literature [1]. These consist of marked collagen deposits within the vascular walls of the postcapillary venules in the superficial dermis [1].

Conclusion

It is likely that cutaneous collagenous vasculopathy is insufficiently diagnosed owing to its clinical similarity to other skin disorders, especially cherry angiomas and generalized essential telangiectasia. But these diseases show dermatoscopic, histopathologic, and unique ultrastructural characteristics different from cutaneous collagenous vasculopathy [4, 5].

It is important to know that cutaneous collagenous vasculopathy not only manifests itself in a telangiectatic form, but also in a papular form. Additional investigations and new cases of the papular form of cutaneous collagenous vasculopathy can shed light on the pathogenesis of cutaneous collagenous vasculopathy.

Potential conflicts of interest

The authors declare no conflicts of interests.

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