Unilateral nevoid telangiectesia: report of two pediatric cases

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Unilateral nevoid telangiectasia (UNT) is characterized by the dermatomal distribution of telangiectasia, which are distributed unilaterally and frequently found in C3-C4 or the trigeminal dermatomal areas. It may be congenital or acquired and has a 2:1 female: male ratio. Several theories where developed in order to explain its etiopathogenesis. The most widely accepted is the one which establishes its probable association with an increase in estrogen levels. Its incidence is unknown, although it's probably subdiagnosed and more frequent than previously recognized. We present two pediatric cases of UNT followed at our instution during the past year.

Key words: telangiectasia, unilateral, child, nevus, unilateral nevoid telangiectasia.

Unilateral nevoid telangiectasia (UNT) is a rare cutaneous disorder first described by Blaschko in 1899 and counted among the primary telangiectasias. UNT is characterized by superficial dermal telangiectasias that occur unilaterally in a dermatomal distrubution and commonly involve the trigeminal nerve as well as cervical and upper thoracic nerve distrubutions (C3-T2 dermatomal areas).[1]

Unilateral nevoid telangiectasia may be congenital or acquired. The rare congenital form occurs more frequently in males in an autosomal dominant fashion and appears during or shortly after the neonatal period. On the other hand, the acquired form occurs almost exclusively in young female patients associated with physiologic conditions; however, it may develop at any age.[2, 3]

There are slightly fewer than 100 reported cases of UNT in the literature available for review.[1] UNT may be more common than previously believed. Herein, we report two pediatric cases of UNT followed at our instution during the past year. This article underlines the importance of physicians awareness.

Case Reports

Case 1

A 8.5-year-old girl presented with a 1 month history of multiple, scattered, asymptomatic red spots, distributed in a linear and unilateral pattern that involved her left of face, neck, and upper arm (C6, C7, and C8 dermatomes). On closer inspection, blanchable telangiectasias were evident surrounded by a pale ring (Fig. 1). Apart from this, the patient was healthy. The patient showed no signs of puberty. No family history of any similar disease. Laboratory results revealed no abnormalities in complete blood cell count, serum chemistry panel and liver function test. Testosterone, progesterone and estrogen levels were within normal limits and antibodies to hepatitis B, and C were absent. A skin biopsy was performed, which revealed multiple, dilated vessels in the reticular dermis (Fig. 2). Immunohistochemical stains for ER and PR were negative. The histopathologic findings were consistent with UNT in the appropriate clinical setting.

Case 2

The 12-year-old boy visited our clinic on presentation of upper respiratory tract infection.

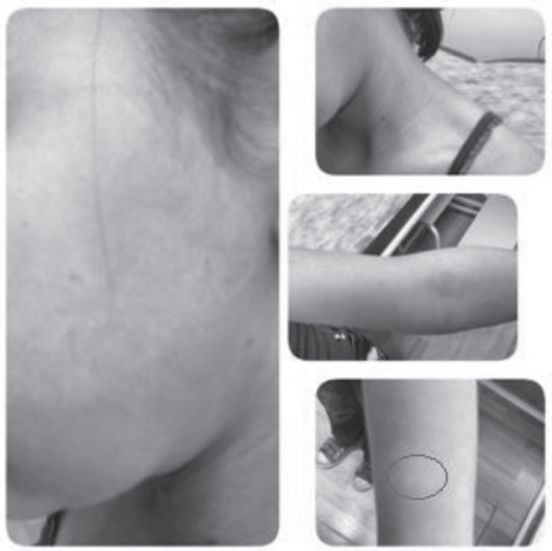

Fig. 1. Telangiectases distributed along face, neck, and C6-C7-C8 dermatomes. Charecteristic anemic halo (red ring).

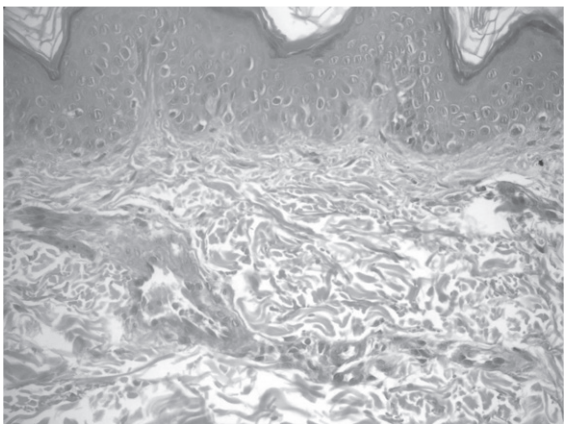

Fig. 2. Dilated dermal capillaries (HEx40).

Fig. 3. Close up of telangiectasias on left hand.

Careful physical examination revealed bright red macules in a blaschkoid distrubution involving the left dorsal hand (C6 dermatome). On closer inspection, blanchable telangiectasias were evident surrounded by a pale ring (Fig. 3). The patient showed no signs of puberty or feminization. His past medical history was not significant. Laboratory results revealed no abnormalities in complete blood cell count, serum chemistry panel and liver function test. Hormone levels were within normal limits and antibodies to hepatitis B, and C were absent. The patient and his parent refused skin biopsy.

Discussion

Blaschko's first description of UNT dates from 1899. Since then many alternative designations have been proposed. Nowadays, however the two terms most frequently used are "unilateral nevoid telangiectasia" and "unilateral dermatomal superficial telangiectasia," this last one described by Wilkin in 1978.[4] Although it is thought to be a rare condition with only fewer than 100 reported cases.[1, 5], we believe that it is more common. Because our patients discussed in this article were observed for a short period of time, the diagnosis requires careful evaluation of patient. Although diagnosis is fairly easy with the clinical findings, it may be missed when the lack of physician and patient awareness. Healthcare professionals should be aware of this condition, and the diagnosis of UNT must be kept in mind. As case reports of UNT increase, awareness among physicians is thought to increase as well.

The exact pathogenesis still remains unknown, although some authors believe in a strong association with a hyperestrogenic state. Endothelial cells possess estrogen receptors, and estrogens are felt to play a role in angiogenesis.[6] In some cases, an increased number of estrogen receptors are observed in the involved skin.[7] Indeed UNT is commonly reported in association with puberty, pregnancy, the use of oral contraceptives, hormonal therapy, chronic alcoholism with or without cirrhosis, chronic liver diseases, hepatitis B and C infections, portal hypertension, carcinoid syndrome with liver metastases, and hyperthyroidism. However, there have been reported cases in otherwise healthy individuals, especially male patients. Independently of the underlying mechanism,

there seems to be a somatic mosaicism, which becomes obvious in environments with excess of the feminine hormone. Other theories claim that hyperestrogenemic states or estrogen receptor abnormalities may not play a major role in the pathogenesis of UNT, especially in adult male patients.[8, 9] These theories propose hemodynamic disturbances, neural alterations, angiogenic factors, and aberrations in perivascular supportive connective tissue as causative agents.[4]

There are a few cases of UNT with serologic evidence of hepatitis C supporting the hormonal theory, which is based on hyperestrogenemic state, but this theory does not apply in all UNT cases.[8, 10] Cases of UNT occurring in the presence of normal serum estrogens and liver function are documented in literature.[2, 3, 11, 12] Tok et al.[13] reported a case of UNT related to pregnancy with no estrogen and progesterone receptors, and suggested that the receptor assays were not sensitive enough to detect estrogen and progesterone levels in the skin when compared with the breast tissue. Our case 1 showed no increased ER and PR in the biopsy of lesional skin using immunoperoxidase staining methods.

Unilateral nevoid telangiectasia arise in linear patterns, can be either small or big in size, and pale with applied pressure. Pale ring, referred as an anemic halo, may be observed surrounding the telangiectasias. Predilection sites are the face, neck, shoulder-arm region, and thorax.[14] Some authors have noted that the distrubution is not dermatomal, but rather follows Blaschko's lines, along which cells migrate during embryonic development. It is thought that a localized increase in estrogen receptors caused by a chromosomal mosaicism, that is unmasked at times of relative estrogen excess, accounts for the distribution that follows Blaschko's lines. Jucas et al.[11] proposed that an as-yet-unknown epidermal angiogenic factor may be the causal agent of the telangiectasia development in UNT. Further studies are needed to better understand this unique condition. Histologically, there are multiple, dilated, thin-walled vessels lined by plump endothelial cells in the papillary and upper reticular dermis, characteristic of UNT.[8] No signs of endothelial cell proliferation or neo-angiogenesis are seen. Without endothelial proliferation UNT has been reported to be associated with hyperestrogenemic states such as puberty, pregnancy and alcoholism. This is explained by the hypothesis that a humoral agent, probably estrogen, stimulates the end target organs that are distributed congenitally in a dermatomal pattern in order to produce telangiectasia.[15]

Other laboratory findings recently described are the functional impairments at the blood vessel level in laser Doppler imaging. This method is able to record subclinical functional defects before they become clinically evident. Lastly, capillaroscopy showed morphologically altered vascular structures with megacapillaries in the fingers' nail grooves.[2]

Unilateral nevoid telangiectasia typically persists but rarely, in acquired cases, resolves spontaneously if the eliciting factor is removed. Treatment begins with cosmetic camouflage. The use of pulsed dye laser has been a useful alternative in aesthetic improvement of the condition;[1, 2] however, none of the patients or parents accepted to be submitted to such treatment, advocating the lack of symptoms of the disease.

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