

Acquired Unilateral Nevoid Telangiectasia Syndrome: A Case Report and Review of Literature

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Abstract

Unilateral nevoid telangiectasia syndrome (UNTS) can be classified into two major categories: congenital and acquired. There have been reports showing an increase in skin estrogen and progesterone receptors in patients with this syndrome. We report a male patient with acquired unilateral nevoid telangiectasia syndrome without any associated physiological or pathological conditions. (*Iran J Dermatol* 2009;12:67-68)

Keywords: acquired, telangiectasia, nevoid, unilateral

Case Report

A 41-year-old man was referred to the outpatient clinic complaining about asymptomatic erythematous eruptions from 4 years ago. He had no history of liver diseases or alcohol abuse or drug consumption. Physical examination was normal. Dermatological examination revealed a patch of telangiectases confined to the left lower eyelid area (Figure 1). The rest of the skin, scalp, genitalia, nails and mucous membranes were without lesions and intact. The results of laboratory investigations, including whole blood count, sedimentation rate, urinalysis, blood glucose, renal and liver function tests, and TSH were within normal limits. Tests for hepatitis B surface antigen (HbsAg), anti-HIV, anti-hepatitis C virus (anti-HCV) gave negative results. A skin punch biopsy specimen taken from the involved area showed dilated small vessels in the dermis without proliferation of endothelial cells.

Discussion

Unilateral nevoid telangiectasia syndrome (UNTS) is a congenital or acquired disorder. Several reports have suggested an increase in skin

estrogen and progesterone receptors in UNTS. The acquired form is associated with physiological conditions such as pregnancy, puberty or hormonal therapy. However, the condition is also described in cirrhosis, alcoholism without cirrhosis, carcinoma metastatic to liver and in hepatitis B infection, hepatitis C infections and hyperthyroidism ^{1,8}.

Lucas et al. reported a case of a pre-pubertal



Figure 1. A patch of telangiectases confined to the left lower eyelid

boy with UNTS. He was the first case without any evidence of increased estrogen. Jucas suggested that the causative factors of this condition were not hormonal ⁴.

In 1997, the first healthy adult male with acquired UNTS was reported who had no demonstrable underlying diseases, alcohol abuse or physiological conditions causing hormonal changes. In addition, estrogen and progesterone receptor studies yielded negative results in this case ⁹. The patient presented with UNTS characterized by no underlying disease or estrogen receptor abnormalities. They suggested that hyperestrogenemic states or estrogen receptor abnormalities may not play a major role in the pathogenesis of UNTS, especially in adult male patients. This case report casts doubt on the commonly held view that unilateral nevoid telangiectasia syndrome is an estrogen-sensitive nevoid anomaly ⁹.

Tok J et al, reported a case of acquired unilateral nevoid telangiectasia syndrome related to pregnancy with no estrogen and progesterone receptors ¹⁰.

Sanchez Conejo-Mir J et al. presented four observations of unilateral nevoid telangiectasia syndrome, pointing out its common cervicothoracic distribution and its relation with hyper-estrogenic conditions, both physiological (puberty, pregnancy, use of contraceptive pills), and pathologic (post-alcoholic hepatic cirrhosis) ¹¹.

The pathogenesis of UNTS has not been completely understood. Moreover, it remains unclear why telangiectasia occurs in strict unilateral distribution. This phenomenon might be explained with abnormal estrogen-sensitive cells that are congenitally distributed in dermatomal pattern, and stimulated by a humoral agent, probably estrogen. Additionally, it was suggested that estrogen might stimulate an angiogenic factor that mediates ectatic formation of vessels ¹².

Nonetheless, more studies are needed to demonstrate the correlation of systemic estrogen levels with the presence of estrogen and progesterone receptors in the skin.

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