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UNILATERAL EXTENDED LINEAR NAEVUS VERRUCOSUS (NEVUS UNIUS LATERIS) – FIRST CASE REPORTED IN ROMANIA

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Abstract: *One of the most interesting and certainly the most unusual of the varieties of nevi is the so-called naevus unius lateris or unilateral extended linear naevus verrucosus. It is a disembryoplasia or verrucous congenital tumor having a unilateral or nearly unilateral distribution in linear streaks or bands, following the long axis of the limbs or extending transversely around the trunk. This unique disease is so striking that it is readily recognized as a distinct clinic-pathologic entity. We present a clinical case of a 10 year old patient. The peculiarity of this case resume in the rarity and special clinical form and in that, it is the first clinical case published in Romania, at 100 years after the first photodocumented case was published worldwide.*

Key words: *naevus verrucosus, disembryoplasia.*

1. Introduction

The verrucous epidermal nevus is a disembryoplasia characterized by the excessive development of epidermal structures especially on the surface. The appearance of extended forms located on the half body area, with the inclusion of the trunk and limbs is extremely rare, and is called nevus unius lateris. When this nevus covers a large portion of the body, it can be described as a systemic verrucous nevus. The first described case, as well as the naming of nevus unius lateris is connected to von Baerensprung in 1863. [17] Similar cases were published by Galloway in 1902 [3], Reckzek in 1903 [14] and Weber in 1911 [18]. Kudish, in 1914 [8], presented the first case of pigmented nevus lateris, and Sibley, in 1914, presents the first systemic or bilateral case [15]. The first photodocumented case of linear verrucous nevus extended on the half body area is from 1915, published by Corbett (Fig. 1) [2]. The rate of occurrence of the disease is unknown, there have been 200 such cases published worldwide so far.

2. Clinical case

We present the case of a young patient of 10 years of age, presenting the symptoms of linear verrucous nevus with striations and stripes which covers the whole right half body area, with the lesions ending suddenly in mid-body area (Fig. 2, 3). Beside the mentioned unaesthetic

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appearance, the patient does not exhibit other subjective symptoms. The routine laboratory investigations, imagistic (ultrasonography, radiology, CT), and others (EEG, EKG), performed in order to investigate possible comorbidities associated with this disembryoplasia, were within normal limits. The possible treatments which were recommended, also with a disappointing effect, were refused by the parents.

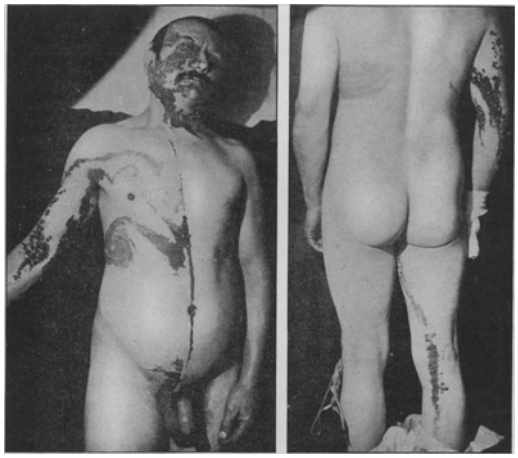

Fig. 1. *The first photodocumented case of linear verrucous nevus extended on the half body published by Corbett in 1915 [2]*

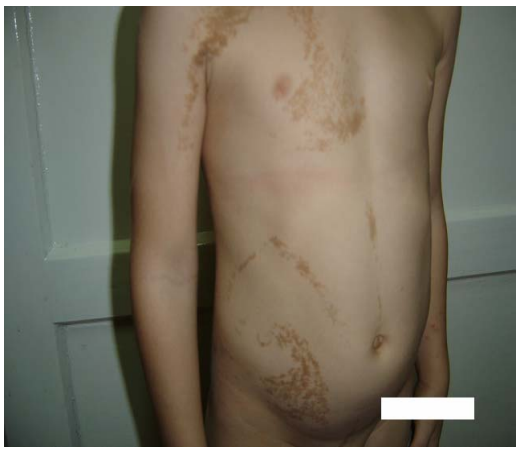

Fig. 2. *Unilateral extended linear naevus verrucosus - anterior aspect*

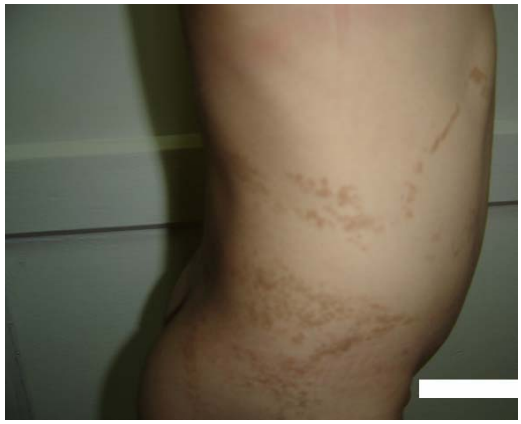

Fig. 3. *Unilateral extended linear naevus verrucosus - lateral aspect*

3. Discussion

The most common clinical aspect consists of a unique plaque of a variable size, having a surface covered with dry and rough hyperkeratotic deposits, separated by depressions and trenches deep on the surface. The color may be dark or pigmented, grey-brown or black. Their most frequent location is on the trunk or limbs, with a frequent topographic placement in striations or bands. They may also take on other patterns, such as: undulating, angular, S-shaped, water jet-like [13]. The sudden stopping of the lesion at the median line is an important sign of the nevoid origin of the lesion. [9] The nevus appears at birth or only in the first years of life, and persists throughout the life. This unique disease is so striking, being easily recognizable, as a distinct clinicopathological entity [4]. The lesions are not usually accompanied by inflammation or pruritus but sometimes they may appear [10]. They may also degenerate in very seldom cases.

The histopathological examination reveals the existence of ortokeratotic hyperkeratosis, associated with hyperacantosis and papilomatosis. The granulous layer is thickened in an irregular

pattern. The upper limit of the malpighian layer, with the granulous layer and the inferior part of cornous layer, form the so-called sawteeth aspect. The basal layer of epidermis may be hyperpigmented [12]. In rare cases the disease has been associated with other comorbidities such as: disturbances of the nervous system, diencephalic syndrome or cranial tumors [5], [6], [11].

As differential diagnoses we may discuss conditions such as: sebaceous nevus, linear syringoadenomatosis, linear lichen plan, localized scleroderma, pityriasis rubra pilaris, psoriasis vulgar, etc. [1], [7].

The treatment is disappointing. Partial results are obtained with systemic etretrinatum, topical tretinonum, 5-fluroruracil and calcipotriolum. Surgical treatment is applied only in unaesthetic cases [16].

4. Conclusion

Nevus verrucous unius lateris is a rare dysembrioplasia, with typical clinical aspect and easy diagnosis. The particularity of this condition results from the rarity and special clinical form, being the first such case published in Romania, presented at 100 years after the first photodocumented case in the world.

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