Porokeratosis of Mibelli: Giant variant

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A 45-year-old female presented with hyperpigmented, solitary, annular plaque on her right buttock since 12 years. She also gave a history of gradual increase in the size of the lesion [Figure 1].

Physical examination showed a single erythematous, circular, scaly plaque 18 cm x 15 cm with characteristic peripheral ridge measuring 12 mm. Histopathology showed stacked parakeratosis within epidermal invagination and underlying absent granular layer, which was suggestive of coronoid lamella [Figure 2].

Porokeratosis is a benign, rare, genetically determined autosomal dominant disorder of epidermal keratinization, characterized clinically by hyperkeratotic papules or plaques surrounded by a thread like elevated border that expands centrifugally. Multiple etiologies are proposed in the clonal proliferation of keratinocytes, like chronic sun exposure, Hepatitis B and C infection, HIV and immunosuppresion.

The classic lesion of porokeratosis[1] was first described by Mibelli in 1893. It is usually seen during childhood as one or multiple annular plaques with central atrophy and elevated keratotic borders usually greater than 1 mm in height that have a longitudinal furrow typically seen in the center of the ridge. This ridge expands over a period of time. It affects men twice as often as women. The lesion may be hypopigmented or hyperpigmented, scaly, atrophic, hairless and anhidrotic. It commonly develops on the extremities but has also been known to occur on the face, buccal mucosa, genitalia, palms and soles.[2]

Giant porokeratosis is considered to be a morphological variant of porokeratosis of Mibelli with a diameter of up to 20 cm and surrounding wall of 1 cm.[3]

As there is risk of development of squamous cell carcinoma in giant porokeratosis (10%), early diagnosis and treatment is necessary.[4]

There are various modalities of treatment like topical 5-fluorouracil, imiquimod, oral retinoid, CO2 laser ablation, 585-nm pulsed dye laser radiation, Grenz ray radiation, Nd:YAG laser radiation, cryotherapy, dermabrasion, surgical excision and electrodesiccation.[5]

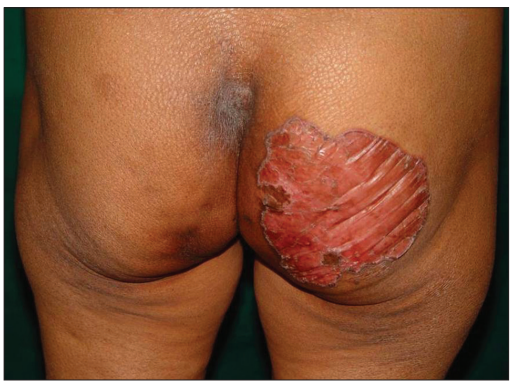

Figure 1: Single erythematous, oval, scaly plaque 18 cm X 15 cm with characteristic peripheral ridge with central atrophy

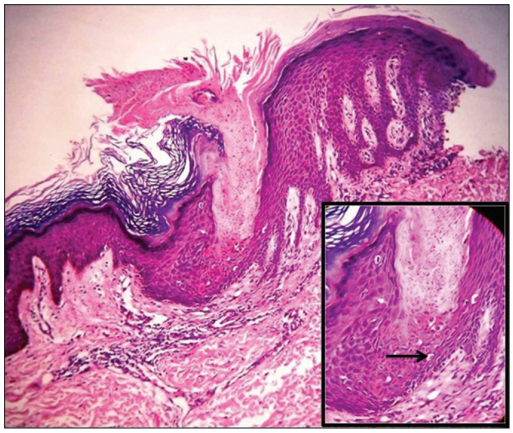

Figure 2: Histopathology of plaque showing invagination of epidermis with prominent cornoid lamella and underlying loss of the granular layer, inset showing dyskeratotic cells (shown by arrow) seen beneath the coronoid lamella (100X)

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