



POROKERATOSIS OF MIBELLI

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Porokeratosis of Mibelli is a chronic and slowly progressive skin lesion due to excessive keratinization. Porokeratoses (singular: porokeratosis) comprise a class of skin conditions of which porokeratosis of Mibelli is one of six distinct variants. The term porokeratosis defines this family and refers to a characteristic skin lesion with a thinned center and a surrounding ring-like border.

In certain cases a hereditary nature of this condition is seen suggesting a genetic component. In such cases, the disease is transmitted in an autosomal dominant fashion. Most commonly, porokeratosis of Mibelli manifests in infancy and early childhood. Adult patients may develop the disease if they become immunosuppressed from medications or illnesses.

Porokeratosis of Mibelli typically appears as a small, asymptomatic skin lesion that may slowly enlarge over time. Lesions usually measure a few centimeters in size however, giant porokeratosis can grow up to 20cm in size. This disorder is characterized clinically by increased keratin deposition by skin surface cells known as keratinocytes. Keratin, a form of cellular protein, is deposited in a well-circumscribed manner defining the borders of the lesion. The ridge-like border, called cornoid lamella, is usually greater than 1mm in height and serves as the clinical and histologic feature of the disease. At the center of porokeratosis of Mibelli, the lesion demonstrates a thin, mildly atrophic patch of skin with minimal hair and scale. Interestingly, areas of the body affected by porokeratosis of Mibelli lose the ability to sweat. Affected areas most commonly include the hand, feet, arms and legs although any part of the body can be affected. In a minority of cases, porokeratosis of Mibelli can develop into a cancerous lesion, namely **squamous cell** and **basal cell carcinoma**.

Given the relative chronic and progressive nature of the disease, intervention is not always necessary. If porokeratosis of Mibelli becomes problematic or cosmetically unappealing, a number of therapeutic interventions may be employed including:

- **5-fluorouracil**
- **Imiquimod**
- **Vitamin D analogs**
- Keratolytics like salicylic acid
- **Cryosurgery**
- **Lasers**
- **Photodynamic therapy**

Finally, due to the small potential for malignant transformation, protection from UV radiation with **sunscreen** and protective clothing is highly recommended.

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