Papular Acantholytic Dyskeratosis of the Inguinal Area in a 49-Year-Old Man

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Dear Editor:

A 49-year-old man presented with recalcitrant pruritic papular eruption on both inguinal areas that had persisted for 6 months. The lesion failed to show improvement, although he had applied a topical steroid on the lesions for 6 months. He had neither personal history nor family history of any skin disorder. Multiple whitish pruritic papules with underlying erythematous patches were noted on both inguinal areas upon physical examination (Fig. 1). No lesions were found on other locations, and his nails were intact. The KOH smear showed negative results for fungus. Skin biopsy showed suprabasal cleft with villi and numerous typical acantholytic dyskeratotic cells in the stratum corneum and spinosum. Hyperkeratosis and irregular acanthosis were also present in the epidermis, with a lymphocytic infiltration in the underlying dermis (Fig. 2).

Direct immunofluorescence or genetic study was not performed. The patient was diagnosed as papular acantholytic dyskeratosis (PAD) of the inguinal area. He was treated with 2.5~15-mg oral prednisolone, 10-mg acitretin, and 0.1% topical mometasone furoate for a year, with unsatisfactory improvement.

PAD is a very rare disease, characterized by multiple small pruritic papules. Lesions have been reported on the genitalia, perineum, inguinal area, and upper thigh¹. PAD shows very similar histopathologic findings with that of both Hailey-Hailey and Darier diseases (HHD and DD, respectively), although HHD tends to show more diffuse and DD shows more prominent dyskeratosis than PAD². HHD and DD are autosomal dominant diseases caused by loss of function mutation of the *ATP2C1* and *ATP2A2* calcium pump genes, respectively. Therefore, familial histor-

Fig. 1. Multiple whitish pruritic papules with underlying erythematous patches were noted on the right (A) and left (B) inguinal areas. Steroid-induced erythematous atrophic patches were also found.

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Fig. 2. Skin biopsy showed suprabasal cleft with villi, and numerous typical acantholytic dyskeratotic cells in the stratum corneum and spinosum. Hyperkeratosis and irregular acanthosis were also present in the epidermis, with a lymphocytic infiltration in the underlying dermis (H&E; A: $\times 40$, B: $\times 200$).

ies are often present, unlike our case. Moreover, our case is distinguished from HHD and DD by clinical manifestation. HHD usually occurs in young adults and shows painful oozing erosions, rather than papules, in the flexural area, including the axilla and submammary fold. DD usually develops in adolescence and is characterized by greasy keratotic papuloplaques in seborrheic regions with hand and/or nail involvement.

Recently, heterozygous mutations in the *ATP2C1* and *ATP2A2* genes were found in three cases and one case of PAD, respectively^{1,2}. It is speculated that segmental mosaic mutations in *ATP2C1* or *ATP2A2* gene, whether somatic or germinal mutation, can result in the distinct clinical phenotype referred to as PAD¹. Since cases of localized HHD or DD to genitocrural area have been rarely reported, genetic study might be helpful to rule-out HHD or DD².

An effective therapy has not been defined yet. Cryotherapy, topical steroids, topical tretinoin, topical tacrolimus, oral isotretinoin, and laser ablation have been reported with variable results².

To date, thirty-nine cases of PAD have been documented, with eleven male cases, in English literature³. Among them, two cases were Korean patients³. Additional two cases were reported in Korean literature^{4,5}. This suggests that PAD may be more common in men than has been previously been described³. It is difficult to differentiate it with condyloma, eczema, and tinea cruris clinically. Dermatologists should consider PAD as a differential diagnosis of inguinal papular eruption and perform a skin biopsy to avoid delaying the diagnosis.

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CONFLICTS OF INTEREST

The authors have nothing to disclose.

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