

CASE REPORT

A case report of dyshidrotic bullous pemphigoid developing after partial anterior circulation ischaemic stroke

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Abstract

Dyshidrotic bullous pemphigoid is a rare form of bullous pemphigoid that affects predominantly a patient's hands and feet. It has been associated in the literature with neurologic, psychiatric and cerebrovascular disorders. We present an interesting case of this rare skin condition developing in a patient following a diagnosis of partial anterior circulation stroke.

INTRODUCTION

Dyshidrotic bullous pemphigoid (DBP) is a unique variant of bullous pemphigoid (BP) that affects predominantly a patient's hands and feet. This report presents an interesting case of this rare skin condition developing in a male patient following a diagnosis of stroke.

CASE REPORT

An 87-year-old gentleman was admitted to our stroke rehabilitation unit following a right partial anterior circulation ischaemic stroke. The patient had a past medical history of hip osteoarthritis, atrial fibrillation, hypertension, asthma and angina; no previous dermatological conditions were noted.

One month after the stroke, the patient developed a widespread maculopapular, blanching rash to the trunk and

limbs. This was accompanied by petechial lesions on the dorsum of the feet. It was thought to represent a possible allergic rash and spontaneously resolved within 3 weeks.

A month following this episode the patient developed vesicular lesions on the palmar aspect of both hands (Fig. 1). He then went on to develop two large bullae measuring 5 × 3 cm and 8 × 4 cm over his left foot and over the following days more bullae appeared over both feet. These were tense and most were haemorrhagic (Fig. 2). He did not develop any blisters elsewhere and there was no mucosal involvement. Punch biopsy of a vesicle appeared to show an intra-epidermal lesion; however, the roof comprised of necrotic keratinocytes and it was commented that this could represent re-epithelialization of a sub-epidermal blister (Fig. 3). Prominent eosinophilic inflammatory infiltrate was noted in the superficial dermis, with extravasation of red blood cells. No sample for direct immunofluorescence (DIF) was taken as the patient was reviewed at a peripheral site out of hours. Serum skin autoantibodies were found to be positive at a titre

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Figure 1: Vesicles appearing 1 month after the stroke.



Figure 2: Tense and haemorrhagic bullae appearing on feet.

of $>1/320$ for pemphigoid and supported the clinical diagnosis of BP.

The patient was started on an ultra-potent steroid cream (1 application once daily), emollients and long-term treatment with oral doxycycline (100 mg twice daily). Within a month the

skin lesions resolved and the patient was able to once again participate in stroke-specific rehabilitation. Post-treatment titre was not evaluated as this is not routinely done at our institution.

DISCUSSION

The immunopathogenesis of BP involves the presence of circulating autoantibodies directed against two proteins: dystonin, also known as BP antigen 1 (BPAG1), and type XVII collagen, known also as BP antigen 2 (BPAG2), of 230 and 180 kDa, respectively [1]. Both are adhesion junction plaque proteins within the hemidesmosome, which anchors the basal keratinocytes to the basement membrane. When these proteins are targeted there is separation at the dermo-epidermal junction, with the formation of sub-epidermal blisters [1].

DBP is a clinical variant of BP distinguishing feature of which is the initial or persistent palmoplantar localization of the lesions. Bullae or vesicles can be haemorrhagic in nature, and purpuric lesions are common [2]; as in the case described, some patients might have prodromal symptoms, mainly eczematous or papular eruptions [3].

Differential diagnosis includes mainly dyshidrotic eczema, which presents with smaller and typically non-haemorrhagic vesicles, but also palmoplantar pustulosis, bullous tinea infection, scabies infection, contact dermatitis, bullous lichen planus and T-cell lymphoma. DBP is typically confirmed by skin biopsy, demonstrating sub-epidermal bullae and eosinophil infiltration, and DIF to show linear deposits of IgG along the dermo-epidermal junction. Treatment is dependant on disease severity. Topical steroids are often sufficient in mild disease and can be used in combination with oral tetracyclines as in this case. Oral steroid therapy is often required and systemic immunosuppression may be needed to wean steroid dose. Although skin lesions are not usually life-threatening, BP is associated with a 6–41% mortality at 1 year [4].

DBP has been associated with neurologic, psychiatric and cerebrovascular disorders. There are only three cases reported in the literature of DBP in patients with a past history of stroke [3]; however, to the best of our knowledge, no previous case has been reported of DBP developing immediately after a cerebrovascular event. Stroke as a risk factor for developing BP has been studied in the literature. Shen *et al.* [5] demonstrated with a large-scale population-based study that the hazard ratio of BP was significantly higher in stroke patients compared with the control group. The hypothesis behind this correlation is that damage to the blood-brain barrier in stroke generates autoantibodies that cross-react with BPAG1/BPAG2 [5]. Interestingly, the inverse relationship might also be true: patients with a history of BP have a 2-fold increased risk of developing stroke [6]. BP patients seems in fact to have a dysfunctioning endothelium, as demonstrated by their higher serum concentration of pro-inflammatory cytokines such as the E-selectin, and this has been considered a risk factor for developing cerebrovascular events [7].

In conclusions, we presented a case of DBP developing after stroke. More studies are necessary to clarify the relationship between these two clinical entities. DBP should be considered

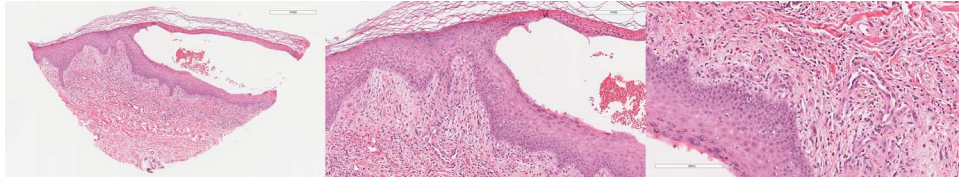


Figure 3: The biopsy showed an intra-epidermal vesicle with a necrotic roof; the underlying dermis showed numerous eosinophils.

in older patients presenting with blistering lesions to the hands and feet, particularly when haemorrhagic.

CONFLICT OF INTEREST

The authors declare that they have no conflict of interest.

FUNDING

None.

CONSENT

Written consent obtained.

ETHICAL APPROVAL

Authors confirm that the research meets ethical guidelines and adheres to the local legal requirements.

GUARANTOR

Daniele Fanelli.

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