Giant Porokeratosis: Report of Three Cases

Abstract:
Giant porokeratosis (PK) is a rare entity.Many consider it as a variant of PK of Mibelli, while others describe it as a separate variant. The diameter may range between 10 and 12 cm and the wall of hyperkeratotic ridge may be upto 1 cm. High tropical temperature and long duration of outdoor activities without adequate clothing are known influencing factors of PK. To the best of our knowledge, only five cases of solitary giant PK and a case of disseminated giant PK have been documented from India. We report three cases of giant PK for their rarity.

Keywords: *Giant porokeratosis, high temperature, porokeratosis of Mibelli*

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Introduction

Porokeratosis (PK) is characterized clinically by one or more annular plaques with central atrophy and surrounding elevated hyperkeratotic ridges that correspond to cornoid lamellae in histopathology. Five important clinical variants are recognized: disseminated superficial PK comprising both actinic and nonactinic variants (most common), classic PK of Mibelli (second most common), linear PK, punctate PK, and PK palmaris et plantaris disseminata (thought to be a subtype of punctate PK). Other rare variants are giant PK, PK ptychotropica (verrucous lesions affecting buttocks),[1] reticulated PK,[2] pruritic papular type,[3] pigmented porokeratosis,[4] prurigo nodularis-like PK,[5] etc. We report three rare cases of giant PK.

Case History

Case 1

A 45-year-old woman, farmer by profession, presented with 20 cm × 9 cm plaque on sole [Figure 1] and lateral borders of the right leg [Figure 2]. It had distinct ridge at periphery. The lesion started on right ankle as small coin-shaped annular lesion about 15 years back which gradually spread to involve the plantar and lateral surfaces of right foot. She used to work in fields with bare feet. There was no family history of this disease. With this presentation, the patient was provisionally diagnosed to have giant PK.

Case 2

A 51-year-old man, rickshaw driver by profession, presented with a 41 cm × 9 cm plaque with a distinct peripheral ridge on right lower leg. The lesion started on the dorsum of foot about 12 years ago. Gradually, it progressed upwards and involved the whole of right leg below the knee [Figure 3]. Everyday he used to work for 10hours in the sun. He was provisionally diagnosed to have giant PK.

Case 3

A 58-year-old male laborer , presented with rough scaly 23 cm × 13 cm plaque on right lower back [Figure 4]. Ten years back, he noted a small annular lesion. In the first year, it spread peripherally to a 2 cm × 2 cm plaque with elevated ridge. Then, it remained static for next 7 years. In last 2 years, it started to spread suddenly. He had to work with bare upper body for 8–10 hours daily and often felt itching and burning sensation when mercury got high. He was also provisionally diagnosed to have giant PK.

These three cases presented in dermatology department of a government hospital in Bankura, West Bengal, India. None of them had received any treatment before. The first two cases were completely asymptomatic while the third patient experienced itching and burning sensation under the sun. The lesions increased in summer.

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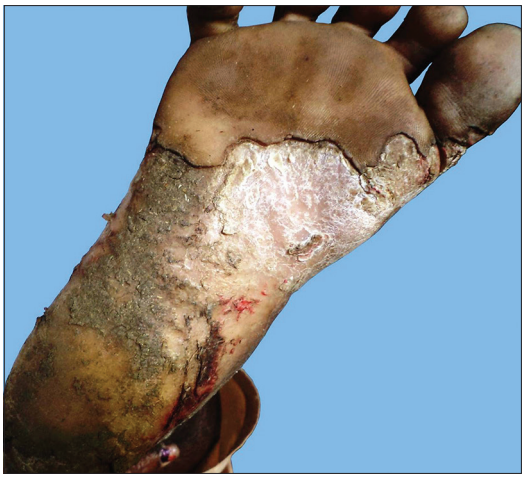

Figure 1: Case 1: porokeratosis involving plantar surface of right leg

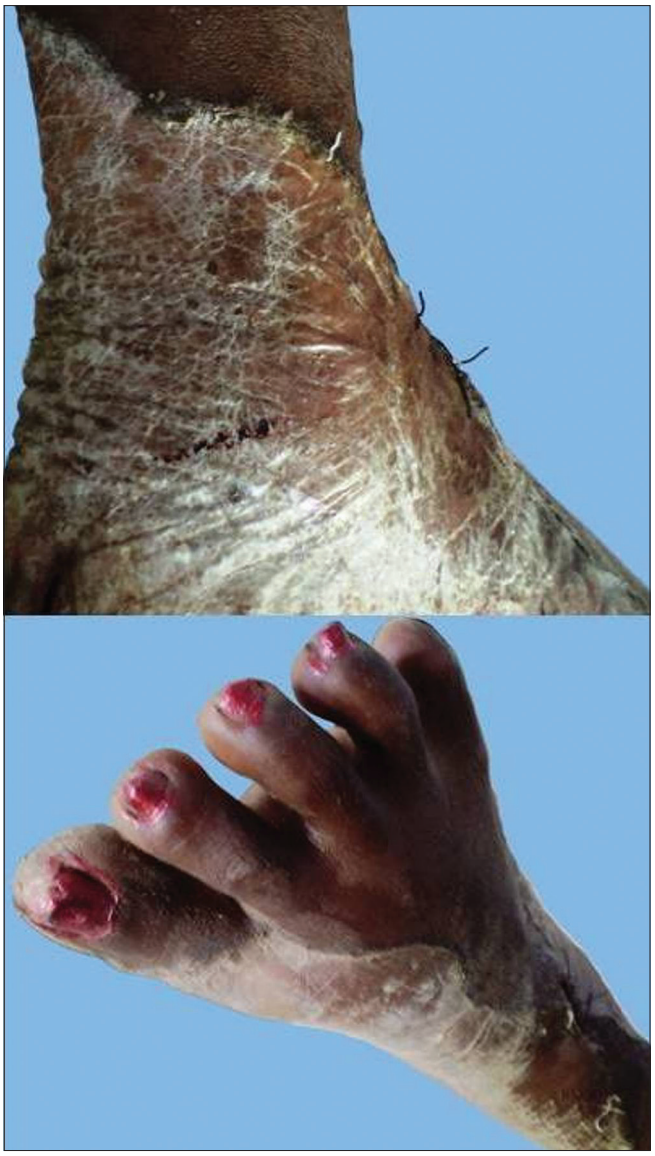

Figure 2: Case 1: porokeratosis involving right lateral foot

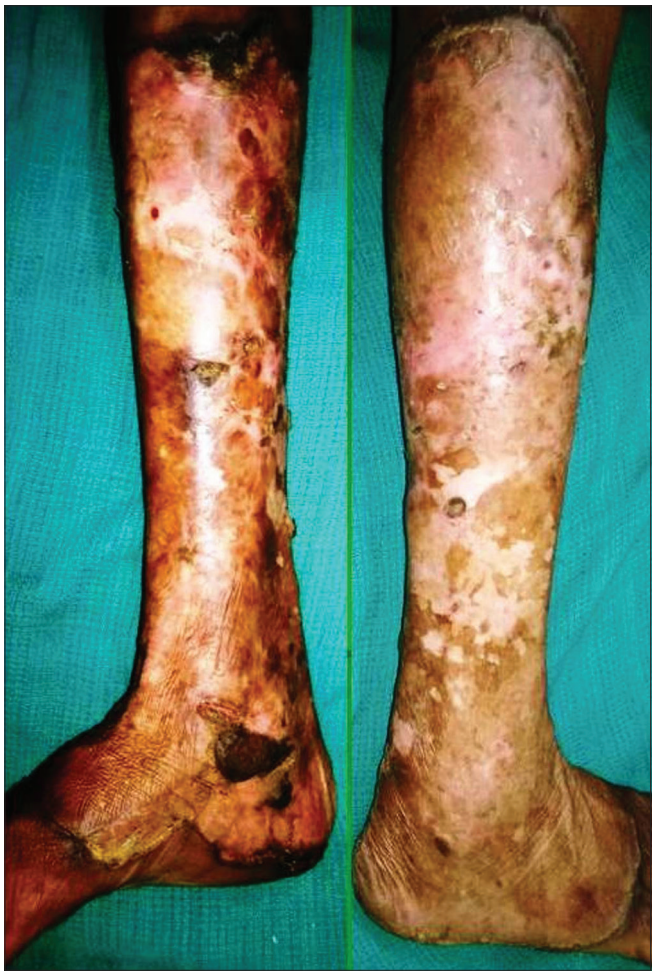

Figure 3: Case 2: porokeratosis involving right lower leg and extending to foot

The patients were investigated thoroughly. Complete blood count, erythrocytic sedimentation rate, fasting lipid profile, liver function tests, renal function tests, chest X-ray, and blood sugar tests (fasting and postprandial) were all within normal limits. HIV status was negative and they were alls immunocompetent. We also considered plaque type of lupus vulgaris, psoriasis, DLE, and intraepidermal squamous cell carcinoma in our differential diagnoses. Biopsy specimens from all patients revealed cornoid lamellae, underlying absent granular and chronic inflammatory cells in papillary dermis [Figures 5a, b, 6a, b and 7], confirming our provisional diagnosis of PK.

They were advised photoprotection, capsule isotretinoin 20 mg daily and topical halobetasol 0.05% application at night daily. However, the first two cases were lost to follow up. In the third case, we did partial excision, cryotherapy, and subsequently applied 0.05% tretinoin cream and 5% fluorouracil cream alternate nights. There were side effects like erythema, pruritus, oozing, crustation, and edema after every 3–4 weeks. Then, topical 5-fluorouracil had to be stopped temporarily and restarted after healing of the wound. After three episodes like these, the patient was lost to follow up. However, a

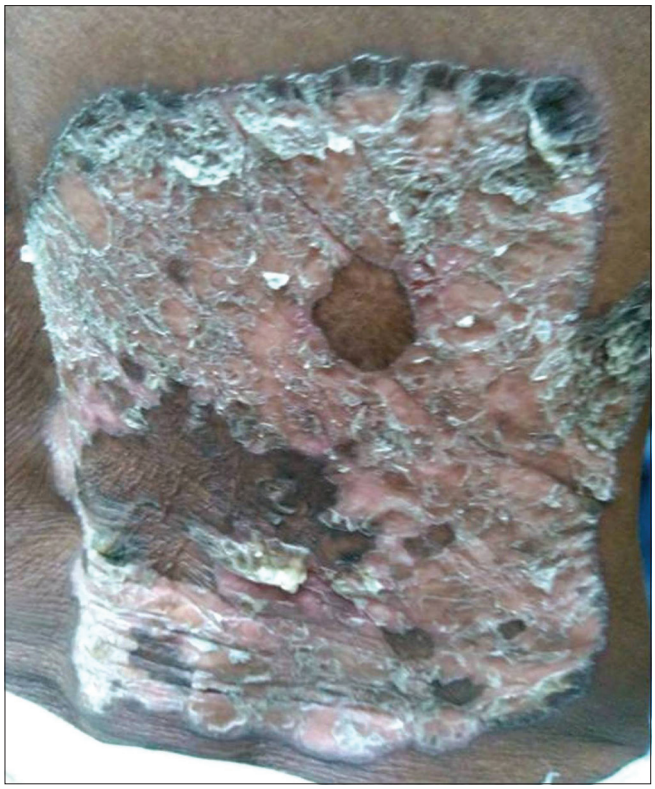

Figure 4: Case 3: porokeratosis on right lower back

moderate improvement could be appreciated over a span of 5 months of treatment.

Discussion

Giant PK is a rare entity.[6-8] Many consider it as a variant of PK of Mibelli,[9] while others describe it as a separate variant. The diameter may range between 10 and 12 cm and the wall of hyperkeratotic ridge may be upto 1 cm.[7] Interestingly the histopathology slide of case no 3 revealed three adjacent cornoid lamellae. Finding of multiple cornoid lamellae is not uncommon; may be underreported. When PK spreads by centrifugal expansion, probably it leaves its footprint behind as extra cornoid lamella.

Dermoscopy, not done in our cases, is a great tool in diagnosis. It reveals double white tracks with brownish discoloration within the furrow and a white homogeneous area at the center, corresponding to an atrophic epithelium. Red dots, globules, and lines (corresponding to enlarged capillary vessels) can be seen through the atrophic epithelium.[10] Staining of the skin surface with whiteboard marker (the furrow ink test) will accentuate the dermoscopic findings and highlight hair follicles and acrosyringia (seen as multiple open pores), some with keratotic plugs.[11] Noninvasive, high-resolution imaging techniques such as optical coherence tomography and reflectance and fluorescence confocal microscopy may identify characteristic cornoid lamella[12] or differentiate between actinic keratosis and disseminated superficial actinic porokeratosis.[13]

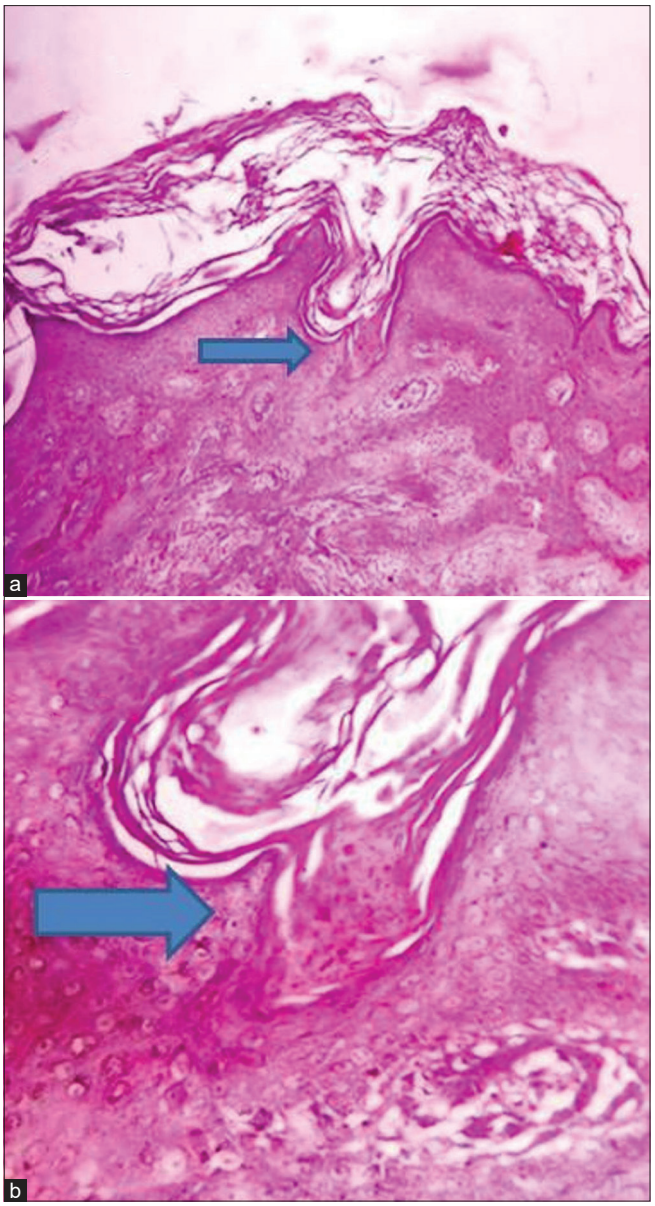

Figure 5: (a): Arrow showing cornoid lamella, absent granular layer with mild dermal dyskeratotic cells (hematoxylin and eosin, ×40); (b): magnified view of (a) (hematoxylin and eosin, ×400)

It is essential that a biopsy specimen be taken from the peripheral, raised, hyperkeratotic ridge to demonstrate the cornoid lamella. An incisional biopsy perpendicular to the border is preferred over punch biopsy as one may lose the coronoid lamellae during processing of the later. Reed *et al*. proposed a new three-step biopsy technique that allowed for proper orientation of the specimen during processing[1]: drawing a line perpendicular to the rim of the lesions,[2] then performing a punch centered at the intersection of the drawn line and coronoid lamellae,[3] then sectioning along the perpendicular line. The laboratory acquisition should mention transection,

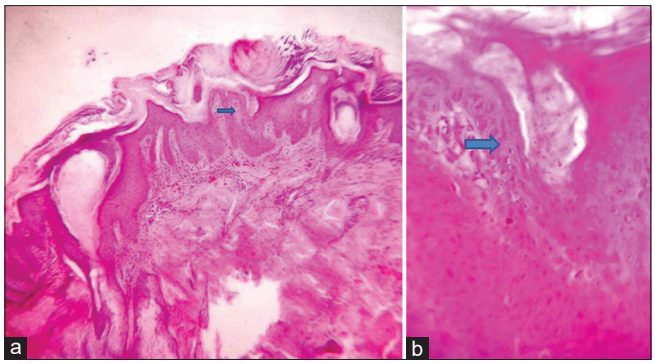

Figure 6: (a): Arrow showing cornoid lamella, diminished granular layer, underlying dermal inflammatory infiltrate (hematoxylin and eosin, ×40); (b): magnified view of (a) (hematoxylin and eosin, ×400)

and the cut sides should be placed down when processed.[14]

PK is a premalignant condition, with reports of malignant transformation from 7.5% to 11.6%.[15] The highest risk is in linear PK followed by giant PK. Giant PK is reported with dysplasia forming cutaneous horns[16,17] or squamous cell carcinoma[8,9,15-19] that might even metastasize.[19] Hence, giant PK should be ideally treated early. There are various modalities of treatment like topical 5-fluorouracil (FU), imiquimod, oral retinoid, CO_2 laser ablation, 585-nm pulsed dye laser radiation, Grenz ray radiation, Nd: YAG laser radiation, cryotherapy, dermabrasion, surgical excision, and electrodesiccation. Of these, surgical excision may be the best possible treatment while topical 5FU[20,21] and cryotherapy[22] are also good options.

To the best of our knowledge, only five cases of solitary giant PK[7,15-17,23] and a unique case of disseminated giant PK[24] have been documented from India, and primary authors of four of them were from West Bengal. Geographic location or race does not play an important role in the distribution of PK. We earlier mentioned that majority of cases of PK in India were reported from West Bengal.[24] The higher incidence of PK may be attributed to the higher tropical temperature. In our patients, the other influencing factor might be their occupation, i.e., long time outdoor activities without proper clothing.

The first case report of hypertrophic PK in India,[25] rare coexistence of different morphological variants of the hypertrophic variant[26] and the first case report of disseminated giant PK as well were made by from Bankura.[24] We report these three cases of giant PK for their rarity.

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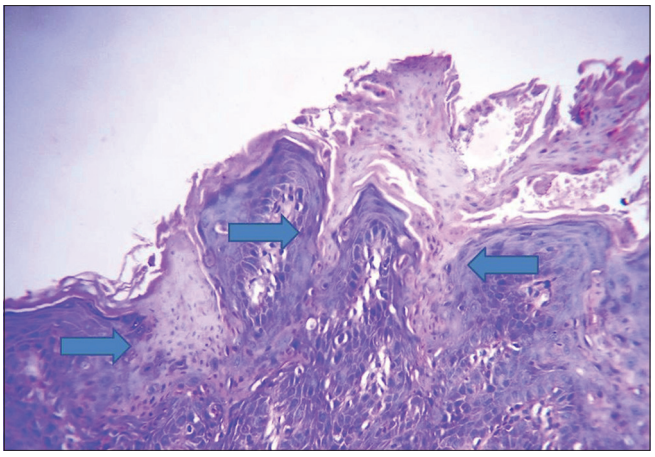

Figure 7: Arrows showing 3 adjacent cornoid lamellae with diminished underlying granular layers and dermal inflammatory infiltrates (hematoxylin and eosin, ×40)

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. McGuigan K, Shurman D, Campanelli C, Lee JB. Porokeratosisptychotropica: A clinically distinct variant of porokeratosis. J Am Acad Dermatol 2009;60:501-3.
2. Helfman RJ, Poulos EG. Reticulated porokeratosis. A unique variant of porokeratosis. Arch Dermatol 1985;121:1542-3.
3. Kanekura T, Yoshii N. Eruptive pruritic papularporokeratosis: A pruritic variant of porokeratosis. J Dermatol 2006;33:813-6.
4. Tan TS, Tallon B. Pigmented porokeratosis. A further variant? Am J Dermatopathol 2016;38:218-21.
5. Kang BD, Kye YC, Kim SN. Disseminated superficial actinic porokeratosis with both typical and prurigo nodularis-like lesions. J Dermatol2001;28:81-5.
6. Bozdağ KE, Biçakçi H, Ermete M. Giant porokeratosis. Int J Dermatol2004;43:518-20.
7. Raychaudhury T, Valsamma DP. Giant porokeratosis. Indian J Dermatol Venereol Leprol 2011;77:601-2.
8. Li JH, Yang ZH, Li B, Chen HD. Squamous cell carcinoma arising from giant porokeratosis. Dermatol Surg 2011;37:855-7.
9. Gotz A, Kopera D, Wach F, Hohenleutner U, Landthaler M. Porokeratosis Mibelli gigantea: Case report and literature review. Hautarzt 1999;50:435-8.
10. Zaballos P, Puig S, Malvehy J. Dermoscopy of disseminated

superficial actinic porokeratosis. Arch Dermatol 2004;140:1410.
11. Uhara H, Kamijo F, Okuyama R, Saida T. Open pores with plugs in porokeratosis clearly visualized with the dermoscopic furrow ink test: Report of 3 cases. Arch Dermatol 2011;147:866-8.
12. Welzel J. Optical coherence tomography in dermatology: A review. Skin Res Technol 2001;7:1-9.
13. Ulrich M, Forschner T, Röwert-Huber J, González S, Stockfleth E, Sterry W, *et al*. Differentiation between actinic keratoses and disseminated superficial actinic porokeratoses with reflectance confocal microscopy. Br J Dermatol 2007;156(Suppl 3):47-52.
14. Reed C, Reddy R, Brodell RT. Diagnosing porokeratosis of Mibelli every time: A novel biopsy technique to maximize histopathologic confirmation. Cutis 2016;97:188-90.
15. Sengupta S, Das JK, Gangopadhyay A. Multicentric squamous cell carcinoma over lesions of porokeratosis palmaris et plantaris disseminata and giant porokeratosis. Indian J Dermatol Venereol Leprol 2005;71:414-6.
16. Sarma N, Boler AK, Bhattacharya SR. Familial disseminated plaque type porokeratosis with multiple horns and squamous cell carcinoma involving anal skin. Indian J Dermatol Venereol Leprol 2009;75:551.
17. Mandal RK, Das A, Kumar P. Giant porokeratosis with overlying cutaneous horn and squamous cell carcinoma. Indian J Dermatol Venereol Leprol 2017;83:66-7.
18. Vence L, Thompson CB, Callen JP, Brown TS. Giant porokeratosis with malignant transformation to squamous cell carcinoma. Dermatol Surg 2018;44:580-1.
19. Zhang F, Bai W, Sun S, Li N, Zhang X. Squamous cell carcinoma arising from giant porokeratosis and rare postoperative recurrence and metastasis: A case report. Medicine (Baltimore) 2020;99:e18697.
20. Nahm WK, Donohue KG, Danahy JF, Badiavas E, Falanga V. Systemic 5-fluorouracil producing an inflammatory response in porokeratosis. J Eur Acad Dermatol Venereol 2003;17:190-2.
21. McDonald SG, Peterka ES. Porokeratosis (Mibelli): Treatment with topical 5-fluorouracil. J Am Acad Dermatol 1983;8:107-10.
22. Dereli T, Ozyurt S, Ozturk G. Porokeratosis of Mibelli: Successful treatment with cryosurgery. J Dermatol 2004;31:223-7.
23. Vasudevan B, Chatterjee M, Grewal R, Rana V, Lodha N. A case of disseminated superficial porokeratosis associated with giant porokeratosis in pregnancy. Indian J Dermatol 2014;59:492-4.
24. Koley S, Mandal RK, Bar C. Disseminated giant porokeratosis and porokeratosis of Mibelli in Bankura and Bardhaman districts, West Bengal, India. Int J Dermatol 2014;53:1119-24.
25. Das S, Banerjee G. Porokeratosis masquerading as lupus vulgaris. Indian J Dermatol 2004;49:212-3.
26. Koley S, Sarkar J, Choudhary S, Dhara S, Choudhury M, Bhattacharya S. Different morphological variants of hypertrophic porokeratosis and disseminated lesions of porokeratosis of Mibelli: A rare co-existence. Indian J Dermatol Venereol Leprol 2011;77:199-202.