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Case Report

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Unilateral Nevoid Telangiectasia: A Clue for Hyperestrogenemia or Hepatic Dysfunction

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Abstract

Observation: Unilateral nevoid telangiectasia(UNT) is a rare cutaneous disease characterized by superficial telangiectasias frequently involving the trigeminal, cervical, and upper thoracic dermatomes. The disease may be congenital or acquired. High estrogenic conditions or hepatic pathologies are usually associated with UNT. Herein, we present a 18-year-old woman with UNT.

Introduction

Unilateral nevoid telangiectasia is a rare cutaneous disease characterized by superficial telangiectasias in a unilateral dermatomal distribution.[1] The disease frequently involves the trigeminal, cervical, and upper thoracic dermatomes.[1] It can be either congenital or acquired.[1] The acquired form affects females twice as often as males and is associated with high estrogen levels, especially during puberty, pregnancy, and oral contraceptive use.[2] The congenital form, which is less common (15% of reported cases), usually affects males and is thought to result from elevated maternal hormones in utero.[2]

Case Report

An 18-year-old woman presented with erythematous macules on her right upper extremity that had been present for two years and had not resolved spontaneously. There was no history of drug use or any other disease. Her parents did not have similar cutaneous findings. On dermatological examination, the macules were distributed in the right C5 and C8 dermatomes (**Figure 1**). Blanchable telangiectasias were more evident on der-

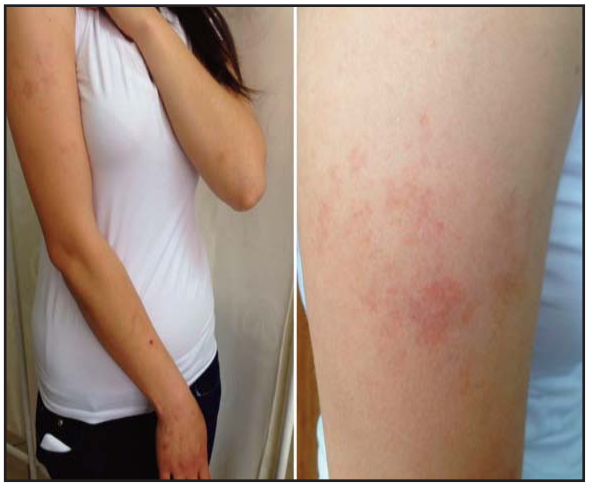

Figure 1. Maculer lesions on right arm

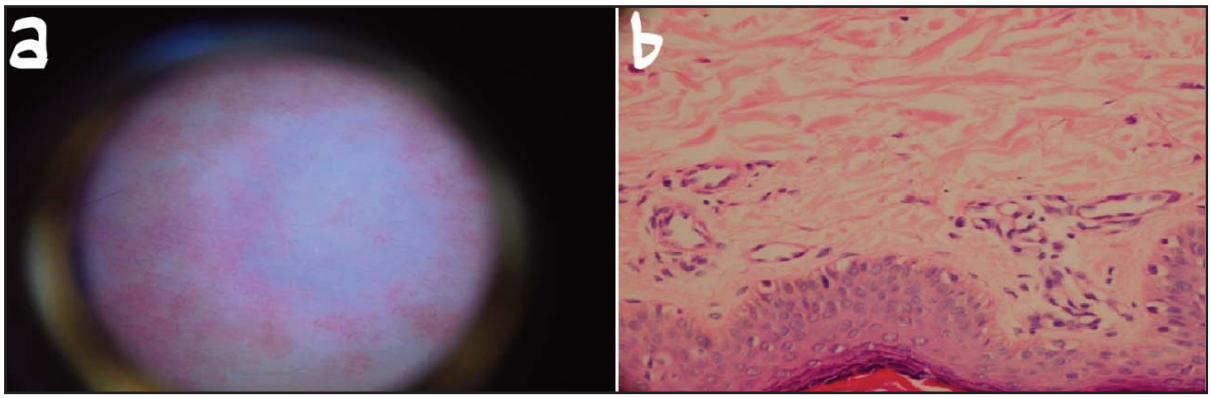

Figure 2. a. Telangiectatic vessels on dermoscopy, b. multiple, thin walled, dilated capillaries in dermiş (H&E x400)

moscopy (**Figure 2a**). The complete blood count and serum estrogen, progesterone, and follicle-stimulating hormone levels were within normal limits. The serological tests for hepatitis B and C were negative. Histologic examination revealed thin-walled, dilated capillaries in the papillary and reticular dermis without endothelial cell proliferation, characteristic of unilateral nevoid telangiectasia (UNT) (**Figure 2b**). Immunohistochemical staining for estrogen and progesterone receptors was negative. The patient was diagnosed with UNT based on the clinical, dermoscopical, and histological findings. Treatment with a pulse dye laser was recommended, but she refused the treatment.

Discussion

Although the pathogenesis of UNT remains unknown, some investigators claim that hyperestrogenic states such as puberty, pregnancy, and oral contraceptives are responsible for the disease.[2] UNT has also been reported to be associated with hyperthyroidism, carcinoid synd rome, hepatitis, cirrhosis, and portal hypertension.[2,3] Some authors believe that the unilateral distribution of the lesions is due to the congenital distribution of estrogen-sensitive cells along a dermatome.[4] However, the lesional skin in most of the reported cases was negative for estrogen and progesterone receptors.[1] Our patient showed no increase in estrogen or progesterone receptor levels in the lesional skin. It has been theorized that hemodynamic disturbances, neurological changes, angiogenic factors, and changes in the connective tissue play roles in the pathogenesis of UNT.[4] In addition, vascular endothelial growth factor is thought to be a causative factor, especially in patients with hepatic disease.[2]

The histopathology reveals multiple, thin-walled, dilated capillaries in the papillary and reticular dermis. No endothelial cell proliferation or neo-angiogenesis is seen.[5] The differential diagnosis includes hemangioma, angioma serpiginosum, and nevus flammeus.[1] UNT usually persists and rarely resolves spon taneo usly. Treatment options include cosmetic camouflage and pulsed dye laser therapy.[5]

Unilateral nevoid telangiectasia is a rare vascular disease that is usually asymptomatic and does not require treatment. However, possible hyperestrogenic conditions and hepatic pathologies must be investigated once the diagnosis is made.

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