

CASE REPORT

Anti-p200 pemphigoid responding to dapsone



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ABSTRACT

Anti-p200 pemphigoid is a rare autoimmune subepidermal blistering disease. Clinical presentation is similar to standard bullous pemphigoid (BP) but mucous membranes and cephalic lesions are more frequent. Histology and direct immunofluorescence (IF) are identical to BP but indirect IF discloses linear deposits of immunoglobulin G (IgG) on the dermal side of artificial salt-split skin. Specific diagnosis is based on western immunoblotting that shows circulating IgG recognizing a 200-kDa protein localized on the dermal extract. The 200-kDa antigen was recently identified as laminin γ 1. Anti-p200 pemphigoid should be considered before all atypical or topical steroid-resistant bullous disease, as well as mucous membranes pemphigoid or epidermolysis bullosa acquisita. Dapsone appears to be the most effective treatment and should be used as the first option in combination with topical steroids. In this report, we describe the case of a patient with a typical clinical and immunopathological anti-p200 pemphigoid, responding to a combination of topical steroids and dapsone.

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Introduction

Anti-laminin γ 1 pemphigoid is a recently-described autoimmune subepidermal bullous disease that has been characterized by autoantibodies against a 200-kDa protein (p200) of the dermal–epidermal junction (DEJ).

Here, we describe the typical clinical, histopathological, and immunopathological features in a patient with anti-p200 pemphigoid and his favorable response to treatment with dapsone.

Case report

A 74-year-old man presented with a blistering pruriginous eruption evolving for 3 months. Skin lesions were mainly localized on palms and soles with erythematous plaques and vesicles or tense blisters (Figure 1A). Oral and genital mucosae were involved with large painful erosions (Figure 1B). The trunk and limbs showed polymorphic erythematous plaques (Figure 1C). The patient was treated

for several years with nebivolol and amlodipine for hypertension. There was no new medication introduced. A personal history for cutaneous disease was negative. Standard blood analysis showed no specificity and particularly no hypereosinophilia.

Histopathological analysis of the lesional skin revealed a slight dermal–epidermal separation and a neutrophilic and eosinophilic infiltrate in the dermal papillae (Figure 2A). Direct immunofluorescence (IF) showed the presence of linear immunoglobulin G (IgG) and complement 3 (C3) at the DEJ (Figure 2B). Diagnosis of bullous pemphigoid (BP) was established and the patient was successfully treated by clobetasol propionate 0.05% cream 40 g daily. However, skin lesions reappeared when the treatment was tapered. Because of this steroid resistance, new analyses were realized. Secondary histopathological analysis and direct IF confirmed the diagnosis of BP. Indirect IF disclosed circulating antibodies at a titer of 1:10, which is not significant. Those antibodies reacted with the floor of an artificial blister created by the salt-split skin technique (Figure 3). Enzyme-linked immunosorbent assay (ELISA) with basal membrane zone proteins of 230 kDa (BP230) and 180 kDa (BP180) were negative. Western immunoblot analysis using a dermal extract showed the reactivity of circulating IgG4 antibodies with the 200-kDa antigen, suggesting the diagnosis of anti-p200 pemphigoid (Figure 4).

The patient was successfully treated by combining clobetasol propionate 0.05% cream 30 g daily and dapsone 100 mg daily, with a progressive healing of erosions without scarring and milia

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Figure 1 Clinical characterization of a patient with anti-p200 pemphigoid. (A) Tense blisters and erythematous plaques on the sole of the foot. (B) Mucous membrane lesions and tense blister on the finger. (C) Pruriginous polymorphic eruption on the trunk and limbs. (D) Complete healing after 2 weeks of topical clobetasol propionate 30 g and dapsone 100 mg daily.

formation (Figure 1D). His disease is currently controlled with dapsone 100 mg daily and without the use of corticosteroids.

Discussion

In 1996, Zillikens et al¹ described the first case of a new sub-epidermal immunobullous disease called anti-p200 pemphigoid. Approximately 70 cases of anti-p200 pemphigoid mediated by IgG antibodies have been reported in the literature to date,² and only one was associated exclusively with immunoglobulin A (IgA) antibodies.³ In 2009, Dainichi et al⁴ identified the 200-kDa protein as laminin γ 1 and renamed the disease anti-laminin γ 1 pemphigoid.

Anti-p200 pemphigoid usually occurs at a younger age than that observed in patients with BP and appears to be more frequent in male patients.⁵ The presentation is rather heterogenous and may

mimic BP, linear IgA dermatosis, dermatitis herpetiformis,² or epidermolysis bullosa acquisita (EBA).⁶ The BP-like type is the most common and is characterized by itchy urticarial papules and plaques and tense blisters on the trunk and extremities.⁵ Oral and genital mucous membranes are affected in approximately 20% of patients with anti-p200 pemphigoid and cephalic involvement is common.⁵ Blood analysis does not disclose the hyper eosinophilia found in classic BP.

In our patient, we found acral predominance of the lesions and considerable mucous membrane involvement. Lesions usually heal without scarring; milia formation has been observed in three previously reported cases.^{7–9}

Except for psoriasis in one-third of cases, no other cutaneous disease or tumor seems to be associated with anti-laminin γ 1 pemphigoid. There is only one case report about penicillin-induced anti-p200 pemphigoid.¹⁰

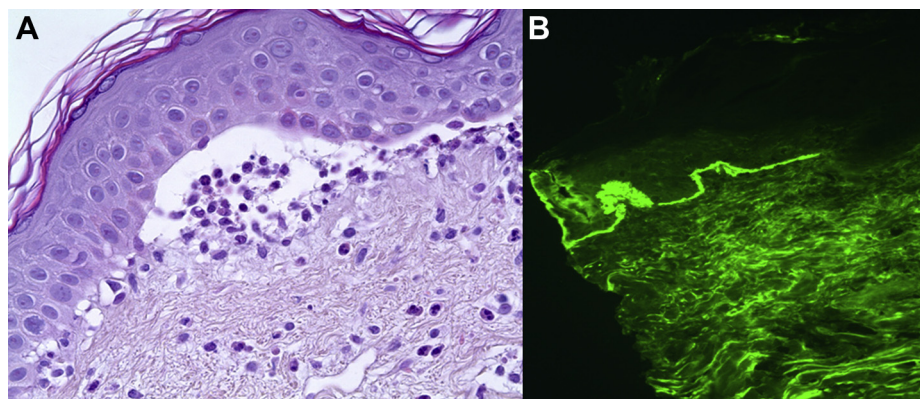


Figure 2 (A) Histological analysis: hematoxylin–eosin staining of the lesional skin ($\times 20$) showing slight dermal–epidermal separation and a neutrophilic and eosinophilic infiltrate in the dermal papillae. (B) Direct immunofluorescence shows immunoglobulin G and complement 3 deposits along the basement membrane zone.

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