

Vaginal Involvement in Familial Benign Chronic Pemphigus (Morbus Hailey-Hailey)

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Abstract. Vaginal pain over a period of 3 years has been shown to be based on a vesiculo-bullous acantholytic disorder suggesting familial chronic benign pemphigus (Morbus Hailey-Hailey). Local application of 5 ml 1% hydrocortisone-entérochinol cream resulted in an almost total regression of all the symptoms, including dyspareunia.

Only a few cases of histologically verified familial benign chronic pemphigus (FBCP) involving the stratified mucous membranes have been reported. The following findings regarding location have been noted: oral (2, 4), ocular (1), oesophageal (5), laryngeal (6) and vulvar (3). No cases with a vaginal localization have been described in the literature available.

CASE REPORT

In 1977 L. J., a 36-year-old woman complaining of a smarting pain in the vagina, but without any significant dis-

charge, consulted one of the authors. Due to a variety of bacteriological findings (*Escherichia coli*, *Lactobacillus* and, on one occasion, *Candida albicans*) corresponding treatment was given locally and systemically, from which the patient did not benefit, however. During the last 2 years she has complained of an aggravation of her vaginal pain, leading to dyspareunia to such an extent that even the pressure of close-fitting pants could provoke pain. Her father had for many years had a chronic dermatosis in the perianal region which unfortunately was not verified diagnostically.

Colposcopic inspection showed a swollen, bright red vaginal mucosa with many smooth erosions and shiny white-spotted patches about 0.2–1.8 cm in size. A biopsy specimen showed a large bulla (Fig. 1) and tiny vesicles in a predominantly suprabasal position, containing individual acantholytic cells as well as aggregations of cells, exhibiting largely the same staining and morphological properties as the epithelial cells in the compact epidermis. The acantholytic process was more obvious in the lower part of the overlying epithelium, exhibiting the appearance of a dilapidated brick wall. The basal layer was coherent, with only tiny cellular defects in a few places. Only a slight oedema was found in the submucosa. These histological findings render a diagnosis of FBCP the most likely one.

The specimen for fluorescence examination has unfortunately been damaged, but it was estimated to be probably negative for deposits of immunoglobulins and complement.

TREATMENT

5 ml of 1% hydrocortisone-entérochinol in a cream base was daily applied vaginally for a period of 7 days and an almost total regression of all

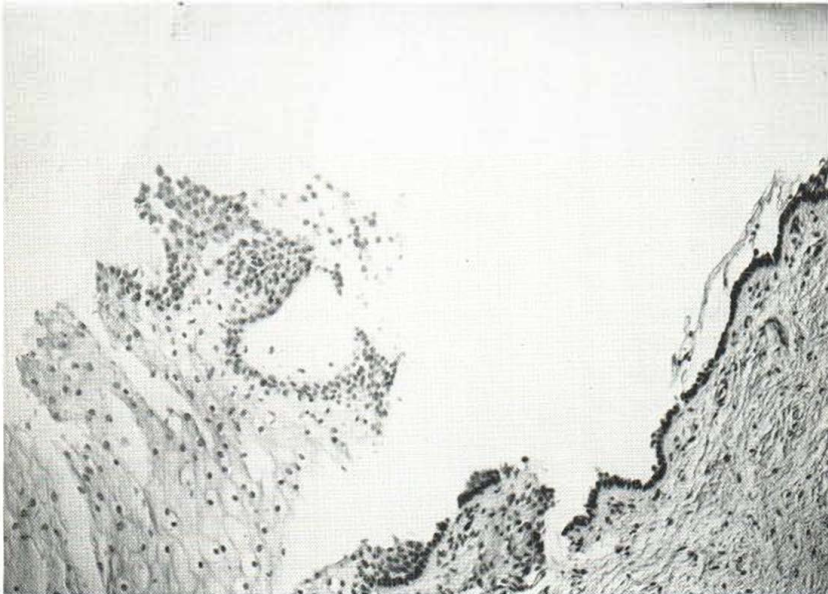


Fig. 1. Part of an intra-epithelial bulla with a mostly coherent basal layer. A few acantholytic cells can be seen in the suprabasal position. In the lower part of the overlying epithelium a pronounced acantholysis in the form of intra-epithelial clefts, detached individual cells, and clumps of cells can be seen.

symptoms, including dyspareunia, was achieved. Slight recurrences—about 4–5 times a year—subsided after a few days' treatment with hydrocortisone–enterochinol cream.

COMMENTS

This is the first described vesiculo-bullous disease in the vagina with histological changes suggesting FBCP. The prompt, rapid benefit afforded by local corticosteroid treatment is explained by the extraordinary favourable conditions for resorption through the damaged vaginal mucosa.

The purpose of this paper is not only to point out the possibility of a vaginal localization of FBCP but to encourage histological examination in cases of vaginal complaints combined with dyspareunia refractory to conventional treatment.

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Generalized Morphoea Successfully Treated with Salazopyrine

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Abstract. A patient with active, generalized morphoea was treated successfully with salazopyrine on two occa-

sions. The first time the trunk was involved and most of the skin softened after the drug therapy was started. After salazopyrine was discontinued, morphoea developed on the thighs but recommencement of therapy resulted in complete resolution.

Key words: Generalized morphoea; Active disease; Salazopyrine

Morphoea and scleroderma are resistant to most forms of treatment. There have been reports of these disorders responding to salazopyrine (1, 2), and with this drug we have successfully treated a patient who had generalized morphoea on two occasions.

CASE REPORT

A 60-year-old woman was first seen in June 1979; she had morphoea involving the entire lower half of the trunk, and also sclerodactyly. The morphoea had evolved over 2 years and was still progressing. She had suffered from Raynaud's phenomenon and tightening of the skin of the hands for a similar period. There were no symptoms of internal involvement. A biopsy confirmed the diagnosis of morphoea. A complete blood count, electrocardiogram and serum creatinine were normal and examination for antinuclear factor was negative.

Salazopyrine, 1 g twice a day, was commenced in July 1979. Within one month there was softening of the sclerotic skin of the trunk. The skin continued to improve over the next few months and, when salazopyrine was discontinued in May 1980, only the lowest quarter of the involved skin was still sclerotic although the remainder remained hyperpigmented. There was no change in the sclerodactyly and in fact, bilateral cervical sympathectomies had to be performed for incipient gangrene. Only temporary relief was obtained from the operation.

In October 1980 the patient returned because morphoea had developed on both thighs. Salazopyrine, again 1 g twice daily, was recommenced and the sclerotic skin began to soften within one month. The thighs had returned completely to normal by January 1981 and the drug was again discontinued. There was no change in the sclerodactyly. The drug was well tolerated and there were no side effects apart from mild nausea occasionally.

DISCUSSION

Clinical trials with salazopyrine in sclerotic disorders have had mixed results. Dover reported a good response in patients with progressive systemic sclerosis (2), while Barnett et al. found it to be ineffective (1). Stava et al. reported it to be effective in generalized morphoea which was still evolving, but not in stable morphoea or in systemic sclerosis (3), similar to the response in this patient. The