Correspondences

Spontaneous Regression of Papular Acantholytic Dyskeratosis of Genitocrural Area: A Case Report with Positive DIF Findings

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Sir,

A 22-year-old Chinese woman who was 2-month postpartum presented to our clinic for slightly pruritic eruptions on her perianal area for 5 months. She denied any family history of similar eruptions and history of inflammatory bowel disease, such as ulcerative colitis or Crohn’s disease. She also denied any high-risk sexual behavior and medical history during pregnancy. Acetic acid test, treponema pallidum particle agglutination assay, toluidine red unheated serum test, and HIV testing results were negative. Cutaneous examination showed multiple smooth, grey-whitish, firm, dome-shaped papules of 2–5 mm in diameter on perianal area, and part of them coalesced into verrucous plaques. There was no similar lesion over other areas. A skin biopsy revealed acantholytic dyskeratosis with suprabasal cleft formation and villi, corps ronds and grains were detected in epidermis, and a superficial perivascular lymphocytic infiltration was displayed in the underlying dermis. DIF examination showed linear deposits of immunoglobulin (Ig) G and IgA in the intercellular space of acantholytic cells and granular deposits of IgG, IgA, IgM, and complement C3 in dermal blood vessel walls. A diagnosis of papular acantholytic dermatosis (PAD) of genitocrural area was made. Considering the patient still needed to breastfeed her baby, she chose to receive treatment after weaning. Two weeks later, most of the skin lesions regressed, and 2 months later, the patient just presented scattered papules over perianal area. A long-term follow-up without any treatment was planned.

PAD of genitocrural area is a rare clinical form of focal acantholytic dyskeratosis, which was first described by Ackerman in 1972. The eruptions are always limited to the anogenital area, while involvement of upper thighs, chest, lips, and palms and soles has also been reported. Skin lesions could be pruritic, painful, or burning, but most of them are asymptomatic.

A warm, moist, high friction environment is recognized as a predisposing factor of PAD of genitocrural area, while the pathogenesis is still uncertain. Somatic ATP2C1 (ATPase gene associated with Hailey-Hailey disease) and ATP2A2 (ATPase gene associated with Darier’s disease) mutations in skin lesions of PAD of genitocrural area were identified. Some reports of positive DIF of intercellular IgG and C3 within the epidermis and premenstrual itching and burning of a female patient suggested that immune dysfunction and ovarian hormones might participate in the pathogenesis of this condition. The treatment of PAD of genitocrural area is challenging. Topical potent steroid, pimecrolimus 1%, tacrolimus 0.1%, maxacalcitol and oral viaminate, acitretin had been used to treat this disease, and results were variable. For those patients with limited skin lesions, surgery might be a good choice.

To the best of our knowledge, this is the first time to report a woman suffering from PAD of genitocrural area with positive DIF findings in both epidermis and dermal blood vessel walls during the late pregnancy which regressed spontaneously after delivery; it reminded us a DIF examination could be used as a routine test and a further examination of anti-desmoglein (Dsg) 1 or anti-Dsg3 antibody should be performed. Although the relationship between PAD of genitocrural area and ovarian hormone needs to be confirmed by more studies, it still provides another clue to the pathogenesis of PAD.
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.

References
1. Ackerman AB. Focal acantholytic dyskeratosis. Arch Dermatol 1972;106:702-6.
2. Lee JH, Kim YC, Lew W. A case of focal acantholytic dyskeratosis occurring on both the lip and the anal canal. Yonsei Med J 2003;44:166-8.
3. Knopp EA, Saraceni C, Moss J, McNiff JM, Choate KA. Somatic ATP2A2 mutation in a case of papular acantholytic dyskeratosis: Mosaic Darier disease. J Cutan Pathol 2015;42:853-7.
4. Wan C, Zheng Y, Su X. Papular acantholytic dyskeratosis occurring on both the vulvar and palmoplantar areas. J Dermatol 2015;42:533-4.
5. Verma SB. Papular acantholytic dyskeratosis localized to the perineal and perianal area in a young male. Indian J Dermatol 2013;58:393-5.

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