CASE REPORT

Unilateral Nevoid Telangiectasia: Case report and literature review

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ABSTRACT

Unilateral nevoid telangiectasia (UNT) is a benign, rare dermatosis, which primarily involves the face and neck on the trigeminal and cervical dermatomes (third and fourth).

It is characterized by telangiectatic, unilateral vascular lesions of linear, segmental or metameric distribution. It may be congenital or acquired, such as in the case of physiological or pathological estrogen excess presented in puberty, pregnancy or chronic liver disease. Such condition is related to an increase of estrogen and progesterone receptors in the involved skin.

A 22-year-old male patient presents with dermatosis on the right thigh's inner face, clinically and histologically compatible with UNT of rare localization.

INTRODUCTION

Unilateral nevoid telangiectasia (UNT) is a benign cutaneous vascular condition, characterized by the presence of multiple linear and non-coalescent arborizing telangiectasias of unilateral distribution, without a central vessel. Though it is most frequently located at the cephalic region, it may also manifest in the torso, limbs, and even in one half of the body. It may be congenital. However, it is more commonly acquired, which may be associated with a physiological or pathological hyperestrogenic state. The condition mainly manifests in fertile females.

Diagnosis of this entity derives from clinical-pathological correlation. Therapy implementation is not essential. Nonetheless, several alternatives are available for cases which require a cosmetic condition improvement.

A male patient presents with a UNT diagnosis after full physical examination of the skin and complementary exams. A brief review of this entity was performed.

CLINICAL CASE

The case of a 22-year-old male patient with no relevant family or personal medical history is presented. He refers to the presence of 8-month lesions on the right thigh's inner face which grew over time without presenting any symptoms. In addition, by increased heat, they become more evident.

At physical examination, multiple telangiectasias of segmental distribution were observed. These involved all of the right thigh's inner face, extending past the proximal third of the ipsilateral leg (Figure 1, 2, 3). These lesions were completely removed by vitropression.

Dermoscopy showed the presence of dilated and tortuous capillary vessels. (Figure 4).

Histopathological examination of one of the lesions was performed in conjunction with clinical diagnosis and presumptive dermoscopy of UNT. This revealed vasodilation without endothelial proliferation in papillary

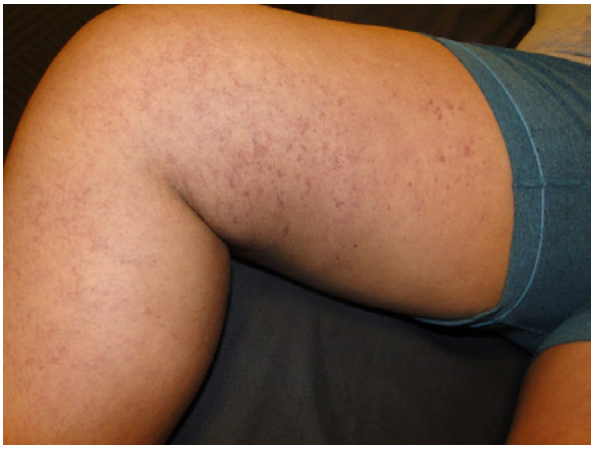

Fig 1. Multiple telangiectasias at right thigh's inner face and proximal third of the ipsilateral leg.

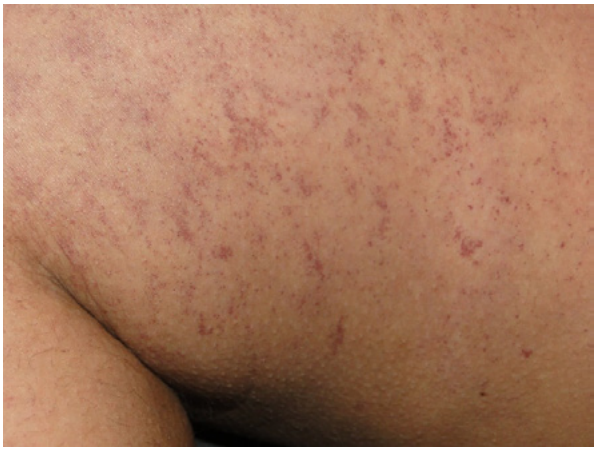

Fig 2. Arboriform telangiectasias without central vessel.

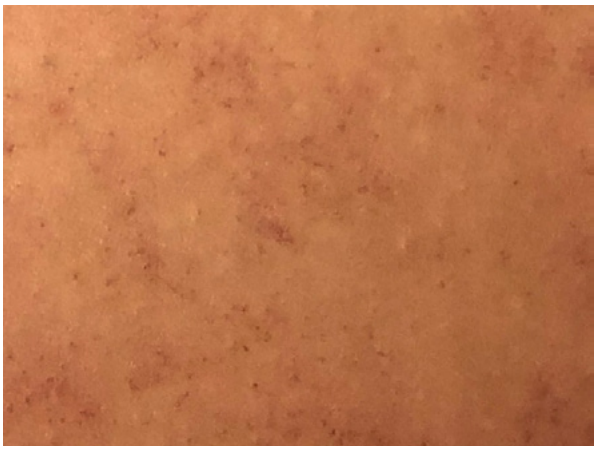

Fig 3.

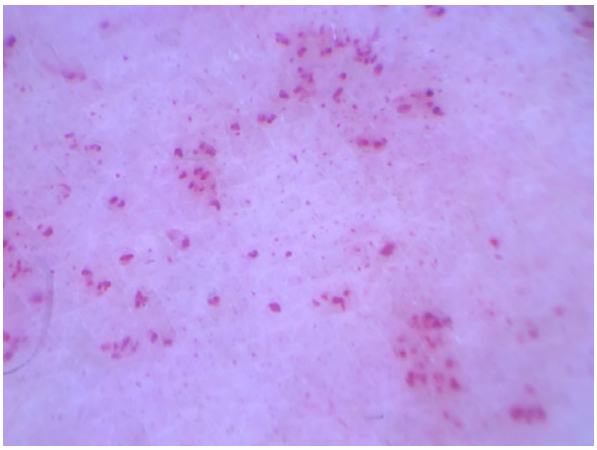

Fig 4. Dermoscopy: dilated and tortuous capillary vessels.

dermis, as well as findings which are compatible with the aforementioned entity (Figure 5).

Routine laboratory tests and the thyroid profile were classified as normal.

DISCUSSION

The disease was firstly described by Blaschkno in 1899. In 1931, Pautrier and Ullmo employed the term "acquired spider telangiectasia." Around 1964, Bowen published a case titled "essential progressive unilateral microtelangiectasia." The name "unilateral nevoid telangiectasia" was suggested by Selmanowitz for the first time in 1970. A case of "linear telangiectasia" was reported by Aram and Solomon that same year. In 1972, there were other cases, by Cunliffe and collaborators, denominated "unilateral spider nevus." Moreover, Koop-manns-van Dorp presented a case known as "unilateral telangiectasia."

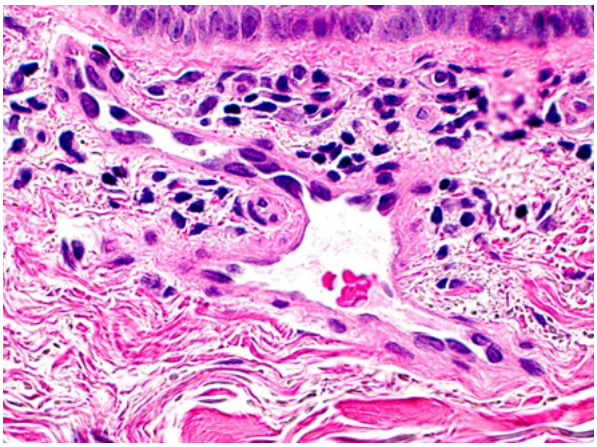

Fig 5. Dermis: dilated and congested capillary vessels (H/E 20x).

By 1978, Wilkin coined the term "unilateral dermatomal superficial telangiectasia."[2,3]

This condition may be congenital. However, it is more frequently acquired throughout a patient's life.

Congenital cases are rare, and they manifest during or after the neonatal period. They are more common amidst males.[3,4]

Acquired cases occur more frequently among fertile females. Nonetheless, they can be developed at any stage in life. They are commonly related to a physiological or pathological hyperestrogenic state.

Puberty, pregnancy and anovulation medication intake are recognized physiological causes of hyperestrogenism.[5,6]

Alcohol-related or infectious liver disease (HCV, HBV) and primitive or metastatic tumors of the liver classify within the condition's pathological causes.[3,7-9]

Medical literature presents UNT cases associated to hyperthyroidism10. However, not all cases reveal associated anomalies.[11,12]

Estrogenic action liberates nitric oxide, causing capillary vessels to enter a relaxed state. Such theory is supported by the predominance of teenage females and chronic liver disease patients presenting with this dermatosis. Nevertheless, the role of estrogen is not clear due to the lack of skin receptors and normal estrogen and progesterone levels in some UNT patients.[13]

In 2015, Happle reported a detailed capillary malformation classification, which included UNT; suggesting it may represent a nevus. In addition, he described two different types of UNT: the punctate and patchy types; erythematous macules distributed in a segmental pattern at one side of the body.[14]

Diagnosis is performed by clinical-pathological correlation, which reveals arborizing telangiectatic vascular spots, without a central vessel and variable number, size and shape.

Dermoscopy is an essential and helpful tool for diagnosis, revealing the presence of tortuous dilated capillary vessels distributed in a reticulated pattern, which is a distinctive feature which corroborates the differential diagnosis with other entities, such as the serpiginosum angioma.[15]

Histopathology showed numerous small-caliber dilated vessels in the superficial, mid-depth and deep dermis to a minor occurrence. They are produced by dilated post-capillary venules of the superficial horizontal plexus.[16,17]

Potential differential diagnoses are Hutchinson's angioma serpiginosum, hereditary hemorrhagic telangiectasia or Rendu-Osler-Weber syndrome, generalized essential telangiectasia, telangiectasia macularis eruptiva perstans, simple stellar angiomas and pigmented purpuric dermatosis.[8,18-20]

Occasionally, lesions involute spontaneously, as in pregnancy-related cases. They disappear after birth. However, in general, lesions are persistent, with no specific treatment.

Therapeutic alternatives for this solely cosmetic disorder include electrocoagulation, radiofrequency, cryosurgery or CO2 laser, argon, Nd: YAG laser, dye laser or intense pulsed light (IPL), which presents a variable response.[7,8,21-23]

CONCLUSIONS

This presentation aims to inform about the case of a rare entity manifested in a male patient, whose diagnosis refers to punctate acquired unilateral nevoid telangiectasia; a dermatosis which manifests more commonly in females. This anomaly presented with no associated condition. In addition, no treatment was undertaken. The case is currently undergoing observation

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