CASE REPORT

Cutaneous Collagenous Vasculopathy: A Case Report and Review of the Literature

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KEYWORDS
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PALABRAS CLAVE
Telangiectasia esencial generalizada; Telangiectasias; Vasculopatía colágena cutánea

Abstract
Cutaneous collagenous vasculopathy is an idiopathic microangiopathy first described in 2000 by Salama and Rosenthal. It must not be confused with generalized essential telangiectasia. To date, all patients have been white men over the age of 50 years, most of whom had multiple pathologies, were taking multiple drugs, and had no family history of similar conditions or hemorrhagic disorders. The disease is characterized by the development of various numbers of telangiectases on the limbs, lower abdomen, chest, or back, with no mucosal or nail bed involvement. Histopathology shows dilated superficial cutaneous vessels with perivascular deposits of periodic acid-Schiff diastase-positive, eosinophilic hyaline material that exhibits positive immunoreactivity to collagen IV. We report a new case in a 68-year-old man with symmetrically distributed telangiectases on his forearms, lower abdomen, posterior thighs, lower legs, and dorsum of the feet.

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Vasculopatía colágena cutánea: aportación de un caso y revisión de la literatura

Resumen
La vasculopatía colágena cutánea es una microangiopatía idiopática descrita por Salama y Rosenthal en el año 2000. Debe distinguirse de la telangiectasia esencial general. Hasta la fecha, todos los pacientes son varones caucásicos de más de 50 años de edad, en su mayoría con pluripatología y multimedicatedos, sin evidencia de historia familiar de procesos similares ni trastornos hemorrágicos. Se caracteriza por el desarrollo de múltiples telangiectasias cutáneas dispuestas de forma variable en las extremidades, la zona baja del abdomen, el pecho o la espalda, sin afectación de mucosas ni del lecho ungueal.
Introduction

Cutaneous collagenous vasculopathy (CCV) is an idiopathic microangiopathy first described in 2000 by Salama and Rosenthal.\(^1\) They reported the case of a 54-year-old man with multiple asymptomatic telangiectases distributed symmetrically on the lower legs, thighs, trunk, and upper limbs, the histopathology of which showed dilated superficial dermal blood vessels with perivascular deposits of a periodic acid-Schiff (PAS)-diastase-positive eosinophilic hyaline material that exhibited immunoreactivity to collagen IV. We have only found 3 cases published since that time, described by Davis et al.\(^2\) We report a new case of CCV recently seen in our department.

Case Report

The patient was a 68-year-old man with a past history of hypertension, hypercholesterolemia, hyperuricemia, benign prostatic hyperplasia, and alopecia areata universalis who was receiving treatment with atenolol, atorvastatin, allopurinol, and tamsulosin hydrochloride. He was referred to our department for the evaluation of asymptomatic reddish lesions on his limbs that had started to appear 15 years earlier. He reported no family history of similar lesions and no personal or family history of mucosal bleeding.

Physical examination showed multiple telangiectases that blanched when pressed under glass. They were distributed symmetrically on the forearms, lower abdomen, posterior thighs, lower legs, and dorsum of the feet (Figures 1 and 2). There was no involvement of the mucosas or nail bed, and no spread of the telangiectases to the rest of the skin.

Generalized essential telangiectasia was suspected, and a biopsy of the lesions of the external aspect of the left forearm was performed. Histopathology showed no alterations of the epidermis and a telangiectatic dilation of the blood vessels with a variable distribution and luminal diameter in the papillar dermis. Deposits of a homogeneous eosinophilic material of irregular thickness, suggestive of amyloid substance, were observed around the largest vessels (Figures 3 and 4). These deposits stained positive with PAS-diastase stain (Figure 5) and Masson’s trichrome stain (Figure 6), but not with Congo red. Immunostaining was strongly positive for collagen IV.

Additional tests—biochemistry, rheumatoid factor, antinuclear antibody, antibodies to extractable nuclear antigen, anticientromere and anti-topoisomerase (anti-ScL70) antibodies, complement fractions C3 and C4, cryoglobulins, thyroid-stimulating hormone, protein electrophoresis, immunoglobulin concentrations, and serology for hepatitis B and C viruses and human immunodeficiency virus—were normal or negative.

A diagnosis of CCV was established, and the patient was offered the possibility of laser therapy. However, due to the absence of symptoms or systemic repercussions, the patient decided not to undergo treatment.

La histopatología evidencia vasos cutáneos superficiales dilatados, con depósito peri vascular de un material hialino eosinófilo que se tiñe con PAS-diastasa y que muestra inmunorreactividad de anticuerpos frente al colágeno IV. Describimos un nuevo caso en un varón de 68 años de edad con telangiectasias distribuidas de forma simétrica en los antebrazos, la región baja del abdomen, la cara posterior de los muslos, las piernas y el dorso de los pies.

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Discussion

All cases of CCV reported to date have been similar to the present case (Table). They were characterized by the appearance of variable numbers of cutaneous telangiectases on the limbs, lower abdomen, chest, or back, with no mucosal or nail bed involvement. In most cases, the tentative clinical diagnosis was generalized essential telangiectasia. All 5 patients were white males aged over 50 years (range, 54-80 years). Four of them had multiple diagnoses, were taking several drugs, and had no family history of similar conditions or hemorrhagic disorders. \(^1,^2\)

Histopathology showed dilated blood vessels of the superficial dermal plexus, with marked thickening of their walls due to deposition of an amorphous eosinophilic material that exhibited staining characteristics of collagen and that stained positive with PAS-diastase and colloidal iron. Immunohistochemically, the material exhibited positive immunoreactivity to collagen IV, fibronectin, and laminin, but was negative for muscle actin. Ultrastructural studies suggested that the vessels were postcapillary venules with abundant deposits of collagen around the basal lamina and with focal splitting or reduplication of the basement membrane.\(^2\) Fibers with abnormally spaced transverse bands (Luse bodies) were observed.\(^1\) Occasional lymphocytes,
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Table Cases of Cutaneous Collagenous Vasculopathy

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Abbreviations: AA, alopecia areata; AP, actinic poikiloderma; BPH, benign prostatic hyperplasia; DM, diabetes mellitus; GE, gastroesophageal reflux; VI, venous insufficiency.

1. Although its etiology is unknown, it has been speculated that it might be due to a primary disease caused by a genetic defect that alters the collagen production of small blood vessels of the skin. It has also been suggested that it might be a manifestation of a systemic disease or caused by some medication, although these possibilities have yet to be confirmed. Theoretically, the cutaneous telangiectases of CCV could be treated with laser therapy.

2. The differential diagnosis should include those conditions that present with telangiectasia, such as autoimmune connective tissue diseases, cutaneous mastocytosis, graft versus host disease, liver disease, ataxia-telangiectasia, traumatic injuries, chronic solar damage, radiation dermatitis, rosacea, unilateral nevus telangiectasia, hereditary hemorrhagic telangiectasia, benign hereditary telangiectasia, many genodermatoses, vascular malformations, and hormonal disorders. However, there are 2 conditions in which differentiation can be more difficult. First, it must be distinguished from generalized essential telangiectasia, a condition that affects a different demographic population (more common in middle-aged women) and does not show the distinctive histological characteristics of CVV, but that is clinically very similar, as both conditions present telangiectases that begin on the lower limbs and progress in a cephalad direction, spreading symmetrically to the trunk and the upper limbs. In some cases there may be conjunctival and oral mucosal involvement. Second, the possibility of an iatrogenic origin needs to be ruled out, given the numerous drugs taken by patients with CVV. Cases of telangiectasia secondary to certain drugs—corticosteroids, lithium, tiotixene, interferon α, isotretinoin—have been reported, as have cases of CCV on photoexposed areas after taking calcium channel blockers (nifedipine or amlodipine), antibiotics such as cefotaxime, or antidepressants such as venlafaxine.

Conflicts of Interest

The authors declare that they have no conflicts of interest.

References