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Classic Juvenile Pityriasis Rubra Pilaris Treated with Oral Alitretinoin

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Dear Editor:

Pityriasis rubra pilaris (PRP) is a papulosquamous disorder of unknown etiology. Although treatment of PRP is challenging, oral retinoids, including acitretin and isotretinoin, are used as first-line treatment. Recently, alitretinoin, a novel panagonist retinoid, has been reported to be effective for PRP. Here, we described a case of refractory PRP that showed remarkable response to alitretinoin.

A 37-year-old man, presenting with relapsing skin lesions over the whole body since 1 year of age, had been diagnosed with and treated for PRP since he was 10 years old. He had a familial PRP history on his mother's side. Physical examination at the first visit revealed erythematous pruritic scaly papuloplaques on the trunk and lower extremities, which was not distinctive due to previous treatments in other hospitals (Fig. 1A, B). Palmoplantar keratoderma was mildly observed. Neither follicular hyperkeratosis nor generalized erythroderma with island of normal skin was seen. The skin biopsy from the left thigh revealed regular acanthosis with broad rete ridge, parakeratosis, and perivascular lymphocytic infiltration with occasional eosinophils (Fig. 1C). There was no confluent hypergranulosis or alternating orthokeratosis and parakeratosis. Classic juvenile PRP was diagnosed. Several treatment options were administered for 1 year in our hospital, which included neotigasone 20～30 mg for 13 months, narrow-band ultraviolet-B phototherapy for 6 months, methotrexate 12.5 mg per week for 1 month, prednisolone 0.5～5 mg for 12 months, and superpotent topical steroid for 6 months. However, none of these treatments led to stable remission. Therefore, oral alitretinoin (30 mg daily) was initiated and continued for 5 months, combined with superpotent topical steroid. The skin lesions and pruritus significantly disappeared in 4 weeks and were not aggravated for 5 months (Fig. 2). Also, there were no side effects except mild dryness. Laboratory test did not show any abnormalities including serum cholesterol. The patient stated that alitretinoin was most effective and tolerable compared with other therapies. Therefore, we decided to continue the same dosage of alitretinoin as far as it was well-tolerated.

PRP can be clinically divided into 6 subtypes[1]. It comprises an adult-onset (classic or atypical adult type), juvenile-onset (classic, circumscribed or atypical juvenile type) and human immunodeficiency virus-associated form. Classic juvenile PRP begins in the first 2 years after birth, showing similar features of classic adult type.

Systemic retinoids such as acitretin and isotretinoin have antiproliferative, immuno-modulatory, and anti-inflammatory effects that are mediated by intranuclear retinoid acid receptors (RARs). However, alitretinoin (9-cis-retinoic acid) uniquely binds not only to RARs, but also to retinoid X receptors with high affinity. Owing to this dual interaction, alitretinoin may be more effective than conventional retinoids[2]. Moreover, alitretinoin inhibits production of nitric oxide and proinflammatory cytokines such as tumor necrosis factor-α, interleukin-1β and interleukin-12p40[3]. In addition, alitretinoin results in fewer mucocutaneous side effects than other retinoids[2].

To our knowledge, only few cases of adult-onset PRP treated with alitretinoin have been reported[2-5]. This is the first case of refractory classical juvenile PRP that re-

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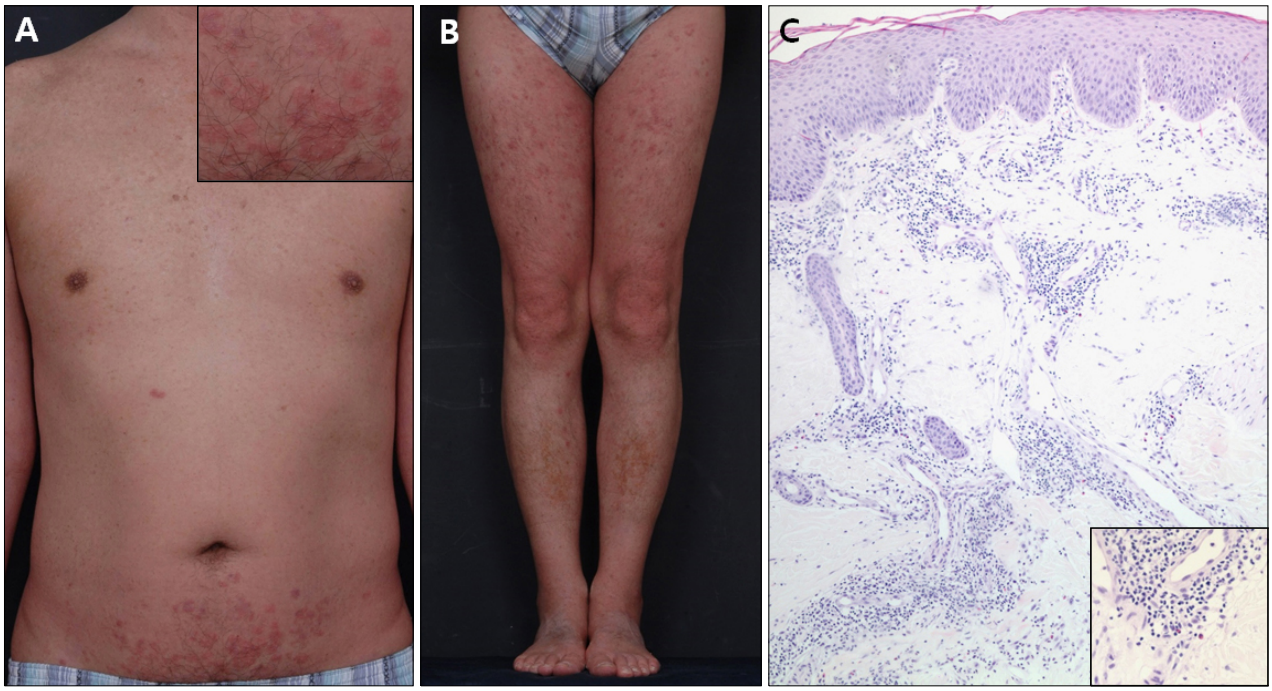

Fig. 1. Clinical findings of pityriasis rubra pilaris presented at the first visit to our hospital. Multiple erythematous scaly papuloplaques on (A) the trunk and (B) the lower extremities. (C) Histopathologic findings revealed regular acanthosis with broad rete ridge, parakeratosis, and perivascular lymphocytic infiltration with occasional eosinophils (H&E; C: ×100, inset: ×200).

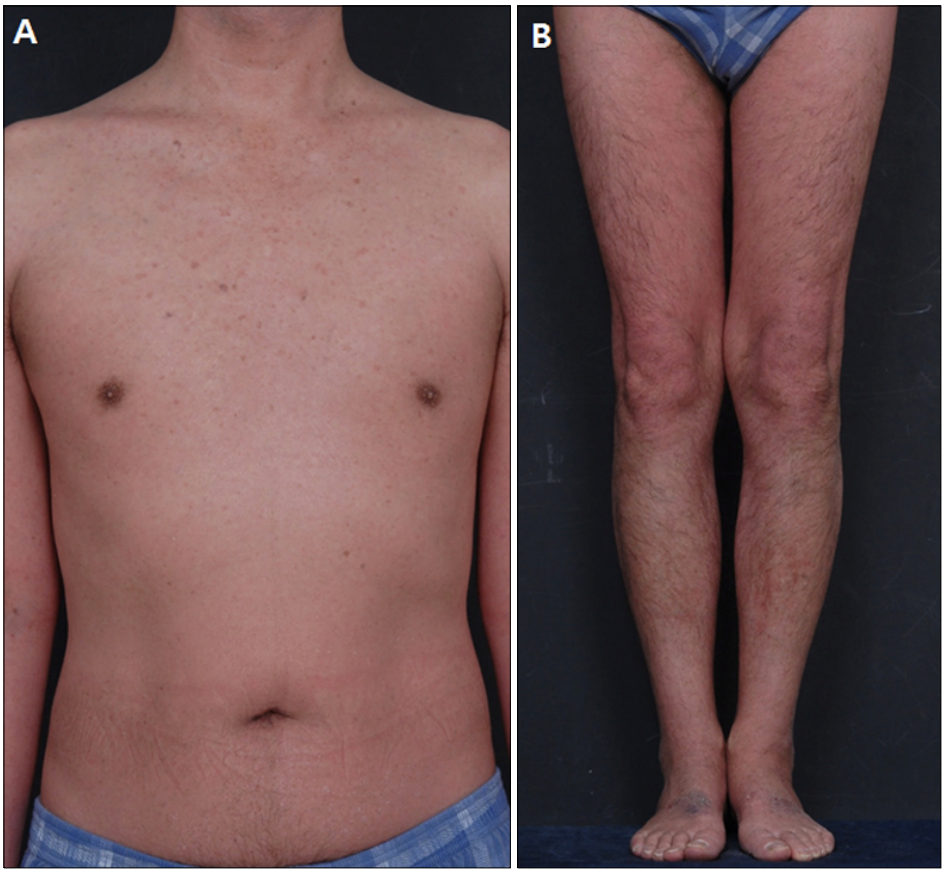

Fig. 2. No skin lesion was seen on the (A) trunk and (B) lower extremities after the 5 months course of oral alitretinoin.

sponded well to alitretinoin. In conclusion, alitretinoin could be a promising alternative for treating refractory classic juvenile PRP.

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