CASE REPORT

Angina bullosa haemorrhagica

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SUMMARY

A woman in her early 40s presented with a painless ulceration on the right side of the posterior palate, she had an earlier history of similar lesions after the rupturing of blood filled blisters in the oral cavity. On examination, a diffuse erythematous area and ulcers covered with necrotic slough were noticed on the right and left side of the posterior palate and on the right buccal mucosa. On follow-up visit, a large blood filled blister was noticed in the buccal vestibule. We advised routine haematological investigations to rule out any bleeding disorders and direct immunofluorescence of the affected tissue and perilesional areas to rule out autoimmune blistering conditions. Haematological investigations revealed no abnormalities and immunofluorescence testing was negative. A diagnosis of angina bullosa haemorrhagica was made by excluding all other conditions. Symptomatic treatment was given, patient was educated about the condition and reassured.

BACKGROUND

Formation of blood filled blisters in the oral mucosa is not an uncommon occurrence and is generally attributed to trauma. Such blisters resolve on their own and are of no major consequence. However, acute and sometimes painful onset of oral blood blisters not attributable to blood dyscrasias, vesiculobullous disorders, systemic diseases or other known causes is not very common and is termed 'Angina bullosa hemorrhagica' (ABH). The pathogenesis of ABH is not clear. Exaggerated host susceptibility to dental or functional trauma may be a significant element in causation of this condition. The diagnosis of this condition is based on its typical presentation, particularly the presence of blood as the blister fluid and exclusion of other conditions.

CASE PRESENTATION

A woman in her early 40s presented with painless ulceration on the right side of the posterior palate since 1 week. She previously had similar lesions in other parts of her mouth which developed after rupturing of blood filled blisters but they had heal within a week. She had no lesions in any other parts of her body. Detailed medical history was elicited from the patient but it was not significant for any systemic disorder or drug intake. Oral examination revealed a diffuse erythematous area and ulcers covered with necrotic slough on the right and left side of the posterior palate and on the right buccal mucosa (figure 1). A provisional diagnosis of pemphigus vulgaris was given as it is the most common blistering disease affecting the oral cavity in middle aged to elderly patients, which

presents with repeated fluid filled vesicles or bullae that rupture and leave an ulcerated area.

Symptomatic treatment with soothing mouthwash was advised and the patient was recalled after a week for further investigations. On follow-up, the palatal lesion had healed without scarring but a blood filled blister was noticed in the buccal vestibule with respect to the left lower posterior teeth (figure 2) and on the gingiva corresponding to the upper right canine.

INVESTIGATIONS

The patient was advised to undergo routine haematological investigations on the first visit (to rule out any bleeding disorders), which were in normal limits. On subsequent visit, direct immunofluorescence (DIF) of the affected tissue and perilesional areas, was advised to rule out any autoimmune blistering conditions, which was negative.

DIFFERENTIAL DIAGNOSIS

On the basis of the patient's history, the clinical examination and investigations, we ruled out most of the disorders in which we can notice blood filled blisters. Patients with bleeding disorders like thrombocytopenia and Von Willebrand's disease can present with intraoral blood-filled lesions but coagulation and haemostatic function tests will show some amount of fluctuations in such cases, all the haematological tests were within the normal range in our patient. Apart from this, subepithelial bullous diseases were also considered like benign mucous membrane pemphigoid or bullous pemphigoid but direct immunofluorescence in such cases show linear deposits of IgG and/or C3. Also, patients clinically present with other signs such as desquamative gingivitis and nasal or conjunctival mucosal

Figure 1 Diffuse erythematous area and ulcers covered with necrotic slough on the right and left side of the posterior palate and on the right buccal mucosa.

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Figure 2 Blood-filled blister in the buccal vestibule corresponding to lower left mandibular molars.

involvement, which were not present in our case. Linear IgA disease was also ruled out as there were no IgA deposits (linear and granular patterns) along the basement membrane zone in perilesional tissue in DIF. Although, it is very uncommon to see oral manifestations in dermatitis herpetiformis but erythematous erosions, purpura and vesicular eruptions had been reported in some patients, but generally this entity is accompanied with skin lesions. Oral Bullous lichen planus was differentiated through immunofluorescence, which shows linear deposits of fibrinogen along the basement membrane zone in such cases. The target-like lesions of the skin in erythema multiforme, the multiple erosions of lips and their crusting haemorrhagic appearance ruled out this condition. History and the presence of bullous skin lesions differentiated the oral blood blisters found in our case from those observed in epidermolysis bullosa. A final diagnosis of angina bullosa haemorrhagica was given.

TREATMENT

The patient was educated about the condition and reassured. Symptomatic treatment in the form of soothing mouthwash (0.2% chlorhexidine gluconate) and topical anaesthetic was prescribed on the appearance of similar lesions.

OUTCOME AND FOLLOW-UP

The patient was kept under follow-up and we noticed similar ulcerations and blood filled blisters at subsequent visits for which the same treatment was given.

DISCUSSION

The term ABH was first introduced in 1967 by Badham.¹ ABH is also known as localised oral purpura² stomatopompholyx haemorrhagica³ and recurrent oral hemophlyctenosis.⁴ The pathogenesis of this condition is not known but according to Hopkins and Walker⁵ there is weakness in the attachment of the oral epithelium to the underlying connective tissue in susceptible patients which ruptures the junction on mild trauma and leads to subepithelial bulla. Local trauma seems to be a major provoking factor. Norio *et al*⁶ had carried out a study in which they investigated the presence or absence of local factors as well as systemic background disease in 16 patients with ABH arising in the soft palate. Scratching of oral mucosa (minor trauma) was present in 75% of the patients and hypertension was most common with regard to systemic conditions among the patients in that study. Several studies have indicated a possible association between steroid inhalers and the occurrence of ABH. One of the studies conducted by High and Main⁷ provided evidence of occurrence of ABH in patients using steroid-based inhaler preparations for periods longer than 5 years. Grinspan *et al* advocated the

contribution of an altered glucose metabolism in the pathogenesis of ABH because they found a positive family history of diabetes mellitus or a sign of abnormal glucose metabolism in 44% of their patients. However, it is unclear if the altered glucose metabolism or injury to the oral mucosa was a contributing factor in the pathogenesis of ABH in their cases.⁴ ABH is commonly seen in elderly patients. Stephenson *et al*⁸ recorded 30 cases of ABH, median age at presentation was 54 years and 63% of patients were in the age group of 45–70 years. It is uncommon to have recurrent episodes of ABH like in the current case. Giuliani *et al*⁹ reported eight cases out of which six patients had one episode of such lesions, one case was sporadic and one case had frequent episodes in different sites. ABH is most commonly seen in the soft palate region. Yamamoto *et al*¹⁰ reported 11 cases which occurred on different sites of the soft palate. Lesions can also develop on other sites of the oral cavity including the buccal mucosa, lip, and the lateral surface of the tongue; masticatory mucosa of the hard palate and gingiva do not seem to be affected. Peculiarly, the lesions were noted on the gingiva in the current case. Curran *et al*¹¹ reported a case of ABH on the gingiva induced after routine scaling and root planning treatment. The current case however, presented with lesions on the gingiva even though no dental procedures were carried out.

Blood-filled blisters may occur in the oral cavity in association with disorders such as mucosal pemphigoid, epidermolysis bullosa and amyloidosis. Oral mucosal pemphigoid may be differentiated from ABH on the basis of direct immunofluorescence which shows linear deposits of IgG or C3. Moreover, the absence of desquamative gingivitis and nasal or conjunctival mucosal involvement will differentiate ABH from benign mucous membrane pemphigoid. In epidermolysis bullosa there is the presence of bullous skin lesions and it is an inherited disorder which begins at birth or soon after. The presence of IgA deposits (linear and granular patterns) along the basement membrane zone in the perilesional tissue helps to distinguish Linear IgA disease and dermatitis herpetiformis (DH) from ABH.^{12 13} In DH oral manifestations are uncommon but erythematous erosions; purpura and pseudovesicular lesions have been reported. Differentiation of ABH from haemorrhagic bullae found in amyloidosis may be made by examining the mouth for other features such as macroglossia, petechiae or ecchymosis and moreover, haemorrhagic bullae in amyloidosis are persistent. Management of the patient with suspected ABH should include a careful history and examination to exclude other conditions in which blood blisters are common.

The patient should be reassured and prescribed 0.2% chlorhexidine gluconate mouth wash. Topical anaesthetic may be used in painful lesions. Long-term follow-up is recommended to assess the healing of the lesions and to exclude other causes of blood blisters. Although ABH is a benign condition, some authors report that lesions located in the posterior pharynx and in the epiglottis may cause a choking or gagging sensation and in rare cases could result in acute airway obstruction.⁸ Hence, any large intact blood blister should be incised to prevent further extension that could cause airway obstruction.

Learning points

- ▶ Angina bullosa haemorrhagica is a self-limiting condition.
- ▶ Although it is a rare disease it should be included in the differential diagnosis of blood-filled blistering conditions.
- ▶ It should be treated symptomatically.

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Competing interests None.

Patient consent Obtained.

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REFERENCES

- 1 Badham NJ. Blood blisters and the oesophageal cast. *J Laryngol Otol* 1967;81:791–803.
- 2 Scully C. The oral cavity. In: Rook AJ, ed. *Textbook of dermatology*. 5th edn. Oxford: Blackwell Scientific Publications, 1992:2732–3.
- 3 Deblauwe BM, van der Waal I. Blood blisters of the oral mucosa (angina bullosa haemorrhagica). *J Am Acad Dermatol* 1994;31:(2 Pt 2):341–4.
- 4 Grinspan D, Abulafia J, Lanfranchi H. Angina bullosa hemorrhagica. *Int J Dermatol* 1999;38:525–8.
- 5 Hopkins R, Walker DM. Oral blood blisters: angina bullosa haemorrhagica. *Br J Oral Maxillofac Surg* 1985;23:9–16.
- 6 Horie N, Kawano R, Inaba J, et al. Angina bullosa hemorrhagica of the soft palate: a clinical study of 16 cases. *J Oral Sci* 2008;50:33–6.
- 7 High AS, Main DM. Angina bullosa haemorrhagica: a complication of long-term steroid inhaler use. *Br Dent J* 1988;165:176–9.
- 8 Stephenson P, Lamey PJ, Scully C, et al. Angina bullosa haemorrhagica: clinical and laboratory features in 30 patients. *Oral Surg Oral Med Oral Pathol* 1987;63:560–5.
- 9 Giuliani M, Favia GF, Lajolo C, et al. Angina bullosa haemorrhagica: presentation of eight new cases and a review of the literature. *Oral Dis* 2002;8:54–8.
- 10 Yamamoto K, Fujimoto M, Inoue M, et al. Angina bullosa hemorrhagica of the soft palate: report of 11 cases and literature review. *J Oral Maxillofac Surg* 2006;64:1433–6.
- 11 Curran AE, Rives RW. Angina bullosa hemorrhagica: an unusual problem following periodontal therapy. *J Periodontol* 2000;71:1770–3.
- 12 Kilpi AM, Rich AM, Radden BG, et al. Direct immunofluorescence in the diagnosis of oral mucosal diseases. *Int J Oral Maxillofac Surg* 1988;17:6–10.
- 13 Korman N. Bullous pemphigoid. *J Am Acad Dermatol* 1987;16(5 Pt 1):907–24.

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