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Clinical features, diagnosis and treatment of bullous pemphigoid- A case report

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Abstract

Bullous pemphigoid (BP) is an acquired autoimmune disease that affects mainly the elderly and is characterized by subepidermal blistering. Various treatment modalities are available but still corticosteroids are mainstay of the treatment. Here we are presenting a case in which systemic and local corticosteroids are used for treatment of bullous pemphigoid.

Key words: Pemphigoid, Blisters, Corticosteroids

Introduction

The pemphigoid group of diseases is characterized by subepidermal blisters due to autoantibody-induced disruption of the components of the dermal– epidermal anchoring complex.[1]

Bullous pemphigoid is the most common blistering disorders affecting elderly. It presents clinically with diffuse eczematous, pruritic, urticaria-like lesions, with the later appearance of tense bullae or blistering lesions typically filled with clear fluid.[2] BP is an autoimmune disease defined immunologically by the existence of autoantibodies directed against 2 structural proteins found in the hemidesmosomes of the dermal-epidermal junction. These proteins, called BP antigen 1 (BPAG1) or AgBP230, and BPAG2 (or AgBP180 or collagen XVII).[3]

Mucous membranes are involved in about 50% of patients, with the oral mucosa most frequently affected. Oral lesions consist of small blisters or erosions and are found mainly on the palatal mucosa. The blister formation may be preceded by an urticarial or eczematous rash. The degree of itch varies from none to intense and may precede the appearance of blisters by weeks, months or occasionally years.[4]

Case report

A 48 year old female patient reported with a chief complaint of burning sensation in her mouth and difficulty to eat food from the past 1 month. She further told that she started experiencing burning sensation and difficulty in having food 2-3 years back. At that time she went to a local doctor where she was prescribed some medication that provided her relief for short period only. Since then she has repeated exacerbation & remission of this burning sensation. Now since 1 month patient reports of excessive burning sensation in her mouth because of which there is difficulty in having even salty food.

On Inspection - Diffuse erythematous ulcerative lesion evident on the right and left buccal mucosa,palate and floor of the mouth. On the right buccal mucosa lesion extended 3cm anterior to pterygomandibular raphe and

4cm away from the right corner of the mouth. Superoinferiorly it was 2cm from the maxillary buccal vestibule and 3 cm above mandibular buccal vestibule. The lesion was of approximately 5 X 3cm in size. The ulcer was having reddish yellow floor. On left buccal mucosa single diffuse ulcer was evident which extended 1cm from the left maxillary buccal vestibule to 2cm occlusally. The ulcer was adjacent to 25, 26 and was of approximately 3 X 2.5cm in size. The ulcer was having reddish yellow floor. On the Floor of the mouth diffuse ulcers were evident bilaterally on the both sides of the lingual frenum. On the right side it was from the lingual aspect of 41 i.e mesial to lingual frenum & was of approximately 2 X 1 cm in size. The ulcer was having yellowish base. On the left side it was approximately 2 cm away from lingual frenum and was of 3 cm mesiodistally and 1.5 cm anteroposteriorly. The ulcers was having reddish floor. Hard Palate showed irregular ulcer of approximately 2.5 X 1 cm in size was evident on the left side of hard palate in the region adjacent to 28 and which was extending to distal side of left maxillary tuberosity region. The ulcer was having yellowish floor and pinkish red borders. (fig 1,2,3,4)

On palpation lesions were tender, floor was not indurated, bleeds on palpation and no discharge was evident.

On the basis of history of a long duration and clinical examination, provisional diagnosis of Pemphigus involving right & left buccal mucosa, hard palate and floor of the mouth was put forth.

INVESTIGATIONS – Random Blood Sugar, Haemoglobin, Bleeding Time, Clotting Time, Total Leucocyte Count & Differential Leucocyte Count, HIV, HCV HBsSg was advised and were within normal limits. Incisional biopsy under LA was done wrt right buccal mucosa

Microscopic features - H & E stained section reveals stratified squamous epithelium exhibiting features of parakeratinization and mild features of mild hyperplasia along with spongiosis. Epithelial – connective tissue interface is flat and shows sub - epithelial split in the superficial lamina propria region of the connective tissue. Underlying connective tissue is chiefly fibrous and showed mixed inflammatory infiltrate along with few blood capillaries.

Based on the history, clinical features and histopathology report, a final diagnosis of Bullous Pemphigoid involving right & left buccal mucosa, palate and floor of the mouth was made. Oint Tenovate 0.5%, local application after meal - TDS,Tab Wysolone 10 mg TDS and Mucopain gel, local application before meal – TDS was prescribed. Patient was recalled after 1 week and reported relief. Lesion was completely healed on palate but still persists with respect to right & left buccal mucosa and floor of the mouth. (fig 5,6,7,8) Patient was advised to continue the medication. Oint Tenovate 0.5%, local application after meal - TDS,Tab Wysolone 10 mg BD and Mucopain gel, local application before meal –TDS. The dosage of systemic and topical corticosteroids was gradually withdrawl.

Discussion

Bullous pemphigoid (BP) is an autoimmune, sub-epidermal blistering disorder that results in large tense bullae, which is triggered by autoantibodies against the dermo epidermal junction of the skin and adjacent mucous membrane.[4] It majorly affects geriatric population in the fifth to seventh decade of life, with average age of onset being 65 years. About 1 in 10,000 people are affected by this condition in the UK each year. In India the incidence of subepidermal auto immune disease is comparatively low. The incidence rate of BP was high in swiss population.[5]

The clinical characteristics of BP may resemble a variety of other blistering conditions. It is important to use clinical, histologic, and immunologic direct immunofluorescence (DIF) findings to differentiate BP from other blistering disorders like dermatitis herpetiform, epidermolysis bullosa acquisita, pemphigus vulgaris, etc.[2]

High-potency topical corticosteroids remain the mainstay of treatment. Typically prescribed agents include fluocinonide, clobetasol propionate and betamethasone dipropionate. The duration of glucocorticoid therapy and the dose should be as low as possible.[6]

Conclusion: Bullous pemphigoid is a rare auto immune disease in India with no permanent cure, but treatment helps to control the disease. Patient quality of life can be improved by appropriate diagnosis and treatment.

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Legends Figure

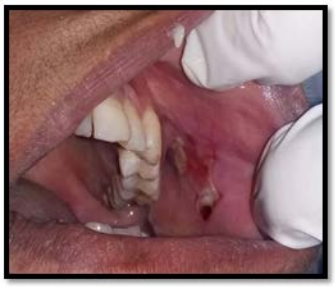
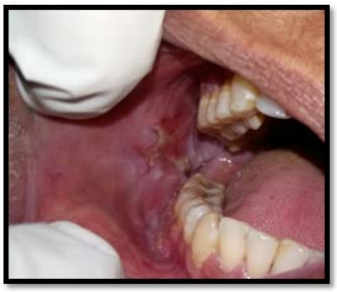


Figure 1



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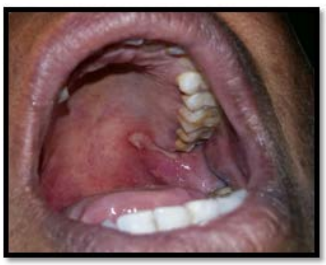

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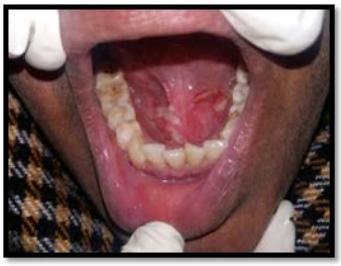

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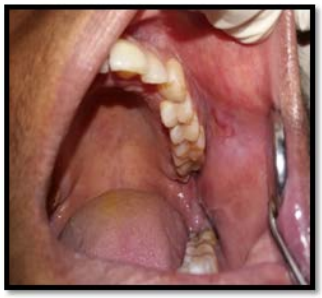

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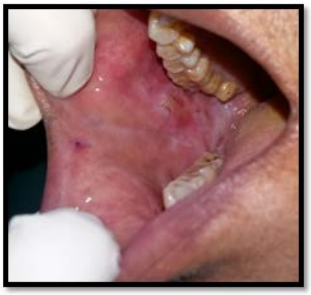

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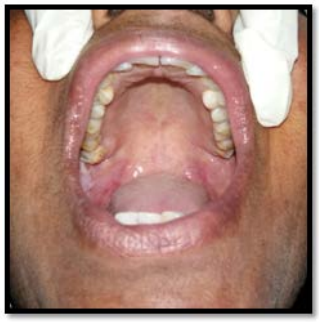

Figure 7

Figure 8