Original Article

Unilateral nevoid telangiectasia.
Report of four cases

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Abstract

Unilateral nevoid telangiectasia is an uncommon congenital or acquired condition characterized by small, unilateral vascular dilations in a linear, segmentary or metameric distribution, usually located on the face and cervical areas, although they may also be found in other locations.
We report four female patients between 22 and 49 years old, who share clinical and histological features of unilateral nevoid telangiectasia. One of the cases showed association with secondary biliary cirrhosis, and another patient had flares during pregnancies (Dermatol Argent 2009;15(1):44-48).

Key words: *unilateral nevoid telangiectasia, telangiectases.*

Introduction

Unilateral nevoid telangiectasia (UNT) is a dermatosis characterized by the presence of multiple linear telangiectasias of arborizing aspect, without central vessel, non-confluent, and unilateral in distribution. Preferably, it appears on the head, but may also be located on trunk, limbs, and even one side of the body.
It may be congenital, or more frequently acquired, and in the latter case it usually associates with physiological or pathological states of hypoestrogenism. It predominates in females, and in the fertile years of the life.
Diagnosis of this entity is eminently clinical, since the histopathology is not pathognomonic.
No therapeutic implementation is indispensable, but several alternatives are available for the cases where improvement is required, from a cosmetic point of view.
We present four female patients, where UNT diagnosis was reached based on complete physical skin examination and supplementary studies. This entity was briefly reviewed.

Clinical cases

Case 1

A 33-year-old female, without a relevant pathological history, who consulted for lip dryness of about two years evolution.
The physical examination showed scaling of the lower lip semimucosa compatible with exfoliative cheilitis. When examining the perioral skin, segmentary distribution telangiectasias were identified in that area, on the right half of the nose, the right malar area, and the right retroauricular area (**Figure 1**).

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She also showed similar lesions on the back of the right hand.

With presumptive clinical diagnosis of UNT, histopathological examination was performed of the lesions, which resulted in vascular dilation without endothelial proliferation in papilar and intermediate dermis; these findings are compatible with this entity. In addition, routine laboratory tests were requested, with liver function tests, thyroid profile, and dosage of estrogens and progesterone, with values within normal ranges.

The patient was cosmetically concerned, and treatment was started. Two sessions of intense pulsed light were applied (filter: 550 nm; fluency: 42 J; spot: 8 × 35 mm, triple shots: T3.5 msec D10 msec) and CO_2 laser 1 watt on some punctual lesions, with moderate improvement after the 2 sessions.

Case 2

A 22-year-old female, without a relevant pathological history, consulted for the presence of facial lesions. She referred gradual appearance of lesions between 13 and 15 years of age, and subsequent stabilization.

The physical examination showed segmentary distribution telangiectasias on the right half of the face with nose, malar and lip dominance (**Figure 2**).

With presumptive clinical diagnosis of UNT, a skin biopsy was performed and resulted compatible with this dermatosis.

Supplementary studies (laboratory routine, thyroid profile, dosage of estrogens and progesterone) gave results within normal ranges.

Case 3

A 28-year-old female, 20-week pregnant was hospitalizad in the Obstetrics Department of our hospital due to extramembranous pregnancy. Interconsultation was requested due to lesions distributed on the left side of the face, chest, and upper limb. She referred that they had appeared during adolescence and showed exacerbations in pregnancies, and no other relevant history.

The physical examination showed segmentary distribution telangiectasias involving the whole face, left chest area, and homolateral upper limb (**Figure 3**).

The histological study was compatible with UNT (**Figure 4**). Laboratory routine and thyroid profiles were reported within normal ranges.

Case 4

A 49-year-old female presented with personal history of psoriasis, secondary biliary cirrhosis and esophageal varices. Complete evaluation showed segmentary distribution telangiectasias located on the right side of the neck, of 6 years evolution (**Figure 5**).

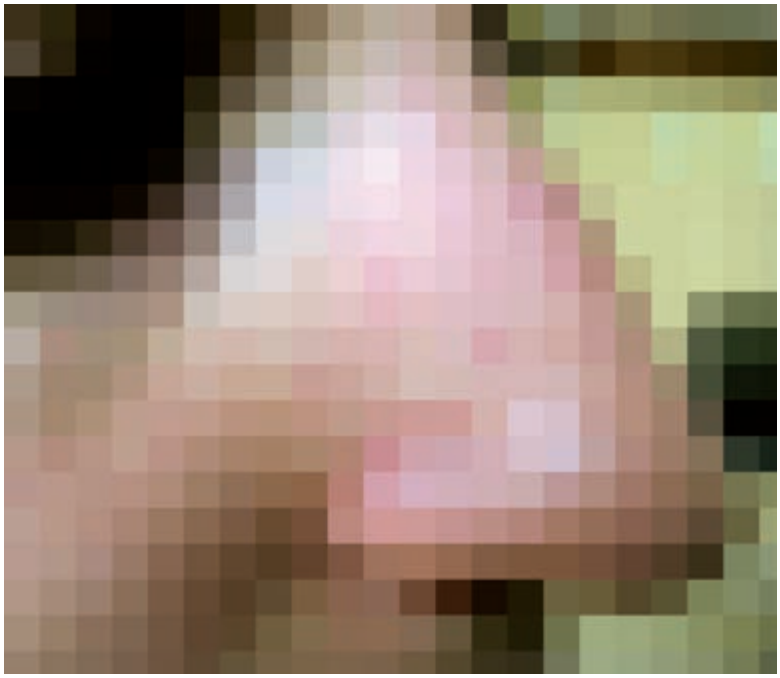

Figure 1. *Case 1. Telangiectasias located on the right side of the nose.*

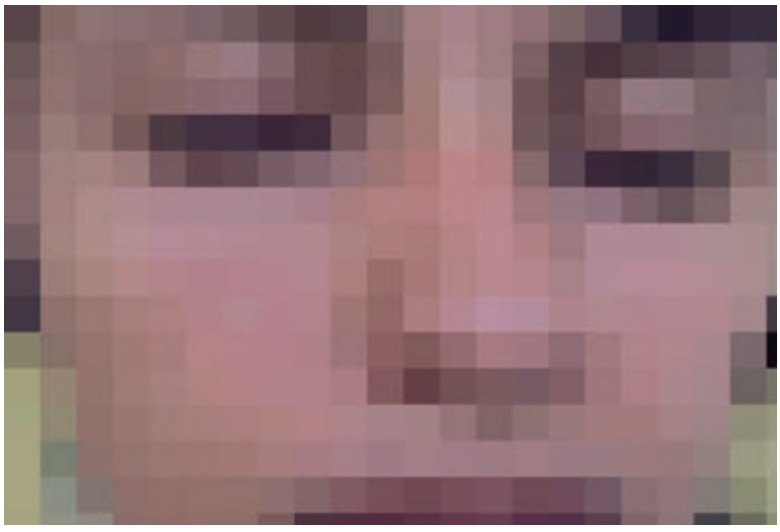

Figure 2. *Case 2. Telangiectasias on the right half of the face.*

With presumptive clinical diagnosis of UNT, a punch biopsy was performed, resulting in ectasia and vascular congestion compatible with this dermatosis. Routine laboratory tests resulted in alterations of liver function tests: TB 3.9 mg/dl; DB 3 mg/dl; ALP 985 U/l (NV 50-250); GGT 349 U/l (NV 7-50); GOT 138 U/l (NV 0-38); GPT 86 U/l (NV 0-41); and the rest, including thyroid profile, within normal ranges.

Discussion

UNT was described initially by Blaschko in 1899, but only in 1970 had Selmanowitz coined the current name.

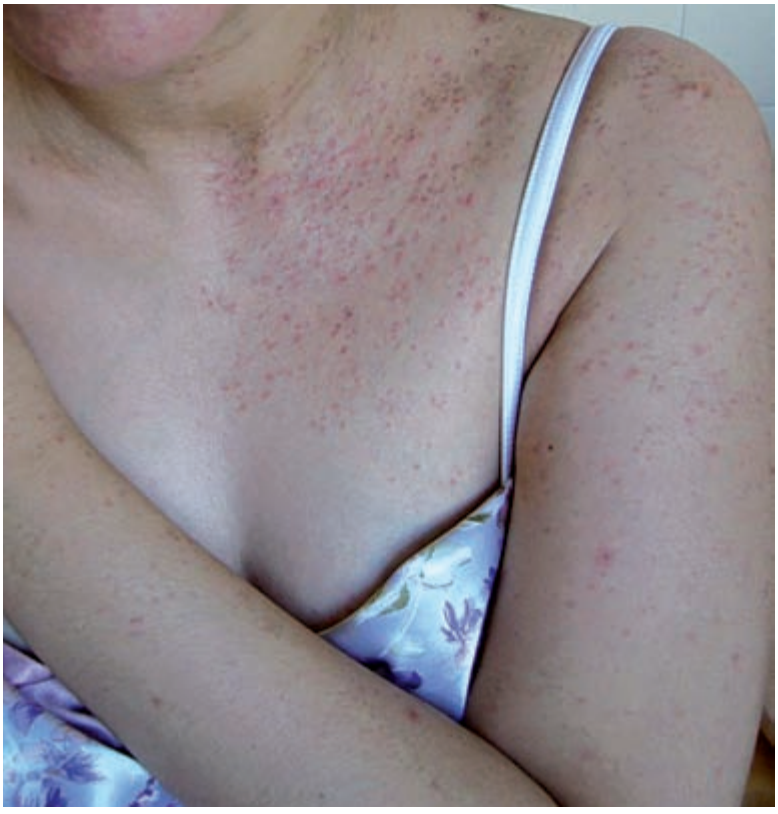

Figure 3. *Case 3. Multiple telangiectasias on the chest area and left upper limb.*

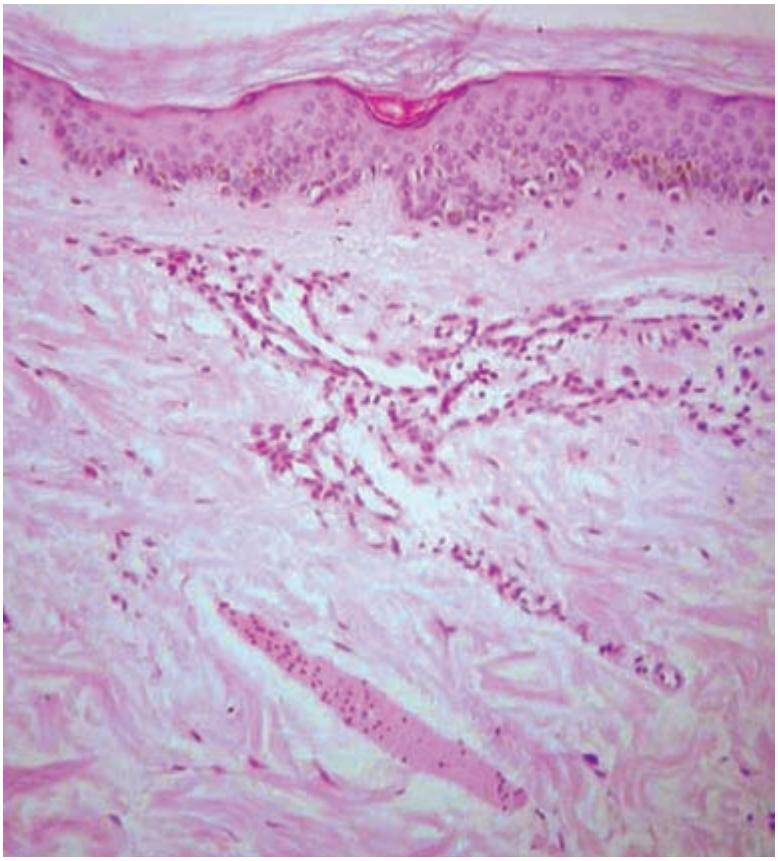

Figure 4. *Case 3. Dilated vessels without endothelial proliferation in dermis (H-E).*

It may be congenital or, more frequently, acquired during lifetime. Congenital cases are rare and occur during or after the perinatal period; they appear more commonly in males.[1,2] The acquired forms are much more frequent, and prefer females during the fertile period of life, but may develop at any age. They usually relate to states of physiologic or pathologic hyperestrogenism (**Table 1**). Mentioned among the physiological causes of hyperestrogenism are puberty, use of contraceptives, and pregnancy.[3,4] In our first three patients, the disease onset occurred during puberty; the third patient additionally referred lesion exacerbation during pregnancies. The other 2 had not yet been pregnant. Mentioned among the pathological causes are alcohol or infectious hepatopathy (HCV, HBV), and primary or metastatic liver tumors.[1,5-7] Our fourth patient showed association with secondary biliary cirrhosis. The literature includes reports of UNT associated with hyperthyroidism,[8] while other cases showed no associated abnormalities.[9,10]

The etiology is not yet well established, but some research reports increased levels of estrogen and/or progesterone receptors in the involved skin of some patients. Other hypothesis indicate abnormalities of these receptors, and that circulating estrogens may stimulate production of telangiectasias.[2,11-13] Kreft et al.[14] propose subclinical function defects of the cutaneous microvasculature. Some authors believe that in some cases the entity may be related to mosaicisms due to somatic mutations of cell populations during embrionary development, and that in situations with hyperestrogenism the cutaneous lesions may become visible; but the high levels of estrogen should not be the primary cause of the disease.[1]

Clinically, the disease is characterized by the presence of arborizing and non-confluent telangiectasias, without central vessel. They are unilateral, and their distribution may be linear, segmentary or metameric. Preferably, they appear on the head and neck, but are also found in other body areas, and even on one half of the body. Dermatomas C3 to D1 are the most commonly involved sites.[2,6,15-17] Some authors noticed that, more than metameric, the distribution follows Blaschko's lines, a situation which may be related to cell migration during embrionary development.[2]

TABLE 1. CAUSES OF HYPERESTROGENISM.

Physiological	Pathological
Puberty	Alcoholic hepatopathy
Contraceptives	Infectious hepatopathy (HCV, HBV)
Pregnancy	Primitive and metastatic liver tumors

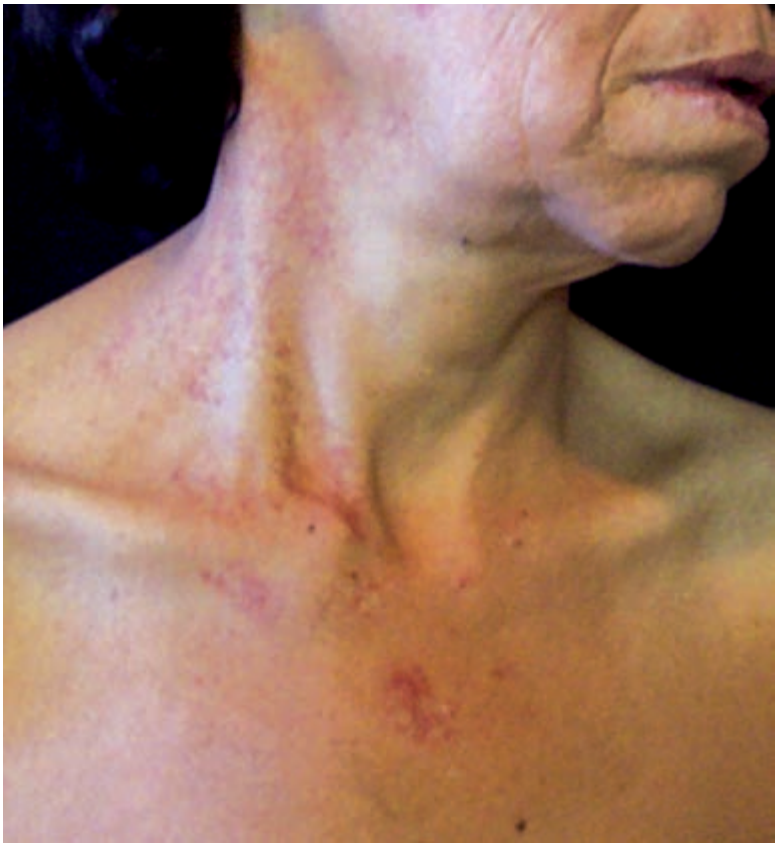

Figure 5. *Case 4. Telangiectasias on the right half of the neck.*

Lesions are asymptomatic, and without mucosal or systemic involvement. The evolution is benign, and in most cases the course is chronic and persistent. Some cases of spontaneous post-partum regressions have been reported. Histopathology is not characteristic, and is usually not necessary. It may show dilated vessels in papillary and intermediate dermis, without endothelial proliferation.[6,17]

Possible differential diagnosis (**Table 2**) are Hutchinson's angioma serpinginosum, familial hemorrhagic telangiectasia or Rendu-Osler-Weber syndrome, generalizad essential telangiectasia, persistent eruptive macular telangiectasia, and simple stellate angioma.[6,13,18,19]

Therapeutic alternatives for this merely cosmetic disorder include electrocoagulation, radiofrequency, cryosurgery, or CO_2 laser, argon, Nd:YAG, dye laser, or intense pulsed light.[5,6,20,21] Intense pulsed light is one of the most often indicated therapeutic methods for telangiectasias, together with pulsed dye laser; laser Nd:YAG may be indicated in the case of greater vessels. We prefer the association with CO_2 laser, because we noticed greater response in combining methods, and without adverse effects or definite sequelae. No treatment was applied in the last three patients.

Conclusion

The interest of this presentation is to communicate four cases of this rare entity, and remark the importance of a complete clinical examination of patients' skin, which may lead us to diagnosis of disorders which are not the reason of consultation.

TABLE 2. UNT DIFFERENTIAL DIAGNOSIS.[6,13,18,19]

Simple stellate angioma: scarce in number or isolated, not adopting metameric distribution. If numerous, a liver disorder must be suspected.
Hutchinson's angioma serpiginosum: predominant in women (90%), it appears in childhood. Lesions are punctiform and aggregate with serpiginous disposition. Slightly papulous surface. Preferred location on pelvic girdle, and progressive growth.
Familial hemorrhagic telangiectasia, or Rendu-Osler-Weber syndrome: hereditary, early manifestation by recurrent epistaxis. Location of telangiectasias is perinasal and peribuccal areas. Associated to internal organ involvement.
Generalized essential telangiectasia: diffuse form, more frequent in lower limbs, may also appear coincident with pregnancy or puberty. Predominant in females. Generally linear telangiectasias.
Persisting eruptive macular telangiectasia: form of cutaneous mastocytosis associated with arborizing disposition telangiectasias and erythematous or hyperpigmented macules, accompanied by itching.

We understand that the therapy, merely cosmetic, represents a challenge. We report our experience with intense pulsed light associated with CO_2 laser.

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