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Papular Acantholytic Dyskeratosis: Report of a Rare Case

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Abstract

Papular acantholytic dyskeratosis (PAD) is a rare dermatosis, characterized by a variety of skin features including solitary discrete lesions, multiple grouped or confluent papules, vesicles, and bullae, and white macerated patches and plaques. The lesions are mostly asymptomatic, and involve the vulva, penis, scrotum, perianal area, and inguinal folds. Herein, we describe a young case with PAD whose lesions were completely cured by cryotherapy.

Keywords: ***Papular acantholytic dyskeratosis; Treatment; Cryotherapy***

1. Introduction

Papular acantholytic dyskeratosis (PAD) is a rare dermatosis with unknown etiology which is poorly defined in the literature [1]. It is a clinical variant of focal acantholytic dyskeratosis, initially described by Ackerman in 1972 [2].

Clinically, PAD manifests with a variety of skin features including solitary discrete lesions, multiple grouped or confluent papules, vesicles, and bullae, and white macerated patches and plaques [3]. Mostly, the lesions are asymptomatic, and involve the vulva, penis, scrotum, perianal area, and inguinal folds [4].

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Herein, we describe a young case with PAD whose lesions were completely eradicated by cryotherapy.

2. Case Report

A 14-year-old boy with cerebral palsy presented to our department of dermatology with multiple pruritic skin-colored papules on his abdominal wall, groin, and the inner side of the thighs. The lesions had been present for 1 year and were gradually increasing in number and size. The patient was the only child of a consanguineous marriage. The parents denied the presence of similar lesions in themselves.

Upon physical examination, multiple, discrete skin-colored 2-3 mm papules that involved the abdominal wall, groin, and the inner side of both thighs were observed. The external genital and anal regions were spared. The lesions had a smooth surface and were firm on palpation (FIG. 1). The examination of the other parts of the body was unremarkable.

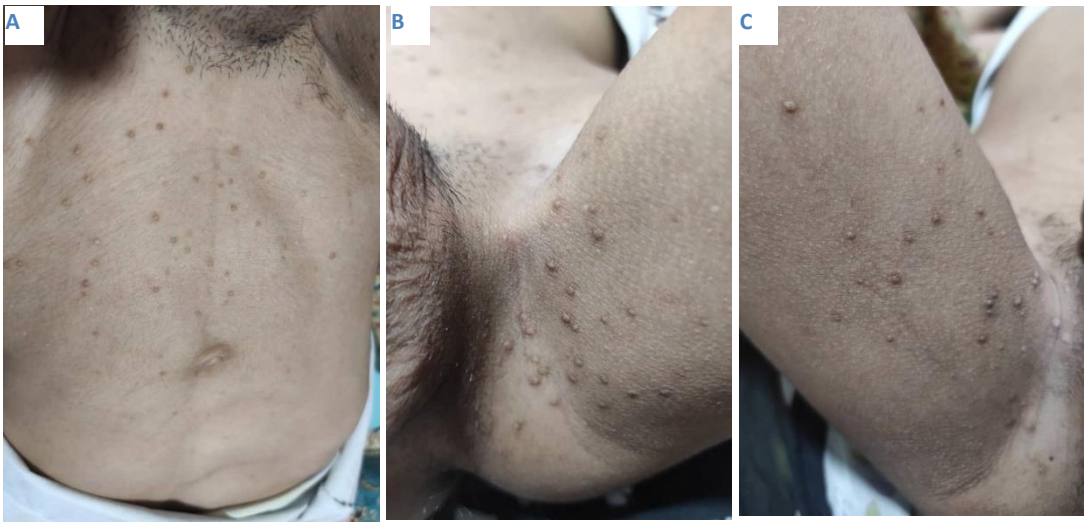

FIG. 1: **A to C; Multiple, discrete skin-colored papules ranging from 2-3 mm in size involving the abdominal wall, groin, and the inner side of both thighs.**

A skin biopsy was taken from one skin papule with the clinical differential diagnoses of molluscum contagiosum, planar wart, lichen planus, and eruptive vellus hair cysts. The histopathological examination showed hyperkeratosis without parakeratosis, focal suprabasal acantholysis, acantholytic cells, classic corps ronds and the presence of a few dyskeratotic keratinocytes. The examination of the dermis was unremarkable.

The histopathologic diagnosis was consistent with focal acantholytic dyskeratosis (FIG. 2). The histologic differential diagnosis also included Darier's disease, warty dyskeratoma and Grover's disease (transient acantholytic dermatosis), none of which fit the clinical presentation. Weekly cryotherapy was performed, which resulted in complete eradication of the lesions after 5 sessions of treatment. The 3 month-follow-up showed no recurrence.

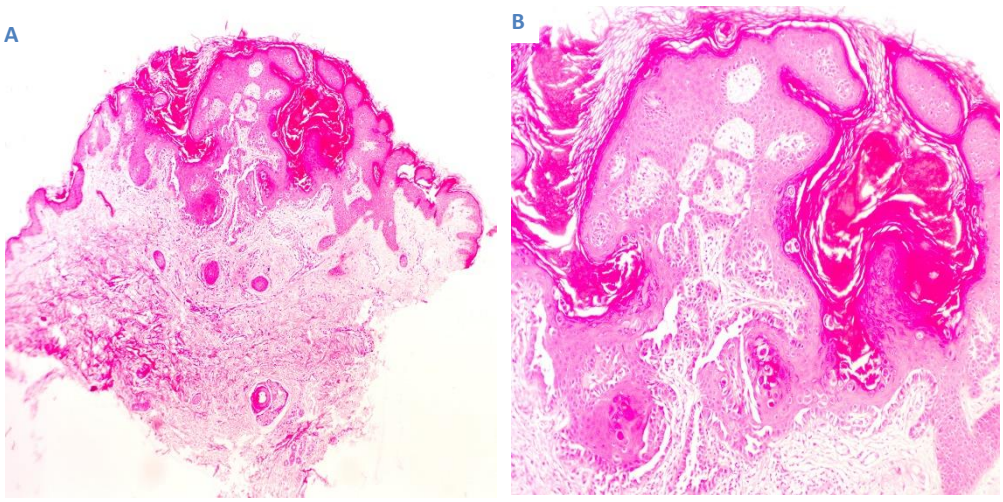

FIG. 2. **A and B; Hyperkeratosis, focal suprabasal acantholysis, acantholytic cells, and a few dyskeratotic cells in the epidermis (H&E staining).**

3. Discussion

Acantholytic dyskeratosis is a histological reaction characterized by the presence of suprabasal clefting, acantholysis, corps ronds, grains, and dyskeratotic cells in the epidermis. In a variety of clinical entities, acantholytic dyskeratosis is a as histologic feature among which Darier's disease, warty dyskeratoma, Haily-Haily disease, Grover's disease, focal acantholytic dyskeratosis, acantholytic acanthoma, and persistent acantholytic dermatosis are the most important [2].

Clinically, PAD is characterized by a range of skin manifestations from solitary discrete lesions to multiple grouped or confluent papules, vesicles, and bullae as well as white macerated patches and plaques [3]. The skin lesions are mostly asymptomatic, but erythema, itching, and burning sensation have been reported in some cases. The usual sites of involvement are the vulva, penis, scrotum, perianal area, and inguinal folds [4]. Conditions like wetness, occlusion and sunlight have been described as triggering factors for appearance of the lesions [1]. In our case, the skin lesions were limited to the abdominal wall, groin, and inner side of upper thighs, and the genital and anal regions were spared.

The prevalence ratio of male-to-female involvement is 0.8:1, and the mean age of patients at the time of presentation is 38.8 years [5]. Mutations in ATP2A2, particularly ATP2C1, which are seen in Darier and Hailey-Hailey diseases, respectively, have been reported in PAD. This observation supports that PAD may be a mild or mosaic variant of these diseases [1].

No effective therapeutic approach has yet been suggested for PAD. Topical agents such as corticosteroids, calcineurin inhibitors, retinoids, and oral isotretinoin have been reported with some success. Simple ablation of skin lesions by procedures including cryotherapy, electro surgery, and surgical excision can be considered as relatively more effective [6]. In our case, a complete cure was seen with cryotherapy.

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