CASE 2

Mohammad Javad Nazemi Tabrizi, MD[1]
Reza Mahmoud Robati, MD[2]
Saeed Amini, MD[1]
Nima Sarrafi-Rad, MD[2]

1- Department of Dermatology, Tehran University of Medical Sciences, Razi Hospital, Tehran, Iran
2- Skin Research Center, Shahid Beheshti University, M.C. Shohada-e Tajrish Hospital, Tehran, Iran

Corresponding author:
Reza Mahmoud Robati, MD
Skin Research Center, Shahid Beheshti University, M.C. Shohada-e Tajrish Hospital, Shahrdari St, 19899341 48, Tehran, Iran
E-mail: rmrobati@gmail.com

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Case

A one-year-old otherwise healthy girl presented with a progressively enlarging patch on the lateral aspect of her right ankle. Her parents stated that the lesion was not remarkable at birth but became more apparent with aging. There was no history of trauma or bleeding and family history for similar lesions was unremarkable.

Physical examination showed an erythematous patch of superficial telangiectases in a linear distribution with a nevoid pattern on her right foot (figure 1). Other parts of the lower limb and general examination were unremarkable.

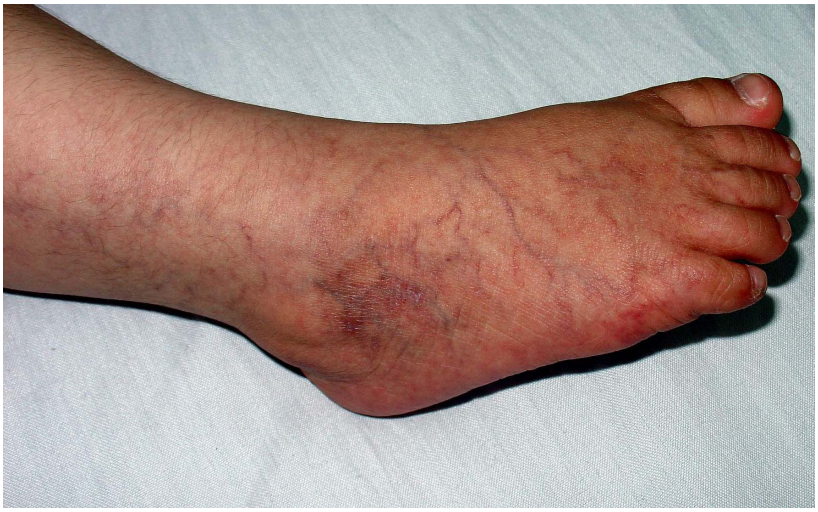

Figure 1

What is your diagnosis?

Move on to next page for the answer and discussion.

DIAGNOSIS: UNILATERAL NEVOID TELANGIECTASIA SYNDROME

Discussion

Telangiectasia (visible dilated blood vessel measuring 0.1–1 mm in diameter) denotes a condition characterized by abnormal, permanent dilatation of end vessels, venules mainly, but also occasionally of capillaries and arterioles of the subpapillary plexus[1].

The common telangiectases can be explained by abnormalities in this organization and ultrastructure rather than by neovascularization or random anastomoses[2]. Telangiectasia may be secondary or primary. Secondary telangiectasia occurs following actinic damage, radiotherapy and trauma or in association with varicose veins, connective tissue disorders, acne rosacea, autoimmune disease and possibly microbial diseases[3].

Primary telangiectases include the following diseases: unilateral nevoid telangiectasia, generalized essential telangiectasia, hereditary hemorrhagic telangiectasia, hereditary benign telangiectasia, and ataxia-telangiectasia.

The unilateral nevoid telangiectasia syndrome (UNTS) is a rare, usually acquired disease counted among the primary telangiectases. UNTS is characterized by unilateral dermatomal distribution of telangiectases. The dermatomes most frequently involved are the trigeminal and the third and fourth cervical nerves. The lesions may be congenital or acquired. Increased numbers of receptors for estrogen and progesterone have been reported in these dilated capillaries compared with normal skin.

Acquired UNTS occurs almost exclusively in states of relative estrogen excess, such as that in pregnancy, puberty, chronic liver disease in alcoholism, and hepatitis C[4-7].

Distribution following the lines of Blaschko in some reports, suggests that a postzygotic somatic mutation leads to a distinct cell population in the affected site (mosaicism). It is slightly more frequent in the right side. Polymorphic light eruption has been described confined to the area of acquired nevoid telangiectasia[8].

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