

Hailey-Hailey Disease with Acrokeratosis Verruciformis Hopf

Sir,

Acrokeratosis verruciformis was described by Gerhard Hopf as a familial dermatosis (1). It is a relatively common symptom in keratosis follicularis (Darier disease) (2) but has never been reported in Hailey-Hailey disease. In our case, acrokeratosis verruciformis Hopf accompanied Hailey-Hailey disease.

CASE REPORT

A 59-year-old male patient was found to have had Hailey-Hailey disease for 30 years. His mother suffered from the same disease, while his father had no skin diseases. When he was admitted to the clinic, the two sides of the neck exhibited symmetrical patches; those on the axillary folds, the anogenitoinguinalis areas and the sacral areas were erythematous and sweaty-smelling, and in some places there were yellow erosions covered by scabs (Fig. 1). On the surface of the feet, many symmetrical, pinhead shaped, white-greyish, hyperkeratotic flat papules were observed. Laboratory examinations led to negative pathological findings. Samples were taken from the papules in the neck area (I) and from the surface of the feet (II).

Histology I: Histological examination revealed typical suprabasal fissures, and the supravascular area was formed by hypergranulosis, hyperkeratosis and parakeratosis. No dyskeratotic cells were observed. In the dermis, there were papillomatosis and a moderate lymphocytic and monocytic infiltrate surrounding the vessels. Direct immunofluorescence studies were performed on lesional skin, but no deposits of immunoglobulin were observed. Diagnosis: Hailey-Hailey disease. *Histology II:* Histological examination showed considerable hyperkeratosis and parakeratosis, the thickness of the epidermal layer rising in the shape of a church tower. Diagnosis: acrokeratosis verruciformis.



Fig. 1. Eczematous-like lesions of Hailey-Hailey disease on the neck.

Local antiseptic, steroid and antibiotic ointment were applied, without significant improvement. Tigason was also given orally (75 mg/day). The patient was readmitted with the same disease, but in a worse condition. Administration was continued with laser therapy. Under local lidocaine anaesthesia, a coherent system 450 carbon dioxide laser was used to vaporize the plaques on the neck and the sacral area. A defocused beam at 10 W on the first, second and third passes vaporized the lesions to a clinically normal plane. The patient had postoperative pain for several weeks, but the treated sites were free and asymptomatic when he was seen after 6 months and after 1 year.

DISCUSSION

Many clinical and pathogenetic similarities suggest a connection between Hailey-Hailey disease and Darier disease (3). Acrokeratosis verruciformis is a common symptom in Darier disease but has not previously been found to be associated with Hailey-Hailey disease, as in our case.

In Hailey-Hailey disease, when the treatment with steroids, antibiotics, Tigason and antiparasitic agents was ineffective, surgical treatment by abrasion and grafting of the skin may be tried. The carbon dioxide laser may be used for precise vaporization of the skin (4). The bloodless field makes clinical evaluation of the disease-free margins easier than with other ablative techniques. Recurrence has not been reported, and in our case, no recurrence of the Hailey-Hailey disease was observed at the 1-year control following carbon dioxide laser vaporization.

The association of acrokeratosis verruciformis with Hailey-Hailey disease is further evidence that there may be a relation between these diseases. Careful examination of patients with Hailey-Hailey disease in order to establish whether they simultaneously have acrokeratosis verruciformis may give more information on the connection between these diseases.

REFERENCES

1. Hopf G. Ueber eine bisher nicht beschriebene disseminierte Keratose (Akrokeratosis verruciformis). *Dermatol Z* 1931; 60: 227-250.
2. Burge SM, Wilkinson JD. Darier-White disease: a review of the clinical features in 163 patients. *J Am Acad Dermatol* 1992; 27: 40-50.
3. Burge SM, Cederholm-Williams SA. Cell adhesion in Hailey-Hailey disease and Darier's disease: immunocytological and explant-tissue-culture studies. *Br J Dermatol* 1991; 125: 426-435.
4. McElroy JA, Mehregan DA, Roenigk RK. Carbon dioxide laser vaporization of recalcitrant symptomatic plaques of Hailey-Hailey disease and Darier's disease. *J Am Acad Dermatol* 1990; 23: 893-897.

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