e - ISSN - 2348 - 2168
Print ISSN - 2348 - 215X

Acta Biomedica Scientia

Journal homepage: www.mcmed.us/journal/abs

ANGINA BULLOSA HEMORRHAGICA; A RARE ENTITY: AN OVERVIEW

Kaushal Luthra[1], Yehoshuva Reddy[1], Richa Wadhawan*[1] and Gaurav Solanki[2]

[1]Institute of Dental Education & Advance Studies, Gwalior, Madhya Pradesh, India.
[2]Jodhpur Dental College General Hospital, Jodhpur, Rajasthan, India.

Article Info

Received 29/07/2014
Revised 16/08/2014
Accepted 19/08/2014

Keywords :-
Angina Bullosa Haemorrhagica, Erosions, Soft Palate, Blood Blisters, Steroid Therapy etc.

ABSTRACT

Angina bullosa haemorrhagica is a rare blood filled vesiculobullous lesion of the oropharynx with the most common site affected being the soft palate. Many etiological factors have been reported including trauma, hypertension, hyperglycemia and even iatrogenic causes. Management is based on the dimension of the lesion. Usually smaller lesions are self-limiting, but larger lesions require surgical intervention. The haemorrhagic bullae spontaneously burst after a short time resulting in ragged, often painless, superficial erosions that heal spontaneously within a week without scarring. The awareness about the clinical presentation of this lession in the field of dentistry is very much necessary as it mimics other lesions leading to misdiagnosis. This paper highlights the diagnostic criteria, essentially based on clinical data, possible treatment approaches of the disorder.

INTRODUCTION

Angina bullosa hemorrhagica is a benign condition affecting mucosa of oropharynx, characterized by sudden appearance of blood filled submucosal blisters of unknown etiology. It was first described in 1933 as 'traumatic oral hemophlyctenosis' then the term 'angina bullosa hemorrhaegica (ABH)' was coined by Badham for the same condition in 1967 and later it was renamed as 'recurrent oral hemophlyctenosis'. It is also less often named as localized oral purpura or stomatopompholyx haemorrhagica [1]. These blisters present a color ranging from dark red to purple and may cause some discomfort. It may occur either as solitary or multiple lesions. These lesions, however, are frequently asymptomatic and they are only observed when their content is spilled over the oral cavity. The most common site affected being the soft palate, but these lesions can also occur in the anterior pillar of the fauces, epiglottis, arytenoids, pharyngeal wall and esophagus.

Corresponding Author

Richa Wadhawan
Email: - richawadhawan@gmail.com

Both middle-aged and elderly individuals are more affected. There is no strong sex predilection [2].

ABH is a benign phenomenon that is characterized by the sudden appearance of a blood blister on the oral mucosa in the absence of an identifiable cause or systemic disorder. Although the pathogenesis is still unclear, ABH seems to be a multifactorial phenomenon: dental or functional trauma seems to be the major provoking factor. Causes that have been mentioned in the literature are related to the minor trauma of hot foods, restorative dentistry, periodontal therapy, endoscopic trauma, dental injections of anesthetics, steroid inhalers, chlorhexidine gluconate mouth rinse & sneezing [3, 4]. Some authors suggested mild trauma as the causative agent in ABH as there is breach in the epithelial-connective-tissue junction, causing bleeding of superficial capillaries and resulting in the formation of a sub epithelial hemorrhagic bullae [5, 6]. Garlic et al [7] in 1988 reported that excessive mechanical forces associated with mastication or minor thermal injuries are possible causes of ABH. Few other researches mentioned that the chronic use of inhaled steroids is known to possibly, causing atrophy of

the mucous epithelium. The continuous use of these medicines may alter the synthesis of collagen and reduce its total content in the mucosa. In addition, tissue elasticity may decrease with the maturation of these fibers. As a result these sequential changes may evoke weakness and breakdown of capillaries, resulting in the formation of ABH even in response to minor traumatic events [8, 9]. Similar morphologic alterations can also be observed in middle aged adults which is the cause of senile atrophy. This fact matches well with incidence of ABH more widely in older individuals [10]. Diabetes mellitus may also be a contributing factor in developing ABH. Hereditary predisposition may be associated in some instances [11].

Appearance

Lesion appears as blood filled blister which is painless, raised, round, dark red to purple in color with ecchymotic halo and measures around 1-3 cm in diameter.(Fig1) The blisters last only short span of time and then spontaneously rupture, leaving a shallow ulcer that heals without scarring, discomfort or pain [12].

Site

Literature review has shown soft palate to be the most common site for ABH. This is so because it is covered by a thin friable squamous epithelium of the non-keratinized type. Considering the fragility of soft palate mucosa, it is easy to speculate that submucosal hemorrhage may be elicited even by subclinical trauma. It is also noteworthy that mastication significantly increases the blood flow rate in the soft palate via parasympathetic reflux vasodilatation. Collectively, the soft palate is easily injured during mastication of hard and crispy food and is therefore prone to ABH. Occasional lesions have been reported in the buccal mucosa and tongue. Similar lesions in other mucous membranes or the skin have not been reported [13, 14]. The diagnosis of ABH essentially is clinical; however, the cases in which a biopsy is done, the microscopic examination reveals a subepithelial bulla filled with blood and an underlying mild and nonspecific mononuclear inflammatory cell infiltrate that generally is limited to the region of the lamina propria. Occasionally, neutrophils may be seen [15]. Direct immunofluorescence staining for IgA, IgG, IgM and fibrin is negative and can demonstrate equivocal staining along the basement membrane zone for complement component. Biopsy and immunofluorescence studies may be useful to exclude other blistering diseases [16]. Stephenson et al [17]. Reviewed the laboratory features of the condition and concluded that, despite the pres-ence of a subepithelial split histologically, there was no evidence of autoimmune mediated damage as with mucous mem-brane pemphigoid. Similarities between angina bullosa haemorrhagica and the acquired form of non-dystrophic epidermo-lysis bullosa have been suggested. The assessment of hematologic and coagulation disorders should be performed so as to exclude the possibility of blood dyscrasia.

Differential Diagnosis

The lesions of ABH can be easily confused with other mucosal or systemic diseases. It is important that the presentation of this benign disorder is distinguished from other more serious disorders with similar presenting features. Differential diagnosis must include pemphigus, bullous pemphigoid, bullous lichen planus, dermatitis herpetiformis, epidermolysis bullosa, oral amyloidosis, linear IgA disease and thrombocytopenia. Patients with bleeding disorders such as thrombocytopenia, Von Willebrand's disease & leukaemia can present with intraoral blood filled lesions. In Rendu- Osler-Weber disease, the angiomatiod lesions mimic bullae, but they are not episodic and other clinical & genetic features may exist. Clinical evidences suggested that the typical presentation, particularly the constant presence of blood as the blister fluid which is usually not found in other cases, itself is sufficient to rule out above mentioned disorders and arrive to a clinical diagnosis of ABH. Biopsy and immunofluorescence further may aid in distinguishing ABH from other vesiculobullous lesions. However isolated nature, rapid healing, rare recurrence of ABH blisters are sufficient findings to rule out vesiculobullous lesions [18].

Figure 1. Image of lesion on soft palate taken at time of onset

Courtesy Anthony Bertram, NZMA J 2010; 123: 1314

Management

Surgical drainage is not necessary in small, localized lesions that do not functionally affect the airways, as these lesions are likely to rupture within some hours. Conversely, it is indicated whenever large blisters are present in the palate, causing a feeling of suffocation. Pahl et al [19] in 2004 have reported a case of ABH in a 56-year old female patient who developed a rapidly growing blood blister on the soft palate that caused oropharyngeal airway obstruction. Because of the large lesion size, nasotracheal intubation was not feasible and tracheostomy was required to allow subsequent ABH drainage. Management usually aims to relieve discomfort caused by blister disruption and to improve the healing of ulceration. Use of benzydamine hydrochloride provides symptomatic relief. Treatment may include the use of anti-inflammatory and antibiotic agents, as well as antisepsis with mouthwash containing 0.25% or 0.12% chlorhexidine digluconate, to help relief painful symptoms and avoid

secondary infections. Because secondary infection of areas of ulceration are not uncommon, it is of paramount importancethat the patient be provided with appropriate information and adequately followed. In patients who use steroid-based inhalers regularly, water gargling following medicine application can be an effective way to prevent ABH. Identification of the etiological factor and its elimination may prove beneficial to the affected individual [20-22].

CONCLUSION

It is possible to conclude that although ABH has been traditionally described as a rare condition but it is utmost important for the clinician to overcome misdiagnosis so as to provide better treatment to the patient. In the secondary stage they may be misinterpreted as a primary ulcerative disease of the oral mucosa. All mucosal bullae and ulcers located in the soft palate area require exact clinical examination of the patient to establish a univocal diagnosis. Thus, oral diagnosticians play a vital role in diagnosing such lesions. Detailed anamnestic data gathering represents an inseparable part of the examination which in turn helps to avoid abundant diagnostic and therapeutic procedures, which may unnecessarily strain the patients.

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