

TREATMENT OF FAMILIAL BENIGN CHRONIC PEMPHIGUS BY SKIN GRAFTING

C. E. Sonck and A. Rintala

From the Dermatological Clinic, University of Turku, and the Finnish Red Cross Hospital for Plastic Surgery, Helsinki, Finland

Abstract. A report on a patient with familial benign chronic pemphigus, resistant to all conservative therapy, treated in several stages by surgical excision of the most affected areas and reconstruction with split skin grafts. The follow-up period of the treated sites varies from two to eight years. All grafted sites have remained totally asymptomatic and there has been no sign of recurrence. Outside the grafted areas the disease is still as active as before.

Key words: Pemphigus, familial benign chronic; Skin grafting; Treatment, surgical

Familial benign chronic pemphigus, originally described by Gougerot & Allée (5) and by Hailey & Hailey (6) has occurred in several families in Finland. A review of 37 cases belonging to 9 Finnish families was presented by Sonck in 1965 (10). One of the patients suffered very severely from lesions in the genitofemoral region and surgical removal of the affected skin area was suggested in April 1953. Because of the long waiting list, 6-8 months, for non-urgent cases, the patient sank into a state of deep depression and committed suicide.

Biopsy scars from cases of Hailey-Hailey's disease mostly remained free from lesions. In the present case (see below) a preliminary transplantation of a single 8 mm punch graft from the anterior surface of the thigh appeared promising, too, and a large grafting procedure was planned. Before this was performed the senior author (C. E. S.) had the opportunity in Budapest in 1965 to examine a Hungarian patient already surgically treated, by Bíró and Máday.

Since the Hungarian authors had published their successful results in 1966 (2, 3), some further reports have appeared in the literature (1, 4, 8, 11, 12).

REPORT OF A CASE

The patient (K. J.), a baker, born 1931, had been severely troubled by skin lesions since the age of 18. His brother,

father, son, aunt, and a paternal cousin had similar cutaneous lesions. Since March 1955 he has been treated by one of us (C. E. S.) and was hospitalized several times at the University Dermatological Clinics in Helsinki and Turku.

The skin condition was worst in both axillae (Figs. 1 and 3), on the side of the neck and in the genitofemoral folds. From time to time, eruptions had also appeared in the cubital folds and in the umbilical and lumbosacral regions. The bullous and oozing areas of the axillae were about 17 × 14 cm in size.

In addition, the patient suffered from hyperhidrosis, blepharitis, and hyperkeratosis of the palms and soles.

Findings from laboratory studies (blood count, ESR etc) were within normal limits. Repeated cultures from the lesions revealed mostly *Staphylococcus aureus* and *Proteus mirabilis*, whereas *Streptococcus β-haemolyticus*, *Pseudomonas aeruginosa*, and *Candida albicans* were found only occasionally. Biopsies showed the typical picture of Hailey and Hailey's disease.

Systemic therapy with corticosteroids was of no value. Much benefit was temporarily afforded, however, by the local use of corticosteroids. Temporary improvement of secondary infections was seen repeatedly after systemic treatment with sulfonamides (Supronal) and antibiotics (Terramycin, Achromycin and Erythromycin), as indicated by the results of the bacterial resistance determinations.

However, as no definite cure could be obtained, despite numerous trials with various systemic and local treatments, it was decided to treat the patient with plastic surgery.

On November 3, 1966, the involved area of the left side of the neck was excised and a split-thickness graft from the anterior surface of the left thigh was applied (Dr Parvinen, at the Department of Surgery, University Central Hospital in Turku). The "take" of the graft was good, and the donor area healed without complication. Histological studies of the excised skin confirmed the diagnosis of familial benign chronic pemphigus. The grafted area of the neck has remained asymptomatic.

In January 1969 the whole thoracic part of the involved skin in the left axilla was excised by one of us (A. R.) and replaced with a thick split-thickness skin graft taken from the anterior surface of the left thigh. A circumscribed lesion in the left cubital fold was treated in a similar way with skin grafting and healed *per primam*. One part of the grafted skin in the axilla, however, became necrotic due to postoperative haemorrhage and a new grafting procedure had to be performed 12 days later. The skin grafting of the left axilla

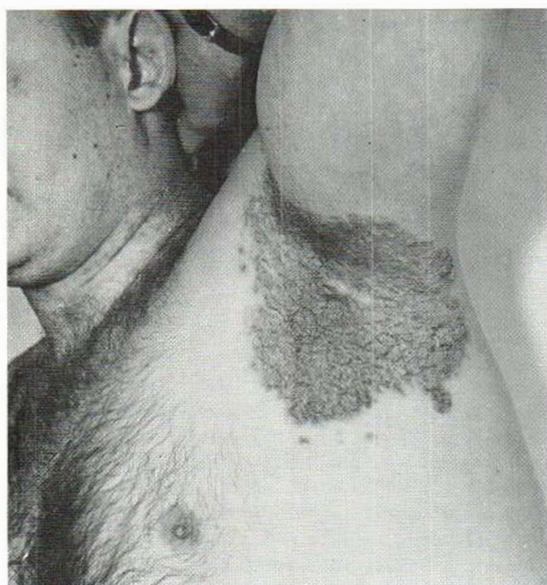


Fig. 1. Hailey-Hailey's disease of nearly 20 years' duration in the left axilla (before surgical intervention).

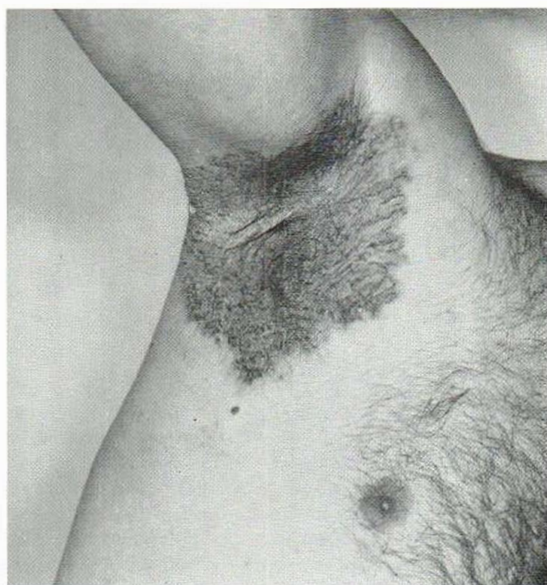


Fig. 3. Right axilla before surgery.

was completed in September 1969, when the involved skin of the brachial surface was excised and replaced with healthy skin from the thigh. The postoperative course was without complication and the result was very satisfactory. The right axilla was treated similarly in 1972.

On re-examination in June 1974, all the grafted areas of the axillary and cubital folds as well as the left side of the neck were found to be in a perfect condition and quite asymptomatic (Figs. 2 and 4).

The patient had no complaints at all as regards his left axilla, but in the right axilla, however, there was a 2-3 cm broad zone showing the typical picture of Hailey and Hailey's disease in the surrounding skin peripheral to the grafted area (Fig. 4). Troublesome lesions of familial benign chronic pemphigus still remain in the untreated parts of the neck and especially in the genito-femoral folds.

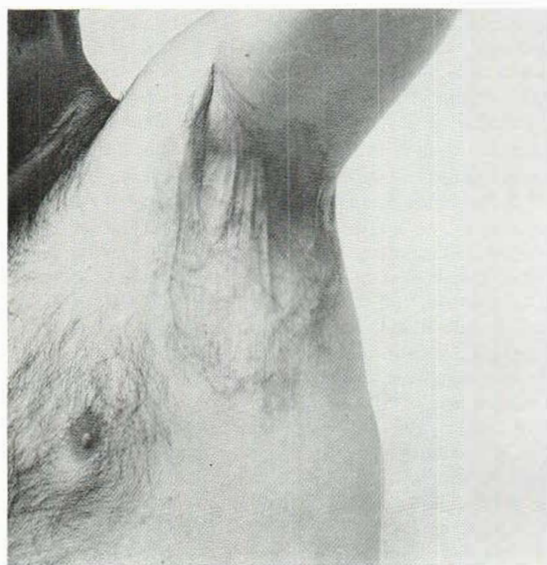


Fig. 2. Left axilla 5 years after adequate excision and reconstruction with a split skin graft (May, 1974).

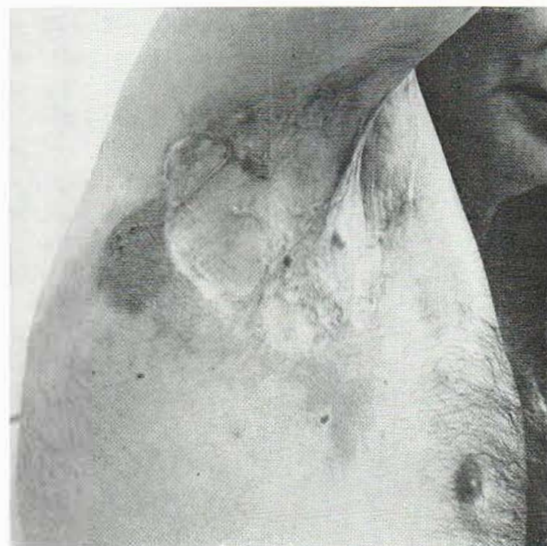


Fig. 4. Right axilla 2 years after surgery. Note the recurrence of the pemphigus lesion immediately outside the grafted area, due to insufficient extension of grafting.

DISCUSSION

The follow-up period of our patient, from 2 to 8 years, is of sufficient length to justify the conclusion that familial benign chronic pemphigus does not recur in a partial thickness skin graft. Thus, combined excision of the affected skin area and reconstruction with a split skin graft will lead to a permanent cure of the treated site.

The explanation for this cure may be of a complex nature. One probable factor is that the graft is taken from an anatomically separate skin region which is not predisposed to pemphigus. If this were the only reason, reconstruction with local flaps would in some instances even be preferable to a free graft. Another possible factor is the absence of the deeper layer of the dermis from the graft. All skin adnexa, for example apocrine as well as eccrine sweat glands, are scarcely present and the innervation regenerates poorly (7). Thus, axillary hyperhidrosis, for example, is successfully treated by excision of the layer containing the sweat glands only (9).

Excision of the lesion should take with it all skin affected during any earlier period. The recurrence of the process in the bordering skin immediately outside the grafted area in the right axilla of our patient is evidence in favour of this theory. As no recent exacerbation of the axillary lesion had preceded the surgical intervention, one may presume that, at this moment, the pemphigus lesions had not reached their maximal extension. The surrounding skin appeared clinically normal. For this reason the area of the skin graft was apparently not large enough. (The recurrence could of course easily be treated later with an additional skin grafting.)

REFERENCES

1. Berger, R. S. & Lynch, P. J.: Familial benign chronic pemphigus. Surgical treatment and pathogenesis. *Arch Dermatol* 104: 380, 1971.
2. Biró, S. & Maday, P.: A pemphigus benignus chronicus familiaris. (Morbus Hailey-Hailey) és annak sikeres műtéti gyógyítása. *Bőrgyógy. Vener Szemle* 42: 225, 1966.
3. Biró, F. A. & Maday, P.: Familial chronic pemphigus (Letter to the Editor). *Arch Dermatol* 100: 385, 1969.
4. Bitar, A. & Giroux, J.-M.: Treatment of benign familial pemphigus (Hailey-Hailey) by skin grafting. *Br J Dermatol* 83: 402, 1970.
5. Gougerot, H. & Allée: Forme de transition entre la dermatite polymorphe douloureuse de Brocq-Dühring, et le pemphigus congénital familial héréditaire. *Arch Derm Syph Clin Hôp* 5: 255, 1933.
6. Hailey, H. & Hailey, H.: Familial benign chronic pemphigus. *Arch Dermatol* 39: 679, 1939.
7. Pontén, B.: Grafted skin. Observations on innervation and other qualities. *Acta Chir Scand*, Suppl. 257, 1960.
8. Shelley, W. B. & Randall, P.: Surgical eradication of familial benign chronic pemphigus from the axillae. *Arch Dermatol* 100: 275, 1969.
9. Skoog, T. & Thyresson, N.: Hyperhidrosis of the axillae. A method of surgical treatment. *Acta Chir Scand* 124: 531, 1962.
10. Sonck, C. E.: On the occurrence of familial benign chronic pemphigus (Hailey and Hailey) in Finland. *Proc Congr Hungar Derm Soc*, pp. 407-414, Budapest 1965.
11. Thorne, F. L.: Letter to the Editor. *Arch Dermatol* 100: 385, 1969.
12. Thorne, F. L., Hall, J. H., & Mladick, R. A.: Surgical treatment of familial chronic pemphigus (Hailey-Hailey Disease). Report of a case. *Arch Dermatol* 98: 522, 1968.

Received November 15, 1974

C. E. Sonck, M.D.
Kauppiaskatu 4 A
SF-20100 Turku 10
Finland