CASE REPORT

Management of a Rare Case of Primary Oral Bullous Pemphigoid

Maria Subash Aaron Muthuraj[1], M.A.J. Mary Kural Ayeni[2].

Abstract

Background: The manifestation of autoimmune bullous disorders as the oral bullous lesion is a common feature. The primary bullous pemphigoid lesion affecting the oral cavity is a very rare entity. Here, we are reporting a case of Primary oral bullous pemphigoid affecting ridge mucosa and palatal mucosa of an 80-year-old female and how we managed it.

Case presentation: The patient complained of blister formation and ulceration in relation to the alveolar ridge mucosa of both arches and palatal mucosa, for the past 5 years and didn't undergo proper test and treatment for that. Lesions are confined intra-orally. Nikolsky's sign was negative. Biopsy showed histopathology features resembling Bullous Pemphigoid and the pathologist recommended an indirect immunofluorescence test which was positive. Diagnosis of Bullous pemphigoid primarily involving oral cavity was made.

Management and prognosis: A topical steroid was prescribed for 1 month and the patient was asked to report after one month. She reported after six months with the resolution of lesions.

Conclusion: Bullous pemphigoid is a rare form of AIBD with predominantly skin lesions. Primary oral bullous pemphigoid is a rare entity. This made us present this case.

Key words: Bullous lesion, Bullous pemphigoid, Pemphigoid, Subepithelial blister, Topical steroids.

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Introduction:

Autoimmune bullous disorders (AIBD) commonly manifest as bullous lesions in the skin and mucous membrane. It is broadly divided into Pemphigus and Pemphigoid.[1,2] Bullous pemphigoid (BP), a form of AIBD affects skin predominantly when compared to the mucous membrane.[3] Sub-epithelial split in BP was due to the formation of IgG antibodies against adhesion complexes 180 and 230-KD. These antibodies bind and induce complement activation followed by leukocyte infiltration into the basement membrane zone and protease secretion, which damages the hemidesmosomes leading to blister formation.[4] Fewer cases of BP in the oral cavity had been reported previously.[5] In our case report, we are reporting a patient with primary oral Bullous pemphigoid in alveolar ridge mucosa and palatal mucosa and how we managed it.

Case history:

An eighty year old female reported to the dental department of a polyclinic complaining of blister formation and ulceration in relation to the alveolar ridge mucosa of both arches and palatal mucosa. (Figs. 1 and 2) Pain aggravated on taking hot and spicy foods. She also reported that the complaint started five years before and increased consistently. She reported to a dental clinic in her native place at that time and the dentist performed total extraction after misdiagnosing it as chronic periodontitis. Even after total extraction and complete denture replacement, symptoms hadn't subsided and it followed an ascending pattern.

[1]Department of Periodontics, Sri Ramakrishna Dental College and Hospital, Coimbatore-641006, Tamilnadu, India.; [2]Consultant, Elan Dental Care, Ganapathy, Coimbatore-641006, Tamilnadu, India.

Corresponding author: Maria Subash Aaron Muthuraj, Department of Periodontics, Sri Ramakrishna Dental College and Hospital, Coimbatore-641006, Tamilnadu, India. Email Id: mariasubashaaronm@srdch.ac.in

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Before two years the patient stopped wearing complete dentures also, due to the same complaint. She was a type II diabetic patient and under medication for the past ten years. Intraoral examination revealed the presence of erosive ulcers in relation to alveolar ridge mucosa of both arches and palatal mucosa on the left side. Nikolsky's sign was checked by applying lateral pressure over the intact ridge mucosa using index finger. Nikolsky's sign was negative as there is no shearing away of epithelium and no formation of intact lesion. A blood investigation was done and was normal. Incision biopsy was planned and performed in the perilesional site. The tissue removed was transported to the laboratory in 10% formalin for histopathological examination. The received specimen was measured at 0.2 x 0.1 cm in size.

Multiple sections from the received biopsy showed a subepithelial blister with hemorrhage. Hyperparakeratinized stratified squamous epithelium was present. Stroma revealed moderately dense inflammatory infiltrate composed of lymphocytes, plasma cells and scattered neutrophils. The basal layer was intact. (Figs. 3 and 4) These features resembled Bullous Pemphigoid and the pathologist recommended an indirect immunofluorescence test. Indirect immunofluorescence test was done using primate oesophagus. Linear basement membrane zone deposit was positive. Salt split skin showed staining only in the roof of bulla. BP 180 and BP 230 were positive, whereas Desmoglein 1 and Desmoglein 3 were negative. A confirmatory diagnosis of Bullous Pemphigoid was made. As the patient had a history of diabetes for the past 10 years and she is of eighty years of age, topical steroids (0.1% triamcinolone) was prescribed for 1 month and the patient was asked to report after one month. But due to personal reasons, she reported after six months with the resolution of lesions in alveolar ridge mucosa and palatal mucosa (Figs. 5A and 5B).

Discussion:

BP is a form of AIBD affecting skin predominantly, with very few numbers of cases reported with intraoral lesions previously.[5] It affects people in the late decades of life.[6] This is the first case in which BP lesions is primarily in the oral cavity with no extraoral manifestations. Many cases with Mucous membrane pemphigoid had been reported with skin lesions,[7,8] whereas only one case of Bullous pemphigoid has been reported with intraoral lesions.[5] This makes our case worthy of documentation.

In our case, clinical finding and histologic findings were similar to Aparna PV et al.[5] An indirect immunofluorescence test was performed and both BP 180 and 230 were positive. Whereas in the previous case report by Aparna PV et al.[5] they had done a direct Immunofluorescence test. Aparna PV et al.[5] treated the patient with systemic steroids as the patient manifested with extraoral lesions also. But in our patient, lesions were exclusively in the oral mucosa and the patient was an eighty-year-old female with Type II diabetic for the past 10 years. Based on the above factors we prescribed a topical steroid (0.1% triamcinolone) for 1 month which was followed by resolving of lesions after 6 months. Differential diagnosis of para neoplastic pemphigus was ruled out as the patient had no relative systemic disorder. Differential diagnosis of mucous membrane pemphigoid and pemphigus were ruled out as Nikolsky's sign was negative. Lichen planus pemphigoides was ruled out due to the absence of lichen planus lesions in association with blisters. Differential diagnosis of Pemphigus lesions was ruled out as histopathological examination showed subepithelial blister and indirect immunofluorescence showed presence of autoantibodies to adhesion complexes 180 and 230-KD. Linear basement membrane zone

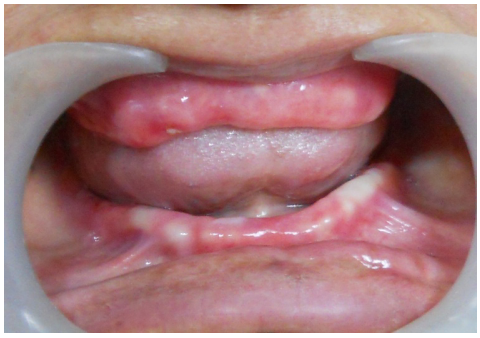

Fig. 1: Ulceration in alveolar mucosa

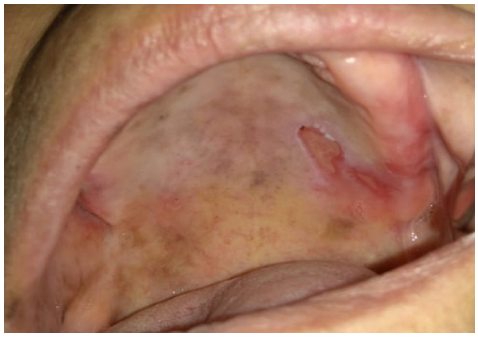

Fig. 2: Ulceration in palatal mucosa

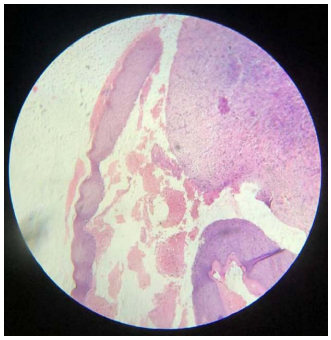

Fig. 3: Subepithelial blister formation and intact basal layer (H and E stain, 10x)

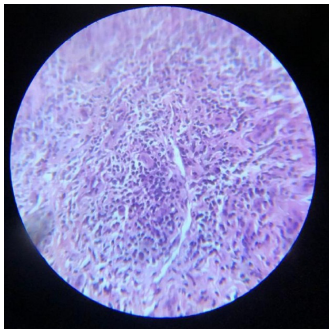

Fig. 4: Moderately dense infiltrate composed of lymphocyte, plasma cells, and scattered neutrophils (H and E stain, 40x)

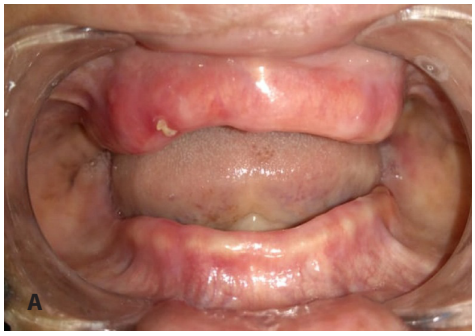

Fig. 5A: Post-operative – at sixth month after application of topical steroids

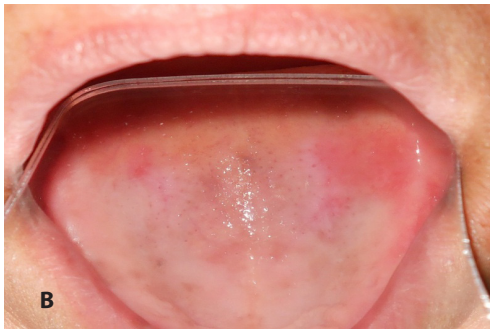

Fig. 5B: Post-operative – at sixth month after application of topical steroids in the palatal mucosa

deposit was positive. Salt split skin showed staining only in the roof of bulla. BP 180 and BP 230 were positive, whereas Desmoglein 1 and Desmoglein 3 were negative.

Conclusion:

Bullous pemphigoid is a rare form of AIBD with predominantly skin lesions. But in our case, the patient reported to our dental department with primary lesions exclusively in alveolar ridge mucosa and palatal mucosa. The rarity of the lesion made us present this case report. As the patient is 80 years old and had a history of diabetes for the past 10 years we opt for topical steroid application rather than for systemic steroids. The patient showed resolved lesions after six months.

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