

A Case of Familial Benign Chronic Pemphigus Misdiagnosed as Eczema and Tinea Cruris

Sir,

A 42-year male patient presented with erythema and small blisters over the skin folds of the armpit and bilateral groins for more than 20 years. He attended many hospitals and was diagnosed with "eczema" and "tinea cruris." Skin lesions were healed gradually after topical hormones and antifungals. However, skin lesions recurred repeatedly.

Upon examination, he had erythema and erosions with yellow crusting on the bilateral axillae, popliteal fossa, and groin (Figure 1). Biopsy and histopathological examination revealed intraepidermal acantholysis with numerous acantholytic cells at places with a tombstone pattern, and a superficial perivascular lymphocytic infiltrate (Figure 2). He was eventually diagnosed with familial benign chronic pemphigus. After 10 days of treatment with 40 mg methylprednisolone tablets and mupirocin ointment, his lesions gradually subsided.

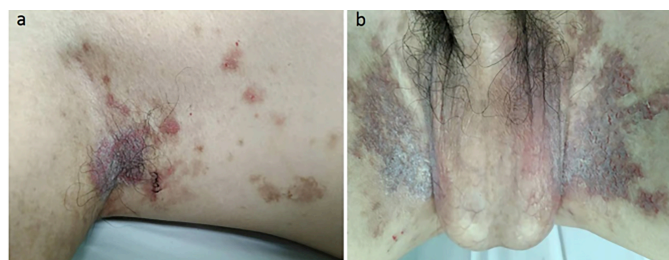


Figure 1: Appearance of the skin lesions of the patient.

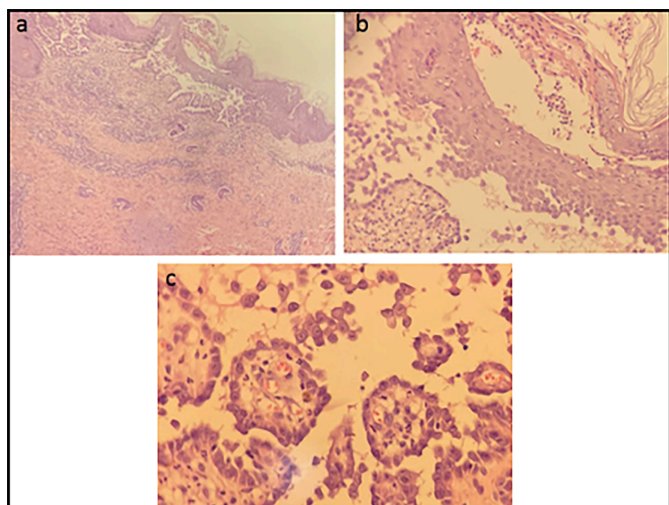


Figure 2: Histopathology of the skin lesion (a x 40, b x 100, c x 400, HE stain).

Familial benign chronic pemphigus is clinically characterised by erythema, blisters, and macules and erosions on the neck, axillae and groin.^{1,2} As the disease is rare, it is easy to be misdiagnosed as eczema, tinea cruris, pemphigus, and keratosis pilaris.³

Familial benign chronic pemphigus is a dominantly inherited condition and previous reports have shown that the incidence is about the same in males and females.^{4,5} Interestingly, the patient indicated at the first visit that all the affected individuals in his family were males, and subsequent detailed investigations revealed that some of the females in this patient's family also had the disease. We speculate that it may be that women tend to conceal diseases that occur in the external genitalia more than men. In addition, men have more severe lesions in the groin and armpit areas due to labour and sports etc. So, they are more inclined to seek medical attention. In addition, among the family members of this case, the patient's father had the disease in middle age, the patient himself had the disease in young adulthood, and the patient's son had the disease since he was a teenager, which seems to indicate that the onset time has advanced generation by generation, which requires further study.

PATIENTS' CONSENT:

This case was granted waiver of informed consent by the Institutional Review Board.

COMPETING INTEREST:

The authors declared no competing interest.

AUTHORS' CONTRIBUTION:

RC: Wrote the manuscript.

KD: Performed the literature review.

CX: Collected the pictures and gathered important clinical information.

All the authors have approved the final version of the manuscript to be published.

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Renhe Chen, Keyun Ding and Chunli Xie

Department of Dermatology, The Affiliated People's Hospital of Jiangsu University, China

Correspondence to: Dr. Keyun Ding, Department of Dermatology, The Affiliated People's Hospital of Jiangsu University, China

E-mail: dky621020@126.com

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