Case report:

Disseminated superficial actinic porokeratosis - a rare case report

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Abstract

Porokeratosis is a heterogeneous group of keratinization disorders characterized by papules and plaques with central atrophy and a hyperkeratotic margin. We describe a woman who had DSAP or disseminated superficial actinic porokeratosis, whose lesions appeared as itchy plaques and papules. Clinical evidence suggested the diagnosis and histologically confirmed. The pruritus brought on by the condition was treated symptomatically, Her skin lesions will be clinically monitored. This rare dermatosis requires clinical and histopathological correlation to diagnose at an early stage and to avoid under-reporting.

Keywords: Cornoid lamella, disseminated superficial porokeratosis, porokeratosis

Introduction

Porokeratosis is a rare keratinization disorder that has the clinical appearance of papules encircled by a clearly defined peripheral keratotic wall, corresponding histologically to a "cornoid lamella".[1] Disseminated superficial actinic porokeratosis (DSAP), disseminated superficial porokeratosis, linear porokeratosis, porokeratosis of Mibelli, porokeratosis palmaris et plantaris disseminata, and punctate porokeratosis are some of the common clinical variations of porokeratosis; less common porokeratosis subtypes are Eruptive bullous, follicular, genitogluteal, lichen planus-like, porokeratotic acanthoma, porokeratotic adnexal ostial nevus, and pruriginous. The most prevalent clinical form of porokeratosis is DSAP, characterized by the bilateral appearance of numerous papules and plaques over sun-exposed areas, particularly on the distal limbs. [2] The lesions are slightly reddish or brownish in colour, and the surrounding ridge usually appears more accentuated than the circumscribed interior. As the lesions centrifugally expand, an annular or irregular configuration is typical.[3] DSAP has been reported in Patients with AIDS, cirrhosis, Crohn's disease, and organ transplant recipients.[1] DSAP is considered a rare disorder; no data exist on the prevalence of the disease in our country.

Case report

A 31-year-old woman, garments worker, presented to the outpatient department with a history of multiple brownish annular macules, patch and plaques over face, trunk, and upper limbs and lower limbs for the past 14 years. The lesions started appearing as patch, brown in colour, then slowly progressed in size as a plaque with a keratotic rim. initially over the face, followed by the chest, back, and upper limbs. The lesions gradually increased in size and number. There was no photosensitivity. Her

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Cite this Article:
Haque AKM R, Mahmud MM, Ashadullah SM, Haque MA, Mia MT. Disseminated superficial actinic porokeratosis - a rare case report. Ban Acad Dermatol. 2023; 03 (02): 81-83

Available at: www.jbadbd.com
An official publication of Bangladesh Academy of Dermatology (B.A.D.)

younger brother has same kind of illness for the last 7 years.

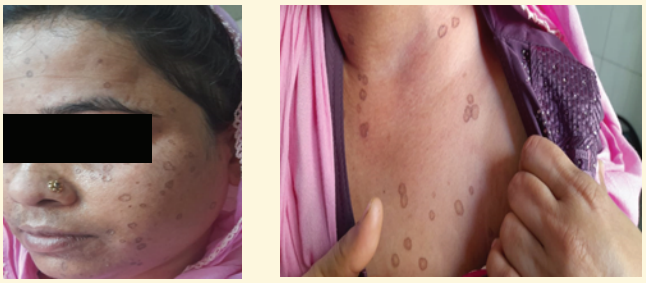

Fig 1: Multiple hyperpigmented annular macules,patchs & plaques over the face,chest

On physical examination the lady appeared to have normal well-being, vitals are within normal limit, cutaneous examination shows hyperkeratotic annular plaques with raised margin over the face, neck, trunk and arms. Other systemic findings were normal. Laboratory findings including Complete Blood Picture, Urine Routine Examination, SGPT, Serum creatinine, HBsAg, Anti HIV (1 and 2), CXR, USG of whole abdomen, were normal. Histopathological study showed epidermis is thin, show mild hyperkeratosis, parakeratosis and focal vacuolar alteration of basal layer. The dermis shows mild perivascular infiltration of chronic inflammatory cells including a few melanophages (Fig. 2).

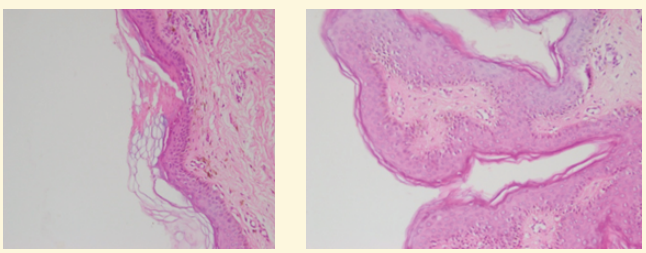

Fig 2: Histopathology of the trunk lesion showing vertical layered parakeratosis, the cornoid lamella and vacuolated keratinocytes

Discussion

Porokeratosis is an abnormal differentiation of keratinocytes rather than hyperproliferation. It can appear as a single lesion or multiple lesions, and as a localized or disseminated form. The cornoid lamella, which histopathologically represents the raised border, is made up of a thin vertical tier of parakeratoses.[4-5] Different forms of porokeratosis frequently appear in particular age groups. Classical porokeratosis of Mibelli and linear porokeratosis typically appear during infancy or childhood, whereas disseminated palmoplantar porokeratosis and punctate palmoplantar porokeratosis typically appear during adolescence. DSAP typically first appears in adulthood. Males are more likely to develop porokeratosis of Mibelli, genital porokeratosis, and punctate porokeratosis, whereas females are more likely to develop DSAP.[6] Our patient is an adult female. Although the pathogenesis of porokeratosis is still not fully understood, DNA polyploidy, as well as keratinocyte and fibroblast sensitivity to ionizing radiation in the affected skin, have been shown to play a role in the pathogenesis.[7] Porokeratosis has been linked to a number of risk factors, including genetic predisposition, exposure to UV rays, and immunosuppression.[8] Our patient has a positive family history. Disseminated superficial actinic porokeratosis lesions begin as asymptomatic or mildly itchy pink to brown papules and macules with raised borders in sun-exposed areas. The most frequently affected body parts are the legs, forearms, shoulders, and back, the face can rarely be affected, the palms and soles are unaffected. When exposed to sunlight, the lesion typically gets worse, and pruritus can get worse.[9] But, in our case, the lesions 1st appear on the face and has no photosensitivity. A skin biopsy should be performed that includes the lesion's border. The cornoid lamella is a column of parakeratotic cells that corresponds to the raised border of the lesion. The granular layer beneath this column can be thin or absent. There is dyskeratosis in the epidermis beneath the cornoid lamella. Spongiosis can be present. Our patient's histopathology report shows a thin epidermis, mild hyperkeratosis, parakeratosis and focal vacuolar alteration of the basal layer. The dermis shows mild perivascular infiltration of chronic inflammatory cells including few melanophages which is suggestive of DSAP. Precancerous status is assigned to these lesions. The likelihood of malignant transformation into basal cell carcinoma or squamous cell carcinoma is between 7.5 and 10%.[10]

Conclusion

The case in this report had late-onset DSAP. Her diagnosis was clinically suspected, and microscopic analysis of a tissue biopsy specimen confirmed it. The lesions run the risk of developing into malignancies. photoprotective measures should be encouraged to reduce the risk of further skin

damage . Patients should also visit frequently so that their lesions can be monitored.

Conflict of Interest

None

Funding source

None

Patient Consent

Taken from parents.

IRB approval status

Not applicable

References

1. Eralp A, Kaymak Y. Disseminated superficial actinic porokeratosis: a case report. Turkish Journal of Medical Sciences. 2009;39(3):491-3.
2. Waqar MU, Cohen PR, Fratila S, Waqar MU. Disseminated superficial actinic porokeratosis (DSAP): a case report highlighting the clinical, dermatoscopic, and pathology features of the condition. Cureus. 2022 Jul 16;14(7).
3. Sotoodian B, Mahmood MN, Salopek TG. Clinical and dermoscopic features of pigmented disseminated superficial actinic porokeratosis: case report and literature review. Journal of Cutaneous Medicine and Surgery. 2018 Mar;22(2):229-31.
4. Rao AG, Lakshmi TS, Haritha S. Disseminated superficial porokeratosis. Indian J Dermatol Venereol Leprol 2002;68:284 5.
5. 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.
6. Ramakrishnan R, Vignesh TA, Durai PC, Narasimhan M. A rare case of disseminated superficial porokeratosis-Case report. Journal of Family Medicine and Primary Care. 2022 Mar;11(3):1195.
7. Imakado S, Otsuka F, Ishibashi Y, Ohara K. Abnormal DNA ploidy in cells of the epidermis in a case of porokeratosis. Arch Dermatol 1988; 124: 331-2.
8. Kamal T, Mufti S, Ahmad TJ. Disseminated superficial actinic porokeratosis: A case report. Journal of Pakistan Association of Dermatologists. 2015;25(1):73-5.
9. Li X, Zhou Q, Zhu L, Zhao Z, Wang P, Zhang L, Zhang G, Wang X. Analysis of clinical and genetic features of nine patients with disseminated superfacial actinic porokeratosis. Zhonghua yi xue yi Chuan xue za zhi= Zhonghua Yixue Yichuanxue Zazhi= Chinese Journal of Medical Genetics. 2017 Aug 1;34(4):481-5.
10. Le C, Bedocs PM. Disseminated Superficial Actinic Porokeratosis. [Updated 2023 Jun 25]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2023 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK459202/