Case Report

Cicatricial Pemphigoid: The Rare Bullous Disease: A Report of Two Cases

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ABSTRACT

Diagnosing a bullous lesion in the oral cavity has always been a difficult task for the diagnostician. Here, we present two cases of cicatricial pemphigoid which were treated very successfully by a short course of systemic steroids. Cicatricial pemphigoid is an autoimmune blistering disease that primarily affects the mucous membranes including the oral cavity, oropharynx, conjunctiva, and the genitalia. Autoantibodies are directed against basement membrane zone target antigens. The antibodies involved are mainly IgG and sometimes IgA. The condition presents itself as tense blisters and erosions which heal with scarring and pigmentation. The treatment objective is to suppress extensive blister formation, to promote healing, and to prevent scarring. The above objectives were fortunately met in both our cases.

Keywords: Autoimmunity, Bullous diseases, Cicatricial pemphigoid, Skin lesion, Systemic steroids

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INTRODUCTION

Many of the blistering diseases of the oral cavity are autoimmune in nature. The most commonly encountered autoimmune blistering diseases include pemphigus vulgaris, paraneoplastic pemphigus, bullous pemphigoid, cicatricial pemphigoid, dermatitis herpetiformis, and IgA dermatosis.¹ In 1953, Lever was the first dermatologist from Boston to make a distinction between pemphigus and pemphigoid.²

Cicatricial pemphigoid is a vesiculo-bullous disease of the skin mainly but may also involve the oral cavity. It can be called commonly by other names such as benign mucous membrane pemphigoid (MMP), desquamative gingivitis, oral pemphigoid, and ocular cicatricial pemphigoid.³

Cicatricial pemphigoid is characterized by severe blistering of the oral mucosa and conjunctiva primarily, but it can also involve mucous membrane of the oropharynx, genitalia, etc.³ It is of considerable importance in ophthalmology because of the high incidence of legal blindness seen in patients suffering with this condition due to corneal scarring.² Only about one-quarter of the patients has skin involvement limited to the upper torso, head, and neck regions.⁴

It is more commonly seen in the fifth or sixth decade though incidence in childhood has also been reported. In the oral mucosa, the most common sites involved are the gingiva followed by the soft and hard palate, buccal mucosa, and tongue.³ Females are reportedly 1.5-2 times more commonly affected than males.⁵ No known racial predilection seems to play a part in the pathogenesis though some studies have shown an association of cicatricial pemphigoid with some immunogenetic haplotypes such as HLA-DQB1.⁶ This condition presents itself clinically as tense bullae which rupture leaving behind painful erosions which can get secondarily infected.¹ Occasionally, gingival inflammation is seen without any presence of obvious bacterial plaque, but bone loss and periodontal pockets are found to be absent.³ This erosion may result in the formation of a scar (cicatrix) on healing.⁷ Early recognition of this condition and express management may reduce the resultant systemic complications.⁴ Diagnosis is based on clinical examination, histopathological studies, and confirmed by immunopathological tests.⁷ MMP may be misdiagnosed as linear IgA bullous dermatosis based on the presence
of IgA class antibasement antibodies. Thorough evaluation both clinically and microscopically is of prime importance. Management mainly revolves around medical therapy with the limited role of surgery. Here are two cases showing excellent results with topical and systemic short-term corticosteroid therapy highlighting the importance of using an open minded approach in diagnosing bullous lesions of the oral cavity. Two cases with the same condition present themselves differently.

**CASE REPORT**

**Case 1**

A 48-year-old female patient reported to the Department of Oral Medicine and Radiology with the chief complaint of burning sensation of the gums from past 6 months which initially started as a feeling of dull continuous pain over the gums for which she visited a dentist nearby and was given medications for the same. She experienced relief for a few days, but the pain returned along with the severe burning sensation of the gums. The patient also visited an ENT specialist for the same and was put on steroid ointment: Triamcinolone acetonide (kenacort) and an antacid (pantoprazole). No changes in symptoms were seen. Her medical, dental, and personal histories were non-contributory.

On the general examination, the patient was moderately built and had a normal gait. The extensor surface of her legs showed multiple irregular shaped hypopigmented areas (Figure 1).

On the extraoral examination, the patient had an apparently symmetrical face with no other abnormalities (Figure 2).

On the intraoral examination, the upper and lower attached gingiva showed severe desquamation on the buccal surface (Figure 3). On palpation, these lesions showed bleeding spots.

On the palatal surface, a well-defined oval shaped shallow ulcer of size 1.5 cm × 1 cm with raised borders was seen extending from the attached gingival of 15-17 with the greatest dimension over the 16 region (Figure 4). The ulcer had pseudo-membranous covering with erythematous borders. On palpation, the ulcer was mildly tender.

The rest of the oral cavity did not show any other abnormality. Nikolski’s sign and Asboe-Hansen’s sign were negative. Generalized periodontitis was present.

Based on history and clinical findings, a provisional diagnosis of pemphigus vulgaris was arrived at with differential diagnosis of cicatricial pemphigoid.

Incisional biopsy was done from the gingival region, and the specimen was sent for histopathological examination and immuno-fluorescence. The histopathological report showed tissue lined by stratified-squamous epithelium with acanthosis and hyperkeratosis. Subepithelial clefting...
noted focally with mild inflammatory infiltrate composed of neutrophils within the cleft. Subepithelial tissue showed a moderate amount of inflammatory infiltrate. Direct immuno-fluorescence picture showed IgG and C3 staining in the dermoepidermal junction (in the roof of the cleft). It was negative for IgA, IgM, and C1q.

On the basis of complete clinical examination, history, histopathological report, and direct immuno-fluorescence test, we arrived at the final diagnosis of cicatricial pemphigoid.

The patient was put on systemic prednisolone on the tapered dose for a week along with triamcinolone acetonide ointment for topical application intraorally, clobetasole propionate ointment for the skin lesions and antacids. Systemic prednisolone was started at the dosage of 20 mg 3 times a day for a week followed by 10 mg 3 times a day for a week and 10 mg twice a day for a week. Good results were seen within 2 weeks. The lesions subsided completely within 3 weeks (Figures 5 and 6). The patient is on a 6-month follow-up.

Case 2

A 46-year-old female patient reported to the Department of Oral medicine and Radiology with the chief complaint of dull continuous pain and ulcers on the right cheek area, which started as a reddish lesion around 9 months ago and this progressed to an ulcer 8 months ago for which she consulted a general surgeon. A biopsy was done, and the result declared the lesion as a lichenoid reaction with focal ulceration, and the patient was given multivitamin capsules. However, the lesions did not subside. The patient was on the thyroid medication for hypothyroidism from past 1 year. On extraoral examination, her lower lip appeared to be swollen with desquamation present over the vermilion border with areas of melanosis (Figure 7).

On the intraoral examination, multiple ulcers were seen over the right buccal mucosa extending about 2 cm posterior to the lip commissure until the retromolar pad, along the occlusal plane. The ulcer had yellowish slough over the floor with no surrounding halo. Generalized erythema was seen around the ulcers (Figure 8).
The ventral surface of the tongue shows the presence of bullae of size 1 cm × 1 cm and 1 cm × 1.5 cm on either side of the midline surrounded with erythematous mucosa which was tender on palpation (Figure 9).

Generalized desquamation of the gingiva was seen which was most severe over the 25, 26, and 27 region. Severe calculus deposits were seen over the posterior teeth on the right upper and lower arches. On palpation, all the inspectory findings were confirmed. The lesions were tender on palpation. Based on the history and clinical findings, a provisional diagnosis of pemphigus vulgaris was arrived at due to the absence of any dermatological lesions. A differential diagnosis of cicatricial pemphigoid was also discussed.

The patient was referred to the Department of Periodontia for deep scaling. An incisional biopsy was done over the right buccal mucosa and sent for histopathological examination and direct immuno-fluorescence. The histopathological report showed a keratinized stratified squamous epithelium in association with an inflamed fibrovascular connective tissue. The epithelium exhibits a sub-basilar split. The underlying connective tissue exhibits a diffuse chronic inflammatory cell infiltrate. Epithelium in many foci exhibits koilocytic changes, suggestive of pemphigoid. Immuno-fluorescence report showed deposits of IgG and C3 in the dermoepidermal junction suggestive of pemphigoid.

Based on the clinical findings, histopathological report, and direct immuno-fluorescence, we arrived at a final diagnosis of cicatricial pemphigoid.

The patient was put on systemic prednisolone (20 mg three times a day) for a week along with betamethasone mouthwash. The dosage was reduced to 30 mg a day after a week. Further, the patient was advised to use triamcinolone acetonide ointment for topical usage. The dosage of prednisolone was further reduced to 20 mg a day for a week. The lesions subsided within 3 weeks (Figures 10-12). The patient is on a regular 6-month follow-up.

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Figure 8: Multiple ulceration over the right buccal mucosa

Figure 9: The ventral surface of the tongue

Figure 10: Follow-up view of the right buccal mucosa

Figure 11: Follow-up view of the gingiva
DISCUSSION

Bullous dermatosis, if left untreated can be highly debilitating and possibly fatal. The most common autoimmune blistering diseases include pemphigus vulgaris, paraneoplastic pemphigus, bullous pemphigoid, cicatricial pemphigoid, dermatitis herpetiformis, and linear IgA dermatitis. Pemphigus vulgaris usually starts in the oral cavity followed by blistering of the skin.\(^1\) Cicatricial pemphigoid also involves the oral cavity presenting itself as severe erosive lesions of the mucous membranes with skin involvement in one-third of the patients. In our case, Patient II did not have any skin involvement, whereas Patient I had skin involvement before the lesions in the oral cavity appeared. Usually, the skin over the head, neck, and upper trunk is involved but in our case extensor surface of the legs was involved.

Cicatricial pemphigoid, also called MMP, has a varied clinical presentation. The lesions seen most commonly are in the form of vesicles, erosions, desquamations, and scarring, especially of the extraoral sites which contribute to the disease morbidity.\(^4\) All the above findings were seen in the first case in our report. Immune electron microscopy shows that antigens from patients suffering from MMP localizes in the junction between lamina lucida and lamina densa\(^6\) and, therefore, healing lesions in MMP cause scar formation in the majority of the cases, especially lesions on the skin.

MMP can also involve the conjunctival mucosa resulting in conjunctival erosions with subsequent scarring and progressive cicatization with foreshortening of the fornices.\(^4\) The sites more commonly involved are: Oral cavity (90%), eye (65%), nose, nasopharynx, anogenital, skin (20%-30%), larynx (8%-9%), and oesophagus.\(^8\) Cicatricial pemphigoid is typically seen in the elderly, between 40 and 70 years of age and also a female predominance of 2:1 is usually seen. Both these demographics are consistent with our cases. Desquamative gingivitis is the main presenting feature of MMP. It has been established that MMP is an autoimmune reaction where antibodies are directed against proteins in the basement membrane zone.\(^9\) In the majority of cases of MMP, the antibodies are directed against Type XVII collagen and also against epiligrin laminin. According to literature, about 80-97% of cases show positive results in direct immuno-fluorescence, once the clinical diagnosis is established. Both our cases fall into this category, where immuno-fluorescence results showed positive IgG and C3 deposition.\(^10\)

The management of cicatricial pemphigoid depends on its severity and sites of involvement. It rarely goes into spontaneous remission. Topical corticosteroid in the form of gels/occlusive base may be enough in mild cases of MMP.\(^4\) In cases of extensive lesions, systemic corticosteroids in combination with topical steroids may be required. Oral mini-pulse therapy has been shown to be useful in severe cases. MMP has a tendency to relapse. Hence, it is mandatory to keep a patient with cicatricial pemphigoid under regular follow up.\(^6\) Thorough investigation of suspicious lesions must be carried out and appropriately treated to avoid complications such as scarring.

CONCLUSION

Mouth is the mirror of the body. This saying holds true in cases of bullous diseases where multiple mucosal areas are involved. A good diagnostician has to be aware of the variations in every disease. In the above case reports, we saw a case of cicatricial pemphigoid with dermatological manifestations and one without any skin lesions which could be easily mistaken for pemphigus vulgaris. Thorough histopathological and immune-histochemistry is mandatory for diagnosing autoimmune bullous lesion.

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How to cite this article: Raj PR, Nausheen E, Kunjumon RM, Abraham MA, Nalin AS, George GB. Cicatricial Pemphigoid: The Rare Bullous Disease: A Report of 2 Cases. Int J Adv Health Sci 2016;2(9):11-16.

Source of Support: Nil, Conflict of Interest: None declared.