Facial Disseminated Superficial Actinic Porokeratosis

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ABSTRACT

Porokeratosis is a disorder of keratinization and is genetically transmitted. It is characterized by one or more atrophic macules or patches and surrounded by a distinctive hyperkeratotic ridge-like border. Multiple clinical variants of porokeratosis exist. It is histologically characterized by the presence of a cornoid lamella. The cornoid lamella is due to an expanding number of unusual keratinocytes. Here, we present a case report of porokeratosis over the nose in a 22-year-old female.

Keywords: Porokeratosis, Cornoid lamella, Ridge-like border.

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INTRODUCTION

Porokeratosis is rare and inherited as an autosomal dominant trait. However, sporadic cases are also known to occur. It was first described by Mibelli and Respighi in 1893. Different clinical types of porokeratosis namely: classic porokeratosis of mibelli, disseminated superficial actinic porokeratosis (DSAP), linear porokeratosis, disseminated superficial porokeratosis (DSP), porokeratosis palmaris et plantaris disseminate and linear porokeratosis. Apart from these five clinical variants, a number of atypical morphological forms, such as facial porokeratosis, giant porokeratosis, punched out porokeratosis, hypertrophic verrucous porokeratosis and reticulate porokeratosis also has been reported.1

CASE REPORT

A 22 years old female presented with lesions over the face of 3 years duration. These lesions were slowly progressing since 3 months and were asymptomatic in nature. The patient was otherwise healthy and appeared to be immunocompetent. There was no similar family history. The patient was housewife and spent most of the time indoors. There was no history of photosensitivity, any systemic complaints, atopy, trauma or any discharge from the lesion. The patient had not taken any medication for the same in the past.

On examination, there were multiple hyperpigmented plaques with central atrophy and raised edges with ridges over the nose of size of about 0.5 × 0.5 cm to 2 × 3 cm. No similar lesions were present elsewhere other than face. Routine laboratory tests were done and they were within normal limits (Fig. 1). Skin biopsy was done and histopathology showed hyperkeratosis, parakeratosis and keratin filled invagination with central parakeratotic column (cornoid lamella), scanty granular cells and dermal inflammatory cell infiltrate. Histopathology report was consistent with porokeratosis (Fig. 2).

The patient was treated with topical tretinoin 0.05% cream since it is known to decrease cohesiveness of abnormal hyperproliferative keratinocytes and modulate keratinocyte differentiation. After 3 months on follow-up, the patient showed improvement. She was asked to continue the same treatment and follow-up after 3 months, but, unfortunately, the patient did not turn up for follow-up after that.

DISCUSSION

Porokeratosis is an inherited autosomal dominant trait. However, sporadic cases are also known to occur. It is disorder of keratinization. Exact pathogenesis is not known but abnormal early keratinocyte apoptosis accompanied by dysregulation and terminal differentiation (keratinization) has been suggested for pathogenesis by cornoid lamella. Disseminated superficial...
actinic porokeratosis is the most common variant. It was first described by Chernosky and Freeman in 1967.\(^2\) Unlike the classic type of mibelli, the incidence of DSAP in females is double that in males. Porokeratosis usually occurs on the trunk or extremities and facial lesions are rare. Only 15% of DSAP patients have facial lesions.\(^3\) Two patients were reported with facial lesions by Nabai et al first in 1979.\(^4\) There have been a few reports of exclusive facial involvement in porokeratosis with skin lesions over or near the nose.\(^5,6\)

Risk factors for porokeratosis include genetic inheritance, ultraviolet radiation and immunosuppression.

Differential diagnosis includes: granuloma annulare, actinic keratosis, linear verrucous epidermal nevi, psoriasis, lichen planus, etc.

Various treatment modalities, like topical retinoids, 5-FU, vitamin D3 analog, imiquimod 5% cream, cryotherapy, photodynamic therapy, CO\(_2\) laser can be considered.\(^7,8\)

**CONCLUSION**

We suggest keeping in mind that porokeratosis as differential diagnosis of any annular facial lesions with distinctive, keratotic border and lesions resistant to topical treatment.

**REFERENCES**