Familial Benign Pemphigus

Synonyms: familial benign chronic pemphigus, Hailey-Hailey disease

Epidemiology and aetiology

This is a rare inherited autosomal dominant skin disorder, caused by a genetic mutation in the ATP2C1 gene located on chromosome 3. Occasionally sporadic cases do occur. There is a defect in keratinocyte cohesion, first described in 1939 by dermatologist brothers Hailey and Hailey.

Presentation

It may start in teenage years but it most commonly presents in the third and fourth decades. Vesicular or bullous rashes and erosions develop first in skin folds (axilla, groin, genitalia and under the breasts). The lesions heal without scarring.

If the lesions persist they may become thickened, with maceration, itching and painful cracks. This can lead to secondary bacterial or candidal infection and malodour.

The trunk and neck can also be affected, with lesions provoked by friction, sun exposure, heat, and trauma.

Fingernails may show white, longitudinal bands. Pits may occur on the palms.

Differential diagnosis

- Rash may be mistaken for pemphigus vulgaris, impetigo or fungal infection.
- Perineal lesions may mimic genital warts.

Investigations

Skin biopsy may be required, although appearance and family history is usually enough. The histology is characteristic.

Management

Supportive:

- Avoid trigger factors - sunburn, friction and sweating; wear soft and absorbent clothing; avoid obesity.

Topical treatments:

- Topical corticosteroids ± antibacterials/antifungals.
- Benzoyl peroxide cream (as antibacterial).
- Wet compresses to dry up oozing patches (eg aluminium acetate 1:40 dilution).
- Calcipotriol cream.
- Tacrolimus ointment.
- Other topical treatments (from recent case reports) are topical cadexomer iodine powder and 5-fluorouracil.

Systemic treatments:

- Prolonged courses of oral antibiotics (eg tetracycline or erythromycin) may help.
- Immunosuppressants have been used, eg retinoids, ciclosporin, dapsone, methotrexate, systemic steroids and alefacept. However, no clinical trials have been performed.

Other treatments:
- Phototherapy.
- Low-dose botulinum toxin - to reduce sweating.
- Carbon dioxide laser ablation[9] - although problems with hypertrophic scars have been reported.
- Photodynamic therapy ± other treatments - with varying success. [10, 11]
- In severe cases, affected areas can be removed surgically, but skin grafts may be required to close the skin deficit and scarring may be a problem.
- Radiotherapy has been reported as successful in local disease control, although it does not seem to influence the natural course of the disease.

Complications

- Eczema herpeticum (disseminated herpes simplex infection of pre-existing skin disease) is a rare complication, requiring systemic antiviral treatment. [12]
- A single case report describes squamous cell carcinoma developing in a vulval lesion after tacrolimus treatment. [13]

Prognosis[3]

- For most patients the condition is a 'nuisance' rather than a serious problem.
- May have long remissions.
- May improve with age.

Further reading & references

- Benign Familial Chronic Pemphigus, DermIS (Dermatology Information System)
- 1. Benign chronic pemphigus (Hailey-Hailey disease), Online Mendelian Inheritance in Man (OMIM)
- 3. Halley-Hailey Disease, DermIS, DermNet NZ, with images
- 4. Helm TN et al; Familial Benign Pemphigus (Hailey-Hailey Disease), eMedicine, Mar 2010

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