Circumscribed Juvenile Pityriasis Rubra Pilaris (Type 4) Koebnerising after a Hot Water Burn: Mild Disease with Maximum Koebner Response

Dear Editor,

Pityriasis rubra pilaris (PRP) is a chronic, inflammatory, papulosquamous skin disorder that is characterized by follicular hyperkeratosis and reddish-orange, scaling dermatitis with islands of normal skin (1,2). PRP is classified into 5 groups based on clinical features. Type 4 PRP is characterized by well-demarcated, hyperkeratotic erythematous plaques localized on the elbows and knees with palmoplantar keratoderma (1,2).

An 8-year-old girl presented to our clinic with erythematous plaques on both elbows, the legs, and the knees. Plantar keratoderma was noticed on clinical examination. The lesions had started on the elbows and knees about a year ago. The lesions on the leg were surrounded by an irregular, hyperpigmented border. On close inspection, the plaques were formed by follicular papules and mild desquamation was noticed. Upon questioning, it was learned that the lesions on the leg with hyperpigmented borders had emerged after a hot water burn three months ago and that they were localized exactly on the burned areas of the skin (Figure 1).

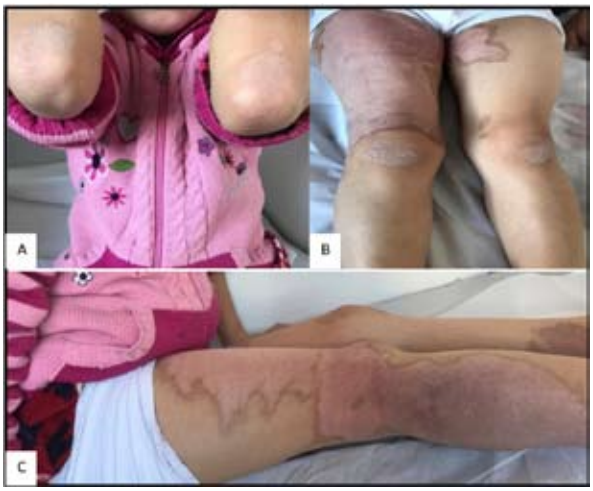

Figure 1. (A) Erythematous plaques formed by follicular papules and mild desquamation on both elbows, the knees, and legs. Koebnerized plaques surrounded by hyperpigmented borders. (B) Image of the lesions on the leg from the lateral aspect. The irregular shape of hyperpigmented borders due to a splash of water.

A biopsy was performed on the new lesions, and histopathological evaluation revealed parakeratosis with alternating orthokeratosis, irregular hyperkeratosis, keratotic plugs, and a mild perivascular lymphocytic infiltration around the blood vessels (Figure 2). A diagnosis of PRP was established.

The Koebner phenomenon (KP) is described as the development of lesions in previously normal skin after exposure to internal or external trauma such as surgical incisions, burns, friction, insect bites, and allergic and irritant reactions (3). The pathogenesis of KP is not fully understood, but epidermal cell injury and dermal inflammation have been proposed as having a role in the pathophysiology (4). Experimental studies on the mechanism of KP have been performed mostly on patients with psoriasis (3). Disease severity, early age of disease onset, and multiple previous therapies have been found to be associated with KP (5,6). KP has previously been reported after injury with the sharp end of a stick in type 3 PRP, a generalized PRP form (7). However, our patient was diagnosed with type 4 PRP, which is a localized form of the disorder. Griffiths reported type 4 PRP does not evolve to generalized forms (8). In this respect, our case was interesting as maximum Koebner response was observed despite the mild PRP. We therefore believe that disease severity is not a determining factor in KP and that the severity of skin damage plays a crucial role. We also think that changes in the cytokine milieu in the burn area may be responsible for KP, as levels of IL-17 and IL-22, which have been shown to be upregulated in burns, also play a role in PRP pathogenesis (9,10). The disease onset at an early age might have also had a contributing role in the Koebner response in this patient. The hyperpigmented borders of the Koebnerized plaques were also notable as they were spared from KP. Some spared areas were also seen within the Koebnerized plaques themselves. A threshold level of trauma is thought to be necessary for inducing KP (3).

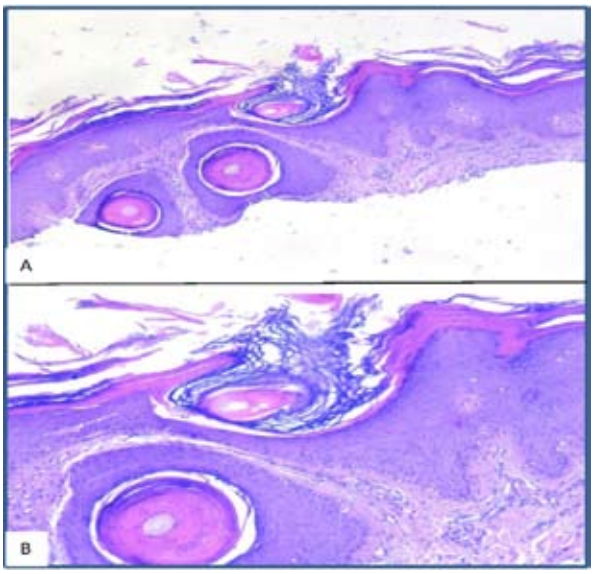

Figure 2. (A) Keratotic plugs, irregular hyperkeratosis, alternating orthokeratosis and parakeratosis (hematoxylin and eosin, ×50); (B) perivascular lymphocytic infiltration around the blood vessels on histopathological examination (hematoxylin and eosin, ×100).

The clinical picture of our patient may indicate that the skin damage was much less severe in some areas of the burn, especially in the periphery, and that KP was therefore not observed in these areas.

Our case clearly demonstrates that the Koebner response is not related to disease severity. We believe that the type of trauma is an important factor in determining the severity of skin damage and the changes in the cytokine milieu in the involved skin. Early disease onset also seems to contribute to the development of KP. Further studies investigating the mechanism of KP in various skin disorders are necessary. As far as we are aware, this is the first case reporting Koebnerization in the circumscribed juvenile form of PRP.

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Successful Treatment with Fusidic Acid in a Patient with Folliculitis Decalvans

Dear Editor,

Folliculitis decalvans (FD) is a rare form of primary neutrophilic cicatricial alopecia. It is a highly distressing disease that affects young and middle-aged adults, with a slight male predominance (1).

The most frequent clinical manifestations are follicular pustules and diffuse and perifollicular erythema that heal with centrifugal scarring. Follicular tufting, erosions, and hemorrhagic crusts can also be present, and this alopecia is most often located at the vertex and occipital area. Patients frequently complain about pain, itching, or burning sensations, and the involvement of other body areas is rare (2).

The pathogenesis of this disease remains unclear. *Staphylococcus aureus* and other hair follicle bacteria can often be isolated from the pustules, suggesting the role of a bacterial infection in its etiology. A defect in the host's immune response can also be postulated by reports of familial cases and the appearance of FD in patients with immunity dysfunctions. Other mechanical factors have been suggested, such as structural abnormalities of the follicle or local inflammation (2).

Management of this alopecia is difficult and its course is typically chronic and relapsing. The treatment aim is to stop inflammation and further irreversible destruction of hair follicles. Antibiotics remain the first-line therapy, due both to their anti-inflammatory and antimicrobial properties (1). Although topical fusidic acid is widely used as adjuvant treatment, there are few data regarding its oral use.

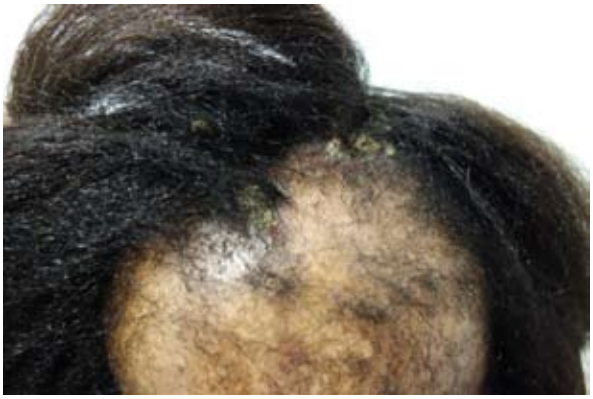

Figure 1. Large atrophic alopecic patch at the occipital region with areas of follicular tufting and peripheral crusts.

We report a case of folliculitis decalvans successfully treated with oral fusidic acid. Our patient was a 41-year old Cape Verdean woman with a two month history of alopecia with painful, purulent discharge at the vertex of the scalp. The patient was diagnosed with human immunodeficiency virus type 1 (HIV-1) infection 5 years prior and was stable on her regimen of efavirenz, tenofovir, and emtricitabine, with undetectable viral load. She denied application of topical or capillary products. Dermatological examination revealed a patch of cicatricial alopecia with crusts and follicular pustules (Figure 1). Direct microscopic examination and mycological culture showed no fungal element. A diagnosis of folliculitis decalvans was established and the patient was started on oral fusidic acid at a dose of 500 mg three times a day. Betamethasone dipropionate 0.05% and salicylic acid 3% lotion as well as azelaic acid 5% lotion were also applied to the affected area once daily. After two months of treatment, the patient showed clinical improvement, with less erythema and suppuration of the affected scalp. A partial hair regrowth was noted, mainly at the periphery. Subsequently the patient maintained only topical therapy, and no recurrences were observed after 6-months of follow-up.

Fusidic acid is useful in the treatment of skin and soft tissue infections, particularly those due to *S. aureus*, as shown by randomized controlled studies (3). The clinical efficacy of fusidic acid in the treatment of folliculitis decalvans has been reported previously. Bogg was the first to describe this useful effect (4). Sutter also reported good results with fusidic acid used both topically and orally (500 mg three times a day) (5). However, both failed to report the treatment duration or the outcome on discontinuation. Abeck described three patients that responded to a three