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Bullous pemphigoid mimicking granulomatous inflammation

Abhilasha Williams, Emy Abi Thomas.

Department of Dermatology, Christian Medical College and Hospital, Ludhiana, Punjab, India.

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Corresponding author:

Dr. Abhilasha Williams

Email: abhilasha.williams@gmail.com

emyabi@gmail.com

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Abstract

Bullous pemphigoid is typically a disease of the elderly, with an onset after 60 years of age. It is the most common autoimmune subepidermal blistering disease of the skin which presents with generalized pruritic bullous eruption. The spectrum of clinical presentation is extremely broad and rare clinical variants have been reported in literature. We describe a patient who presented with ulceration of the nose of 3 months duration. He denied history of vesicles, bullae or urticarial lesions. The clinical diagnosis of granulomatous inflammation was considered. Biopsy done from the nose and buccal mucosa showed subepithelial eosinophil and neutrophil rich bullous lesion suggestive of bullous pemphigoid. On immuno-histochemistry, the floor of the bulla showed positive staining for laminin. The patient later developed tense vesicles and bullae over the trunk and thighs. He showed complete resolution of the skin lesions within a week of starting oral steroid therapy. These findings suggest our patient had an atypical, non-bullous presentation of the pemphigoid spectrum.

Introduction

Bullous pemphigoid (BP) is the most common autoimmune blistering disorder occurring in the elderly population presenting with tense erythematous blisters occurring on healthy or normal skin.^[1] It is usually a chronic disease, with spontaneous exacerbations and remissions, which may

be accompanied by significant morbidity. The presentation of BP is polymorphic and initially misdiagnosed and in early or atypical cases, full-blown bullous lesions may be completely absent. In these cases, establishing the diagnosis of BP requires a high degree of suspicion and it is important for prompt institution of appropriate treatment.

We report an unusual presentation of BP which clinically resembled a granulomatous inflammation. The diagnosis of BP was made on the basis of skin biopsy.

Case report

A 65 year old gentleman presented with an asymptomatic, gradually progressive ulcer over the tip of the nose of 3 months duration. He denied the appearance of vesicles, bullae or urticarial lesions and also denied trauma to the site of the lesion. There was no history of travel outside his state and he was treated for pulmonary tuberculosis 15 years ago. There was no history of photosensitivity, joint pains or oral ulcers and he did not have any systemic complaints.

Examination revealed an infiltrated, crusted, plaque with erythematous margins over the tip of the nose (**Fig 1**). Mucosal infiltrations were noted on the buccal mucosae (**Fig 2**). Palms and soles were spared and rest of the cutaneous and systemic examination was normal. A clinical diagnosis of granulomatous inflammation was made.



Fig 1: Well defined crusted plaques over the nose, lips and moustache area.



Fig 2: Mucosal infiltration over the hard palate and buccal mucosa.

Tissue cultures from the site did not grow fungal or bacterial elements. Biopsy done from the nose and buccal mucosa showed subepithelial eosinophil and neutrophil rich bullous lesion with spongiosis and occasional necrotic keratinocytes (**Fig 3**). On immuno-histochemistry, the floor of the bulla showed positive staining for laminin. Immuno-histochemistry for collagen 4 was non-contributory. A possibility of bullous pemphigoid was considered.

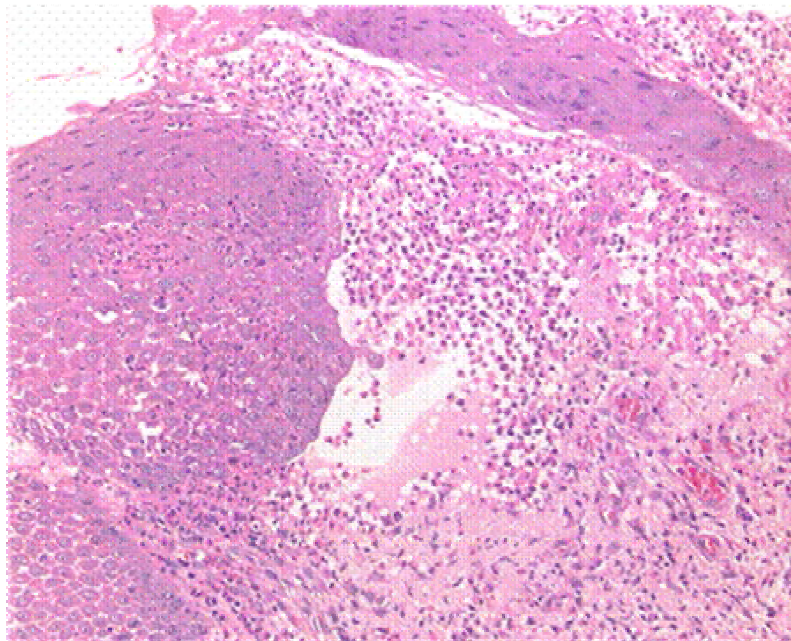


Fig 3: On H & E stained section (40X): Subepithelial eosinophil and neutrophil rich bullous lesion with spongiosis and occasional necrotic keratinocytes.

This patient re-presented after 2 weeks with new lesions on the lips and chin. Examination revealed yellowish crusted erosions over the lips and moustache area. A single yellowish plaque studded with pustules was present on the right mandibular area. A week later he developed tense vesicles and bullae over the groins and axillae. Since the histopathological diagnosis was established, repeat skin biopsy or direct immunofluorescence study from the new lesions was not done.

Patient started oral steroids at 1mg/kg/day and resolution of the cutaneous and mucosal lesions was noted within a week of initiation of therapy (**Fig 4**).



Fig 4: One week after treatment with oral steroids.

Discussion

Bullous pemphigoid, when presenting with tense pruritic blisters developing on either healthy or erythematous skin, is easily recognized. There may be mucosal involvement with blisters and erosions.[2] It is twice more common in males.[3]

The clinical hallmark of BP is the presence of widespread tense bullae, which may arise from normal (non-inflamed bullae) or erythematous (inflammatory bullae). They can occur anywhere, but there is a predilection for the groin, lower abdomen and the flexural surface of limbs.[2]

Rarely, non bullous manifestations of BP such as non bullous form, those presenting with exfoliative erythroderma, dyshydrotic eczema, prurigo nodularis, subacute simple prurigo, ecthyma gangrenosum are documented in literature.[4,5,6,7,8]

Our patient's presentation was a diagnostic challenge due to the lack of the characteristic

morphologic features of BP, namely vesicles or bullae at the time of presentation. It re emphasizes the need for diagnostic biopsies and immuno-histochemistry in elderly patients with atypical presentations. These patients should be followed up regularly as they are at risk for developing a generalized eruption later in life.

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