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SHORT COMMUNICATION

Peristomal Epidermolysis Bullosa Acquisita in a Patient with Crohn's Disease

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Intestinal stomas consist of a section of gastrointestinal tract that is opened to the skin in order to drain the stoma effluents. Most frequent indications are malignancy and inflammatory bowel disease (IBD). Peristomal skin plays an important role in the adequate use of stoma appliance, being essential to maintain a normal lifestyle (1). It is therefore imperative to correctly diagnose the skin problems in patients with a stoma (2).

CASE REPORT

A 56-year-old man who had undergone an ileostomy 9 years before, noticed erosions at the site of the stoma for the last 10 months. He had a medical history of Crohn's disease (CD) diagnosed 25 years ago that required a proctocolectomy 9 years previously but he had not received immunossuppresive therapy for over one year. Twenty years earlier a diagnosis of epidermolysis bullosa acquisita (EBA) was made in our department, but the patient had remained cutaneous disease-free for the last 15 years.

Physical examination revealed an eroded and indurated erythematous plaque along with isolated blisters surrounding the ileostomy on the right lower quadrant of his abdomen (Fig. 1A). There were no other skin or mucous membrane lesions. Histological examination of one of the erosions revealed a subepidermal blister with a poor inflammatory infiltrate (Fig. 1B). Standard Spanish patch testing and also patch testing with antiseptics and stomal devices showed a negative reaction. A diagnosis of peristomal EBA was established by exclusion of other diagnoses. Initially local treatment with dressings was prescribed, achieving little improvement. Instead, we performed periodical steroid infiltrations.

DISCUSSION

The differential diagnosis of a blistering peristomal lesion includes contact irritation, allergic contact dermatitis, EBA, bullous pemphigoid (BP), pemphigus vulgaris (PV), infection, pyoderma gangrenosum (PG) and squamous cell carcinoma. In our patient, irritant and contact dermatitis could be ruled out as there were no leakages, the skin was not exposed to stoma effluents and the patch test was negative. There are a few cases of peristomal BP (3–7) and PV (8) described in the literature. In these cases indirect immunofluorescence studies lead to the correct diagnosis. Peristomal pyoderma gangrenosum is a variant of classic PG. Although it is a rare condition, it is more common than expected, most likely due to its association to IBD. The lack of neutrophils in the biopsy excluded this diagnosis. Infection of the peristomal skin can be due to bacteria, fungi or viruses, and this may be caused by a combination of factors such as loss of the integrity of the peristomal skin, colonisation by enteric flora, or immunosuppression due to treatment of inflammatory BD. Our biopsy was inconsistent with this diagnosis. Malignancy could also be excluded based on the histology. Metastatic CD is an extension of the inflammatory pathology to other sites than the gastrointestinal tract. This option should also be taken into account, but the biopsy in this case was not consistent with it. Any generalised or pre-existing dermatosis may affect the skin around the stoma. The diagnosis is then generally made by assessment of the clinical signs in places unrelated to the stoma. Since our patient had been EBA disease-free for the last 10 years, we did not consider EBA as the first diagnostic option. Nevertheless, peristomal EBA was finally confirmed by the biopsies that demonstrated its classical features.

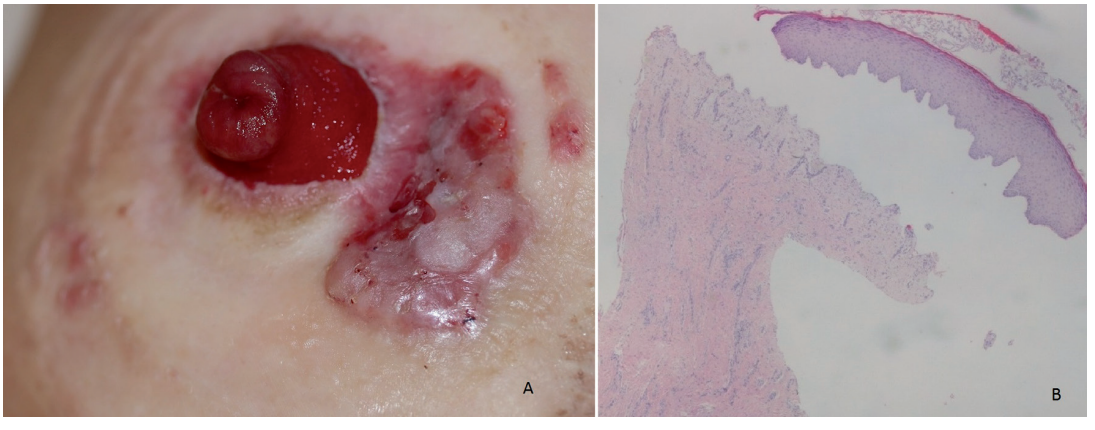

Fig. 1. Indurated erythematous plaque and blisters surrounding the ileostomy (A). Subepidermal blister with a poor inflammatory infiltrate (B).

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Regarding the pathogenesis of peristomal EBA, Koebner phenomenon has been suggested (3). Moreover, it is considered that trauma could lead to the development of blisters through modification of the basement membrane zone antigens, which would allow antigen presentation followed by autoimmunisation (5, 7). In our case, the affected area was exposed to continuous trauma, which could explain the pathogenic mechanism of blister formation.

An intriguing association between EBA and CD has been extensively documented over the last decades. CD has been described in approximately 30% of EBA patients, and auto-antibodies against type VII collagen have been detected in up to 68% of IBD patients (9). These findings suggest that chronic intestinal inflammation in IBD predisposes to autoimmunity against type VII collagen. It is proposed that the inflammation in IBD exposes type VII collagen within the basement membrane of the gut, stimulating the production of auto-antibodies. In susceptible individuals, these antibodies cross-react with the collagen type VII of the skin, provoking blisters and erosions that constitute EBA (10). Despite this frequent association, peristomal EBA in patients with underlying IBD has not been reported.

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