



## Case Report

# Porokeratosis mibelli- A Case Report

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### Abstract

A twelve years old girl of class VII reported at skin & VD out patient department of Rajshahi Medical college Hospital in November 2003 with multiple small pink to violeceous non pruritic patches extending from medial aspect of left ankle joint to the medial aspect of left thigh in a linear fashion. The individual lesions are small-circumscribed patches having hyperkeratotic margin and central atrophy. The raised hyperkeratotic margin contains on its top a furrow along the course of the wall. The surface was verrucous and many of the lesions have serpiginous border. Near the ankle joint and below the ankle joint individual lesion forms zosteriform groups. On the thigh the lesions are linear. Some small satellite lesions are also present. All the lesions are hairless and anhidrotic. The onset of the individual lesions was at the age of six years, near the ankle joint. Lesions enlarge slowly and progressed to upward direction in a linear fashion. Skin biopsy for histopathology revealed the picture of porokeratosis.

TAJ 2004; 17(2): 106-107

### Introduction

Porokeratosis is a heterogeneous group of disorders that are inherited as an autosomal dominant fashion. Clinically, the basic lesion is sharply demarcated and hyperkeratotic. The clinical lesion of porokeratosis is unique. The lesion has an annular railed keratotic border with central furrow or depression. Lesions may be annular, linear and punctuate with central atrophy. Five clinical variants are recognized.

- 1) Classic porokeratosis mibelli
- 2) Disseminated superficial prokeratosis and disseminated superficial actinic porokratois
- 3) Porokeratosis palmaris et plantaris disseminata
- 4) Linear porokeratosis
- 5) Punctate porokeratosis

Immunosuppression, Immunosuppressive diseases such as AIDS, ultraviolet exposure and radiation therapy may exacerbate porokeratosis and promote the development of skin cancers. There is 7.5% risk of developing cutaneous malignancies and patients with linear type are at great risk.

### Case report

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**Investigations:** All routine examinations were done. Histopathology of skin revealed the diagnostic clues in favor of porokeratosis.

**Histopathology:** The histopathological picture is distinctive because of an invaginating parakeratotic column, the coronoid lamella – a thin column at closely stacked parakeratotic cells extending through the stratum corneum. The coronoid lamella may be due to an abnormal clone of keratinocytes exhibiting accelerated proliferation or premature keratinization or both.

### Discussion

Porokeratosis has a wide variety of manifestations, but with the exception of the punctuate type; it is characterized by a distinct peripheral keratotic ridge that corresponds histologically coronoid lamella.

In porokeratosis of mibelli, the lesions occur anywhere on the body and are usually well

developed, possess a sharp, elevated border and central atrophy. There may be a single annular plaque of varying size or multiple plaques arranged in a linear or segmental nevoid distribution. The onset is in childhood, usually before age 10. In our case the lesions are unilateral, linear and zosteriform in morphology, slowly progressive hyperkeratotic plaque with central atrophy. The raised border of individual lesions with a furrow on its top was distinctive. The lesions were asymptomatic and the onset was at the age 6. Linear epidermal naevus, lichen striatus and lichen planus were excluded by histopathology. As squamous cell carcinoma may develop on lesions of porokeratosis of Mibelli, careful follow up is warranted.

### References

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