Sir,

Bullous pemphigoid (BP) is the most frequent autoimmune bullous disorder. BP is associated with antibodies to two hemidesmosomal proteins, BP180 (type XVII collagen) and BP230 (1). The disorder usually affects elderly people, and severe pruritus is typically present. In the majority of patients, lesions are generalized and are most common on flexural aspects of the limbs and lower abdomen (2). By contrast, localized forms have also been reported at particular sites, including the pre-tibial area, palmo-plantar region, and genital, perineal, and perianal area, or confined to sites previously affected by, for example, radiotherapy, surgery, trauma, and burns, as well as around stomata and haemodialysis fistulae (2–4). Localized lesions may remain localized or develop into classical BP.

CASE REPORT

An otherwise healthy 61-year-old Caucasian woman presented with a 2–3-year history of itchy erythema and erosions in the umbilical area, which had occasionally lead to a burning sensation and blood-tinged exudation. Treatment with topical antiseptics had not been effective, but the lesions sometimes improved spontaneously. Ultrasound examination had been unremarkable one year previously. Most bacterial swabs had been negative, and candidosis had been excluded repeatedly. No type IV allergy against nickel was reported. The patient was taking fenofibrate, captopril, hydrochlorothiazide, and citalopram for hypercholesterolaemia, arterial hypertension, and depression, respectively, all of which treatments had been initiated without relation to the onset of the skin lesion. On examination, a sharply demarcated erythema, erosions, and a tense vesicle were found in the umbilical area (Fig. 1). The remaining skin and mucous membranes were unaffected.

Histopathology of a lesional skin biopsy showed that the epidermis was completely detached from the dermis and revealed a dense, almost band-like, infiltrate of lymphocytes and eosinophil granulocytes in the underlying papillary dermis. Direct immunofluorescence microscopy of a perilesional skin biopsy showed strong linear staining of C3 and IgG (Fig. 2a) and weaker labelling of IgA at the dermal-epidermal junction. The n-serrated pattern seen in some areas is typically found in diseases with lamina lucida autoantibody reactivity and differentiates them from epidermolysis bullosa acquisita (5). By indirect immunofluo-
rescence (IF) on human 1 M NaCl-split human skin, weak binding of IgA at the epidermal side of the arti-

cicial split was shown to be present (Fig. 2b), while no

IgG reactivity was found. By immunoblotting of the recombinant 16th non-collagenous domain of BP180

(BP180 NC16A) (6), the immunodominant region in classical BP, serum IgA antibodies were detected (Fig.

2d), whereas IgG reactivity was absent. In addition, the patient’s serum contained IgG antibodies to the soluble

cetodomain of BP180 (LAD-1) by immunoblotting of concentrated conditioned medium of cultured HaCaT

cells (Fig. 2c) (6). The stronger IgG staining by direct

IF microscopy and the strong labelling of C3, that can only be weakly induced by IgA autoantibodies, favoured

a diagnosis of BP.

The combined use of crystal violet solution 0.1% and

betamethasone 0.1% cream resulted in rapid resolution of

the lesion. After withdrawal of the topical cortico-

steroid the lesion recurred, but this was controlled by

its re-application once per week. After 2 months, the

lesion finally healed completely and the topical cortico-

steroid was tapered off.

Our patient presented with immunopathological features of BP; however, there were several interesting find-
ings in this case, including the peculiar clinical picture, the combined IgA and IgG response against BP180,

and the patient’s relatively young age. Interestingly, her

lesions were restricted to the umbilicus. While the umbi-

lical area is a predilection site for patients with classical

BP (2), BP limited to this region has not been described

previously. Periumbilical lesions are also typical for

patients with pemphigoid gestationis. Our patient had had

two pregnancies without skin lesions or pruritus and gave

birth to two healthy children. The specificity of serum

autoantibodies has been reported rarely in patients with

localized BP. In two patients with lesions limited to the

pretibial area and face and scalp, respectively, reactivity

against the BP180 NC16A domain was described (7, 8)

and in a child with vulval BP, reactivity against LAD-1

was observed (9). In these localized variants including our

patient, the autoantibody response is comparable with

classical BP and favours the hypothesis that localized

forms of BP are not proper entities, but rather represent

forms of classical BP with lower disease activity.

The generation of both IgG and IgA autoantibodies

against BP180 is also frequently observed in classical

BP. Indeed, the majority of BP patients develop, in addition to IgG autoantibodies, IgA reactivity against BP180

(10). It may be speculated that the IgA autoantibodies are due to the relatively young age of our patient. We

have previously shown that patients with IgG antibodies to the basal membrane zone are significantly older

compared with patients with IgA reactivity (11).

This report describes an unusual case of localized

BP and highlights the importance of suspecting BP in chronic pruritic erythema and erosions, even when

lesions are restricted to small areas and no obvious blistering is present.

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REFERENCES


