Clinical Cases in Dermatology
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Dédée F. Murrell *Editor*

Clinical Cases in Autoimmune Blistering Diseases

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Clinical Cases in Autoimmune Blistering Diseases

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Chapter 1
An Elderly Patient with a Generalized Pruritic Eruption

Stefanie Häfliger and Luca Borradori

A 86-year-old patient presented with generalized pruritic eruption of 3 month duration. The patient had a past history of type 2 insulin-dependent diabetes mellitus, dyslipidemia and arterial hypertension. On examination, the patient showed widespread excoriations, prurigo-like lesions, post-inflammatory hypopigmentations, with atrophic scarring distributed predominantly over his trunk, upper limbs, neck, and scalp (Fig. 1.1a–c). On his lower limbs, some scratched lesions were also observed. The patient also had isolated erosions on his buccal mucosa (Fig. 1.1d). Light microscopy studies showed changes consistent with chronic prurigo. Direct immunofluorescence microscopy studies obtained from perilesional skin showed linear deposits of IgG and C3 along the epidermal basement membrane zone. By indirect IF microscopy using NaCl-separated normal human skin, there were circulating IgG autoantibodies binding the epidermal side of the split. The search of circulating anti-BP180 antibodies by ELISA was positive (41.7 U/ml; N: 9 < U/ml).

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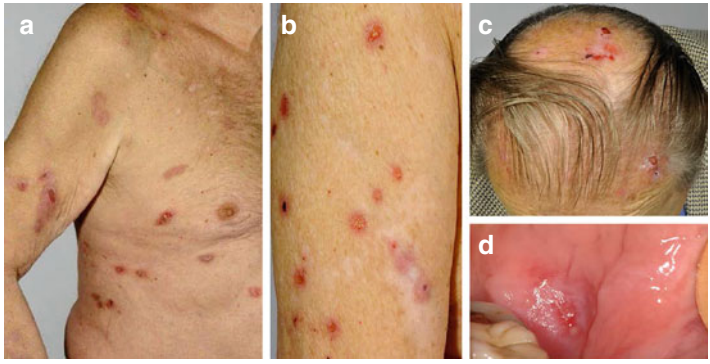

FIGURE 1.1 (**a**) Excoriations, postinflammatory hypopigmentations and atrophic scarring on trunk and arms; (**b**) Close-up view of excoriated lesions and scarring with isolated milia on arm; (**c**) Erosions, crusting and atrophic scarring on the scalp; (**d**) Erosion on buccal mucosa

What Is Your Diagnosis?

- Bullous pemphigoid (BP)
- Mucous membrane pemphigoid
- Brunsting-Perry pemphigoid

Discussion (1)

Diagnosis of BP is not always easy and straightforward. Manifestations of BP might resemble those of a variety of dermatoses, including drug reactions, contact dermatitis, prurigo, fixed urticaria, vasculitis, arthropod reaction and scabies (Table 1.1). Although the recent availability of ELISAs have facilitated the search of circulating autoantibodies, diagnosis still relies on a combination of clinical, histopathological and immunopathological features, particularly direct IF microscopy findings [5].

Our patient presented with chronic excoriated lesions and post-inflammatory changes predominantly localized on the

Table 1.1 Clinical presentations of bullous pemphigoid

Chronic prurigo, prurigo nodularis-like features
Papular pemphigoid
Eczematous lesions
Erythema multiforme-like and Lyell-like pemphigoid
Lymphomatoid papulosis-like
Ecthyma-like
Palmo-plantar lesions (dysidrosiform pemphigoid)
Intertrigo (vegetating pemphigoid)
Vesicular pemphigoid
Erythrodermic pemphigoid
Brunsting-Perry form (variant of cicatricial pemphigoid with skin lesions)
Localized forms
Pretibial
Peristomal
Umbilical
"Stump" pemphigoid
On paralyzed body sites
On irradiated/traumatised body sites

upper trunk and his head and as well as isolated lesions of the buccal mucosa. Immunopathological findings were consistent with the pemphigoid group of autoimmune bullous disorders. We favor the diagnosis of an unusual form of chronic prurigo-like BP [5]. Nevertheless, our case presented also with features of the so called Brunsting-Perry variant of mucous membrane pemphigoid [2] with a peculiar extensive cutaneous involvement. In the absence of well recognized criteria, a conclusive classification of our case is not possible.

How Do You Manage This Patient?

- Topical corticosteroids
- Tetracyclines and nicotinamide
- Systemic steroids
- Systemic Steroids and immunsuppressants

Our BP patient was first treated with topical clobetasol propionate 0.05 % combined with doxyciclin, 200 mg daily and nicotinamide, 2 g daily. Since this regimen did not sufficiently control his skin disease, the patient was first given sulfasalazine and later oral prednisolone, 0.5 mg per kg body weight. The latter treatment resulted in control of the disease, but lead to a severe weight gain and a decompensation of his diabetes. Finally, topical steroids were initiated in combination with azathioprine 75 mg daily which lead to a complete remission of the skin disease.

Discussion (2)

BP has frequently a chronic evolution with remissions and relapses. It is associated with significant morbidity, such as severe itch, bullous and eroded lesions, and impetiginisation. The impact on the quality of life is significant [5].

Prior starting a therapy in patients with bullous pemphigoid, the overall clinical context and the evidence about the available therapeutic intervention should be considered: (1) affected patients have usually an advanced age, older than 75 years of age, (2) they have frequently additional comorbidities, such as neurological or cardiovascular diseases (3) they show a significantly increased mortality patients during the first year of treatment. Finally, (4) so far, except for topical and systemic steroids, there are no studies which have validated the use of the other drugs, which have been commonly used in BP [1].

The first line therapeutic option in localized, mild bullous pemphigoid consists of high potency topical corticosteroids