

Segmental Porokeratosis Pigmentosa - An Unusual Presentation of Porokeratosis or a New Entity?

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Abstract: A 3 year old boy presented with complaints of darkening of skin along the right side of body for the past 2 years, progressively increasing in size and pigmentation. Cutaneous examination revealed multiple hyperpigmented linearly arranged macules and annular plaques following the lines of Blaschko anteriorly running along the right side of the body extending from the axillae to the inguinal and face. The macules and plaques on the body and extremities were hyperpigmented with atrophy in the centre with raised hyperkeratotic borders. We made a clinical diagnose of Linear Porokeratosis and a skin biopsy confirmed the diagnosis, which showed hyperkeratosis and typical coronoid lamella tightly stacked with parakeratotic cells along with increased amount of melanin in the basal layer and mild lymphocytic infiltration into the dermis. The diagnosis of Porokeratosis was confirmed and the boy has been under our treatment ever since for the past 1 year. We report this case for its rarity and even though this case if classified in the existing classification fits more into the Linear variant of porokeratosis, hyperpigmentation in porokeratosis has not been reported before. With the histological findings showing increased amount of melanin in the basal layer along with other findings of porokeratosis we could also consider this case as a new clinical variant which we have named as segmental porokeratosis pigmentosa.

Keywords: Porokeratosis, Linear porokeratosis, Pigmentation in porokeratosis

1. Introduction

Porokeratosis is a morphologically distinct disorder of keratinisation, characterized clinically by hyperkeratotic papules or plaques surrounded by a thread-like elevated border that expands centrifugally.¹ The peripheral keratotic ridge corresponds histologically to the coronoid lamella. Five different forms has been distinguished: 1) The plaque type classical porokeratosis of Mibelli, 2) Disseminated superficial actinic porokeratosis (DSAP), 3) Linear porokeratosis, 4) Porokeratosis plantaris, palmaris et disseminata and 5) Punctate porokeratosis.² Various cases of coexistence of the different variants of porokeratosis in a single patient has been reported but they are regarded as a rare occurrence.^{3, 4} Here we present a case of Linear porokeratosis presenting with hyperpigmentation.

2. Case Report

A 3 year old boy presented with complaints of darkening of skin along the right side of body and both sides of the face since the past 2 years, progressively increasing in size and pigmentation. There was no similar history in the family. To start with the lesions evolved on the right arm extensor aspect to slowly involve the entire right arm and gradually spread to the right side of body, bilaterally both sides of the face and later the groins and inguinal region. There was no associated exacerbating factors and the lesions were asymptomatic.

Systemic examination of the patient was normal. In cutaneous examination we saw multiple hyperpigmented linearly arranged macules and annular plaques following the lines of Blaschko anteriorly running along the right side of the body extending from the axillae to the middle of right thigh, bilateral involvement of the inguinal region and genitals, extensor aspect of right arm, bilateral involvement of the face and minimal involvement of posterior aspect of right side of body. The macules and plaques on the body and extremities were hyperpigmented with atrophy in the centre with raised hyperkeratotic borders and while the macules on the face were hyperpigmented and irregularly spread. We

made a clinical diagnose of Linear porokeratosis as it fitted more into this variant even though few features of Porokeratosis of Mibelli and Disseminated superficial porokeratosis were present. A skin biopsy was done and histopathological examination confirmed the diagnosis with the presence of hyperkeratosis and typical coronoid lamella tightly stacked with parakeratotic cells. There was also increased amount of melanin in the basal layer and mild lymphocytic infiltration was present in the dermis. The diagnosis of Porokeratosis was confirmed and the boy has been under our treatment ever since for the past 1 year.

3. Discussion

Linear Porokeratosis is an uncommon variant of Porokeratosis and by itself is a rare disease listed by the Office of Rare Diseases (ORD) of the National Institute of Health, USA. It can be present at birth following the lines of Blaschko or may not develop until adult life. Occasionally, there is family history of linear porokeratosis or other kind of porokeratosis such as disseminated superficial actinic porokeratosis, suggesting genetic predisposition.^{5, 6, 7, 8} Clinically the lesions may be confined to one extremity which is more common than the generalized form which may involve several extremities and the trunk.^{1, 9} A skin cancer can develop within a linear porokeratosis patch. This may be either a basal or squamous cell carcinoma, and is more likely to occur in older adults. If a lump or sore appears within a porokeratosis lesion, it has to be carefully reviewed,^{1, 7, 8, 10} and may require a biopsy or cutting out.

We have been regularly following up the patient and the patient's parents have been counseled regarding the possibilities of malignant degeneration. We report this case for its rarity and even though this case if classified in the existing classification fits more into the Linear variant of porokeratosis, hyperpigmentation in porokeratosis has not been reported before. With the histological findings showing increased amount of melanin in the basal layer along with other findings of porokeratosis we propose to consider this case as a new clinical variant which we have named as segmental porokeratosis pigmentosa.

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