Verrucous Porokeratosis of Mibelli on the Buttocks Mimicking Psoriasis

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The typical presentation of porokeratosis of Mibelli is of a solitary plaque with a prominent raised border cleaved by a central furrow. The central portion of the plaque is usually slightly atrophic. The plaques vary in size from a few millimeters to several centimeters in diameter and tend to be acrally distributed, though they can occur on any part of the body. We report an unusual case of verrucous porokeratosis of Mibelli, localized to the natal cleft, that mimicked psoriasis. This entity, though unusual, is not unique. Two similar cases of verrucous porokeratosis of Mibelli limited to the natal cleft region and resembling psoriasis have been reported in the British literature. Verrucous porokeratosis of Mibelli localized to the natal cleft appears to be a distinct clinical entity that can mimic psoriasis. Better recognition of this form of porokeratosis of Mibelli may result in earlier diagnosis and initiation of appropriate therapy.

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Porokeratosis is a heterogeneous group of disorders inherited in an autosomal-dominant fashion and characterized histologically by the presence of a cornoid lamella. The plaque type, termed *porokeratosis of Mibelli*, has a predilection for the acral areas and perigenital region. Porokeratosis of Mibelli is characterized clinically by one or more plaques, with a prominent raised border cleaved by a central furrow. The center of the plaque is often atrophic, hairless, and either hyperpigmented or hypopigmented. However, lesions can be psoriasiform or verrucous, with varying degrees of hyperkeratosis. The lesions begin as small keratotic papules and enlarge centrifugally to form annular or serpiginous lesions, ranging from a few millimeters to several centimeters in diameter. Lesions are usually asymptomatic but can be pruritic. Although onset can occur at any age, porokeratosis usually appears during childhood. Sporadic cases, however, generally occur later in life. Men are affected more often than women.

Lucker et al[1] and Stone et al[2] both described individual cases of a verrucous type of porokeratosis of Mibelli that was localized to the natal cleft region. We report a similar case of verrucous porokeratosis of Mibelli limited to the natal cleft that mimicked psoriasis.

Case Report

A 44-year-old Asian man presented with a 28-year history of pruritic plaques on the buttocks. These plaques had been gradually enlarging. The patient had seen several dermatologists and undergone various treatments for psoriasis, including topical steroids, coal tar, calcipotriene, tazarotene, psoralen-UVA, and topical tacrolimus, all of which were unsuccessful. He had a history of essential hypertension and glaucoma but was otherwise healthy. His medications included losartan, hydrochlorothiazide, triamterene, and brimonidine eyedrops. Family history of skin disease included only atopic dermatitis. Results from a review of systems were within reference range.

On physical examination, several well-demarcated, scaly, red-brown, verrucous plaques with hyperpigmented raised borders distributed bilaterally on the buttocks (Figure 1) were noted. No other cutaneous lesions were evident, and there was no nail pitting.

Two biopsies were obtained. The first, a 4-mm punch biopsy of the center of a plaque, demonstrated psoriasiform hyperplasia of the epidermis with focal parakeratosis suggestive of a cornoid lamella. The second, a 6-mm punch biopsy of the border of a plaque, revealed a thin column of parakeratotic cells with underlying absence of the granular layer and dyskeratotic and vacuolated

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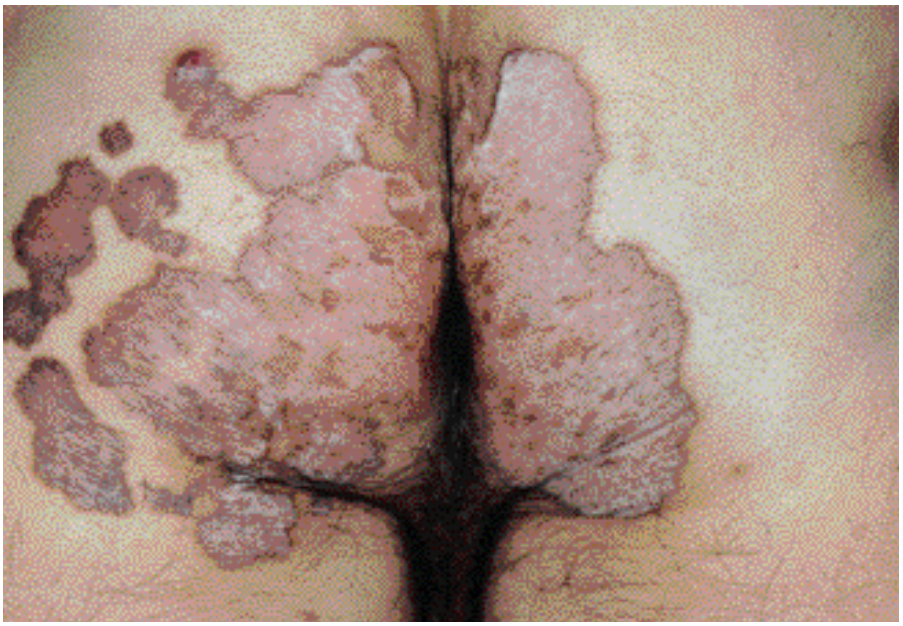

Figure 1. Scaly verrucous plaques with hyperpigmented raised borders on the buttocks.

cells in the spinous layer, which is characteristic of a cornoid lamella (Figure 2). Although there was some psoriasiform hyperplasia, the rete ridges were not club shaped, and no thinning of the suprapapillary plate or platelike parakeratosis and no collections of neutrophils in the epidermis or stratum corneum were noted, as seen in psoriasis. This was consistent with porokeratosis, making the diagnosis of psoriasis unlikely.

The patient underwent a trial of imiquimod cream applied once a day to one side of the buttocks and 5-fluorouracil cream applied twice a day to the other side. The imiquimod cream caused severe irritation without much improvement and was discontinued. Subsequently, the 5-fluorouracil cream was applied to the entire lesion for 2 months, with resolution of the scaling and pruritus but with no decrease in the verrucous component or size of the plaques. After this therapy, treatment consisted of a trial of carbon dioxide (CO_2) laser resurfacing on one side and dermabrasion on the other side. The porokeratosis recurred on the side treated with CO_2 laser resurfacing. However, dermabrasion successfully removed the porokeratosis, and, at the 6-month follow-up, no clinical evidence of porokeratosis in the dermabrasion-treated areas was seen.

Comment

Porokeratosis of Mibelli localized to the genital region alone is rare, with only 7 reported cases.[1-7] Two of these cases are clinically and histologically identical to ours. In 1995, Lucker et al[1] described a 34-year-old man with a 9-year history of pruritic dermatosis confined to the natal cleft. They determined this to be a novel form of porokeratosis, which they termed *porokeratosis ptychotropica* from the Greek words *ptyché* (fold) and *tropé* (a turning). In 1999, Stone et al[2] described a 32-year-old man with a 13-year history of pruritic dermatosis confined to the natal cleft, which they termed *verrucous porokeratosis*. As with our case, this second case also had been mistaken for and treated as psoriasis. All 3 cases presented in young men and were intensely pruritic, progressive, and confined to the natal cleft. As suggested in the previous case reports, this may represent a distinct clinical variant of porokeratosis.

Porokeratosis is known to undergo malignant transformation to Bowen disease, squamous cell carcinoma, and basal cell carcinoma; the latest review revealed at least 56 published cases.[8] This review found that approximately 11% of patients with porokeratosis have lesions that undergo malignant transformation. Malignant transformation in porokeratosis has been associated with an overexpression of p53.[9-11] This potential for malignant transformation makes distinguishing porokeratosis from psoriasis even more important.

Treatment of porokeratosis has been largely anecdotal. There are isolated case reports of successful treatment using topical 5-fluorouracil (with and without occlusion), isotretinoin and acitretin, CO_2 laser ablation, 585-nm pulsed dye laser radiation, grenz ray radiation, frequency-doubled Q-switched Nd:YAG laser radiation, cryotherapy, dermabrasion, surgical excision, and

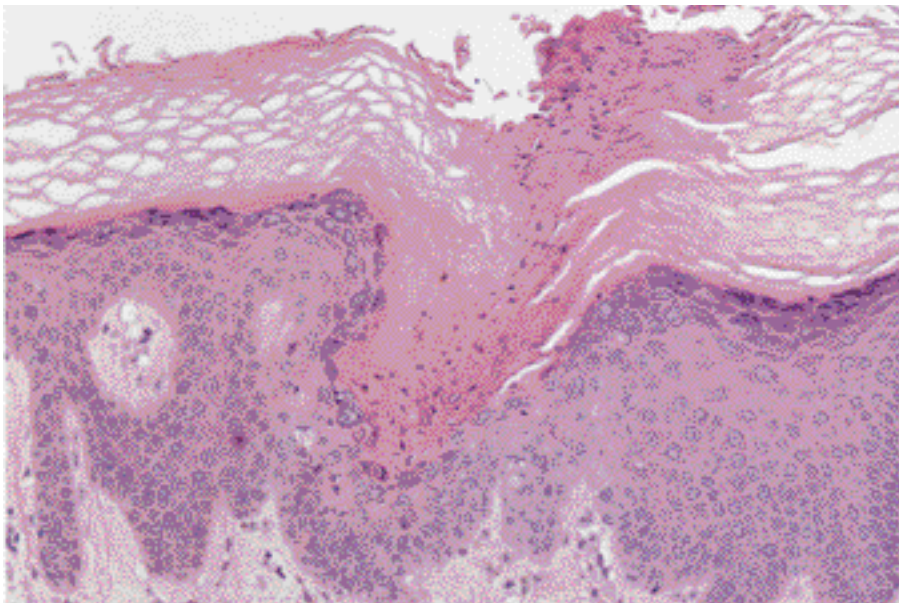

Figure 2. Cornoid lamella formation with the characteristic slanted column of coarse parakeratosis and underlying loss of the granular layer (H&E, original magnification ×60).

electrodesiccation. In this case, topical 5-fluorouracil, imiquimod cream, and CO_2 laser resurfacing were all ineffective in clearing the porokeratosis. However, we found dermabrasion to be a potentially long-term and effective treatment for porokeratosis of Mibelli.

In summary, verrucous porokeratosis limited to the natal cleft appears to be a distinct clinical entity that may mimic localized psoriasis. Some of the treatment modalities used for psoriasis are inappropriate and ineffective for porokeratosis (eg, UV light). However, unlike psoriasis, verrucous lesions of porokeratosis should be monitored for malignant change because correct diagnosis is important for optimal patient care. This stresses the importance of considering a biopsy in cases where the skin disease is not responsive to appropriate therapy.

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