Case Report

Pseudocarcinomatous changes in giant porokeratosis of mibelli': Histopathology as a valuable asset for early recognition of neoplastic changes – A case report with review

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ABSTRACT

Porokeratosis is defined as a rare disorder of keratinization caused by clonal expansion of keratinocytes and characterized by an expanding thread-like border of hyperkeratotic plaques or papules with atrophic center. It is a genetically heterogeneous disorder that is mostly inherited as autosomal dominant trait. It is characterized histologically by the presence of a thin column of parakeratotic cells "cornoid lamella" representing the active border. This is a case report of a patient suffering from giant porokeratosis of Mibelli who developed secondary changes over the lesion in due course of time arousing suspicion of malignancy which turned out to be benign histopathologically. The risk factors for malignant changes are usually persistence of the lesion for long time, radiation therapy, long-term exposure to ultraviolet radiation, electron beam therapy, and immunosuppression associated with malignancies such as lymphoma, HIV infection, or due to immunomodulatory drugs.

Key words: Malignancy, Porokeratosis of Mibelli, Pseudocarcinomatous changes

Porokeratosis is a rare mostly inherited disorder of keratinization caused by clonal expansion of keratinocytes and characterized by an expanding thread-like border of hyperkeratotic plaques or papules with atrophic center.

Neoplastic changes are not uncommon in porokeratotic lesions as reported in the literature and it is often referred to as a pre-malignant condition [1]. This case report shows the importance of histopathology in long-term lesions of porokeratosis as it helps in the early detection of neoplastic changes.

CASE REPORT

A 60-year-old male presented with hyperkeratotic irregular plaque with central atrophy extending from the right thigh and knee to lower leg for 20 years. The lesion started as small patch which spread gradually and peripherally with atrophy in the center, eventually forming a circinate and well-defined plaque surrounded by a hyperkeratotic wall. Gradually, several papules and pustules appeared in the margin of lesion which had a history of bleeding on and off. On examination, he had a 26 cm×15 cm^2 lesion over thigh and thinner lesions on the leg with central atrophy surrounded by a 0.5 cm raised hyperkeratosis wall (Figs. 1 and 2). We kept differential diagnosis of porokeratosis of Mibelli, Bowen's disease, and Lupus vulgaris. His routine investigations were within normal limits.

Histopathological examination of the lesion sample revealed a moderately dense superficial perivascular patchy lichenoid lymphocytic infiltrate with focal interface vacuolar change. In two small foci, the epidermis had an invagination of the floor which lacks granular layer, while the walls show hypergranulosis. Rising from the center of this invagination is a column of parakeratotic cells (cornoid lamella). Occasional dyskeratotic cells can be seen at the bottom of this parakeratotic column giving the impression of porokeratosis (Figs. 3 and 4).

Another section showed moderately dense superficial perivascular lymphocytic infiltrate along with irregular epidermal pseudocarcinomatous hyperplasia centered around follicular infundibula. The infiltrate tends to be denser around the hyperplastic appendages and encroaches on them obscuring the dermoepidermal junction. Wedge-shaped hypergranulosis is seen within hyperplastic appendages. Follicular infundibula are dilated and plugged by compact orthokeratotic corneocytes. One of the follicular infundibula shows focal dyskeratosis and parakeratosis. Pseudocarcinomatous hyperplasia reaches the base of the specimen giving the impression of plaque-type porokeratosis with pseudocarcinomatous hyperplasia (Fig. 5).

Access this article online	
Received - 24 July 2020 Initial Review - 10 August 2020 Accepted - 18 August 2020	Quick Response code
DOI: 10.32677/IJCR.2020.v06.i08.016	

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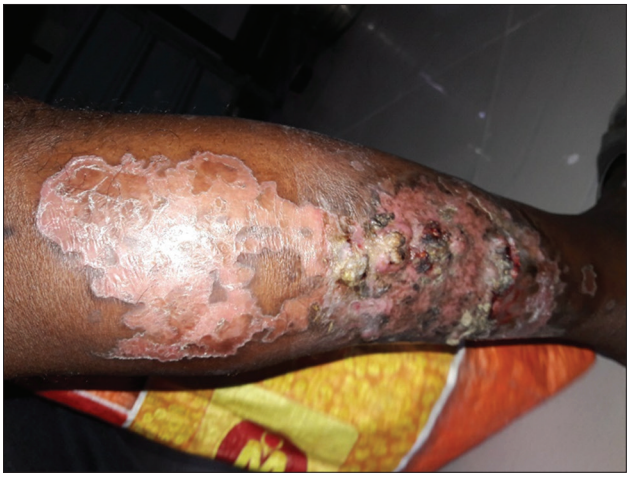

Figure 1: Atrophic patch of porokeratosis with secondary changes on lower side of the right leg

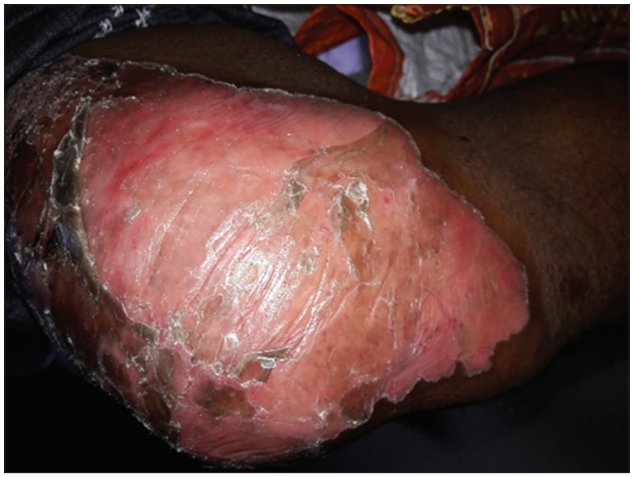

Figure 2: Atrophic patch of porokeratosis extending on the right knee joint

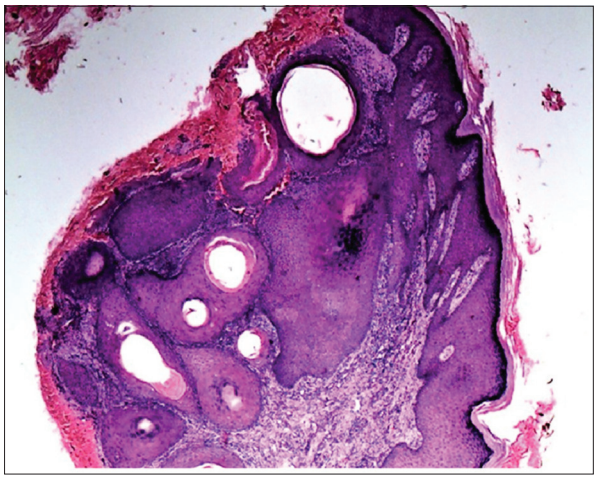

Figure 3: A moderately dense superficial perivascular patchy lichenoid lymphocytic infiltrate with focal interface vacuolar change and in two small foci the epidermis had an invagination of the floor which lacks granular layer, while the walls show hypergranulosis. (H&E Stain, ×40)

The patient was prescribed oral isotretinoin (0.5 mg/kg/day) and topical 1% 5-Fluorouracil alternate day for 1 month. After his lesion improved a bit, he was referred to the plastic surgery department for excision and graft replacement of the lesion after which he was lost to follow-up.

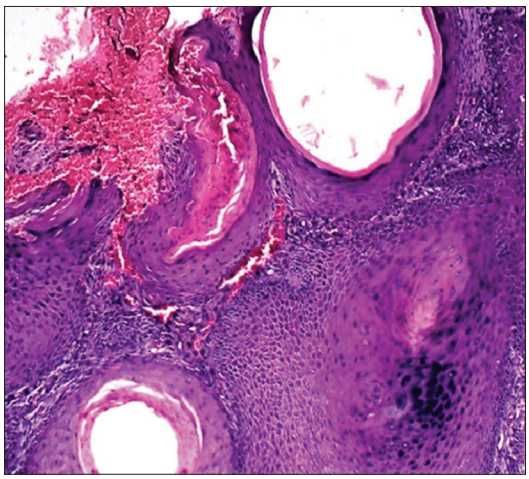

Figure 4: The center of invagination has a column of parakeratotic cells (cornoid lamella). Occasional dyskeratotic cells can be seen at the bottom of this parakeratotic column giving impression of porokeratosis. (H&E Stain, ×40)

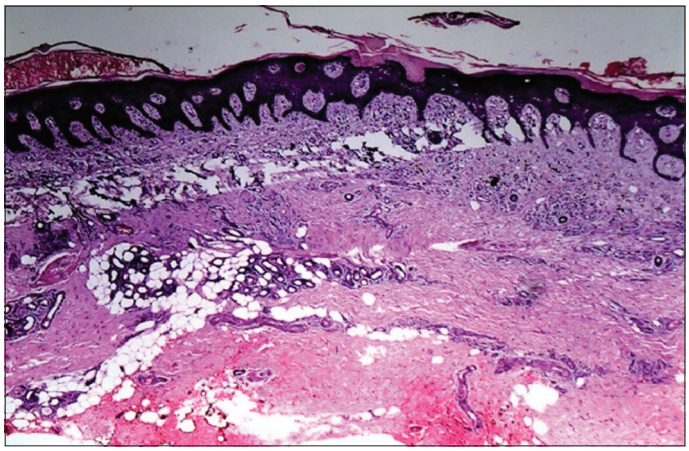

Figure 5: Pseudocarcinomatous hyperplasia reaches the base of the specimen giving the impression of Plaque type porokeratosis with pseudocarcinomatous hyperplasia.(H&E Stain, ×40)

DISCUSSION

Porokeratosis is considered a pre-malignant condition which may develop neoplastic changes in 6.9%–30% of cases, most frequently squamous cell carcinoma, and less frequently basal cell carcinoma [2]. Heterozygous mutations in the *MVK* gene have been reported as causative factor that encodes mevalonate kinases which are involved in the biosynthesis of cholesterol and isoprenoid and regulate keratinocyte differentiation [3]. The centrifugal progress of individual lesions is thought to reflect the migration of a clone of abnormal cells. The tumor suppressor protein p53 is overexpressed in the cornoid lamella.

The exact risk of cutaneous malignancy developing in porokeratosis is unknown, but approximate estimates of type of cancer and their prevalence in relation to type of porokeratosis is tabulated (Table 1). Some of the risk factors are persisting lesion

Table 1: List of malignant changes associated with various types of porokeratosis

S. No.	Type of porokeratosis	Chances of malignant transformation	Malignancy reported (% of total reported cases)
1.	Porokeratosis of mibelli	7.6%	*SCC-about 67% BCC- about 20% Bowen's disease-about 13%
2.	Disseminated superficial actinic porokeratosis	3.4%	*SCC-about 50% Bowen's disease-about 37% Amelanotic melanoma-about 12% (only one reported case)
3.	Linear porokeratosis	19%	*SCC-about 80% Bowen's disease- about 20%
4.	Porokeratosis palmaris ET plantaris disseminata	9.5%	SCC (all cases reported)
5.	Porokeratosis ptychotropica	--	SCC (only one case reported)
6.	Porokeratotic eccrine ostial dermal duct nevus	--	SCC (one case reported) Bowen's disease (one case reported)

#SCC: Squamous cell carcinoma, # BCC: Basal cell carcinoma, *-- Most cases of neoplasia are reported on sun-exposed areas or lower limb

of long time, radiation therapy, long-term exposure to ultraviolet radiation, electron beam therapy, and immunosuppression associated with malignancies such as lymphoma, HIV infection, or due to immunomodulatory drugs or following organ transplantation.

Any patient who presents with a long history of porokeratosis must be evaluated histopathologically to rule out early neoplastic changes. As in this case, pseudocarcinomatous changes may be considered benign, removal of the lesion is considered important to prevent development of frank carcinoma.

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Funding: None; Conflicts of Interest: None Stated.

How to cite this article: Sushantika S, Gupta SK. Pseudocarcinomatous changes in giant porokeratosis of mibelli': Histopathology as a valuable asset for early recognition of neoplastic changes – A case report with review. Indian J Case Reports. 2020;6(8):468-470.