Unilateral nevoid telangiectasia (UNT) is a rare, benign, acquired disorder presenting with cutaneous telangiectasias distributed in a predominately unilateral and dermatomal fashion. The etiology of UNT is unknown.

Case

We present a 28-year-old male with an asymptomatic blanchable dermatosis over his bilateral shoulders and upper arms. At the age of 19, he first developed unilateral segmental telangiectasias across his left shoulder and upper arm. The rash stabilized in distribution by the age of 21. A biopsy was done by an outside institution, which showed superficial telangiectasias and sparse perivascular inflammation with extravasated red blood cells. He was given a diagnosis of Schamberg disease, and underwent five rounds of laser treatment without significant benefit. Patient then presented to our clinic for evaluation of a two-year history of similar but less prominent lesions on his contralateral side involving the right upper arm. Patient denied a history of flushing, palpitations, diarrhea, pruritus, or syncope.

On physical exam, the patient had blanchable erythematous coalescing patches with telangiectatic vessels on dermoscopy. The patches extended over his left shoulder and upper extensor arm (Figure 1).

Similar but sparsely distributed patches were also seen on his right upper arm. The patches did not elevate upon rubbing (negative Darier’s sign), and there was no palmar erythema or other evidence of estrogen excess.

A skin biopsy of the upper arm showed telangiectasias with mild superficial perivascular dermatitis. Periodic acid-Schiff (PAS) staining was negative for depositions around the blood vessels. Laboratory evaluation included: normal comprehensive metabolic panel, thyroid stimulating hormone, tryptase, antinuclear antibodies, ferritin, dehydroepiandrosterone sulfate (DHEA-S), testosterone (free and total), estradiol, prolactin, and luteinizing hormone. Screens for hepatitis B and C were negative. Based on clinical and histologic findings, a diagnosis of UNT was made.

Discussion

The differential diagnosis for acquired segmental telangiectasias includes unilateral nevoid telangiectasia (UNT), telangiectasia macularis eruptiva perstans (TMEP), or angioma serpiginosum. TMEP is an uncommon form of cutaneous mastocytosis. This patient lacked the significant increase of mast cells on skin biopsy for diagnosis, nor had any systemic symptoms to suggest TMEP.

Angioma serpiginosum typically has an onset in childhood (80%), favors women (90%), and is composed of punctate partially blanchable macules that show oval lagoons on dermoscopy. In childhood, as angioma serpiginosum enlarges, it leaves a central clearing, resulting in a serpiginous or ring-like morphology. PAS staining on histology would show deposits around dilated dermal blood vessels, which was not found in this case.

The patient had been given a diagnosis of Schamberg disease by an outside clinic. Schamberg disease is a type of pigmented purpura, which is unlikely in this case. Pigmented purpura is non-blancheable, and dermoscopy would show petechial and pigmented macules instead of telangiectasias. On histology, pigmented purpura would have prominent extravasated red-blood cells and hemosiderin deposits.

Patient’s presentation and histopathologic findings were most consistent with UNT. UNT is an idiopathic segmental proliferation of telangiectasias. In the literature, 15% of UNT cases are congenital, with the remainder being acquired.
etiology of UNT remains unknown. Some cases of UNT have been associated with either an increase in systemic levels of estrogen or a localized increase in the level of estrogen receptors in the involved skin. This has not been a universal finding. Others have reported an upregulation of vascular endothelial growth factor (VEGF) or its receptor in involved skin in patients with hepatic disease. Tanglertsampa et al. in 2013 proposed a classification scheme for UNT: (I) congenital, (II) acquired with association (estrogen-related, liver-related, skin-related, others), or (III) without association.

Given the patient’s negative workup, no association was found. Though most UNTs are unilateral, bilateral cases have been reported. This case of UNT was unusual for it developing a bilateral distribution, seven years after disease onset.

Legend

Figure 1: Erythematous, blanchable patches on the extensor surface of the upper arm.

REFERENCES


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