

## Porokeratosis of Mibelli in a child: A case report

Mohammad Abid Keen

Consulting Dermatologist, Health Services Kashmir.

**Abstract** Porokeratosis are a group of hereditary or acquired disorders characterized by annular plaques with an atrophic centre and a hyperkeratotic peripheral ridge. Pathologically, porokeratosis is characterized by a column of parakeratotic cells, called the cornoid lamella. This report describes a 6 yrs. old girl affected by porokeratosis of Mibelli.

**Key words**

Porokeratosis, parakeratotic cells, cornoid lamella.

### Introduction

Porokeratosis represents a rare, benign, heterogeneous group of genodermatoses having abnormal keratinization, with an autosomal-dominant mode of inheritance.<sup>1,2</sup> Clinically, it presents with annular plaques with atrophic center and hyperkeratotic edge, mainly over the extremities. The cornoid lamella, a column of parakeratotic cells that occupies the small epidermal invaginations, constitutes a characteristic histopathological finding, primordial for its diagnosis.<sup>1,2</sup> Porokeratosis of Mibelli (PM) was first described in 1893 by Vittorio Mibelli. Since then many clinical variants, both localized and generalized forms, have been described.<sup>3</sup> Clinically, porokeratosis is classified into the localized forms, which include PM, linear porokeratosis (LP), and punctate porokeratosis, and the disseminated forms including disseminated superficial actinic porokeratosis (DSAP), superficial disseminated porokeratosis, and porokeratosis palmaris et plantaris disseminata (PPPD). In addition, other uncommon morphological variants such as

facial, giant, punched out, hypertrophic, verrucous, and reticulate porokeratosis have also been reported.<sup>4,5</sup>

Porokeratosis of Mibelli is the most characteristic and distinctive variant of the described forms of Porokeratosis. The classical form of Mibelli consists of a single plaque, or a small number of plaques, of variable size and most often affects the limbs, particularly the hands and feet, the neck and shoulders, although any part of the body may be affected including the mucous membranes and scrotum.<sup>6</sup> The exact etiopathogenesis is unknown but multiple factors are proposed, such as chronic sun exposure, ultraviolet (UV) radiation, trauma, hepatitis B and C infection, human immunodeficiency virus infection (HIV) and immunosuppression.<sup>4,7</sup> Histopathologically, PM has the hallmark cornoid lamella, a column of a parakeratosis arising within the invagination of the epidermis. The granular layer is focally diminished and keratinocytes are dyskeratotic.

### Case report

A 4 year old female child, born of a non-consanguineous marriage presented with a history of erythematous hyperchromic plaques with slightly elevated edges and atrophic centre on the lateral aspect of thenar eminence of her

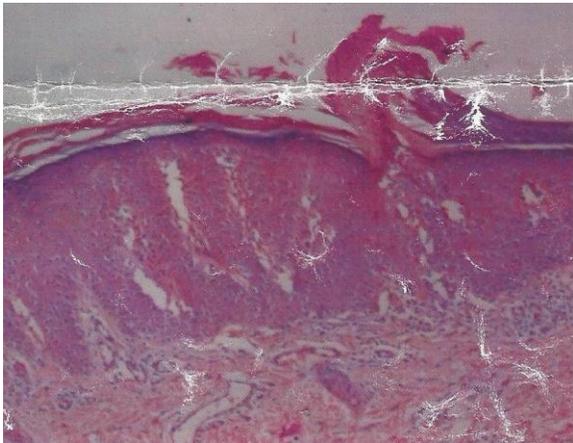
---

**Address for correspondence**

Dr. Mohammad Abid Keen (MD)  
Consulting Dermatologist  
Health Services Kashmir, Pin: 192101.  
Ph: 9419963692  
Email: keenabid31@gmail.com



**Figure 1** Annular plaque with central clearing and raised peripheral edge, measuring about 3×3cm in size, present over dorsum of right hand.



**Figure 2** Histopathology revealing foci of epidermal invagination filled with keratin and parakeratosis cornoid lamella.

right hand since the age of two years. The lesions were slowly progressive with no signs of self-healing. The patient had normal physical milestones of development as a child. She was immunocompetent with no history of recurrent infections or any other skin lesions. Her parents and siblings (1 brother and 1 sister) were all normal and healthy. She had no family history of a similar condition or other skin problems. There was no history of photosensitivity, recurrent fever, Raynaud's phenomenon, or joint pains. Physical examinations of the patient was within normal limits. On systemic examination, no abnormality was revealed. Clinically there was no evidence of sexually transmitted diseases and

laboratory investigations were within normal limits. Dermatological examination revealed a single annular plaque with central clearing and raised peripheral edge, measuring about 3×3 cm in size, present over dorsum of right hand (**Figure 1**). Dermoscopy revealed an annular plaque with a peripheral ridge and a central crater-like depression, the ridge showing scaling in the center. Differential diagnoses of DLE, porokeratosis, lupus vulgaris, and sarcoidosis were considered. A full depth skin biopsy was taken from the outer part of the lesion with a 2 mm disposable skin biopsy punch and subjected to histopathology which revealed foci of epidermal invagination filled with keratin and parakeratosis cornoid lamella (**Figure 2**). On the basis of history, clinical examination and histopathology, a diagnosis of plaque type of Porokeratosis of Mibelli was made. Topical Imiquimod was prescribed for the management of the lesion.

## Discussion

Porokeratosis is a clonal disorder of keratinization characterized by one or more atrophic patches surrounded by a clinically and histologically distinctive hyperkeratotic ridge like border called the coronoid lamella. It is a chronic progressive disorder of keratinization. The mode of inheritance is autosomal dominant, but some sporadic cases have also been reported. It has a slight male preponderance (3:2), usually beginning in childhood or adolescence, and is more prevalent in Caucasians.<sup>8</sup> PM consists of one or more plaques, usually a small number of them, which may occur anywhere in the body, more frequently in extremities, especially hands and feet, with unilateral distribution. Other areas, such as neck, shoulders and genitals may also be affected.<sup>9</sup> Lesions are asymptomatic with an indolent evolution. Various risk factors involved in the development of porokeratosis include genetic inheritance,<sup>10</sup> immunosuppression,<sup>10,11</sup>

ultraviolet radiation and sun exposure,<sup>10</sup> particularly immunosuppression.<sup>12</sup> Porokeratosis may also be associated with diabetes mellitus,<sup>13</sup> HIV infection,<sup>14</sup> liver disease,<sup>15</sup> renal transplant,<sup>16</sup> and hematologic or solid organ malignancy.

Histopathology of Porokeratosis shows hyperkeratotic lesion with discrete Parakeratotic column at margin. Diagnostic feature is Coronoid lamella which represents visibly raised margin of the lesion. It is a Parakeratotic column overlying a small vertical zone of dyskeratotic and vacuolated cells within the epidermis along with focal loss of granular cell layer, mild lymphocytic infiltrate may be seen. The factors demonstrated to be associated with a higher risk of malignant transformation include duration and size of the lesions, age of patient, large lesions on the extremities and linear type<sup>12</sup> as well as multiple patterns of porokeratosis.<sup>17</sup>

Clinically, PM needs to be differentiated from the following: DLE, lupus vulgaris, sarcoidosis, Bowen's disease, annular lichen planus, lichen sclerosus et atrophicus, pityriasis rubra pilaris, acrokeratosis verruciformis, Darier's disease, seborrheic keratosis, linear scleroderma, actinic keratosis, xerosis, elastosis perforans serpigiosa, and inflammatory linear verrucous epidermal nevus.<sup>3,4</sup>

The approach to treatment is individualized and based on many factors, such as lesion size and location, risk of malignant transformation, and functional and aesthetical considerations. Treatment options include topical 5-fluorouracil, topical and systemic retinoids, topical and oral corticosteroids, keratolytics, topical vitamin D3 analogues, 5% imiquimod, 3% topical diclofenac sodium, lasers (CO2 ablation, Nd:YAG, and 585 pulse dye laser), Grenz ray radiation, cryotherapy, dermabrasion, surgical

excision, photodynamic therapy, and electrodesiccation.<sup>3,17,18</sup>

## References

1. Gutierrez EL, Galarza C, Ramos W, Tello M, De Paz PC, Bobbio L, *et al.* Facial porokeratosis: A series of six patients. *Australas J Dermatol* 2010;**51**:191-4.
2. Chen TJ, Chou YC, Chen CH, Kuo TT, Hong HS. Genital porokeratosis: a series of 10 patients and review of the literature. *Br J Dermatol*. 2006;**155**:325-9.
3. Ferreira FR, Santos LD, Tagliarini FA, Lira ML. Porokeratosis of Mibelli — literature review and a case report. *An Bras Dermatol*. 2013;**88**:179-82.
4. Chaudhary RG, Bilimoria F, Katare SK. Large annular plaque with central atrophy on nose. *Indian J Dermatol Venereol Leprol*. 2009;**75**:552-3.
5. Mehta V, Balachandran C. Simultaneous co-occurrence of porokeratosis of Mibelli with disseminated superficial actinic porokeratosis. *Indian J Dermatol* 2009;**54**: 390-1.
6. Dongre A, Adhe V, Sanghavi S. Genital Porokeratosis: A rare entity. *Indian J Dermatol*. 2013;**58**:81-3.
7. Avhad G, Jerajani H. Porokeratosis of Mibelli: Giant variant. *Indian Dermatol Online J*. 2013;**4**:262-3.
8. Ehsani AH, Shakoei S, Ranjbar M. Giant porokeratosis of Mibelli with squamous cell carcinoma. *Indian J Dermatol Venereol Leprol*. 2014;**80**:96.
9. Mukhopadhyay AK. Simultaneous occurrence of disseminated superficial, linear and hypertrophic verrucous forms of porokeratosis in a child. *Indian J Dermatol Venereol Leprol* 2004;**70**:364-6.
10. Odeyinde S, Belcher H. Isolated single digit porokeratosis of mibelli: An unusual case. *Dermatol Online J* 2012;**18**:13.
11. Pérez-Crespo M, Betlloch I, Lucas-Costa A, Bañuls-Roca J, Niveiro de Jaime M, Mataix J. Unusual evolution of giant porokeratosis developing in two renal transplant patients. *Int J Dermatol*. 2008;**47**:759-60.
12. Sasson M, Krain AD. Porokeratosis and cutaneous malignancy. A review. *Dermatol Surg*. 1996;**22**:339-42.
13. Nakamura M, Fukamachi S, Tokura Y. Acute onset disseminated superficial porokeratosis associated with exacerbation

- of diabetes mellitus due to development of anti-insulin antibodies. *Dermatoendocrinol*. 2010;**2**:17-8.
14. Rodriguez EA, Jakubowicz S, Chinchilla DA, Carril A, Viglioglia PA. Porokeratosis of Mibelli and HIV-infection. *Int J Dermatol*. 1996;**35**:402-4.
  15. Hunt SJ, Sharra WG, Abell E. Linear and punctate porokeratosis associated with end-stage liver disease. *J Am Acad Dermatol*. 1991;**25**:937-9.
  16. Herranz P, Pizarro A, De Lucas R, Robayna MG, Rubio FA, Sanz A. High incidence of porokeratosis in renal transplant recipients. *Br J Dermatol*. 1997;**136**:176-9.
  17. Chua IS, Lee JS, Chiam LY. Pruritic papules on the nose in a 25-year-old female. *Indian J Dermatol*. 2014;**59**:106.
  18. Saritha M, Kumari R, Thappa DM, Rajesh NG, Verma SK. Benign giant cutaneous horn formed by giant porokeratosis of Mibelli with dysplasia. *Indian J Dermatol Venereol Leprol*. 2013;**79**:433-5.