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-tingled 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 cilostazol 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-

Fig. 1. Erythema, erosions, and a tense vesicle in the umbilical area, in an area of approximately 2.5 cm².
recurrence (IF) on human 1 M NaCl-split human skin, weak binding of IgA at the epidermal side of the artificial 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 ectodomain 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 corticosteroid the lesion recurred, but this was controlled by its re-application once per week. After 20 months, the lesion finally healed completely and the topical corticosteroid was tapered off.

Our patient presented with immunopathological features of BP; however, there were several interesting findings 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 umbilical area is a predilection site for patients with 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 ectodomain 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.

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.

ACKNOWLEDGEMENT
We thank Silvana Noll, Würzburg, for excellent technical assistance.

The authors declare no conflict of interest.

REFERENCES