CASE REPORT

Verrucous porokeratosis (porokeratosis ptychotropica) with dermal amyloid deposits

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INTRODUCTION

Porokeratosis is a group of diseases characterized by the gross appearance of hyperkeratotic annular plaques with a ridge-like border and microscopically, cornoid lamellae. There are five main variants: porokeratosis of Mibelli, disseminated superficial actinic porokeratosis, porokeratosis palmaris et plantaris, linear porokeratosis, and punctate porokeratosis.

In 1995, Lucker et al described a 34-year-old man with a 9-year history of pruritic lesions confined to the natal cleft, which was termed "porokeratosis ptychotropica".1 In Greek, the word "ptyche" means fold and "trope" means turning. Later in 1999, Stone et al described a similar case, in which a 32-year-old man with a 13-year history of pruritic lesions also confined to the natal cleft. It was named "perianal inflammatory verrucous porokeratosis".2 Until now, this is still a rare variant of the disease. It is also termed porokeratosis ptychotropica, hyperkeratotic porokeratosis, and genitogluteal porokeratosis. In this report, we present a 43-year-old man with this rare disorder.

CASE REPORT

This 43-year-old, otherwise healthy man, presented with long-standing itchy skin lesions on the buttocks for more than 20 years. It first appeared as a single lesion on his right buttock at 15 years old. He went through a surgical excision of the lesion at a dermatology clinic. However, the skin lesion progressed and extended from the natal cleft and buttocks to the penis and scrotum. During the following years, he had been diagnosed as having psoriasis, viral warts, and condyloma. Various treatments were given including medical medications, surgical excision, cryotherapy, and electrocauterization at different hospitals and dermatology clinics. The lesion seemed to disappear initially, but recurred a few months later. During the past 28 years, the lesions had gradually increased both in size and number. There is no associated family history. The skin lesions were itchy and appeared reddish or brownish in color. During physical examination, numerous verrucous to flat papules and plaques were seen on buttocks, natal cleft, penis, and scrotum (Figure 1). Epi- dermodyplasia verruciformis was suspected clinically and a skin biopsy of the hyperkeratotic plaque on the left buttock was performed.

Histopathology from the biopsy specimen revealed compact hyperkeratosis and the presence of several parakeratotic columns with underneath scattered dyskeratotic keratinocytes, forming

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a cornoid lamella (Figure 2A–C). The epidermis was acan-thotic. Heavy pigment incontinence and eosinophilic amorphous globular materials were deposited diffusely in the papillary dermis (Figure 2D). The globular materials were positive for both congo red and high molecular weight cytokeratin (34βE12, for cytokeratin 1, 5, 10, 14; Figure 3). The cornoid lamella was negative for high molecular weight cytokeratin (stained keratin 1, 5, 10, 14 in the epidermis) and cytokeratin 7 (stained keratin in eccrine coil), but weakly positive for cytokeratin 5/6 (stained basal keratinocytes and follicular keratin) (Figure 4). The case was diagnosed as verrucous porokeratosis according to the clinical distribution and pathological presentation. Topical tretinoin cream for keratolysis and topical doxepin cream for relief of itching were prescribed. He was regularly followed-up at the nearby dermatology clinic. Although the itch improved, the lesions persisted. Oral retinoid was suggested if the response to topical therapy was poor but was not given at the time of writing.

**Discussion**

Clinically, it is a challenge to diagnose verrucous porokeratosis at the first examination. Instead of the characteristic rim seen in porokeratosis, the verrucous plaques of verrucous porokeratosis may often be mistaken as lesions caused by other diseases, such as condyloma acuminata, psoriasis, dermatophytosis, candidiasis, chronic eczema, acrodermatitis enteropathica, necrolytic migratory erythema, or epidermodysplasia verruciformis. Coincident with other reports, which required decades to make the definite diagnosis, it took more than 20 years for our patient to be correctly diagnosed. According to a review of 23 cases, verrucous porokeratosis has a predilection for the gluteal cleft, penis, scrotum, and vulva, and therefore often be called porokeratosis ptychotropica to emphasize the specific genitogluteal location. They may sometimes involve wider area including buttocks, thighs, and lower legs; as in our patient, the lesions had extensive butterfly-shaped plaques.
on the buttocks, surrounding the anus, scrotum, and extended to the thighs.

It is interesting that the immunohistochemical stains showed amyloid positive for congo red and high molecular weight cytokeratin (34BE12) in the papillary dermis. 34BE12 is an anticytokeratin antibody against cytokeratins 1, 5, 10, and 14. In a study of cytokeratin profiles in localized cutaneous amyloids, 34BE12 is the best polyclonal anticytokeratin antibody to immunolabel the amyloid deposits of primary or secondary cutaneous amyloidosis. Additionally, all of the four cases of porokeratosis included in this study stained positive by 34BE12. The keratin-based amyloid found in our patient is most likely to be secondary deposition due to an epidermal defect. According to a retrospective histopathologic study of 30 patients with porokeratosis, amyloid deposits were observed in 11 of 34 biopsies. Elderly patients and the chronic nature of the lesion were postulated to be the predisposing factors. The deposits of dermal amyloid in verrucous porokeratosis were previously reported in a few cases. These lesions are located on buttocks with extension to adjacent areas, which are frequent sites of friction. The patients may also scratch the rash due to itching. Therefore, the diffusely distributed amyloid in the papillary dermis may indicate frequent rubbing which contributes to secondary cutaneous amyloid deposition.

In 1893, Mibelli first named porokeratosis based on the prediction that the parakeratosis (keratosis) of the cornoid lamella was originated from the ostia of the eccrine ducts (poro). In fact, cornoid lamella had been found to be associated with hair follicle as well as acrosyringium. Follicular involvement is not that common and has been reported in association with porokeratosis of Mibelli, disseminated superficial actinic porokeratosis, and follicular porokeratosis. In a retrospective study, cornoid lamella related to follicular infundibula was found in 15.7% of 34/72 patients. In another retrospective study, the localization of cornoid lamella within adnexal orifices including the hair follicle and eccrine ostia was present in 41% (25/61) of the cases. The frequent occurrence of cornoid lamella in association with follicular infundibulum suggested this may not be merely coincidence. However, it is still unknown whether there is a secondary involvement of follicular infundibulum or starts primarily from hair follicle. The negative staining results for keratin 1, 5, 7, 10, 14 and weakly positive keratin 6 found in our case also suggested a follicular-origin cornoid lamella instead of epidermal or eccrine glandular origin.

Malignant change has been reported in 7–11% of patients with porokeratosis. Increased risk of malignant transformation was evidently seen in nonexposed skin, large porokeratosis lesions, previous radiation, long standing lesions, older patients, or linear porokeratosis. However, no cases of malignancy have been reported in verrucous porokeratosis to date. The presence of several aforementioned risk factors requires continuous follow-up in our patient.

In conclusion, verrucous porokeratosis represents a distinct variant of porokeratosis. It often progresses slowly in years and forms verrucous plaques over the perianal region with extension to gluteal cleft, buttocks, lower legs, and genital areas. The actual prevalence of verrucous porokeratosis is unknown due to being frequently misdiagnosed. A long-standing genitogluteal lesion should undergo biopsy to rule out verrucous porokeratosis.
References