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Case Report

A Case of Linear Porokeratosis: Dermoscopic and Immunofluorescence Findings

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Abstract

Observations: Porokeratosis is a dyskeratotic disorder of the skin characterized by hyperkeratotic papule or plaque with an annular appearance and usually diagnosed easily, both histopathologically and clinically. We present a case of linear porokeratosis which was most likely suggesting the linear epidermal nevus and linear lichen planus. We observed a tiny brown border in the dermoscopic examination which suggested us the diagnosis might be porokeratosis and a column of cytoid bodies in the direct immunofluorescence examination. We think this case report will guide the clinicians to make the diagnosis of porokeratosis.

Introduction

Porokeratoses are a group of disorders of keratinization characterized by annular lesions surrounded by a characteristic keratotic border which corresponds to a typical histopathologic feature, namely, the cornoid lamella [1]. Linear porokeratosis is a clinical variant of porokeratosis and arises in infancy or in childhood. It consists of one or more plaques that are similar in appearance to classic porokeratosis: however, the plaques follow the lines of *Blaschko* most commonly on the extremities [2, 3]. When linear porokeratotic lesions have a typical clinical appearance, it is easy to diagnose. However, in the lesions which are smaller and having less elevated borders it may be confused with other linear arranged lesions. Differential diagnosis includes inflammatory linear verrucous epidermal nevus, linear lichen planus, incontinentia pigmenti (stage II), and lichen striatus [3, 4, 5].

We report a case of linear porokeratosis which had a tiny brown border in dermoscopy and a column of cytoid bodies in direct immunofluorescence (DIF) examination that was clinically indistinguishable from linear epidermal nevus and linear lichen planus.

Case Report

A 6-year–old girl having multiple, round, slightly erythematous and brownish, slightly itchy, papules and plaques on the left leg for 3 years had been applied to our clinic (**Figure 1**). They were arranged linearly. In some plaques, atrophy was barely seen in the center. The lesions first appeared on the knee and subsequently spread through the thigh and the leg. The clinical appearance was most likely suggesting the linear epidermal nevus and linear lichen planus. She was otherwise healthy and there were no any kinds of developmental problems. No similar lesions were present in other members of her family.

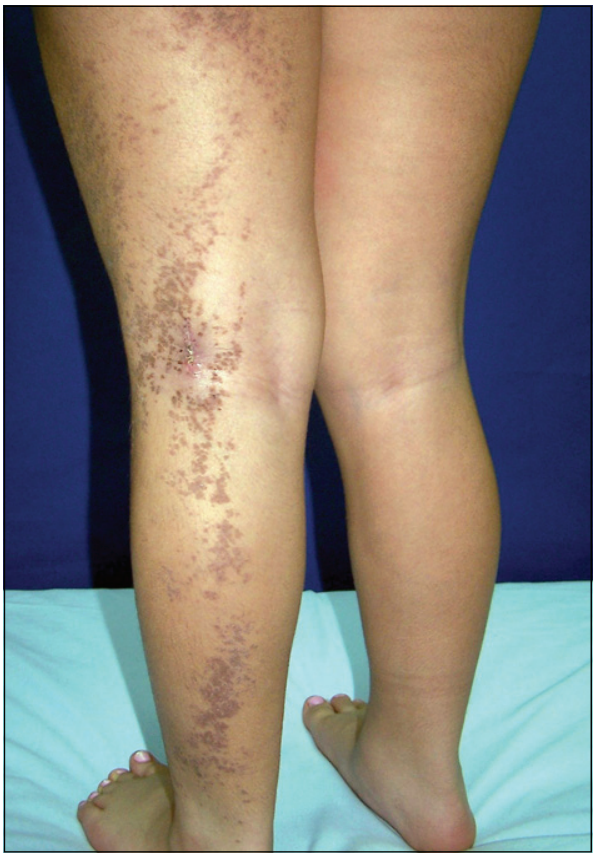

Figure 1. Multiple, round, slightly erythematous and brownish papules and plaques on the left leg

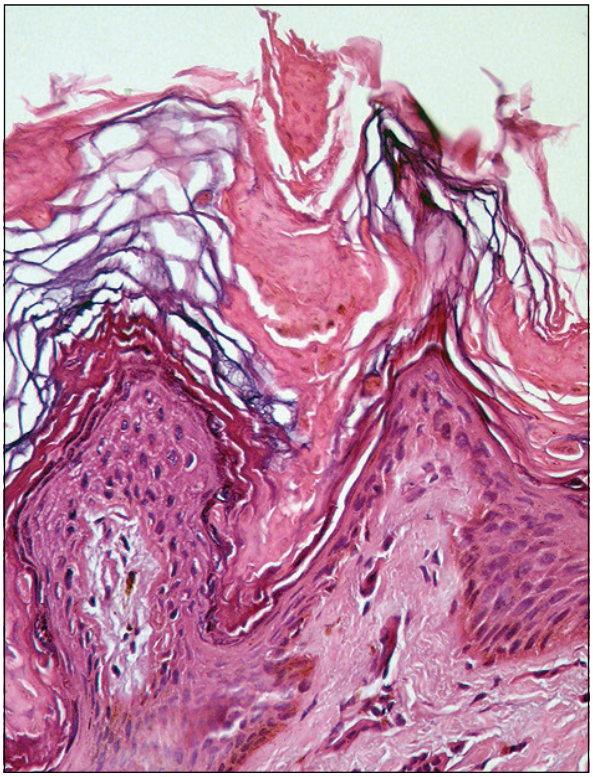

Figure 3. Cornoid lamella with absent granular layer below the parakeratotic column (x 600 magnifications, hematoxylin-eosine stain)

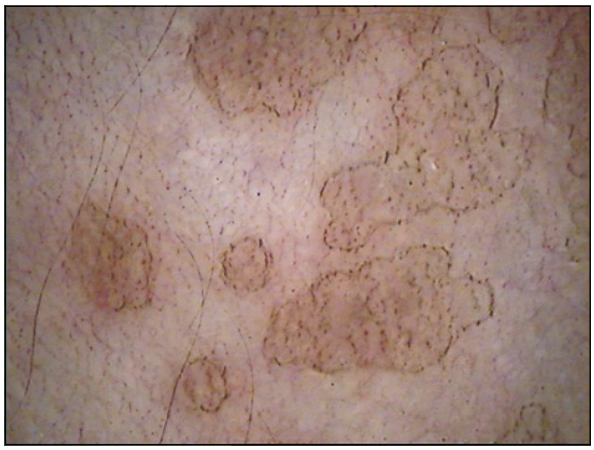

Figure 2. Each lesion was sharply demarcated and surrounded by a hyperpigmented border. Inside of this border, there were dark brown dots and globules on a brownish background (*MoleMaxII*® digital dermatoscop, X30 magnification)

Since linear lichen planus was considered in the differential diagnosis, we performed dermoscopy in order to see the *Wickham* strias: and DIF examination in order to see the fibrinogen deposition. In dermoscopic examination (MoleMaxII® digital dermatoscop), each lesion was sharply demarcated and surrounded by a hyperpigmented border (**Figure 2**). This border was composed of linearly arranged dots and globules. Inside of this border, there were dark brown dots and globules on a brownish background. By these findings, we focused on porokeratosis. For the histopathological and DIF examination, we obtained a biopsy specimen including the border. The histopathological examination of the skin biopsy specimen showed the characteristics of cornoid lamella with absent granular layer below the parakeratotic column (**Figure 3**). In the DIF examination, we observed a column of cytoid bodies staining positive with fibrinogen as a

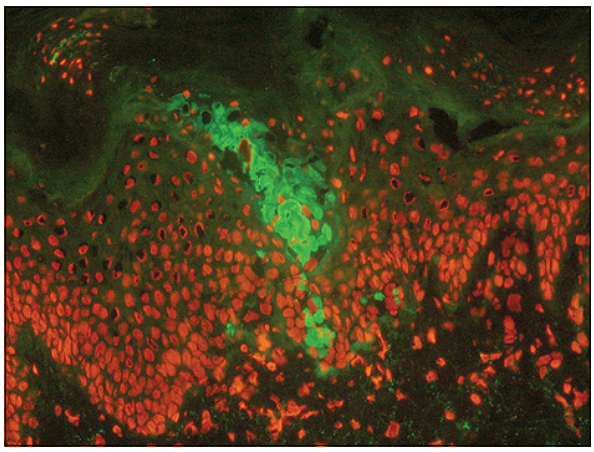

Figure 4. Multiple cytoid bodies, stained positive with fibrinojen were located in the wedge shaped invagination into epidermis densely appearing as a cornoid lamella and in the epidermis below this formation (x 600 magnifications, DIF).

wedge shaped invagination into the epidermis, where cornoid lamella was seen in the histopathological examination and a few cytoid bodies beneath them. A few cytoid bodies in the epidermis and little granular deposition at the basal membrane zone which stained positive with C3 were also seen (**Figure 4**). Altogether: dermoscopy, histopathology and DIF findings, suggested us the diagnosis of porokeratosis. The case had been treated with cryotherapy and followed by periods of 3 months.

Discussion

Linear porokeratosis is caused by a clonal proliferation of keratinocytes distributed along the lines of *Blaschko* [**4**]. Although familial cases with linear porokeratosis have been reported, the occurrence of the linear porokeratosis is sporadic with no definite pattern of inheritance established. They are asymptomatic, but can be associated with pruritus [**5**]. Lesions usually persist throughout life, but resolutions can occur. Genetically defective mutant keratinocytes may be cancer-prone because possible occurrences of malignancies such as *Bowen*'s disease or squamous cell carcinoma have been reported in all variants except the punctate variety [**3, 5**]. There is a 7.5% risk of malignant transformation in all variants of porokeratosis, and the linear porokeratosis carries the highest risk [**4**].

Dermoscopy is a noninvasive diagnostic technique that represents a link between macroscopic clinical dermatology and microscopic dermatopathology. In the last years dermoscopy has been employed also for the evaluation of nonpigmented skin disorders, such as nonpigmented skin tumors, inflammatory and infectious diseases [**6, 7**]. Dermoscopic findings of porokeratosis in some previously reported cases and in our case were shown in **Table 1**.

Although there were sharp borders around the all the lesions in the dermoscopic examination, each lesion had different characteristics, as in previously reported cases and in our case report. Inside the lesions atrophy, red dots, globules, white homogenous areas were seen. We determined dark brown dots and globules on the brownish background which were surrounded by the hyperpigmented border. It will be useful to study the dermoscopic findings of porokeratosis in more cases to determine the evident changes of porokeratosis. In the present case, dermoscopy allowed us to visualize the characteristic morphologic findings of porokeratosis described better as the annular structure was encircled by a raised keratotic border.

We observed a striking finding in DIF examination, a column of cytoid bodies stained positive with fibrinojen, in location where the cornoid lamella was seen in the histopathological examination, and also a few cytoid bodies in the underlying epidermis. Cytoid bodies could be stained positive with immunoglobulin, fibrinogen, and C3 in DIF examination [**10**]. *Shen* et al. showed abnormal early keratinocyte apoptosis in the pathogenesis of porokeratosis [**11**]. So, our finding may support the role of apoptosis in the pathogenesis.

As in our case, if linear porokeratosis does not have a characteristic appearance it may be indistinguishable from other linear arranged dermatoses. In such cases, dermoscopy which is a noninvasive technique,

Table 1. Dermoscopic Characteristics of Porokeratoses in Previously Reported Cases and in Our Case

Authors	Types of porokeratosis	Number of Patient	Dermoscopic Findings
***Delfino* et al**[1]	Disseminated superficial porokeratosis	1	Whitish-yellowish annular structure (appearing as the outlines of a vulcanic crater as observed from a high point), pink-white scar-like area in the center.
***Zaballos* et al**[8]	Disseminated superficial actinic porokeratosis	3	"White track" structure at the periphery (single or double), brownish pigmentation inner side (red dots, globules and lines, white homogenous area).
***D'Amico* et al**[9]	Porokeratosis of *Mibelli*	10	Dark-brown globules/dots circumscribed the central hypopigmented scar-like area. These globules/dots joined to form a continuous line. Some lesions contain brown globules/dots and red dots in the central area.
Our case	Linear porokeratosis	1	Hiperpigmented border (linearly arranged dots and globules), dark brown dots and globules on the brownish background in the center.

may guide the clinicians and the pathologists to make the diagnosis. Besides the histopathologic examination, DIF findings may also aid the diagnosis, because it is easier to see the bright cytoid bodies in dark ground in the DIF examination.

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