A rare case of Cicatricial Pemphigoid (Mucus Membrane Pemphigoid)

Case Report

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ABSTRACT

Cicatricial pemphigoid (CP), refers to a rare chronic autoimmune blister disease that predominantly affect mucosal membranes. We report a case of 60-year-old male with the diagnosis of cicatricial pemphigoid (CP). The pathology involved oral mucosa, Pharynx and esophagus. Diagnosis was confirmed by histopathology, and the patient responded well to oral steroids. It is characterized by progressive bullous skin and mucous membrane lesions that tend toward scarring and involution.CP is commonly seen in elderly age group, but it has been reported in children in much lower frequency. It differs from bullous pemphigoid in its prolonged course and significant scarring.The oral mucosa is the most frequently affected area (85 to 90% of patients), followed by the ocular mucosa (66%).The upper airway and digestive tract are also affected in a considerable number of patients. Nasal involvement has been reported to occur in (15 to 23%) of patients, and laryngeal involvement has been seen in . Clinical diagnosis is made on the basis of clinical presentation established by histology,and confirmed by immunpathological studies.Approximately, 50% of patients with oral pemphigoid have been reported progression to involve extra-oral sites, such as eyes, larynx, pharynx, or oesophagus.

Keywords : Cicatricial pemphigoid , autoimmune, odynphagia, ulcer
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INTRODUCTION

Cicatricial Pemphigoid (CP), also known as mucous membrane pemphigoid (MMP) is a rare, chronic, subepithelial, autoimmune, bullous disease which mainly affects the mucous membranes.[1] It is characterized by progressive bullous skin and mucous membrane lesions that tend toward scarring and involution.[2]

CP is commonly seen in elderly age group, but it has been reported in children in much lower frequncy.[3] It differs from bullous pemphigoid in its prolonged course and significant scarring.[4]

The oral mucosa is the most frequently affected area (85 to 90% of patients), followed by the ocular mucosa (66%).[2] The upper airway and digestive tract are also affected in a considerable number of patients.[5] Nasal involvement has been reported to occur in (15 to 23%) of patients, and laryngeal involvement has been seen in (8 to 21%).[6] Clinical diagnosis is made on the basis of clinical presentation established by histology,and confirmed by immunpathological studies.[7,8] Approximately, 50% of patients with oral pemphigoid have been reported progression to involve extra-oral sites, such as eyes, larynx, pharynx, or oesophagus.[8]

CASE REPORT

A 60 year old male presented with history of sore throat, odynphagia and dysphagia since last month. He was also complaining of change of voice and drooling of saliva for same period of time. The patient was referred to the ear, nose and throat (ENT) department for evaluation. General examination revealed dehydration, pallor, and difficulty talking. ENT examination revealed shallow, irregular,and erythematous erosions over vestibule floor, ventral surface of the tongue, soft palate and uvula.

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There were inflammation of hypopharynx and larynx.

Nose and ears were normal. There were no cutaneous, ophthalmic or genital lesions. Endoscopic examination were done, and it revealed extensive ulcerations extended from the hard palate to down to the first 15 cm of the esophagus. For histopathology, biopsy from the esophagus showed chronic active ulcers and acute esophagitis.

Another Biopsy taken from the oral cavity showed cicatricial pemphigoid. After case conformation, the patient was put on systemic steroids and he improved dramatically.

Figure 1.0 : Clinical photograph showing ulcer covered with slough and surrounding erythema present on the hard palate

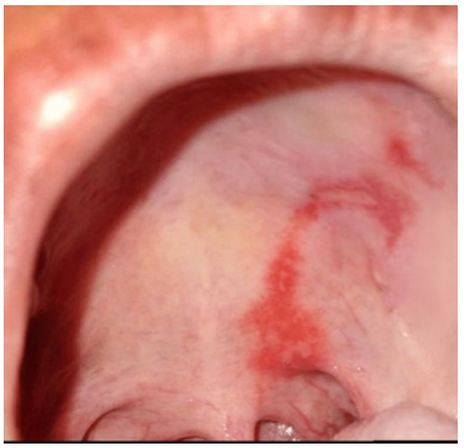
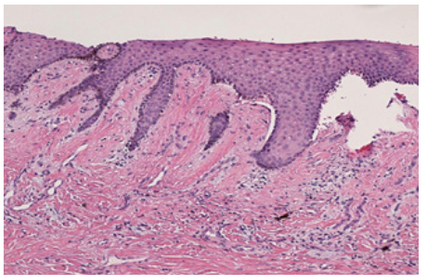


Figure 2.0: Separation of the epithelium from the connective tissue at the basement membrane zone. Numbers of both acute and chronic inflammatory cells are seen in the lesioned areas (hematoxylin-eosin)



DISCUSSION:

Cicatricial pemphigoid (CP) is mainly a disease of elderly. It occurs most frequently during the sixth and seventh decades of life, but it can develop from the third to the ninth decade.[9]

CP may remain localized to the oral cavity, eye or skin or it may be generalized. Early recognition and treatment of the disease can improve prognosis and keep the patient away of surgical interventions.[10]

Pathogenesis of CP is not well understood. However, there is circumstantial evidence to suggest its autoimmune nature.Deposition of immunoglobulins and complement at basement membrane zone (BMZ) in vivo, and presence of circulating anti BMS antibodies have been demonstrated. [12,2]

The oral mucosa is the most frequently affected site (85 to 90%), and the conjunctival mucosa is the second most frequently effected (66%).[2] It also affects other mucosal membranes of the body, like nasal mucosa (15 to 23%), pharynx (20%), larynx (8 to 21%), and esophagus (1 to 8%). Cutaneous lesions are observed in 10 to 30% of patients. [13]

Most patients with CP manifest oral involvement, which can progress to the development of a residual scar in the buccal cavity. Moreover, slowly healed erosions, which can affect the nasopharynx, conjunctiva, larynx, genitalia, and esophagus.2 Nasopharyngeal involvement is characterized by rupture of the vesicles in the nasal mucosa, rhinorrhea, crust formation, and epistaxis, which can eventually culminate in cicatricial stenosis. When pemphigoid affects the pharynx, patients tend to experience sore throat, dysphagia, and odynophagia. Lesions typically appear as multiple shallow ulcers. Pharyngeal involvement can be presented with dysphagia and odynophagia,and it can culminate in glottic stenosis. In addition to laryngeal lesions which usually appear as bullae and ulcers in the epiglottis.[5,6]

Lesions in the respiratory tract can cause dysphonia, dyspnea, and laryngeal stenosis. Esophageal stenosis can be observed in patients with severe form the disease.[6]

CP can lead to blindness If ocular involvement is present14, and it can be fatal if it progresses into stenosis of esophagus or trachea.[13]

Diagnosis of CP is based on its histological characteristics, which are seen as subepidermal blister that contains a mixed infiltrate of numerous eosinophils, mononuclear cells, and some neutrophils. In addition to, direct immunohistologic examination.

Biopsy specimens will detect complement and antibasal-membrane autoantibodies.[2]

Direct immunofluorescence of the epithelium will demonstrate a linear and homogeneous pattern of IgG and C3 deposition in the basal membrane.[2,7]

A presumptive diagnosis of CP can often be made on the basis of history and clinical findings alone. However, it is not unusual for an extended period of time to elapse before the correct diagnosis is reached..[15]

Treatment options of CP include mainly corticosteroids and immunosuppressive therapy.

Topical corticosteroid is indicated as the initial treatment for mild cases of CP. However, most patients require systemic corticosteroids. Immunosuppressants such as azathioprine, methotrexate, and cyclophosphamide have been used with success. Dapsone can be used for recurrences.[2,10,13]

In selected cases, airway intervention in the form of tracheotomy or laser debulking may be necessary to secure the airway.[5] Surgical treatment is indicated for the removal of scars and stenosis.[6]

CONCLUSION:

The diagnosis of cicatricial pemphigoid (CP) may be difficult and requires high index of suspicion, in addition to thorough understanding of the disease process,and dermatology consultation. Immunofluorescent findings in the absence of cutaneous involvement, support the diagnosis of CP and rule out bullous pemphigoid. corticosteroids and immunosuppression are the most effective treatment options. This case illustrate the importance of otorhinolaryngologist awareness of this disease as the first manifestations of CP in this patient were head and neck signs and symptoms.

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