Angina Bullosa Hemorrhagica: A rare and poorly understood entity

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ABSTRACT

Angina bullosa hemorrhagica (ABH) is a rare benign condition characterized by the acute formation of subepithelial blood-filled bullae in the oral or oropharyngeal mucosa, in the absence of dermatological, hematological or systemic disorders (1,5). First described by Badham in 1967, this disorder is generally seen in adults over 30 years of age, with a peak incidence in the 5th decade of life and without a clear gender predilection (1). ABH is characterized clinically by the sudden onset of one or more red-blue blood-filled 1.2 to 2.5 cm subepithelial bullae, mainly on the soft palate, lateral borders of the tongue and buccal mucosa (4-6). While the etiology is unclear, local trauma, long term use of corticosteroids, and diabetes mellitus may precipitate the formation of ABH (5). Lesions may be asymptomatic or may present as a burning or tingling sensation, with occasional reports of discomfort (10). The time of evolution of the bullae is variable but generally short as they rupture leaving a superficial ulcer that heals within one or two weeks without scarring (1,5,7,8). Recurrent lesions are frequent, appearing in the same or in another location (2,6). Our poster documents a detailed evolution of a recurrent case of ABH from acute onset to resolution. At present, no such cases exist in the literature. In addition, we present possible etiologies as well as the formation of a differential diagnosis.

CURRENT LITERATURE / HISTOLOGY

Little is known about Angina Bullosa Hemorrhagica in the current literature, and most publications are case reports. Every patient in the published cases, dating back to 1985, reported no hematologic or coagulative abnormalities, and laboratory studies were performed in most instances to confirm. Most patients were able to figure out their trigger with some being as minor as a sneeze. These lesions were also isolated to the oral cavity mostly on non keratinized moveable mucosa (2,5,6).

Non-specific findings include oral mucosa lining epithelium showing areas of detached connective tissue and hemorrhage as seen below in section B..

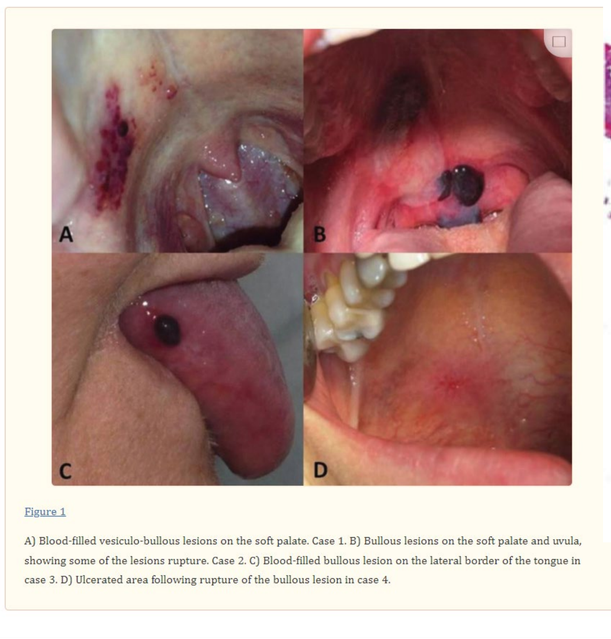


Figure 1

A) Blood-filled vesiculo-bullous lesions on the soft palate. Case 1. B) Bullous lesions on the soft palate and uvula, showing some of the lesions rupture. Case 2. C) Blood-filled bullous lesion on the lateral border of the tongue in case 3. D) Ulcerated area following rupture of the bullous lesion in case 4.

DIAGNOSTIC CRITERIA

There is a diagnostic criteria created by Ordioni *et al*. (2019) to qualify lesions as ABH (7). Cases must fulfill at **least six** of the nine diagnostic criteria. For patients experiencing these lesions, there is a need to identify the trigger to prevent the recurrence. Further studies are needed to understand the pathogenesis of how these blood filled bullae occur.

I	Clinically notable haemorrhagic 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	Favourable evolution without a scar within few days
VII	Painless lesion, tingling or burning sensation
VIII	Normal platelet count and coagulation profile
IX	Negative direct immunofluorescence

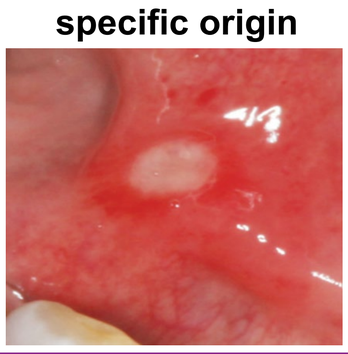
For a positive diagnosis of ABH using these criteria, the case should meet a minimum of 6 out of 9 defined criteria, with criteria I and II as required (6).

DIFFERENTIAL DIAGNOSIS

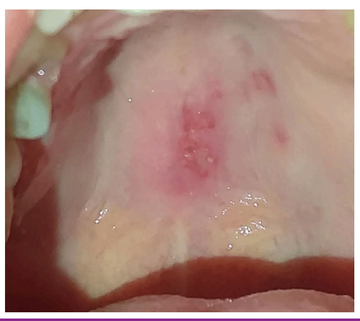
Since Angina Bullosa Hemorrhagica ruptures soon after formation, there is a greater likelihood that oral healthcare providers will encounter patients with the remaining superficial ulcer than the bulla itself. Thus, it is important to be able recognize the later presentations of this lesion especially since ABH often recurs. We have created a differential diagnosis list for the post rupture stages of ABH as a tool.

Diff Dx: recurrent aphthous stomatitis, recurrent HSV, ulcer NOS, muco-cutaneous bullous lesions (pemphigus vulgaris, mucous membrane pemphigoid pemphigoid, bullous lichen planus, etc.), syphilitic chancre, coagulopathies.

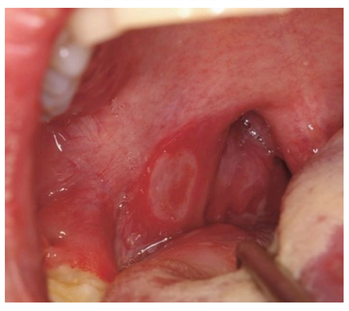
Ulcer of non-specific origin



Recurrent HSV



Recurrent Aphthous Stomatitis



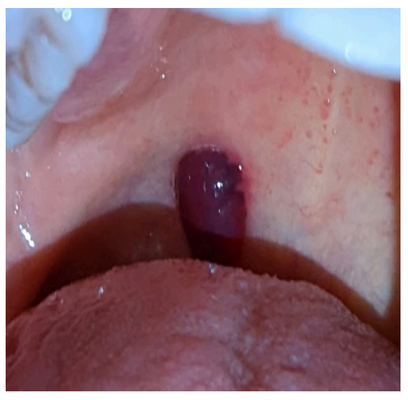
PATIENT PRESENTATION

The patient is a 58 year old female with no significant medical history and no vascular abnormalities. The patient reports that she enjoys drinking very hot beverages, and it is suspected that the hot beverages may be a trigger for this patient.

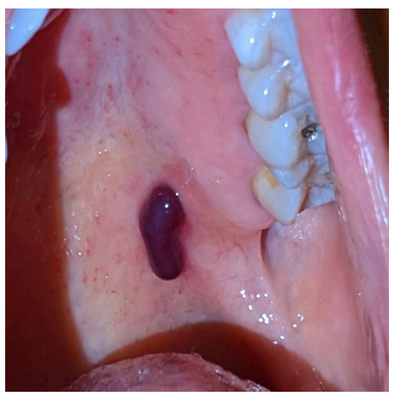
The patient's first lesion was discovered on April 28th 2022. It formed on the right soft palate, measured 2cm x 1cm, with a well defined rounded rectangular shape. It was a blood filled exophytic bulla, and the patient reported that it was asymptomatic. The bulla ruptured and healed without scar formation within 10 days.

The second lesion was followed closely and initially discovered on October 23rd 2022 at 8AM. It recurred on the left side near the junction of the hard and soft palate and measured 1.5cm x 1cm in a slightly triangular shape. Evidence of additional soft tissue trauma can be seen anteriorly on the hard palate. Again, the patient reported that it was asymptomatic. The lesion showed signs of rupture one hour later with a slightly decreased fullness and hazy borders. Twelve hours later, the bulla fully drained and left a superficial ulcerative lesion. Three days later, there were signs of normal wound healing and the ulcer completely healed within 10 days.

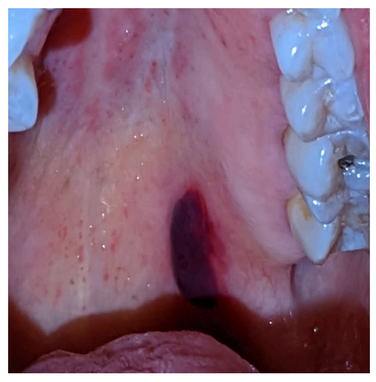
The third lesion was discovered on January 21st 2022 at the junction of the hard and soft palate on the right side. It was smaller than the first two lesions and measured 0.7cm x 0.5cm. This lesion also ruptured soon after discovery, was asymptomatic, and healed without scarring. This lesion was also associated with the consumption of a very hot beverage.



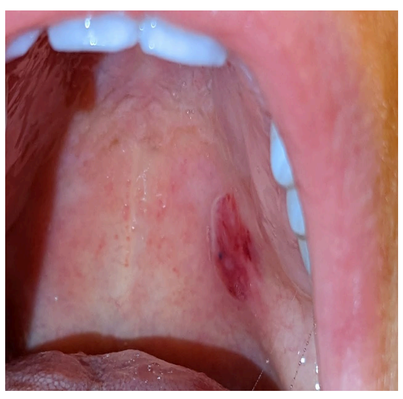
First Lesion: 4/28/2022



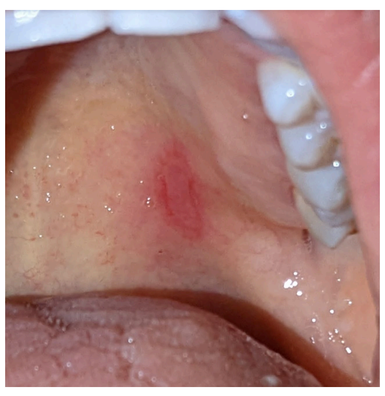
Second Lesion: 10/23/2022 8PM



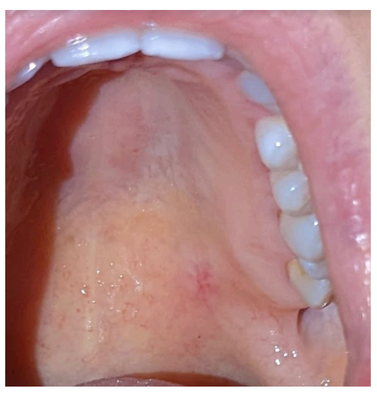
Second Lesion: 10/23/2022 9PM - 1 hour later



Second Lesion: 10/24/2022 8AM- 12 hours after



Second Lesion: 10/26/2022 9AM- 3 days after



Second Lesion: 11/2/2022 9AM- 10 days after

CONCLUSION

Angina bullosa hemorrhagica (ABH) is a rare benign condition characterized by the acute formation of subepithelial blood-filled bullae in the oral or oropharyngeal mucosa, in the absence of dermatological, hematological or systemic disorders (1,5). Lesions are asymptomatic and heal without scarring.

Little is known or found in the literature about Angina Bullosa Hemorrhagica, and most publications offer scant case reports or retrospective descriptive cross-sectional studies. Our poster documents the detailed evolution of a recurrent case of ABH from acute onset to spontaneous resolution in a 58-year old woman. We have presented possible etiologies as well as the formation of a differential diagnosis to aid in the diagnosis and care of the patient who presents with an ABH lesion.

REFERENCES

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