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Type2 Segmental Manifestation Of Disseminated Superficial Actinic Porokeratosis In An 8 Year Old Girl

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Abstract

Porokeratosis is a form of genodermatosis of different clinical types. The combination of different types of porokeratosis in one individual is rare. Here we report a rare case in which two different types of porokeratosis linear and disseminated type coexisted. An 8 year old girl developed a centrifugal lesion on her right chest at the age of 2 years, scattered lesions on her both cheeks at the age of 3 after ultraviolet exposure and linear porokeratosis on her left side of body at the age of 5. No other family member and siblings had similar skin eruptions. Histopathology revealed cornoid lamella, dyskeratosis in the epidermis and scanty dermal perivascular lymphocytic infiltration. We diagnosed her facial lesions as disseminated superficial porokeratosis and linear porokeratosis on left side of body. The combination of two varieties may be a result of the loss of heterozygosity. We consider that this case may represent segmental manifestation of disseminated superficial actinic porokeratosis. We treated the facial lesions with topical tretinoin and the lesions on her legs and hands with cryotherapy and topical halobetasol with salicylic acid.

Introduction

Porokeratosis is a form of genodermatosis characterized clinically by annular hyperkeratotic lesions and histologically by the formation of cornoid lamella. There are different clinical types of porokeratosis namely; classic porokeratosis of Mibelli, linear porokeratosis (LP), disseminated superficial actinic porokeratosis (DSAP) and porokeratosis palmaris *et* plantaris punctuate [1]. The combination of different types of porokeratosis in one individual is rare. Here we describe a patient with two different types of porokeratosis representing type 2 segmental manifestation of disseminated superficial actinic porokeratosis.

Case report

An 8 year old girl from a village of Chhattisgarh presented with widespread eruptions on her face and the left side of her body. When she was 2 years old a centrifugal lesion on her right chest and right arm appeared [figure1]. At the age of 3, she developed scattered lesions on her both cheeks [figure1]. At the age of 5, she developed numerous small keratotic lesions on the left side of her body in a linear fashion [figure 2, 3]. Her medical history was unremarkable. On examination, she presented with light brown annular keratotic lesions, 5-7 mm in diameter, distributed on both cheeks. On her arms, left shoulder, clavicle area and left leg from the inguinal fold, there were numerous small deep brown colored hyperkeratotic annular lesions, up to 8 mm in diameter, distributed linearly along the lines of Blashko. Some lesions are pruritic and due to this there was erosion on right arm. No other family member and siblings had similar skin eruptions. The initial diagnosis was porokeratosis, while epidermal verucae nevus was also considered. Histopathology findings revealed an invagination of the epidermis with a column of parakeratotic cells (cornoid lamella) overlying an absent granular layer and dyskeratosis in the epidermis [figure 4]. Hyperkeratosis, parakeratosis and a lichenoid tissue reaction with a band like lymphocytic infiltration and dilated vessels in the upper dermis were observed [2, 3] [figure 5]. We diagnosed this patient as having linear porokeratosis [5] on left body and disseminated superficial porokeratosis on her face [4]. We advised her parents to avoid sun exposure and use of sunscreens. We treated the lesion on her face with topical tretinoin of 0.25% and the lesions on her legs and hands with cryotherapy and topical halobetasol with salicylic acid. After 3 visits her facial lesion faded and the lesion on her limbs decreased little and is on follow up.

Discussion

Porokeratosis is a clonal disease characterized by keratotic abnormalities with autosomal dominant inheritation. Each type of porokeratosis has distinct clinical features and distribution. Nevertheless, all

these types are associated with cornoid lamellae as a common histological feature [1].

Here, we described a case of porokeratosis in which two different clinical types of porokeratosis existed together in a girl. We diagnosed her facial eruption as DSAP because the small lesions were distributed symmetrically on both cheeks, did not follow the lines of Blashko and appeared after UV exposure at the age of 3. On the other hand we are certain about the LP diagnosis concerning the eruptions on the left and right side of her body surface because of the linear distribution of the eruptions along the lines of Blashko. Happle proposed the loss of heterozygosity caused by somatic recombination as an explanation for this association [6]. In the case of an autosomal dominant trait, at the early stage of embryogenesis, somatic crossing-over, nondisjunction or deletion may occur involving this gene locus and this may cause the loss of heterozygosity. In our patient the combination of LP and DSAP may be a result of the loss of heterozygosity. It is possible that our patient is susceptible to ultraviolet light for some reason and DSAP appeared first after strong ultraviolet exposure at the age of 3.

Various treatments have been applied for porokeratosis, such as keratolytic agents, topical or intralesional corticosteroids, topical tretinoins, topical 5-flurouracil, systemic retinoids, cryotherapy, electrodessication, CO2 laser or derma abrasion. However the results were unsatisfactory. We tried tretinoin and cryotherapy with moderate effect. However long term follow up is necessary to determine the efficacy of these treatments.

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Illustrations

Illustration 1

Figure 1: centrifugal lesions on right chest and scattered lesions on both cheeks.

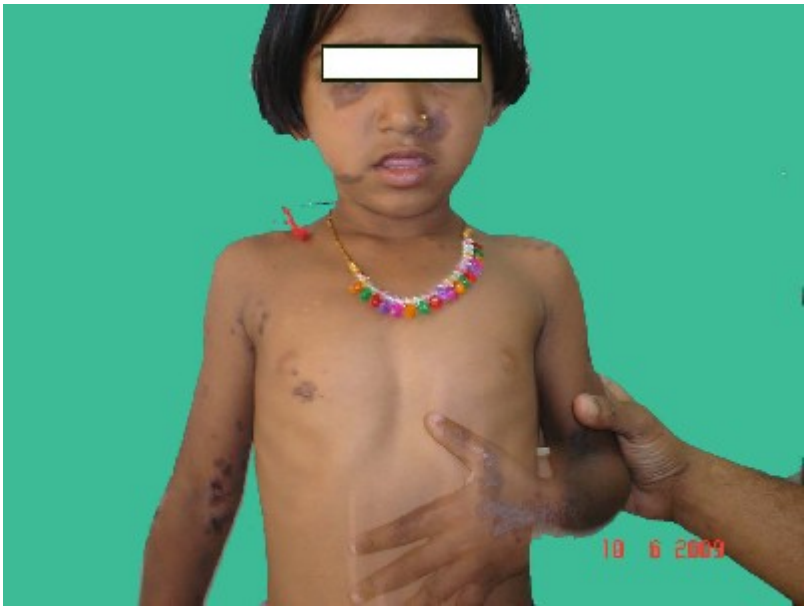


Illustration 2

Figure 2: numerous small keratotic lesions on her left hand in a linear fashion

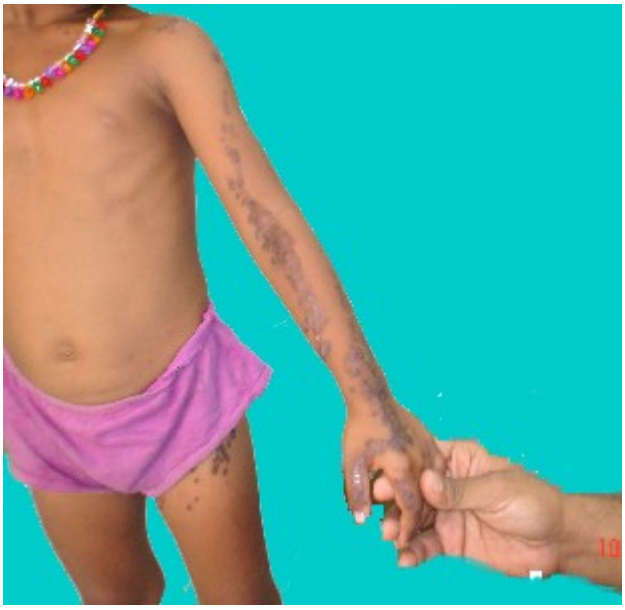


Illustration 3

Figure 3: numerous small keratotic lesions on her left leg in a linear fashion

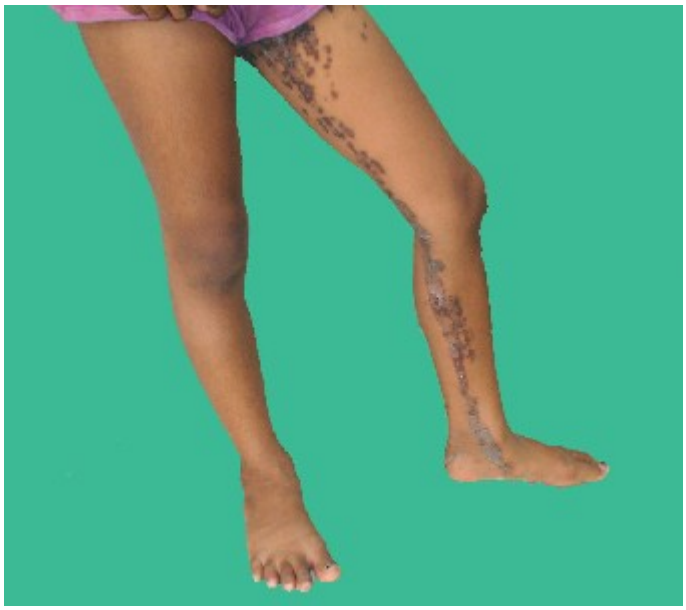
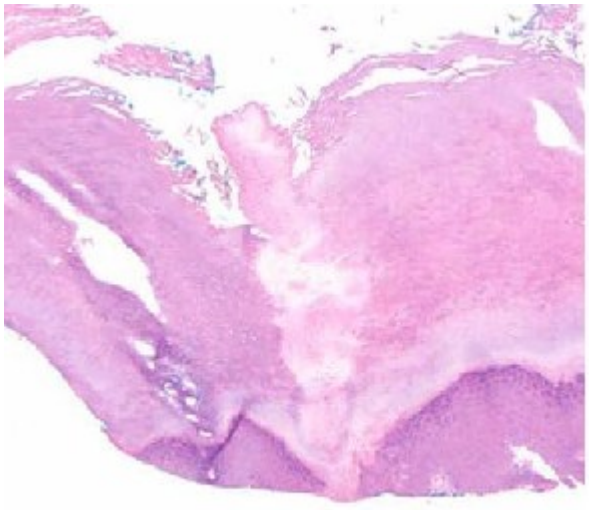


Illustration 4

Figure 4: cornoid lamella and dyskeratosis in the epidermis.



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