

## CIRCUMSCRIBED NEVOID ANGIOKERATOMA

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### Summary

**Introduction.** *Angiokeratomas are telangiectatic papules histologically characterised by vascular ectasias of the superficial dermis and covered by a hyperkeratotic epidermis. Circumscribed nevoid angiokeratoma (ACN) is a rare congenital vascular malformation.*

**Clinical case.** *We report the case of a 20-year-old patient presenting from birth with multiple clustered hyperkeratotic papules located on the anterior thorax. The dermatological examination identified multiple dark red hyperkeratotic papules, varying in size between 2 and 3 mm, located on top a slightly edematous plaque located on the anterior thorax. Histopathological examination revealed a orthokeratotic epidermis and multiple dilated capillaries in the superficial dermis.*

**Discussions.** *ACN was described in 1890, being more common in females. It is characterized by multiple small-sized angiokeratomas located on a limb segment, buttocks, neck and trunk. The lesions may follow a linear distribution. The treatment consists of surgical excision, cyotherapy, electrosurgery and lasertherapy.*

**Keywords:** *angiokeratoma, hyperkeratosis, ectasia.*

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### Introduction

The term „angiokeratoma” originates in the greek words “angio” (vessels) and „kera” (cornified), while as an indicator of the tumoral character of the lesions, the suffix „oma” is added [1]. The angiokeratomas are telangiectatic papules which present with a common histopathological pattern, consisting of ectatic vessels located in the superficial dermis and covered by a hyperkeratotic epidermis [2]. The lesions are usually benign, but in some cases may be an indicator of serious conditions, such as Fabry disease. Circumscribed nevoid angiokeratoma (ACN) is a congenital malformation and represents the rarest of the 5 subtypes of angiokeratomas.

### Clinical case

We present the case of a 20-year-old male patient who was consulted for the existence of multiple clustered keratotic papules located on the anterior thorax. The lesions appeared at birth, enlarging with age. Dermatological examination identified a slightly tumefied plaque covered with multiple telangiectatic keratotic papules, ranging in size between 2 and 3 mm, of dark-red color and located on the anterior thorax [Fig.1]. General examination didn't reveal any hypertrophic or hypotrophic degeneration of the underlying soft tissues or bones. Histopathological examination revealed a hyperkeratotic and orthokeratotic epidermis and dilated capillaries in the superficial dermis. The profound dermis and

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Figure 1. Multiple angiokeratomas located on the anterior thorax

the hypodermis were normal. Therefore, the diagnosis of angiokeratoma was established. Blood examinations and cardiac ultrasound were within limits. Medullary MRI didn't reveal any additional vascular malformations.

Based on the clustered distribution of the angiokeratomas and the laboratory tests within limits, the diagnosis of ACN was established.

## Discussions

Angiokeratomas are formed due to the dilation of the capillaries located in the papillary dermis through three mechanisms: venous hyperpression, vascular malformations or fragility [2]. The exact epidermal reaction is unknown. The neviform disposition of angiokeratomas suggest the possibility of a mosaicism due to an autosomal genetic mutation [3,4], the same mechanism being involved in the appearance of unilateral nevoid teleangiectasias as well [5].

Angiokeratomas are clinically characterized by keratotic teleangiectatic papules of various sizes, which partially blanch when performing diascopy. Depending on their size, distribution, extension or additional systemic signs, they can be divided in 5 categories:

- solitary or multiple angiokeratomas, which represent the most common subtype
- angiokeratoma of the fingers (Mibelli)
- angiokeratoma of the scrotum or the vulva (Fordyce)

- ACN
- angiokeratoma corporis difusus (Fabry disease)

The histopathological examination reveals vascular ectasias in the papillary dermis associated with an acanthotic and hyperkeratotic epidermis. The angiokeratoma can be an isolated tumor or can be associated with vascular malformations and the atrophy or hypertrophy of the underlying soft tissues and bones in Cobb syndrome (where vascular lesions are associated with arteriovenous medullar malformations in the same individual) [6,7], highlighting in such cases the need for additional investigations.

Dermoscopy shows a number of specific features for angiokeratomas: the presence of red ovoid gaps (histologically corresponding to dilated vascular spaces in the superficial dermis) [8], purplish and black ovoid gaps (caused by partial or complete vascular thrombosis) [9], white veil (corresponding to acanthosis or hyperkeratosis) [10], hemorrhagic crusts secondary to bleeding, peripheral erythema caused by inflammation and extravasation of erythrocytes in the papillary dermis [10].

ACN is a rare congenital malformation, first described in 1890. It mainly affects women, with a male/female sex ratio = 1/3 [11]. ACN is represented by multiple lesions with a diameter ranging from 1 to 5 mm, clustered on one or more plaques located mainly on a limb segment or on a buttock, but also on the neck, trunk, face and even on the penis [12] and on the tongue [13]. There is also the possibility of a linear distribution, in such cases the lesions are following the Blaschko lines [3].

The differential diagnosis of ACN is made with Hutchinson's serpiginous angioma, verrucous angioma, superficial lymphangioma, Klippel-Trenaunay syndrome and melanoma. Hutchinson's serpiginous angioma doesn't present with hyperkeratosis and isn't encountered since birth. Verrucous angioma is distinguished by the presence of a proliferation in the deep dermis and hypodermis, with the angiomatous phase preceding the appearance of verruco-keratotic lesions by a few years. Superficial lymphangioma is either congenital or appears in the first years of life and it's clinically characterized by multiple clear liquid content pseudo-vesicles with a diameter ranging from 1

to 5 mm, clustered in bouquets or irregular plaquards. Klippel-Trenaunay syndrome is a rare congenital disease, characterized by the unilateral presence of extensive plane angiomas associated with venous and lymphatic malformations. Melanoma, especially cases presenting with satellite metastases that easily bleed and show discoloration, can be confused with angiokeratoma.

The treatment consists of surgical excision with histopathological examination for limited lesions (11). Other therapeutical options are cryotherapy, electrosurgery and lasertherapy (erbium, diode and CO<sub>2</sub>), especially for extensive lesions [14, 15]. Recurrences are rare. In conclusion, by a proper understanding of this disease, the diagnosis of ACN can be clinically established, without the need for complementary explorations.

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Conflict of interest  
NONE DECLARED

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