

Case Report

Nevoid Porokeratosis Surmounted By Squamous Cell Carcinoma: A Rare Presentation

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Abstract: Porokeratosis is a clonal disorder of keratinization, a premalignant condition, characterized by one or more atrophic patches surrounded by a clinically and histologically distinct ridge-like border called 'cornoid lamella'. Nevoid porokeratosis presents as usually small lesions and this is usually not associated with any malignancy. In this case the patient, 55 year old male, present with unusually large lesions of Nevoid porokeratosis with squamous cell carcinoma changes, having family history and confirmed by biopsy.

Keywords: Porokeratosis, cornoid lamella

INTRODUCTION

Porokeratosis (PK) is a clonal disorder of keratinization, a premalignant condition, showing one or multiple atrophic patches surrounded by a clinically and histologically distinct ridge-like border called 'cornoid lamella'. The common clinical variants include classical plaque-type porokeratosis of Mibelli (PM), disseminated superficial porokeratosis, linear porokeratosis, porokeratosis Palmaris ET plantar is disseminate and punctate porokeratosis. Nevoid porokeratosis is a rare variant presents as usually small lesions and is usually not associated with any

malignancy. We report a case of nevoid porokeratosis with unusually large lesions with squamous cell carcinoma changes.

CASE REPORT

A 55 year old male patient presents with multiple large round to oval 15 to 20 cm in size hyperkeratotic plaques, with atrophic centre and prominent border at right side of the back and right forearm since birth and having 5 to 7 cm large indurated ulcer with elevated margins since around 18 months.



Fig 1, 2, 3: 55 years old male with porokeratosis and squamous cell carcinoma changes

He has family history of similar type of illness in his maternal grandfather.

No abnormality in Chest X- ray and abdominal USG.

Histopathological findings are consistent with porokeratosis and squamous cell carcinoma.

A. Porokeratosis:

Acanthotic epidermis, hyperkeratosis, focal parakeratosis, *cornoid lamellae*, dyskeratotic keratinocytes, focal lichenoid inflammation and mild perivascular inflammatory infiltrate and pigment incontinence in superficial dermis.

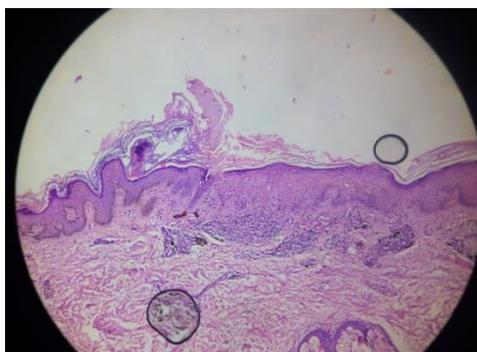


Fig. 4: Histopathology showing Porokeratosis with *cornoid lamellae*

B. Squamous cell carcinoma:

Moderately differentiated squamous cell carcinoma consisting of dysregulated keratinocytes with hyperchromatic atypical nuclei, some atypical mitotic figures, keratinization of individual cells and characteristic *malignant pearls* (horn Pearls).

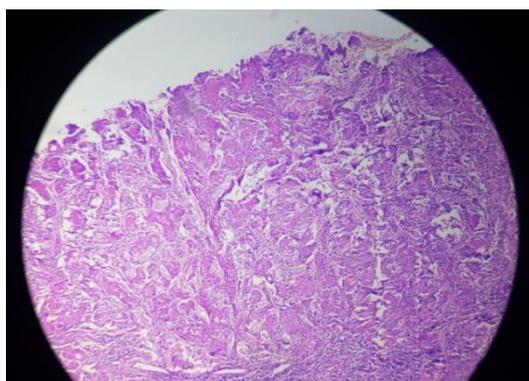


Fig. 5: Histopathology showing squamous cell carcinoma with *malignant pearls*

DISCUSSION

Porokeratosis has several clinical varieties including porokeratosis of Mibelli, giant porokeratosis, DSAP, porokeratosis palmaris ET plantaris; punctuate porokeratosis, and linear porokeratosis [1, 2]. Other rare variants include giant porokeratosis, hyperkeratotic,

verrucous, and nodular, punched out & reticulate type [1, 2]. Malignant transformation into squamous (most common), bowenoid or basal cell carcinoma has been described in all forms of porokeratosis with the highest risk for the linear form followed by giant porokeratosis [3, 4, 5]. Nevoid porokeratosis presenting as unusually large lesions which is a rare feature of Nevoid porokeratosis and this is usually not associated with any malignancy.

CONCLUSION

We report this case because of unusual presentation and rare association with squamous cell carcinoma.

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