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CASE REPORT

Angina bullosa hemorrhagica: a rare or undiagnosed condition?

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Abstract

Angina Bullosa Hemorrhagica (ABH) is a recurrent disorder characterized by the appearance of a blood-filled blister in the oral mucosa, mainly in the soft palate and tongue of middle-aged or older adults. The purpose of the present study is to report an additional case of ABH in conjunction with a comprehensive literature review describing this entity's clinical and therapeutic characteristics. A 68-year-old Caucasian woman with no relevant medical history sought an oral medicine service presenting a blood blister on the lateral surface of the tongue lasting two days. No possible triggering factors were identified. Based on clinical features and exclusion of hematological disorders, the diagnosis was ABH. The treatment was an incision of the blister and a prescription of analgesic drugs. At 15 days of follow-up, the oral mucosa was normal. ABH can be confused with autoimmune and hematological diseases. Therefore, clinicians must recognize ABH to avoid misdiagnosis. The clinical management of ABH is often focused only on the relief of symptoms.

Keywords: Angina bullosa hemorrhagica; diagnostic; oral mucosa blisters.

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Introduction

ABH is a benign disorder characterized by the acute onset of a blood-filled blister in the oropharyngeal and oral mucosa[1,2]. Although it is often described as a rare condition, some authors suggest that ABH is probably underdiagnosed due to the clinicians' lack of knowledge and its self-limited nature and spontaneously favorable evolution[2]. Thus, it is difficult to determine the true incidence and prevalence of this condition.

Although several case reports have been published in the English literature[1-16], the etiology of ABH remains poorly understood, and lesions are often underdiagnosed[1-4]. In addition, the differential clinical diagnosis of ABH can be difficult because it includes several vesiculobullous lesions and hematological diseases[2-4]. After the blister rupture, differential diagnosis becomes broader and challenging and includes several conditions that cause post-bullous erosions[2]. Therefore, clinicians must recognize ABH to avoid misdiagnosis with more serious pathologies.

Herein, we describe a case of ABH involving the tongue of a 68-year-old female. Additionally, we provide a comprehensive literature review describing

the clinical and therapeutic characteristics of ABH and discuss the proposed diagnostic criteria for this condition.

Case report

A 68-year-old Caucasian female with no relevant medical history (no known allergies and no medication use) was referred to an oral medicine service, with a chief complaint of a painless blood blister in the tongue with a duration of 2 days. The past medical history was not contributory. The intraoral examination revealed a blood blister on the lateral surface of the tongue, measuring approximately 1.5 cm in diameter (Figure 1A). The lesion had a flat consistency on palpation and was asymptomatic. No possible triggering factors were identified. Also, the patient reported that a similar episode occurred six months earlier during feeding, and the blood blister ruptured hours later and healed normally within a few days. Platelet counts and coagulation tests were requested to rule out hematological disorders, and the results were normal. Based on clinical features and exclusion of hematological disorders, the diagnosis was ABH. The treatment was an incision of the blister and a prescription of analgesic drugs. At 15 days of follow-up, the oral mucosa was normal (Figure 1B).

Discussion

ABH is a benign condition of uncertain etiology, characterized by the acute onset of a painless blood-filled blister in the oral or oropharyngeal mucosa, which is not related to vesiculobullous diseases, blood dyscrasia, or autoimmune diseases[2,4,15]. ABH tends to occur in the soft palate and tongue's lateral border of female adults between the fifth and the seventh decades of life[2], similar to the present case.

The etiology and pathogenesis of ABH remain unclear[1-5]. However, a previous history of trauma associated with hard, hot, or spicy food intake seems to be the most relevant etiological agent, and it has been the main finding reported in most cases (> 80%)[2], similar to the present case. A possible association

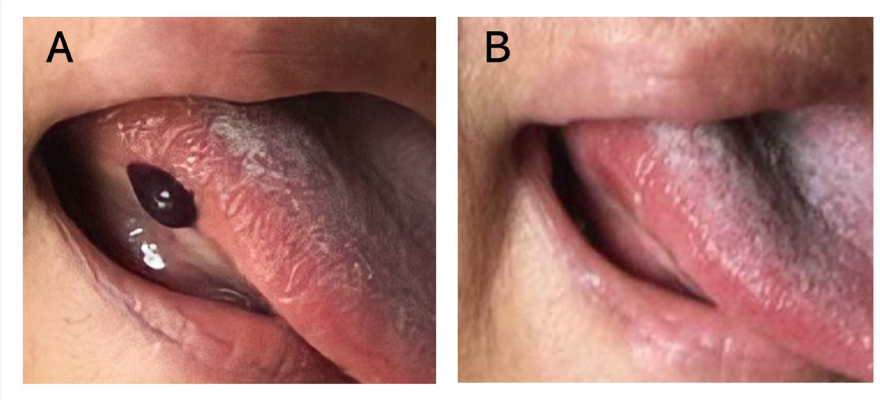

Figure 1. Clinical aspects of angina bullosa hemorrhagica. **A –** Note the blood-filled blisters in the border of the tongue and **B** – clinical aspect 15 days after the incision and drainage of the blood content.

of ABH with diabetes mellitus, hyperglycemia, a family history of diabetes, and hypertension has also been suggested[2-4]. However, these associations appeared to be speculative. Some authors also report an association between ABH and long-term use of inhaled corticosteroids[2-4,9,10]. Chronic use of these drugs may impair collagen formation and cause atrophy of the epithelial tissue, which can cause hemorrhagic lesions after trauma, predisposing patients to these conditions[2,4].

The differential diagnosis of ABH is broad and includes mainly vesiculobullous lesions, such as pemphigus vulgaris, bullous pemphigoid, mucous membrane pemphigoid, linear IgA dermatosis, epidermolysis bullosa acquisita, amyloidosis, dermatitis herpetiformis, and oral bullous lichen planus and hematological diseases, such as thrombocytopenia, von Willebrand's disease, and leukemia[2-4].

According to the criteria recently proposed by Ordioni et al.[2], the diagnosis of ABH is given if the case meets 6 out of 9 of the proposed criteria, with criteria I and II as required (Table 1). These criteria are essentially clinical. In general, a clinically noticeable hemorrhagic blister or an erosion with a history of oral bleeding due to the rupture of a previous blister exclusively located in oral mucosa is essential for the diagnosis. In addition, the presence of a triggering event (chewing), isolated or recurrent lesions on the soft palate with favorable evolution within a few days without leaving scars also favors the diagnosis of ABH[2].

On the other hand, some authors advocate a comprehensive review of the patient's systemic condition, especially in patients with suspected hemostasis abnormalities[2,12]. In these cases, tests such as platelet counts and coagulation tests should be performed[2,4,12]. In order to exclude these disorders, we requested a complete blood count and coagulation profile, and the results were normal, excluding hematological disorders. Biopsy and direct immunofluorescence have also been suggested[2]. However, when a biopsy is performed, a microscopic evaluation of ABH reveals only nonspecific ulceration and chronic inflammatory cell infiltrate in the lamina propria[2,15]. Therefore, clinicopathological correlation is essential for correct diagnosis.

Table 1. Diagnostic criteria for the diagnosis of ABH proposed by Ordioni et al.[2].

I*	Clinically notable hemorrhagic bulla or erosion with a history of bleeding of the oral mucosa
II*	Exclusively oral or oropharyngeal localization
III	Palate localization
IV	Triggering event or food promoting factor (food intake)
V*	Recurrent lesions
VI*	Favorable evolution without a scar within few days
VII*	A painless lesion, tingling, or burning sensation
VIII*	Normal platelet count and coagulation profile
IX	Negative direct immunofluorescence

For ABH diagnosis using these criteria, the case must meet a minimum of 6 out of 9 defined criteria, including criteria I and II; *Criteria met by the patient of the present case.

The presence of hemostatic disorders, antithrombotic treatment, or positive direct immunofluorescence may exclude the diagnosis of ABH[2]. Our case fulfilled 6 out of 9 of the proposed diagnostic criteria for ABH (Table 1).

The clinical management of ABH is often focused only on the relief of symptoms, and the patient should be reassured[2-4]. The prognosis is good. Control of the symptoms with analgesic drugs and the use of topical antiseptics, such as chlorhexidine in concentrations between 0.12% and 0.2%, has been described[2,4]. Large intact lesions, especially on the soft palate, should be incised and drained to avoid a possible obstruction of the upper aerodigestive tract[2,13]. In addition, some authors have suggested combining ascorbic acid and citroflavonoids as a strategy to prevent recurrences[14,17].

In summary, ABH is a poorly understood disorder, and its etiology remains uncertain. Although ABH has a favorable evolution in a few days, it can share some clinical and histological characteristics with more serious diseases, making diagnosis difficult. Therefore, a careful clinical examination is essential to ensure a correct diagnosis.

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Ethical matters

The patient provided an informed consent declaration to permit the use of images and medical information. This case was conducted as part of research previously approved by the Ethical Committee of the School of Dentistry of Piracicaba (CAAE 45415821.7.0000.5418).