Cutaneous collagenous vasculopathy: A rare case report

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ABSTRACT

Cutaneous collagenous vasculopathy (CCV) is a distinct, rare, and underdiagnosed condition. We report a case of CCV in a 50-year-old woman presenting as asymptomatic, erythematous to hyperpigmented nonblanchable macules over both the lower extremities. The clinical differential diagnosis of the lesions was pigmented purpuric dermatoses (Schamberg's purpura) and cutaneous small vessel vasculitis. Histology of the lesions revealed dilated superficial dermal vessels with abundant pink hyaline material in the vessel wall, which stained with periodic acid Schiff stain. The patient was diagnosed as CCV. This condition remains largely underdiagnosed and is commonly mistaken for pigmented purpuric dermatosis or generalized essential telangiectasia. Emphasis on the differentiation of CCV from its clinical and histological mimicks is made.

Key words: Cutaneous collagenous vasculopathy, dilated superficial dermal vessels, hyaline, microangiopathy

INTRODUCTION

Cutaneous collagenous vasculopathy (CCV) is a distinct and rare cutaneous microangiopathy affecting the superficial dermal blood vessels. These patients present with asymptomatic blanchable pink or red macules, telangiectasia, or petechiae distributed symmetrically on bilateral lower extremities with progressive involvement of the upper extremities and trunk. The comorbidities frequently associated in these patients are diabetes mellitus, hyperlipidemia, hypertriglyceridemia, and hypertension. Microscopically, it reveals thickened and dilated superficial dermal vessels. This condition remains largely underdiagnosed and is commonly mistaken for pigmented purpuric dermatosis or generalized essential telengiectasias.

CASE REPORT

A 50-year-old female presented with asymptomatic rash on the lower extremities since 2 years. Cutaneous examination revealed erythematous to hyperpigmented nonblanchable, nontender macules and petechial lesions over both the lower extremities [Figure 1]. There was no edema or warmth of the affected area. There was a history of diabetes mellitus in the patient for which she was currently not on any medications.

Skin biopsy from the erythematous macule showed vasculopathy involving the small vessels of the superficial plexus. The affected vessels showed abundant pink collagenous deposit in the wall and moderately dense lymphocytic infiltrate around them [Figures 2 and 3]. The collagenous deposits in the vessel walls were Periodic Acid Schiff (PAS) positive [Figure 4]. Many of the vessels showed dilatation of the walls resembling telangiectasia. There was neither vasculitis nor any perivascular hemosiderin deposition. The case was diagnosed as cutaneous collagenous vasculopathy (CCV).

DISCUSSION

CCV is a rare distinct idiopathic microangiopathy of superficial dermal blood vessels. The first case of cutaneous collagenous vasculopathy presenting with asymptomatic telangiectases

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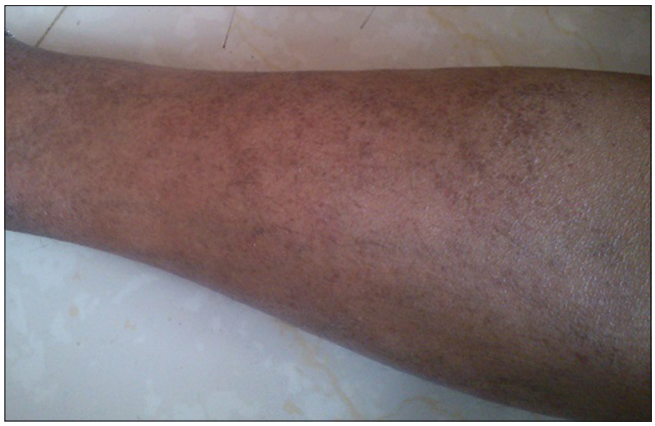

Figure 1: Nonblanchable macules and petechiae on lower extremity

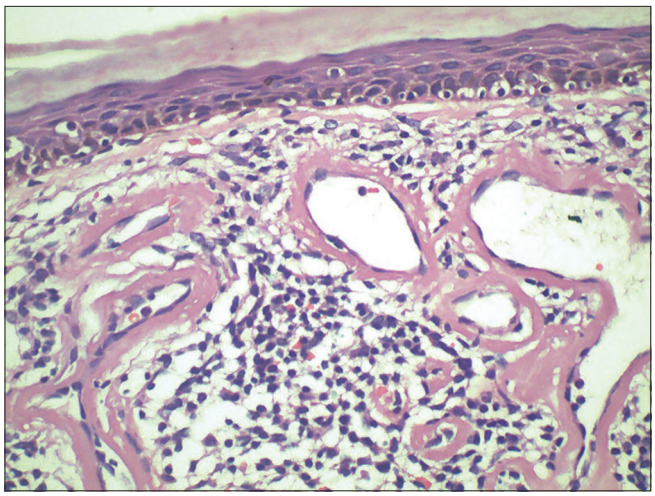

Figure 3: Dilated superficial vessels with pink hyaline deposit in the vessel wall. (Hematoxylin and Eosin ×400)

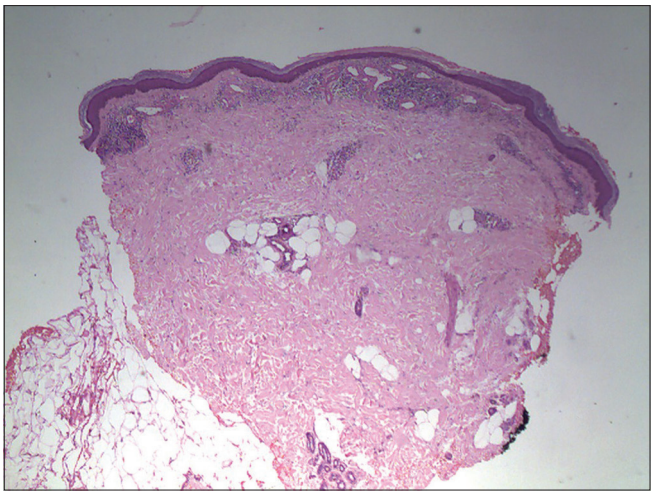

Figure 2: Dilated superficial vessels with pinkish hyaline deposit in the vessel wall. (Hematoxylin and Eosin ×100)

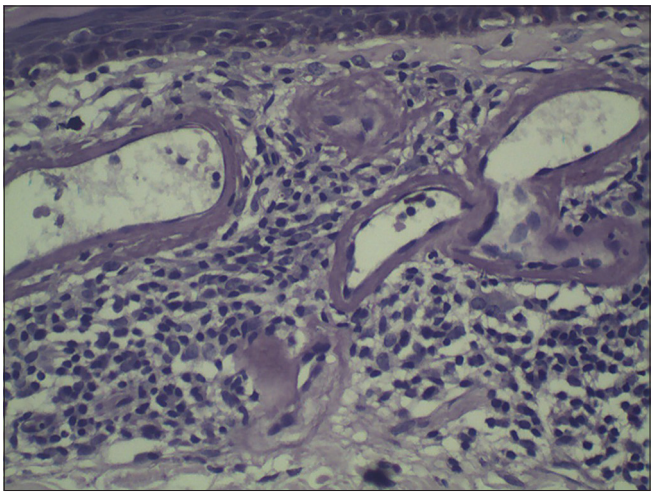

Figure 4: PAS positive hyaline deposit in the vessel wall. (Periodic Acid Schiff ×400)

was described by Salama and Rosenthal in the year 2000.[1] This condition has been described in middle-aged and elderly men. There have been descriptions of this condition in women too.[2] The exact etiology of the condition is not known. Several patients with CCV described in literature had diabetes mellitus. The occurrence of microangiopathy in diabetes mellitus could possibly contribute to the development of CCV.[3] Other factor implicated in the causation for CCV is trauma. It is also speculated that this condition may represent a genetic vascular disorder.[4] Another hypothesis is that CCV may be caused by a genetic defect in collagen synthesis.[5] The comorbidities seen in patients with CCV were diabetes mellitus, hypertension, hyperlipidemia, hypothyroidism, and venous insufficiency.[5]

Clinically, the most common presentation was in the form of asymptomatic bilaterally symmetrical progressive telangiectases on lower extremities with gradual progression to upper extremities and rarely the trunk. These lesions were clinically mistaken for generalized essential telangiectasia. Other clinical morphologies described were discrete dark pink blanchable macules,[4] petechiae,[4] red macules, and ecchymosis[5] in one patient. The patient described in this report had erythematous, petechial lesions, and nonblanching macules.

Histologically, CCV is characterized by dilated and thickened superficial dermal vessels with flat endothelial cells and the vessel wall containing hyaline material. There is paucity of inflammation and the vessel walls show no sign of damage by inflammatory cells. There is no hemorrhage or hemosiderin deposition in the perivascular area. The hyaline material stains with PAS and is diastase resistant.[6] The hyaline material of the thickened walls is highlighted by immunohistochemistry for laminin and type IV collagen but not with elastic stains, which indicates it is a derivative of the basement membrane collagen and is the result of reduplication and splitting of the basement membrane zone surrounding small vessels.[7] Histopathologically, CCV may

be misdiagnosed as telangiectasia (dilated capillaries without inflammatory infiltrate), livedoid vasculopathy (deposits of fibrin in the dermal vessels without significant inflammatory infiltrate) or cutaneous small vessel vasculitis (leukocytoclasia, damage to the vascular endothelium with fibrin deposition). Other conditions showing hyaline material in the vessel walls are lipoid proteinosis (hyaline material around the eccrine gland with atrophy of the glands), porphyrias (subepidermal blister), pseudoporphyrias (subepidermal blister), secondary amyloidosis (hyaline material in papillary dermis and subcutaneous tissue with Congo-red positivity), and thrombotic vasculopathy (occlusion of the lumen by the thrombus and hemorrhage.) Electron microscopy reveals "Luse bodies" representing collagen fibers with abnormally long spaces between electron-dense bands.[8]

The exact pathomechanisms of the condition are not yet elucidated. However, it is proposed that vascular damage is followed by repair leading to a defect in collagen formation and abnormal disorganized collagen deposition in the cutaneous blood vessel wall.[8]

Cutaneous collagenous vasculopathy is an under-recognised and underdiagnosed condition. The current report emphasizes the need for awareness of this condition and its clinical and histological mimicks.

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Conflicts of interest

There are no conflicts of interest.

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