Case report

POROKERATOSIS LINEARIS
A variant of linear epidermal nevus with cornoid lamellae?

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ABSTRACT

An 18-month-old girl presented with a linear verrucous lesion on the right half of the chest following the curved path of lines of Blaschko. A skin biopsy specimen showed features of epidermal nevus with cornoid lamellae. Linear porokeratosis and linear epidermal nevus can be similar both clinically and histologically. In particular, the histologic phenomenon of cornoid lamellae-thought to be pathognomonic for all forms of porokeratosis-have also been described in epidermal nevi (porokeratotic epidermal nevi). Linear porokeratosis and porokeratotic linear epidermal nevus may represent variants of the same disorder.

KEY WORDS
Linear porokeratosis, Cornoid lamella, Linear epidermal nevus, Porokeratotic epidermal nevus

INTRODUCTION

Porokeratosis was first described by the Italian dermatologist Vittorio Mibelli in 1893 (16). It is characterized clinically by solitary or multiple annular lesions with hyperkeratotic border and atrophic centre most commonly localized on the limbs, face and genitalia. A generalized form of porokeratosis was described in 1967 by Chernosky and Freeman (6) under the term "disseminated superficial actinic porokeratosis" (DSAP).

Several other clinical variants have been described in addition to the original picture reported by Mibelli, mainly characterized by the distribution of the lesions (i.e. zosteriform, neviform, or others) (7,8,9,10,14,18,20,21) (Tab. 1).

This report describes an 18-month-old girl with a linear verrucous lesion on the chest clinically suggestive of linear epidermal nevus.

CASE REPORT

An 18-month-old female child presented with numerous tiny, yellow-brownish, verrucous papules in a linear configuration on the right side of the chest (Fig. 1 and 2). The papules followed the curved path of Blaschko's lines. The lesion was first observed about two weeks after birth as "roughening of the skin", and subsequently developed over several months to the actual size. No medical advice was

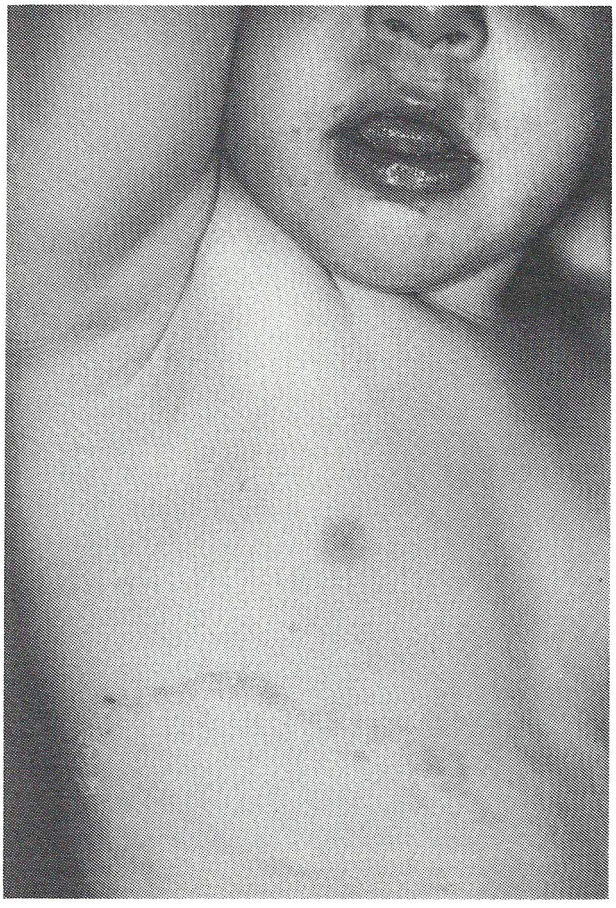

Figure 1
Band-like configuration of papules following the lines of Blaschko on the right half of the chest.

seeked until now. Familial history was negative and the patient was in good general health.

A biopsy specimen was obtained under local anesthesia. Histology showed acanthosis of the epidermis with numerous epidermal invaginations displaying parakeratotic plugs (so called "cornoid lamellae") (Fig. 3 and 4).

Based on clinical and histologic features a diagnosis of linear porokeratotic epidermal nevus was made.

After detailed information about the nature of the lesion, the parents of the patient refused any further treatment.

DISCUSSION

Differential diagnosis of linear dermatoses in childhood include a wide spectrum of diseases (Tab. 2). Despite similar clinical appearance, their characteristic histological features usually allow a specific diagnosis (1). The only exception might be the histopathological differentiation between linear

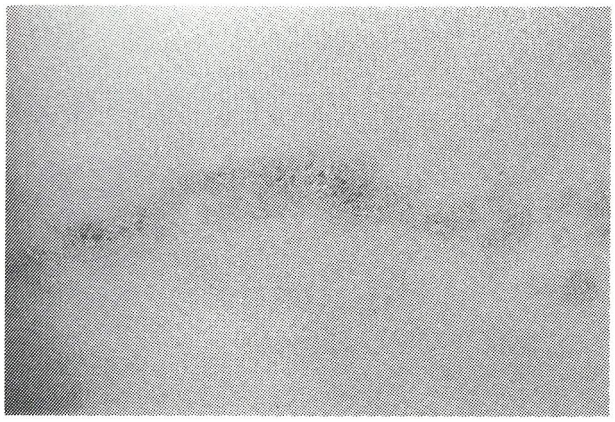

Figure 2.
Verrucous yellow-brownish pin-point sized papules.

epidermal nevus and linear porokeratosis.

Linear epidermal nevi are commonly single and unilateral extending down an extremity as a band of varying width or coursing transversely over the trunk sometimes following the lines of Blaschko. They may be bilateral or even distributed over most of the body, often associated with disorders of the central nervous system and skeletal anomalies (19). The lesion are firm, horny or wartlike, their colour ranging from yellow to grey, brown and black. Histologically hyperkeratosis, acanthosis and papillomatosis are the predominant features. The granular layer is variably thickened or thinned and spotty parakeratosis may occur.

Inflammatory linear verrucous epidermal nevi (ILVEN) are mainly observed on the lower left limb of female patients,

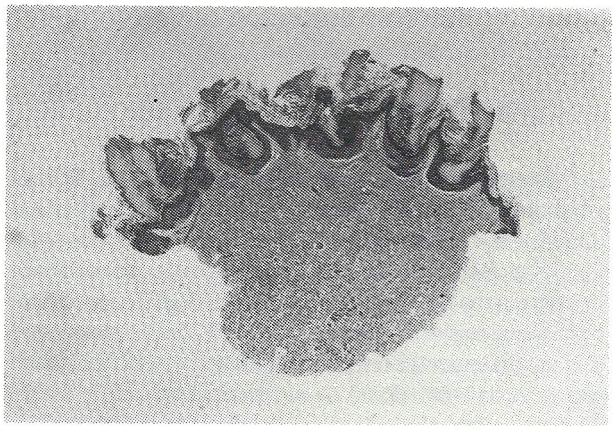

Figure 3.
Akanthotic epidermis with several parakeratotic plugs in epidermal invaginations.

Table 1. Types of porokeratosis and their features

AUTHOR	TYPE	CLINICAL FEATURES	LOCALIZATION
Mibelli 1893		single or few anular lesions with hyperkeratotic border and atrophic centre	limbs, face, genitalia
Freund 1934	linear	linear configurated small papules	unilateral
Rhabari 1977	punctate	pin-point sized follicular keratotic papules	disseminated
Chernosky Freeman 1967	disseminted superficial actinic porokeratosis	multiple uniformly sized (5 mm) lesions with slightly raised border in symmetrical distribution	sun exposed areas, especially extremities
Guss et al. 1971	porokeratosis palmaris plantaris et disseminata	multiple annular lesions with hyperkeratotic border and central atrophy	palmoplantar
Goldner 1971	zosteriform	multiple anular lesions with hyperkeratotic border and central atrophy	corresponding to a nervous territory
McMillan 1976	linear with giant cornoid lamella	small circular plaques, sharply rising edges, central keratinous horn	-
Schramm Bork 1982	neviform	(=syn. for zosteriform)	-
Strani et al. 1983	zoniform	(=syn. for zosteriform)	-

comprising eczematous or psoriasiform papules in linear configuration. The histopathologic findings of ILVEN are characterized by parakeratosis,focal spongiosis, exocytosis of lymphocytes, papillomatosis and a mild perivascular lymphocytic infiltrate in the upper dermis.

Lichen striatus - like ILVEN - is also a gynecotropic disease most commonly located on the upper extremities. It is characterized clinically by discrete, reddish-brown, scaly, slightly verrucous papules. Histological features show a sparse lymphohistiocytic perivascular infiltrate, slight epidermal hyperplasia, necrotic keratinocytes and multiple foci of hypogranulosis overlayed by relatively short, broad zones of parakeratosis.

The linear variant of lichen planus is rare, but could be a clinical pitfall disguising linear porokeratosis of Mibelli. Sometimes lichen planus is spread over a whole limb or even appears in segmental configuration with shiny flat-topped-papules. The histological changes of lichen planus are characterized by a lichenoid lymphoid infiltrate with melanophages, vacuolar alteration at the dermoepithelial

Table 2.

Linear Dermatoses of Childhood
Linear epidermal nevus
Inflammatory linear verrucous epidermal nevus (ILVEN)
Lichen striatus
Linear lichen ruber
Linear psoriasis
Linear porokeratosis

interface, Civatte bodies, irregular epidermal hyperplasia with jagged sawtooth appearance of the rete ridges and hypergranulosis as well as compact orthokeratosis.

Furthermore linear psoriasis can be regarded as an important differential diagnosis of linear porokeratosis of Mibelli. This unusual type of psoriasis shows small erythematosquamous papules in linear bands and may represent a Köbner reaction. Essential histologic features of psoriasis include confluent parakeratosis, hypogranulosis or absence of the granular layer, prominent psoriasiform epidermal hyperplasia with thin dermal papillae, dilated tortuous capillaries in the papillary dermis and sparse lymphohistiocytic infiltrates around the blood vessels.

The least common denominator of all variants of porokeratosis is the presence of the so called "cornoid lamella", which can be regarded as a special identification mark for this group of diseases. Cornoid lamella has been interpreted by Wade and Ackerman in 1980 as a histological reaction

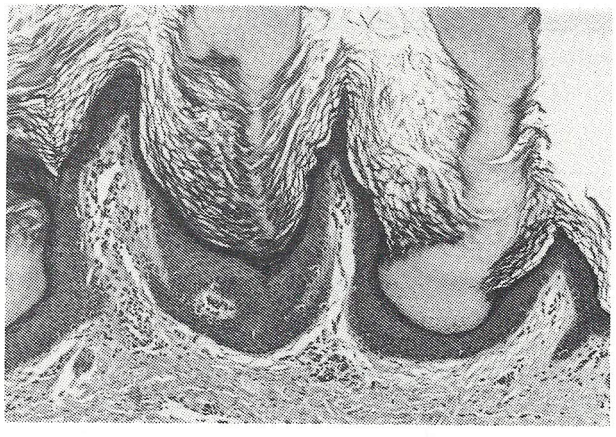

Figure 4.
Higher magnification showing a stereotypical cornoid lamella.

Table 3.

Conditions other than porokeratosis in which cornoid lamellation has been found (according to Wade and Ackerman, 1980) (23).
Solitary inflammatory lesion
Seborrhoic keratosis
Scar
Verruca vulgaris
Milia
Solar keratosis
Squamous cell carcinoma
Basal cell carcinoma

pattern that reflects disordered progression of epidermal cells to cornification (23). According to these authors, however, the presence of cornoid lamellae does not necessarily mean that the condition actually represents porokeratosis. In fact, cornoid lamellae may be found in several other inflammatory, hyperplastic and neoplastic conditions (Tab. 3). In addition, a variant of epidermal nevus with cornoid lamellae was described by Su in 1982 under the name "porokeratotic epidermal nevus" (22).

In 1901 Blaschko discovered the standard pattern of embryonal cell development with the conception of "nevus lines"(3). These S-shaped lines obviously represent the clonal growth of embryonal cell populations in an antero-lateral direction. Disturbances within the embryogenetic evolution are visualized by morphologic alterations following the lines of Blaschko. Modern genetics tried to explain this phenomenon either as a result of an early somatic mutation or of gametic half-chromatid mutations (11,12). The clinical appearance of our patient with verrucous yellow-brownish papules in a linear configuration following the lines of Blaschko on the right side of the chest was strongly suggestive of a linear epidermal nevus. Linear porokeratosis, however,has not yet been observed in a pattern following the lines of Blaschko. Therefore-despite the histological presence of multiple cornoid lamellae - the verrucous skin lesion in our patient are probably best interpreted as a linear epidermal nevus with cornoid lamellatio or as porokeratotic epidermal nevus.

The cause and mechanism of cornoid lamellatio is still unknown. There are, however, reports on activation of genetically determined aberrant cell populations by immunosuppression producing clinical symptoms of porokeratosis (15, 17). A hypothesis on the histogenesis of porokeratosis of Mibelli postulates that the lesion originates from a clone of keratinocytes which are represented by the vacuolated cells underlying the parakeratotic plug (6, 7).

All variants of porokeratosis are regarded as skin diseases

with potential for malignant transformation (2, 4, 5, 13). As it is not clear whether porokeratotic epidermal nevi also have a malignant potential, therapy of such lesion should be conservative. Cryosurgery or CO_2-laser are the treatments of choice.

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