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POROKERATOSIS OF MIBELLI RESEMBLING LUPUS VULGARIS – A RARE CASE REPORT

Dermatology

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ABSTRACT

Porokeratosis of Mibelli presents with annular plaques with atrophied centers along with hyperkeratotic edges, majority of the cases are having autosomal dominant inheritance. The following report describes a 23 year old male with lesions present over the nose.

KEYWORDS

Porokeratosis, Keratinization, Cornoid Lamella.

Introduction:
Porokeratosis is a clonal expansion of keratinocytes which differentiate abnormally, but are not truly neoplastic.[1] The disease may either present as single or multiple lesions; which may be localized or disseminated. Histologically characterized by a thin column of parakeratotic cells extending throughout the stratum corneum, termed as Cornoid Lamella.[2]

Case Report:
A 23-year-old male came to the skin OPD with complaints of a pigmented lesion over the nose for 4 years. Patient was apparently normal 4 years back after which he developed a small papule over the nose which gradually progressed to become a plaque with ulceration. Patient had history of itching present, history of photosensitivity present, no history of similar complaints elsewhere in the body. No history of any personal antecedents. No history of similar lesions among the family members.

On examination, multiple small plaques present over the right, left and central area of the nose, well-circumscribed, pigmented & have a verrucous surface with raised borders.

Skin biopsy was done and histopathological examination revealed hyperkeratosis, parakeratosis, furrow within the epithelium filled with keratin called the cornoid lamella with underlying hypogranulosis, dermal infiltrates of lymphocytes; all of which are consistent with Porokeratosis of Mibelli.

Discussion:
Mibelli described the classical form of porokeratosis in 1893. The term porokeratosis is a misnomer as it does not involve the sweat gland always. Typically appears during infancy or adolescence.

Porokeratosis is mainly divided into 5 types clinically. They are (i) Porokeratosis of Mibelli (ii) Disseminated superficial actinic porokeratosis (DSAP) (iii) Linear porokeratosis (iv) Porokeratosis palmaris et plantaris disseminate (v) Punt ate porokeratosis.

Other types include Porokeratosis ptychotropica, Porokeratoma, Porokeratotic adnexal ostial nevus (PAON), Pigmented Porokeratosis and Pruritic Papular Porokeratosis.

Risk factors involved in the development of Porokeratosis of Mibelli include: genetic inheritance, immunosuppression, sun exposure.[3]

The lesions start as papules or plaques which develop into annular lesions with a thin, often thread like elevated rim occurring frequently in extremities.[4]Other sites include neck, shoulders and genital. Very rarely mucosal and facial lesions are seen.

Factors determining malignant conversion include: Age, Duration, size of the lesions, multiple patterns of Porokeratosis and also the occurrence of large lesions on the extremities.[5]

On Histologic Examination: Keratin filled invagination extending deeply downwards with a parakeratotic column arising from its center called Coronoid Lamella.[6] This is the classic hallmark of diagnosing Porokeratosis of Mibelli. The epidermis beneath the parakeratotic column show hypogranulosis and dyskeratinization.[6]

Differential Diagnosis includes a wide variety of entities including: Psoriasis, Actinic Keratosis, Bowen's Disease, Squamous Cell Carcinoma, and Melanoma. Less commonly: Lichen Planus Annulare, Lichen Sclerosis et Atrophicus, Lupus Vulgaris, Pityriasis Rubra Pilaris, Acrokeratosis Verruciformis, Seborrheic Keratosis, Lupus Erythematosus, Basal Cell Epithelioma and Linear Verrucous Epidermal Nevus(ILVEN).[3]

First line therapy includes: Cryotherapy, 5-Flurouracil, Imiquimod, Curettage and Cautery, Photodynamic therapy, CO2 laser and Vitamin D analogues. Second line therapy includes mainly oral retinoids and also helps in reducing the risk of malignant transformation.

Conclusion:
Though the lesions mimicked Lupus Vulgaris clinically; it was proved histologically to be Porokeratosis of Mibelli.

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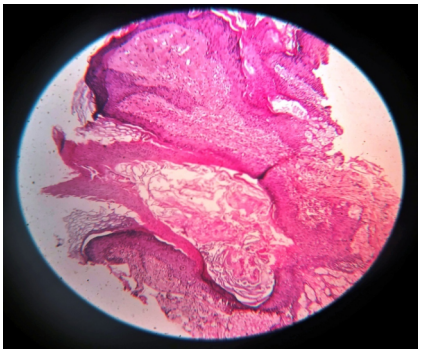
Conflict of Interest: The authors declare that they have no conflict of interest.

Figure 1: Clinical photograph showing a total number of four pigmented plaques of varying sizes, with the largest measuring about 3×2cmpresent over the center of the nose. Lesions appear pigmented, with mild scaling with a verrucous surface, peripheral raised border, and mild ulceration noted over the lesions.

Figure 2: Histopathology picture showing Hypogranulosis (Black arrow), Hyperkeratosis & Parakeratosis (Red arrow) and Cornoid Lamella (Green arrow).



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