

Acquired Unilateral Nevoid Telangiectasia Syndrome

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We describe a man with acquired unilateral nevoid telangiectasia syndrome, in whom no underlying disease, alcohol abuse or physiological conditions causing hormonal changes are demonstrable. To our knowledge, this is the first case of acquired unilateral nevoid telangiectasia syndrome seen in a healthy adult male and not associated with a hyperestrogenaemic state and estrogen receptor abnormality. This case report casts doubt on the commonly held view that unilateral nevoid telangiectasia syndrome is an estrogen-sensitive nevoid anomaly.

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Unilateral nevoid telangiectasia syndrome may be subdivided into two major categories: congenital and acquired (estrogen-related, alcohol/hepatic-related) (1). The condition, characterized by dermatomally distributed fine telangiectases, is usually asymptomatic and is seen in hyperestrogenaemic states (2). Elevation of both estrogen and progesterone receptors has been demonstrated in the lesional skin of a pregnant woman with unilateral nevoid telangiectasia syndrome (3). The condition is also associated with chronic liver diseases, metastatic liver diseases and alcohol-related cirrhosis (4, 5).

We here present a case of acquired unilateral nevoid telangiectasia syndrome with no associated physiologic or pathologic state.

CASE REPORT

A 40-year-old man with a good health record was referred to our outpatient clinic with the complaint of slightly erythematous asymptomatic eruption of 7 years' duration. He had no history of liver disease or alcohol abuse. General physical examination revealed no abnormality. On dermatologic examination, there were fine, blanching, thread-like telangiectases unilaterally localized to the right side of the neck and right upper chest wall in a dermatomal distribution (C3-T1) (Fig. 1). The rest of the skin, genitalia, scalp, nails and mucous membranes were clear.

A biopsy specimen of the skin from the right upper chest wall showed ectatic vessels in the dermis (Fig. 2). Immunohistochemical studies were performed by using an avidin-biotin complex method. Monoclonal antibodies used included: pS2 estrogen inducible protein (BioGenex, San Ramon, Ca) and progesterone receptor protein (Novocastra, Newcastle, UK). In the skin biopsy specimen taken from the involved area, the endothelial cells lining the telangiectatic vessels did not show immunoreactivity for estrogen or progesterone receptors. Uninvolved tissue obtained from a symmetrical biopsy on the opposite side of the upper chest wall was also negative for both receptors.

The results of the laboratory investigations, including whole blood count, urinalysis, sedimentation rate, blood glucose, renal function tests, blood electrolytes, total protein, albumin, globulin, protein electrophoresis, immunoglobulins, prothrombin time, partial throm-

boplastin time and hormone analyses (FSH, LH, prolactin, estradiol, 17-OH-progesterone, beta-HCG, sex hormone binding globulin, cortisole, DHEA-SO₄, free-testosterone), were within normal limits. Liver function tests revealed the following values: AST, 23 U/l (normal 8-40 U/l); ALT, 28 U/l (normal 5-40 U/l); alkaline phosphatase, 86 U/l (normal 41-117 U/l); LDH, 289 U/l (normal 180-390 U/l); direct bilirubin, 0.1 mg/dl (normal 0.0-0.2 mg/dl); indirect bilirubin,

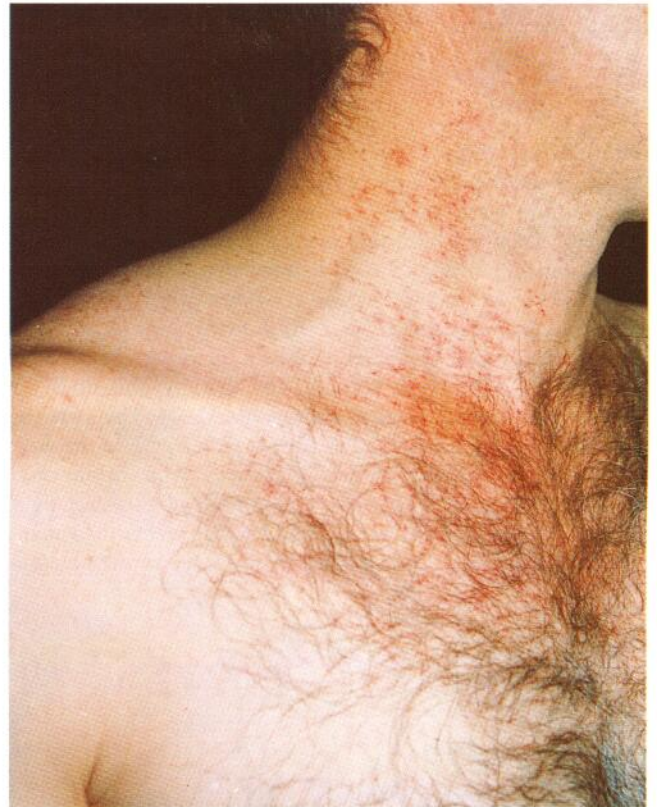


Fig. 1. Unilateral dermatomal distribution of telangiectases.

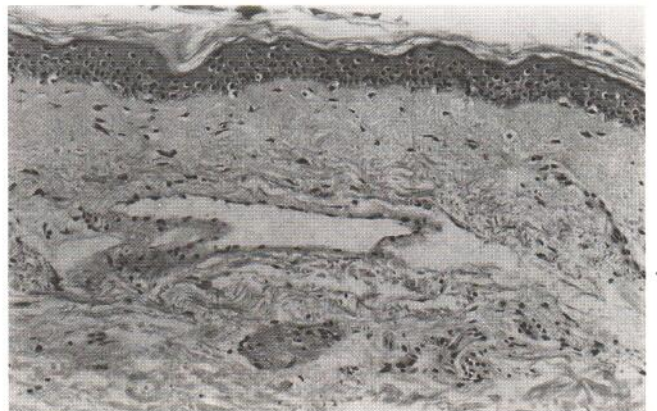


Fig. 2. A dilated vessel in the dermis (H & E).

0.4 mg/dl (normal 0.2–0.8 mg/dl). Tests for HBsAg, ANA and VDRL gave negative results. No pathologic finding was reported in abdominal ultrasonography and abdominal computerized tomography.

DISCUSSION

Unilateral nevoid telangiectasia syndrome is characterized by unilaterally located telangiectatic lesions. It is most often found in the C3–T1 dermatomes (2, 6). Unilateral nevoid telangiectasia syndrome, first described by Zeissler in 1922 (7), was reviewed and classified by Wilkin as congenital and acquired (1). Data obtained from the cases reported so far document that acquired unilateral nevoid telangiectasia syndrome is commonly associated with hyperestrogenaemic conditions such as pregnancy, puberty, liver disease and hormonal therapy (1, 3, 6).

The pathogenesis of unilateral nevoid telangiectasia syndrome is not completely understood. Some authors suggest that the telangiectatic lesions result from a localized estrogen receptor abnormality. According to this theory, abnormal target end organs, congenitally distributed in a dermatomal pattern, are stimulated by a humoral agent, probably estrogen (3, 5, 6, 8). It has also been suggested that estrogen may stimulate an angiogenic factor, which in turn mediates the formation of telangiectases (9). Controversies still exist on the theory that unilateral nevoid telangiectasia syndrome is seen under the conditions of hyperestrogenaemia and is a localized estrogen receptor abnormality, in other words, a dermatomal developmental defect in which estrogen receptors are increased.

In 1979, Jucas et al. reported a case of acquired unilateral nevoid telangiectasia syndrome in a prepubertal boy. He was the first man in whom unilateral nevoid telangiectasia syndrome developed without evidence of increased estrogen. In the article, it was suggested that the factors responsible for this condition were not hormonal (10). But in this case, hormone receptor analysis of the skin was not performed. So, authors supporting the hormonal theory objected to the suggestion made by Jucas et al., asserting that there is some estrogen production in the prepubertal period and that, in addition, the exquisite dermatomal sensitivity of this patient's cutaneous vessels may have resulted in telangiectasia formation (3).

A male patient who developed unilateral nevoid telangiectasia syndrome following liver metastases from a carcinoid tumour of the stomach was presented by Beacham & Kurgansky in 1991. It was reported that the involved skin did not show estrogen receptor activity but showed progesterone receptor activity (5). In 1994, Tok et al. reported a female patient with unilateral nevoid telangiectasia syndrome related to pregnancy with no estrogen and progesterone receptors (6).

In our case of unilateral nevoid telangiectasia syndrome, the patient was absolutely healthy and no associated pathological condition was detected. His telangiectatic lesions developed when he was 33 years old and biopsy specimens did not reveal estrogen or progesterone receptors.

In the light of the results of the clinical studies outlined above, we suggest that a hormonal theory based on hyperestrogenaemic states and estrogen receptor abnormalities is not operative in all cases of unilateral nevoid telangiectasia syndrome.

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