CASE REPORT

Itchy lesions in pigmented skin

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SUMMARY

A 37-year-old woman with type VI skin presented with 1-year history of pruritic lesions affecting her arms, chest and legs. The lesions were approximately 5 mm in diameter, annular and with a raised border. A skin biopsy was performed which showed a diagnosis of disseminated superficial actinic porokeratosis. Porokeratosis is an unusual presentation in pigmented skin and there are very limited reports of this occurrence in the literature.

BACKGROUND

Porokeratosis is a rare group of primary epidermal keratinisation disorder characterised by a cutaneous plaque that progressively enlarges and is surrounded by a raised hyperkeratotic border. They are common in sun-exposed areas of fair skin (Fitzgerald Grading of types I–II) people. There are very few reported occurrences of porokeratosis in pigmented skin. We present a case of disseminated superficial actinic porokeratosis (DSAP) in type VI skin which remains exceedingly rare.

CASE PRESENTATION

A 37-year-old woman presented with 1-year history of new pruritic lesions affecting her arms, chest and legs. She had no relevant medical history or family history. On examination, multiple pigmented macules approximately 5 mm in diameter were present on her upper body. Annular lesions with a raised border were evident on her lower limbs (figures 1 and 2). Evidence of excoriations was also noted.

INVESTIGATIONS

A skin biopsy (figures 3 and 4) showed a hyperkeratotic stratified squamous epithelium and epidermal invagination with an overlying parakeratotic tier. The epithelium deep in the tiers showed vacuolation and was devoid of the granular layer.

DIAGNOSIS

Disseminated superficial actinic porokeratosis.

OUTCOME AND FOLLOW-UP

There is no known effective treatment of DSAP but our patient was treated with dovonex (calcipotriol) and effudix (fluorouracil) ointment and exhibited some flattening of her lesions.

DISCUSSION

Porokeratosis is a rare group of primary epidermal keratinisation disorder characterised by a cutaneous plaque that progressively enlarges and is surrounded by a raised hyperkeratotic border. The pathognomonic hallmark of this disease is the presence of the cornoid lamella found in the raised border on histology. There are many classifications of porokeratosis, but generally they appear in the following clinical forms: linear, isolated lesions (porokeratosis of Mibelli), DSAP and disseminated superficial porokeratosis (a variant similar to DSAP occurring in non-sun-exposed skin).¹

Since Chernosky and Freeman² first described DSAP, almost all reports have been in Caucasian patients. Women are primarily affected, the average age of the onset is 43 years and the average duration of the disorder is 8 years. Mensch and Hazen³ published the first case of DSAP occurring in patients with skin type VI and since then only a few cases have been subsequently reported in the literature.

Figure 1 Annular lesions on the lower limb.

Figure 2 Annular lesions on the lower limb.

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Figure 3 Skin biopsy under high magnification showing a hyperkeratotic stratified squamous epithelium.

DSAP has an autosomal dominant trait with decreased penetrance of 22%. In addition, risk factors include immunosuppression and UV light exposure. Length of exposure to the sun triggering the mutational events can explain the delayed expression of DSAP in adulthood. Studies by Happle⁴ suggest that a homozygous form may lead to earlier, more severe and larger sized porokeratosis and hence increased risk of skin cancer although definitive studies have yet to be conducted.

DSAP has not been reported on the skin that is completely protected from the sun such as the axilla and perineum. The

Figure 4 Skin biopsy under low magnification showing two areas of porokeratosis.

distribution is usually symmetrical with the extremities being most affected. Pruritis occurs in 50% of patients. The number of lesions can be anything from 3 to 700.

The ideal treatment for DSAP remains unknown. Psoralen drug in combination with ultraviolet A light (PUVA), liquid nitrogen, oral and topical retinoids have been effective in some cases.⁵

Our patient was treated with topical dovonex, effudix and emollients. There have been reports of successful treatment with dovonex and effudix in the literature⁶ and our patient similarly exhibited some flattening of the lesions and symptomatic improvement.

It is likely that we will be seeing more cases of DSAP in darker skin types although this presentation remains exceedingly rare.

Learning points

- ▶ Disseminated superficial actinic porokeratosis (DSAP) can occur type VI skin although it remains a rare presentation.
- ▶ Standardised effective treatment for DSAP remains unknown, but our patient responded to the use of dovonex and effudix.
- ▶ In cases where the diagnosis is unclear, a skin biopsy will reveal the presence of the cornoid lamella in the raised border, which is pathognomonic for this disease.

Contributors SH conceived the idea of the case report. RH prepared the draft and all authors were involved in the revising of the final manuscript.

Competing interests None.

Patient consent Obtained.

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