Cukurova Medical Journal
ÇUKUROVA ÜNİVERSİTESİ TIP FAKÜLTESİ

Cukurova Med J 2021;46(2):853-855

EDİTÖRE MEKTUP / LETTER TO THE EDITOR

Intraoral bullous pemphigoid

İntraoral büllöz pemfigoid

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Cukurova Medical Journal 2021;46(2):853-855

To the Editor,

Bullous pemphigoid is a chronic autoimmune disease where subepidermal blistering is seen. It is a rare disease with an incidence of 10 cases per million population in a year. It is a self- limiting disease and rarely involves the mucous membrane. IgG autoantibodies are produced against the hemidesmosomal glycoproteins - called bullous pemphigoid antigen 1 (BP230) and bullous pemphigoid antigen 2 (BP180)[1]. Bullous pemphigoid rarely involves the oral mucosa as skin is the most commonly affected[2]. We present a rare scenario in this case report where a young female presented with bullous pemphigoid of the oral mucosa.

A 20 year old female patient reported to the department of oral medicine and radiology with the chief complaint of blister in the left upper back tooth region which appeared and ruptured within the last 24 hrs. Patient gave a history of similar blisters in the same as well as other regions of the oral cavity. These blisters rapidly increased in size and ruptured within a day, leaving raw painful ulcers which caused burning sensation and discomfort while eating. These ulcers healed within a week without scarring. The blisters were preceded by pruritis of the skin. Patient had no relevant medical history and family history. On extra oral examination, no lesions were seen on the skin or other mucosal surfaces. On intra oral examination, an area of desquamation was seen in the palatal and distal mucosa with respect to 28 measuring about 0.4 × 0.3 cm. The lesion was flat with irregular borders. On palpation, it was soft in consistency and tender. No bleeding or discharge was present. Nikolsky's sign was negative. No other positive intra oral findings were noted. On further investigation the patient had a picture of the blister prior to rupture which was taken one day ago. (Figure 1) The photograph showed a hemorrhagic bullae adjacent to 28 posteriorly

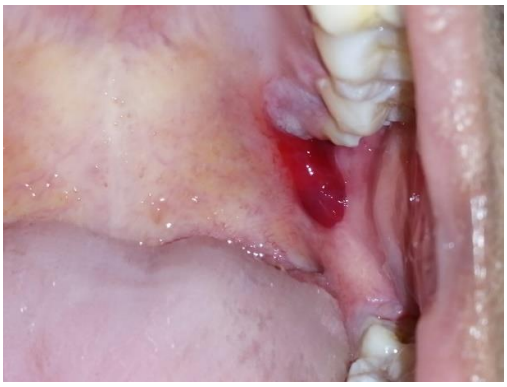

Figure 1. Haemorrhagic bullae seen distal to 28.

The patient was advised to undergo routine blood investigations and referred to a dermatologist for opinion regarding pruritis of the skin. The patient reported the next day with a new lesion in 18 region. Intraoral examination showed an area of desquamation in 18 region similar to that in 28 region. Patient showed a photograph prior to the rupture of the bulla, which showed a hemorrhagic bullae adjacent to 18 distally (Figure 2). Blood investigation showed a normal blood picture. The dermatologist suggested a disease of vesiculobullous origin. Based on the clinical presentation a provisional diagnosis of bullous

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Geliş tarihi/Received: 19.01.2021 Kabul tarihi/Accepted: 18.02.2021 Çevrimiçi yayın/Published online: 20.05.2021

pemphigoid was given. The differential diagnoses that were considered include mucous membrane pemphigoid and bullous lichen planus.

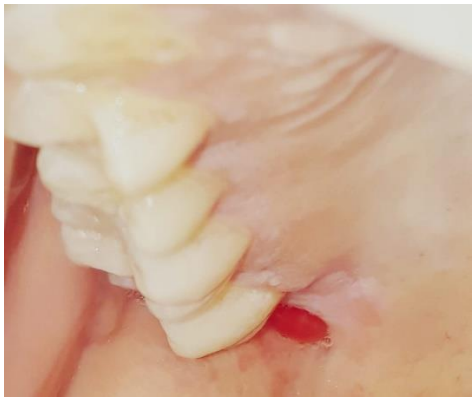

Figure 2. Haemorrhagic bullae seen distal to 18.

Incisional biopsy was performed with the patient's consent, at the perilesional area. Histopathological evaluation of the biopsy tissue revealed, subepithelial split (Figure 3) below the basement membrane with subepithelial bullae formation. The subepithelial space also showed inflammatory cell infiltrate and fibrinous exudate. The underlying connective tissue showed dense chronic inflammatory cells chiefly lymphocytes and plasma cells, with extravasated red blood cells. Based on the clinical and histopathological evaluation a final diagnosis of bullous pemphigoid was given. The patient was prescribed prednisolone 10mg twice daily for 2 weeks. Patient was reviewed after 2 weeks and patient reported no new lesions and a marked reduction in pruritis. Prednisolone was then tapered to once daily for a week and then once daily for alternate days and then stopped. Patient was kept on regular follow-up and educated to report back immediately in case of new lesions in the oral cavity or any other parts of the skin or other mucosal surfaces. Written informed consent was obtained from the patient to share the case in scientific area.

Bullous pemphigoid is usually the disease of the elderly. 80% of the affected individuals are above the age of 60 years.[1] In the presented case the patient is only 20 years old. Bullous pemphigoid shows no gender predilection. It usually begins as cutaneous lesions with rare involvement of the oral mucosa. Only about 10 – 20% of the cases show oral involvement.[3] But our case had only oral involvement with no cutaneous involvement. Oral lesions are usually seen as bullae or vesicles which rapidly rupture to form ulcers or erosions[4]. Our patient also gave a similar history of bullae formation which ruptured within 24 hours to leave painful ulcers. Areas commonly involved include gingiva, buccal mucosa, palate, tongue and floor of the mouth. In our case palate was the most commonly affected site. Sometimes gingiva maybe the only site involved. Gingiva shows generalized edema, inflammation, desquamation with localized areas of vesicle formation. Desquamative gingivitis is the most common oral manifestation of bullous pemphigoid[3]. Our patient did not show any marked changes of the gingiva. They are sometimes seen associated with multiple sclerosis, malignancies or in patients under drug therapy especially diuretics. Our patient had no known associated conditions or previous history of drug intake.

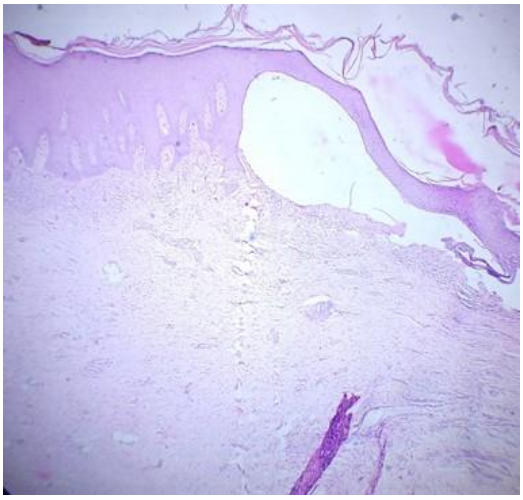

Figure 3. Histological section showing subepithelial split.

Histopathological sections usually show separation of the epithelium from the connective tissue at the basement membrane. Inflammatory infiltrate characteristically rich in eosinophils is seen in the basement membrane zone. Epithelial cells show no acantholysis. The vesicles show fibrinous exudate and inflammatory cells[1]. H&E stained sections of the biopsied perilesional tissue revealed similar features on histopathological examination. Direct immunofluorescence studies show Ig G and C3 bound in a linear band to the basement membrane. Indirect immunofluorescence studies show Ig G antibodies bound to dermal side of salt split skin. ELISA shows circulating autoantibodies against bullous pemphigoid antigens – BP180 and BP 230 – in serum samples of affected individuals. The differential diagnoses to be considered include erosive lichen planus, pemphigus vulgaris and other subepithelial dermatoses.

Localized oral lesions can be treated using high potency topical steroids like clobetasol or betamethasone. Extensive lesions can be treated with systemic corticosteroids in combination with immunosuppressants like azathioprine, cyclophosphamide, mycophenolate or rituximab. Oral prednisolone – 0.3 to 1.25 mg/ kg body weight / day helps control the disease process within one to two weeks.[5] Azathioprine – 0.5 to 2.5 mg / kg body weight/ day is an effective immunosuppressant. Moderate lesions can be treated using immunosuppressants like dapsone,

tetracycline, doxycycline, minocycline in combination with niacinamide. This helps to reduce the usage of systemic corticosteroids. Dexamethasone – 100mg in 500 ml 5% dextrose iv over 2- 3 hours for 3 consecutive days alone or in combination with cyclophosphamide is used in pulse therapy.[4] Our patient responded well to prednisolone and control of the disease was established in two weeks.

Bullous pemphigoid is a rare but potentially life threatening condition. In rare cases where patients present only with oral lesions, dentists need to be keen and perform a complete evaluation and required appropriate investigations. This will aid in arriving at an early diagnosis at the initial stages of the disease which is greatly beneficial. As dentists we should also equip ourselves in managing the disease to achieve control of the disease in its early stages. A multidisciplinary approach involving dentists, dermatologists and ophthalmologists should be taken early on to assess and treat the patients in the initial stages.

Yazar Katkıları: Çalışma konsepti/Tasarımı: SS, RLC, GSB, VA, VS, SG, AB; Veri toplama: SS, RLC, GSB, VA, VS, SG, AB; Veri analizi ve yorumlama: SS, RLC, GSB, VA, VS, SG, AB; Yazı taslağı: SS, RLC, GSB, VA, VS, SG, AB; İçeriğin eleştirel incelenmesi: SS, RLC, GSB, VA, VS, SG, AB; Son onay ve sorumluluk: SS, RLC, GSB, VA, VS, SG, AB; Teknik ve malzeme desteği: SS, RLC, GSB, VA, VS, SG, AB; Süpervizyon: SS, RLC, GSB, VA, VS, SG, AB; Fon sağlama (mevcut ise): yok.
Hakem Değerlendirmesi: Editoryal değerlendirme.
Çıkar Çatışması: Yazarlar çıkar çatışması beyan etmemişlerdir.
Finansal Destek: Yazarlar finansal destek beyan etmemişlerdir.

Author Contributions: Concept/Design : SS, RLC, GSB, VA, VS, SG, AB; Data acquisition: SS, RLC, GSB, VA, VS, SG, AB; Data analysis and interpretation: SS, RLC, GSB, VA, VS, SG, AB; Drafting manuscript: SS, RLC, GSB, VA, VS, SG, AB; Critical revision of manuscript: SS, RLC, GSB, VA, VS, SG, AB; Final approval and accountability: SS, RLC, GSB, VA, VS, SG, AB; Technical or material support: SS, RLC, GSB, VA, VS, SG, AB; Supervision: SS, RLC, GSB, VA, VS, SG, AB; Securing funding (if available): n/a.
Peer-review: Editorial review.
Conflict of Interest: Authors declared no conflict of interest.
Financial Disclosure: Authors declared no financial support

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