A rare case of unilateral nevoid telangiectasia in a Saudi pediatric patient

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Abstract

Unilateral nevoid telangiectasia (UNT) is a rare congenital or acquired benign cutaneous vascular anomaly first identified by Blaschko in 1899. It is characterized by superficial telangiectasias with a reticular or linear pattern on one side of the body, often following a dermatomal distribution. We present the first case of UNT documented in Saudi Arabia in a 12-year-old girl.

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Introduction

Unilateral nevoid telangiectasia (UNT) is a rare congenital or acquired benign cutaneous vascular anomaly. First identified by Blaschko in 1899, it is characterized by grouped superficial telangiectasias with a reticular or linear net-like pattern confined to one side of the body (unilateral) that blanches with diascopy and frequently follows a dermatomal pattern.[1] The condition can present at any age but is most often identified during adolescence or early adulthood. UNT tends to occur more frequently in females than males, mostly on the upper trunk and extremities.[1,2] The precise cause of UNT is not fully understood, though it is believed to involve both genetic and hormonal factors. Hormonal changes during puberty, pregnancy, or the use of oral contraceptives have been linked to the condition, suggesting a role for estrogen sensitivity.[3-5] Additionally, UNT has been associated with systemic conditions such as liver disease and hyperthyroidism in some patients.[2,6-9]

Clinically, UNT presents as clusters of fine, red to purple linear or spider-like vessels that blanch under pressure. These telangiectasias are typically asymptomatic, though some patients may have mild discomfort or cosmetic concerns.[1] Diagnosis is primarily clinical, based on the characteristic appearance and distribution of the lesions. Dermoscopy can assist in the evaluation by revealing a network of non-melanocytic red tortuous dilated capillaries.[10] Despite its benign nature, UNT can sometimes be mistaken for other vascular or dermatologic conditions, highlighting the importance of awareness and accurate diagnosis among dermatologists. Differential diagnoses include port-wine stains, spider angiomas, and other causes of unilateral vascular lesions. In this report, we present the first documented case of a Saudi pediatric patient diagnosed with unilateral nevoid telangiectasia, a condition previously unreported in Saudi Arabia.

Case Report

We report the case of a healthy 12-year-old girl who presented to the Dermatology outpatient clinic complaining of multiple red-violet cutaneous lesions on her right forearm. The lesions began at birth and have grown with her age. They were asymptomatic, with no previous bleeding, ulceration, infections, or change in size. In addition, her personal and family medical history was insignificant. The only concern was their appearance.

Clinical examination revealed multiple ill-defined violaceous blanchable macules that disappear with pressure (Figure 1), involving the lower lateral segment of the right dorsal forearm with some proximal extension (Figure 2). The rest of the examination was normal, with no mucosal lesions. We proposed a diagnosis of unilateral nevoid telangiectasia (UNT). Based on these findings, the patient was given follow-up visits to receive pulsed-dye laser sessions.

Discussion

Unilateral nevoid telangiectasia is a rare and uncommon cutaneous condition with an unknown etiology. It is clinically characterized by multiple blanchable red-violet telangiectasias mainly located unilaterally in a C3-T1 dermatomal pattern.[1,11] The pathophysiology of UNT remains incompletely understood, but several contributing factors have been identified, suggesting a multifactorial etiology.

Genetic predisposition plays a significant role in the development of UNT, which often appears to follow a dermatomal distribution and aligns with the patterns of skin development and innervation. This dermatomal presentation hints at possible mosaicism, where genetic mutations during embryogenesis could lead to localized vascular anomalies.[12] Cases of UNT have also been associated with other rare conditions, such as ipsilateral melorheostosis and vascular twin spotting, further indicating a possible role of somatic mosaicism.[13,14] These genetic changes might result in the abnormal proliferation or dilation of superficial capillaries and venules, manifesting as the characteristic telangiectasias seen in UNT.[12] Hormonal influences are crucial in the pathogenesis of UNT, which is more frequently diagnosed in females and often coincides with hormonal fluctuations such as puberty, pregnancy, and oral contraceptive use. Estrogen, in particular, increases the synthesis of vascular endothelial growth factor (VEGF) and other angiogenic factors, promoting blood vessel formation and dilation.[9]

Systemic conditions may also contribute to UNT development. Associations have been observed with systemic diseases like liver cirrhosis and malignancy, chronic alcoholism, hepatitis B and C virus infection, and hyperthyroidism.[2,6-9] These conditions may worsen vascular abnormalities by altering blood flow dynamics, affecting hormone metabolism, or directly impacting vascular integrity. Liver diseases can elevate circulating estrogen levels due to impaired metabolism, hence increasing the levels of VEGF, which potentially triggers telangiectatic changes in susceptible individuals.[9] Moreover, UNT has been observed in conjunction with neurological disorders in a subset of patients, suggesting a possible neurocutaneous link. A study involving eight patients with UNT reported hypoesthesia over the skin lesions in all patients. Additionally, cranial magnetic resonance imaging revealed subcortical hamartomatous lesions in one patient and demyelinized plaques in another. Neurological symptoms in these patients included mild mental retardation, dysarthric speech, hemiparesis, and dysmetria, indicating significant neurological involvement. In one case, UNT was accompanied by findings suggestive of multiple sclerosis.[15] A rare instance reported the occurrence of UNT in a healthy male following chemotherapy for Hodgkin's disease. This case suggests a potential new clinical association between chemotherapy and UNT.[16] In another instance reported, UNT occurred in a healthy male without any evident predisposing factors or lab abnormalities.[17] Our case revealed the occurrence of congenital UNT in a young girl, as the lesions have been present since birth. Treatment of UNT is often sought for cosmetic reasons, and several options are available. Pulsed dye laser (PDL) is the primary and effective therapeutic option that showed significant improvement in the appearance of telangiectasias with minimal adverse effects.[18,19]

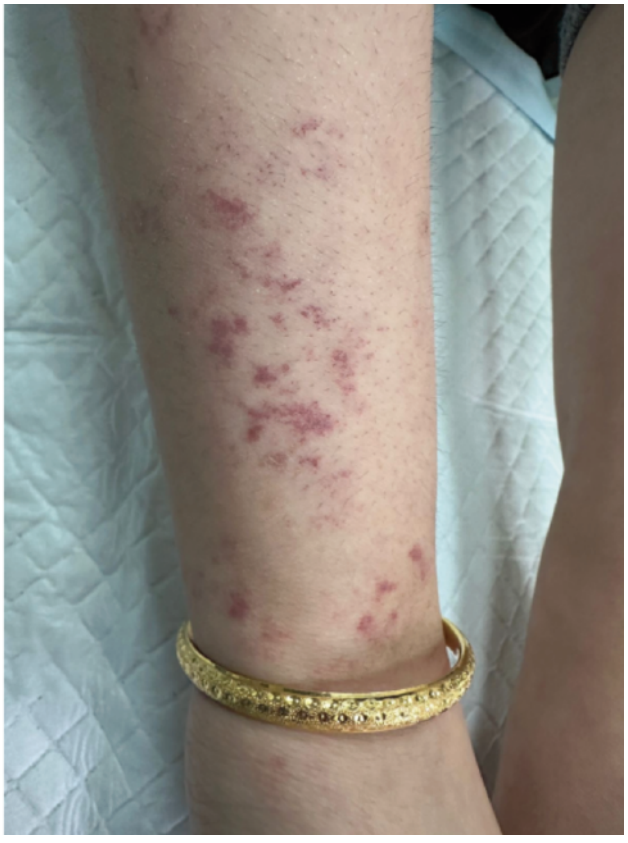

Figure 1. Multiple ill-defined violaceous macules.

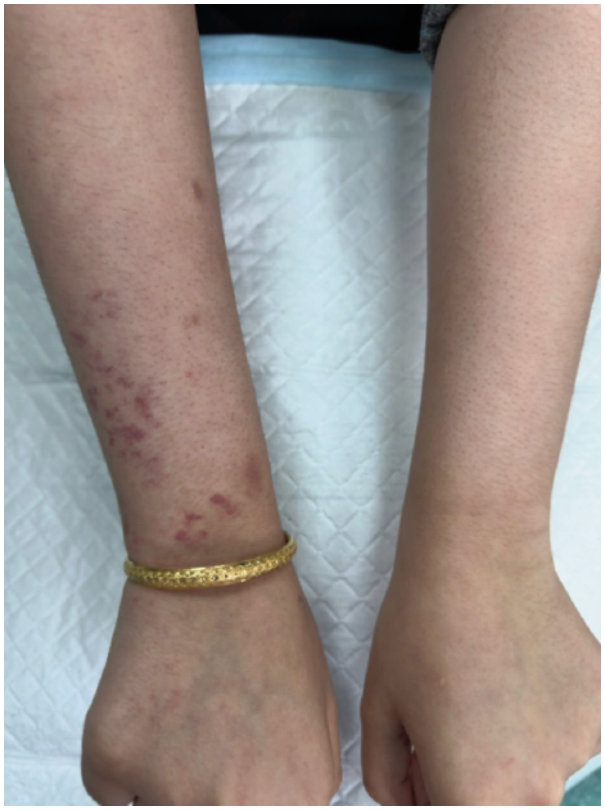

Figure 2. Unilateral nevoid telangiectasia involves the lower lateral segment of the forearm and extends proximally.

Conclusions

UNT is a complex and multifactorial vascular dermatosis. Genetic mosaicism likely sets the stage for this localized condition, which is then influenced by hormonal factors and potentially exacerbated by systemic conditions.

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