Papular acantholytic dyskeratosis (PAD) is a subtype of focal acantholytic dyskeratosis, typically presenting in the second to fifth decade of life. Patients often present with a chronic, asymptomatic skin-colored to whitish papular eruption, confined to the perineal region and genitalia occasionally extending to the groin or the upper thighs. This disease carries a great diagnostic challenge and lacks a standard algorithm for therapy due to its rare incidence as well as its complex clinical and pathological presentation. Herein, we present the case of a 36-year-old male suffering from PAD who well responded to oral isotretinoin.

CASE REPORT
A 36-year-old man with no premorbid illness presented to the outpatient dermatology clinic with recurrent pruritic and painful eruption on his groin and perineal area for the past 7 years. He did not report any contact allergies or risky sexual behaviors. The lesions were persistent in nature but worsened during summer. He previously received oral fluconazole, itraconazole, prednisone, antihistamines and antibiotics, topical antifungals, potent corticosteroids, and pimecrolimus without significant improvement. Physical examination revealed hyperkeratotic, white smooth cobblestoned papules with an underlying erythematous base in the scrotum, inguinal and intergluteal folds [Figures 1 and 2]. Lesions were macerated and excoriated. No vesicles/bullae formation was noted. Nail, scalp, and mucosal physical examination was insignificant.

A 4-mm punch biopsy was obtained from the left inguinal area [Figures 3 and 4]; it showed mild epidermal hyperplasia with hyperkeratosis, foci of acantholytic dyskeratosis, and suprabasal cleft formation. The epidermis showed scattered acantholytic and dyskeratotic cells (scarce corps ronds), without spongiotic vesicles. Underlying dermis showed vascular ectasia with sparse superficial perivascular mononuclear inflammatory infiltrate. Gomori methenamine silver (GMS), periodic acid-Schiff (PAS), and acid-fast bacillus (AFB) stains were negative for mycobacteria and fungi. A diagnosis of papular acantholytic dyskeratosis (PAD) was made.

The patient refused consent for genetic testing. He was started on 40 mg/day of oral isotretinoin for 8 weeks. Eight weeks later, the patient’s condition improved significantly.

DISCUSSION
PAD is a focal cutaneous disease sometimes called “acantholytic dermatosis of the genitocrural area.” The eruption can sometimes be accompanied by an intense pruritus. It mainly

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affects women; few cases have been reported in men. Histologically, PAD shares signs of both Hailey–Hailey disease (HHD) and Darier’s disease (DD), with slight differences including acantholytic and dyskeratotic cells with hyperkeratosis and focal parakeratosis, negative direct and indirect immunofluorescence.\(^\text{[1-3]}\)

Given the chronic course of the disease in our patient and the long duration of treatment with different antimicrobials and corticosteroids, GMS, PAS, and AFB stains were performed to exclude grafted mycobacterial and fungal infections.

Histopathological similarities with transient acantholytic dermatosis (Grover’s disease) can mislead the diagnosis. Nevertheless, lesion type and location are different in Grover’s disease where transient, confluent macules and papules appear on the trunk, neck, and proximal limbs.\(^\text{[4]}\)

Clinically, the absence of truncal lesions and distribution in seborrheic areas, lack of palmoplantar keratoderma, verrucous papules on extremities, nail changes as well as onset in adult age rule out DD. Moreover, upon histological examination in DD, dyskeratotic cells are found throughout the epidermis with more prominent hyperkeratosis.\(^\text{[2-5]}\)

Although the patient presented erosions in the intertriginous area, HHD could be ruled-out because of the absence of blistering, sparing of axillary and neck fold, absence of diffuse acantholysis with dilapidated brick wall appearance on histology and lack of family history.\(^\text{[6]}\)

However, recent evidence indicates that PAD could be caused by genetic mutations that are closely related to HHD contrary to what was previously thought. The disease is sporadic in nature and the etiology is still unclear.\(^\text{[7]}\)

Although HHD is known to be inherited in an autosomal dominant way, nearly one-third of HHD cases occur randomly from \textit{de novo} mutations.\(^\text{[8]}\) Heterozygous germline mutations of the ATP2C1 gene were recently identified in two patients from the same family, one having PAD and the other one HHD. However, mutations in PAD are distinct and not specific which can highlight the allelic nature of both diseases (different mutations of the same gene) and the existence of other factors (genetic or environmental) influencing their clinical
expression. Lipoff et al. considered PAD a variant of the same disease due to a spontaneous mutation. As the patient declined consent for genetic testing, a common genetic etiology could not be confirmed.

The treatment of PAD is not well defined and it yields various results. Oral and topical steroids or retinoids may be helpful in controlling the disease. Ablative treatments such as surgical excision, cryotherapy, or electrocautery are also interesting options to consider.

PAD is a chronic acantholytic dermatosis. Clinical and histological characteristics can overlap with those of DD and HHD. Whether it is a benign subtype of HHD or a distinct clinical entity remains to be confirmed. Larger case series are required to establish a clear management plan.

**Declaration of patient consent**
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**
There are no conflicts of interest.

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