



Acquired unilateral nevoid telangiectasia in a healthy men*

Telangiectasia nevoide unilateral adquirida em homem hígido

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Abstract: Unilateral nevoid telangiectasia is a rare vascular dermatosis, characterized by areas of superficial telangiectases distributed in a linear unilateral pattern. It was described in 1899 by Zeisler and Blascko. Several theories were developed in order to explain its etiopathogenesis. The most widely accepted is the one which establishes its probable association with an increase in the estrogen levels. There are two types: congenital and acquired. The latter is associated with hepatopathies in male patients. The acquired form is rarely observed in healthy men, with a few cases reported in the medical literature, and its etiology is unknown. This study reports the case of a healthy young man with acquired unilateral nevoid telangiectasia, without any comorbidities, clinical and /or laboratory findings indicative of hyperestrogenism.

Keywords: Estrogens; Liver Diseases; Telangiectasis

Resumo: A Telangiectasia Nevoide Unilateral é uma dermatose vascular rara, caracterizada por áreas de telangiectasia superficial, em uma distribuição linear unilateral, descrita, em 1899, por Zeisler e Blaschko. Diversas teorias foram desenvolvidas para explicar sua etiopatogenia, sendo a provável (e a mais fundamentada) a relação com elevação dos níveis de estrogênio. Há duas formas: a congênita e a adquirida. Esta, geralmente, se relaciona a hepatopatias, em pacientes do sexo masculino. Em homens hígidos, a ocorrência da forma adquirida é rara, com poucos relatos na literatura, sendo sua causa desconhecida. Este trabalho tem como objetivo relatar um caso de telangiectasia nevoide unilateral adquirida em homem jovem, sem comorbidades, alterações clínicas e/ou laboratoriais sugestivas de hiperestrogenismo.

Palavras-chave: Estrogênios; Hepatopatias; Telangiectasia

INTRODUÇÃO

Unilateral nevoid telangiectasia is a rare vascular dermatosis, characterized by areas of superficial telangiectases distributed in a linear unilateral pattern, described in 1899 by Zeisler e Blaschko.¹ Since then, less than a 100 cases have been described in the medical literature². Several theories were developed in order to explain its etiopathogenesis. The most widely accepted is the one which establishes its probable association with an

increase in the estrogen levels. There are two types: congenital and acquired. The first is prevalent in men through dominant autosomal inheritance and the second in women, in association with physiological and pathological conditions of hyperestrogenism.¹ The acquired type, in male patients, is generally related to hepatopathies. The occurrence of this type among healthy men is rare and its cause remains unknown.

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CASE REPORT

Male patient, aged 19, with history of asymptomatic violaceous macula growing progressively in the cervical and pectoral regions as well as on the right upper limb. The lesion had been progressing for 2 years. The patient denied comorbidities, the use of medicines or family history of liver, cutaneous or vascular diseases. During the exam, erythematous-violaceous maculae were observed in the areas mentioned above (Figures 1-3). There were not palpable lesions, hypertrophy of the limb, changes in the skin appendages or in the local temperature

CBC (complete blood count), hepatic function tests and serum sex hormones were normal. Serology for hepatitis, HIV and syphilis were negative. Ecodoppler and capillaroscopy of the right upper limb did not show abnormalities. Histopathology on hematoxylin and eosin staining showed normal epidermis with dilated thin-walled capillaries on the superficial dermis (Figure 4).

DISCUSSION

Unilateral nevoid telangiectasia is a rare disease and its etiopathogenesis is still unclear. The main hypothesis is based on the probable relation with an increase in the estrogen levels due to a higher prevalence of the disease in states of physiological hyperestrogenism (such as pregnancy and puberty) and pathological hyperestrogenism (such as hepatopathie). Wilkin and col. refer to increase in cellular receptors for estrogen and progesterone in the lesions^{1,3}. It is believed that the unilaterality of the lesion is due to the presence of estrogen-sensitive cells distributed congenitally along a dermatome path.⁴ Some authors propose that the segmental



FIGURE 2: Erythematous-violaceous maculae on the right pectoral region

nature of the disease results from somatic mosaicism which is evident in the presence of hyperestrogenism.⁵ The association with other lesions resulting from mosaicism, such as inflammatory linear verrucous epidermal nevus strengthens this hypothesis.⁶ It could be included among the other mechanisms studied, hemodynamic disturbances, neurological changes, influence of angiogenic factors and changes in the connective tissue¹. Sharma and col. call the attention for the presence of cutaneous telangiectasia concomitant with retinal telangiectasia, subglossal, oral and gastric as an incomplete manifestation of Hereditary Hemorrhagic Telangiectasia.⁷ However, the absence of mucosal lesions and family history in the majority of the patients refute this possibility.⁷ The disease can be congenital or acquired. The first type which prevails among men is transmitted by dominant autosomal



FIGURE 1: Erythematous-violaceous maculae on the right hand



FIGURE 3: Erythematous-violaceous maculae on the cervical and pectoral regions and on the right upper limb

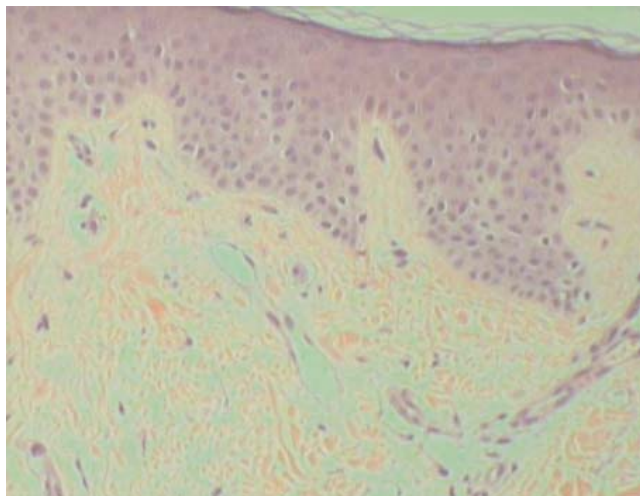


FIGURE 4: Histopathological exam showing normal epidermis with dilated thin-walled capillaries in the superficial dermis

inheritance with possible influence of maternal estrogen. The second type occurs more frequently in women, in association with hyperestrogenism, being rare before puberty¹. In 1997 Karabudak and col. described the first acquired case in a young man without comorbidities.⁴ Bilateral cutaneous lesions, oral and gastric were reported in a few number of

patients.⁵ Normally the lesions are persistent but, in some acquired cases, they involute spontaneously. This is more frequent in cases associated with pregnancy and, in the postpartum period, there is spontaneous involution of the lesions⁸. The diagnosis can be confirmed by the observation, in histopathologic exams, of dilated capillaries and thin walls in the papillary dermis. As for the Serpiginous Angioma, main differential diagnosis, there is proliferation of dilated capillaries with thickened walls in the papillary dermis.⁸ Unilateral nevoid telangiectasia can be detected still in the sub-clinical phase through laser Doppler, method that allows the analysis of microcirculation. There might be hyperfusion of the affected areas, evidenced by erythema, besides increase in the caliber of capillaries, characteristic also present in scleroderma. Ungual capillaroscopy can evidence the presence of giant capillaries, also seen in some collagenolysis.⁹ Such alterations were not seen in our patient. Treatment with *pulsed dye laser* is highly used in nevoid telangiectasia. However, there are reports in the medical literature of important hyperpigmentation and hypopigmentation after the treatment, which turn to be an aggravator since the stigma of the disease is also aesthetic.⁷ □

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