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Case Report

Porokeratosis palmaris et plantaris disseminata - a rare entity

Kar Chinmay, Sarkar Prodip, Banerjee Sabyasachi, Biswas Rabindranath, Dutta Pallab*

Department of Dermatology, Malda Medical College and Hospital, West Bengal, India
* Centre for Healthcare Science and Technology, Indian Institute of Engineering Science and Technology, Shibpur, Howrah, West Bengal, India.

Abstract Porokeratosis is a well-known keratinization disorder where cornoid lamella is characteristically seen. There are seven clinical variants of which porokeratosis palmaris et plantaris disseminata (PPPD) is a very rare entity. Our PPPD case was sporadic and diagnosed early at 35 years of age. The lesions started on trunk and disseminated to extremities, face, oral cavity, palms and soles. Involvement of oral mucosa made easier to differentiate our case from disseminated superficial porokeratosis. The response was good with isotretinoin therapy.

Key words
Porokeratosis, porokeratosis palmaris et plantaris disseminata, cornoid lamella.

Introduction

Porokeratosis is a morphologically distinct disorder of keratinisation. It is characterised clinically by hyperkeratotic papules or plaques with raised and advancing edge, which histologically corresponds to a column of parakeratotic cells. There are seven clinical variants.[1] Among those, porokeratosis palmaris et plantaris disseminata (PPPD) is a very rare variety, originally described by Guss *et al.*[2] in 1971. This variety is usually inherited in an autosomal dominant manner, though some sporadic cases have been reported. More than one type of porokeratosis may be found in same patient.[3] Different types of porokeratosis may also be found in multiple members of an affected family.[4] Malignant transformation has been reported in all forms except punctate variety. The incidence of malignant transformation is 6.8-11% of all porokeratosis patients.[5] Older and linear variants have higher risk and actinic variant has lowest risk of malignant transformation.[6] Here we report a case of PPPD because of its atypicality and rarity.

Address for correspondence
Dr. Sabyasachi Banerjee,
Associate Professor, Department of Dermatology, Malda Medical College, Malda, West Bengal.
E-mail: sabyasachibanerjee2007@yahoo.co.in

Case Report

A 35-year-old male presented with numerous blackish brown papules and plaques all over the body including oral mucosa since last fifteen years. The lesions of trunk appeared first, followed by lesions on extremities, face, oral cavity, palms and soles, respectively. All the lesions appeared within one-year duration. The patient was farmer by profession. There was no history of long-term drug intake. None of his family was affected by similar disease.

On clinical examination, the lesions of trunk, extremities and face 2-7mm in diameter **(Figure 1a)**. The lesions were discrete, annular or polycyclic in shape. On careful examination at the periphery of lesion, well-demarcated ridge with longitudinal furrow was seen. There was slight depression at center without scaling. The lesions of palms and soles were discrete wart-like papule of 2-6mm

in diameter **(Figure 1b, 1d)**. A few of these lesions had characteristic border with furrow.

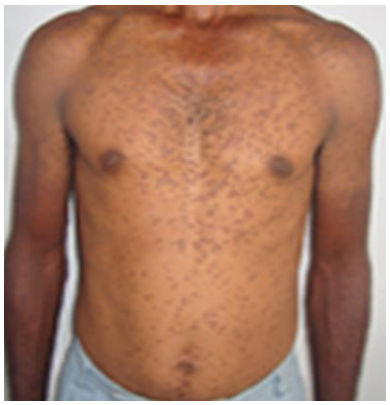

Figure 1 a) Porokeratosis of trunk (front) and upper limbs.

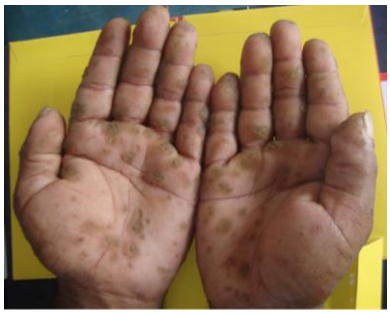

Figure 1 b) Porokeratosis of palms.

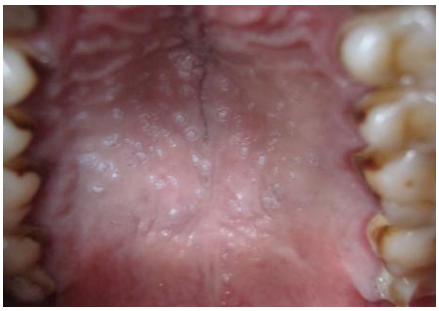

Figure 1 c) Porokeratosis of palate.

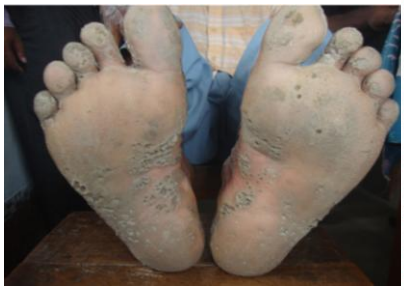

Figure 1 d) Porokeratosis of soles.

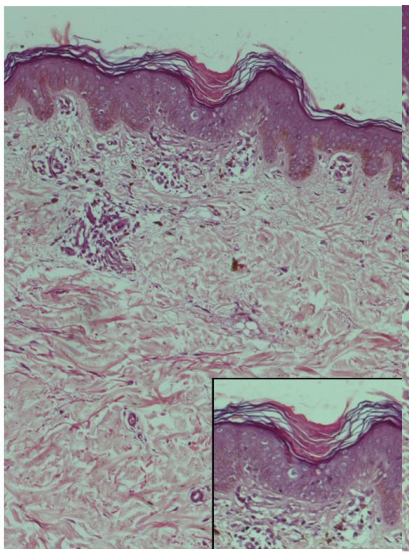

Figure 2 a) HPE of PPPD (back) showed basket weave orthokeratosis, the cornoid lamella with hypogranulosis or agranulosis in subcornoid lamella region in epidermis (X10). Inset: HPE focussing cornoid lamella, H&E stain (X20). (Nikon Eclipse Ti microscope).

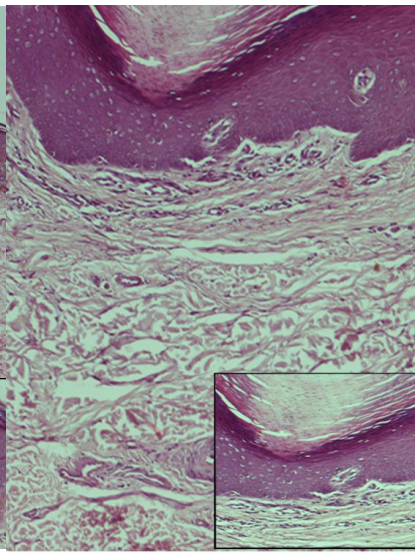

Figure 2 b) HPE of PPPD (palm) showed compact orthokeratosis with cornoid lamella in epidermis (X10). Inset: HPE focussing cornoid lamella (X20).

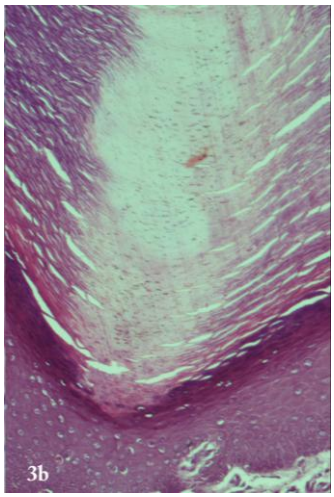

Figure 3 a) HPE of back focussing cornoid lamella, H & E stain, X80

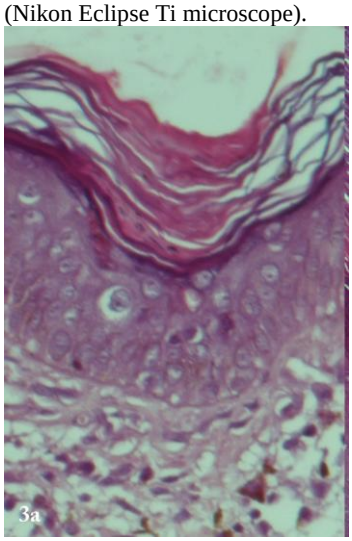

Figure 3 b) HPE of palm focussing cornoid lamella, H & E stain, X80

magnification. magnification.

Oral lesions with characteristic border were clearly seen over hard palate **(Figure 1c)**.

All basic blood investigations were within normal limit. Estimation of arsenic level from hair and nail were also normal. Punch biopsy was taken from back and histopathological examination (HPE) of hematoxylin and eosin-stained (H&E) section revealed basket weave orthokeratosis with a column of parakeratotic cells, the cornoid lamella, characteristic of porokeratosis. There was hypogranulosis or agranulosis in subcornoid lamella region along with mild dyskeratosis and a few vacuolar cells. Dermis showed chronic inflammatory infiltrates in perivascular regions **(Figure 2a, 3a)**. Punch biopsy from palm showed marked compact orthokeratosis with characteristic cornoid lamella **(Figure 2b, 3b)**. Other findings were similar to trunk lesion. So clinical and histopathological examination of our case revealed rare variant of porokeratosis palmaris et plantaris disseminata There was no malignant change in our case.

Our case was treated with isotretinoin as 30 milligram per day and about 30% lesional improvement was observed within four weeks. Some lesions became hyperpigmented macules following treatment. Mostly the lesions of trunk, extremities and face improved without any visible improvement of palmoplantar lesions. Regular follow-up is needed to note whether there is any complete clearance of disease, the duration of treatment and further recurrence.

Discussion

In 1893, Mibelli first described the classic form of porokeratosis. After that, various types of porokeratosis were discovered. After introduction of PPPD by Guss *et al*.[2] a few cases were reported. In PPPD, it is usually observed that porokeratosis starts from palms and soles and then disseminates to other areas including mucous membrane. It is mostly autosomal dominant with a few sporadic occurrences. Males are affected more than females and it usually starts in late teens to early twenties.[2] But due to very slow progress of the disease, diagnosis of PPPD is usually confirmed at a much later age. It also has malignant potential due to abnormal DNA ploidy in lesional epidermis like other porokeratosis.[7] The lesions of our case started typically at the age of twenty but appeared first on trunk. Then the lesions disseminated to other areas including oral mucosa within one year. The palms and soles were the last sites to be involved. Though it was not typical, a few reports were found with this sequence.[8] Clinically and histopathologically, our case had no abnormal change.

Lesions of trunk, extremities and face are similar to lesions of disseminated superficial porokeratosis (DSP) and disseminated actinic superficial porokeratosis (DSAP) clinically, morphologically and even histopathologically.[9] But palms, soles and mucosae are usually spared in DSP and DSAP.[10] Palmoplantar lesions of PPPD are close to palmoplantar porokeratosis (PP).[11] So PPPD is difficult to differentiate from DSP or DSAP with PP. But with involvement of any mucosa, PPPD can easily be differentiated from DSP or DSAP with PP. So our case is a classical case of PPPD. This PPPD should also be differentiated from systematized, widespread form of porokeratotic eccrine ostial and dermal ductal nevus (PEODDN). This PEODDN is a rare, benign hamartoma of eccrine sweat gland with porokeratotic histology and clinically presents as keratotic papules or plaques with central plugged pits.[12] The sporadic occurrence of PPPD is also rare. We have found only eight previous sporadic cases of PPPD in English literature **(Table 1)**. Our sporadic case was diagnosed at the age of thirty five, which is, most probably the second youngest reported age.[8,13] Successful treatment

of PPPD by isotretinoin has been reported earlier.[14] Our case also showed visible improvement with isotretinoin.

Table 1 Sporadic cases of PPPD

Sr. No.	*Authors, year of publication*	*Age of presentation (years)*	*Sex*	*First site of involvement*
1	Kaur and Singh (1993) [13]	30	F	Shins
2	Sawheny *et al*. (1995) [15]	65	M	Trunk
3	Lucke *et al*. (1998) [16]	38	F	Palms and soles
4	Seishima *et al*. (2000) [17]	63	M	Palms
5	Jih (2003) [18]	66	F	Trunk and extremities
6	Jensen *et al*. (2005) [19]	65	M	Distal arms and legs
7	Sengupta *et al*. (2005) [20]	40	M	Groins
8	Hartman *et al*. (2010) [21]	73	F	Palms and soles
9	Kar *et al*. (2016)	35	M	Trunk

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