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Case Report

Unusual Spontaneous Hematoma on the Floor of the Mouth Associated with Acquired Hemophilia A

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Abstract

Acquired hemophilia is rare and the presentation of a spontaneous hematoma on the floor of the mouth is very unusual. We present an unusual case of an older adult patient who presented a spontaneous hematoma on the floor of the mouth without apparent cause, with acquired hemophilia A. The data source for the case was the patient's medical history. When reviewing the literature only 13 reported cases of upper airway bleeding have been reported in patients with diagnosed acquired hemophilia, withmales the most affected and the mean age of patients being 69.53 years. Differential diagnoses, imaging, radiographs, scans and laboratory tests help to establish a timely diagnosis and act quickly.

***Keywords:** Hematoma, Airway obstruction, Hemophilia A*

Introduction

There are numerous causes of bleeding in patients who go to the emergency department or the intensive care unit. Example are platelet disorders, different types of hemophilia, uremia, liver failure, myeloma, myeloproliferative disorders, lymphoproliferative conditions, medications, reduced production of coagulation factors, liver dysfunction, vitamin K deficiency, broad-spectrum antibiotics that cause dilution of coagulation factors, trauma, hemodilution for preoperative fluid resuscitation, massive preoperative transfusion and cardiopulmonary bypass for complex cardiac surgery.[1,2]

Acquired hemophilia is an uncommon serious condition in which inhibiting antibodies develop against coagulation factors. Most often affected is factor VIII of coagulation, leading to significant bleeding episodes that carry high morbidity and mortality.[1]

Acquired hemophilia can occur in previously healthy men and women of any age. The incidence of this condition is 1.4 per million and it is most frequent in elderly patients. It should be suspected in patients with a new onset of bleeding and an isolated prolongation of activated partial thromboplastin time. Diagnosis of acquired hemophilia is confirmed by demonstrating markedly reduced factor VIII activity (FVIII:C) and neutralizing anti-FVIII antibodies, so-called inhibitors. Several limitations and pitfalls exist with the laboratory tests used to diagnose acquired hemophilia.[3]

The priority is to control acute bleeding. The first-line therapy is bypassing agents such as recombinant activated FVII or activated prothrombin complex concentrate. Once the diagnosis has been achieved, immediate autoantibody eradication to reduce subsequent bleeding risk should be performed.[4]

A case report of a patient with no pathological history presented with an unusual spontaneous hematoma on the floor of the mouth and tongue. The patient on the first day of hospitalization did not present significant alterations in the laboratories, but on the second day, he presented an increase in partial thromboplastin time that was impossible to quantify. The aim of this study is to report on an unusual spontaneous hematoma on the floor of the mouth associated with acquired hemophilia A.

Case Report

A 95-year-old male patient was admitted to the emergency department with a spontaneous hematoma on the floor of the mouth and tongue. The patient and relatives reported that he woke up with that condition, and they immediately attended the consultation. The patient and family members denied important personal pathological history and denied a history of trauma or the use of dental prostheses. Clinically, an increase in volume was observed in the tongue and a blackish-purple color (Figure 1). Laboratories tests were indicated on admission to the hospital (Table 1).

Table 1. Laboratory results taken on the first day of the patient's admission to hospitalization.

Laboratory tests of first day	Results	Reference value	Interpretation
Leukocytes	5.80 x 10^9/L	5.0-10.0 x 10^9/L	Normal
Neutrophils	56.9 %	50-65%	Normal
Lymphocytes	13%	20-45%	Low
Hemoglobin	10.5 g/dl	16 ± 2. g/dl	Low
Hematocrit	34.4 %	37-47%	Low
Platelets	248 x 10^9/L	150-400 x 10^9/L	Normal
Blood urea nitrogen	21.1 mg/dl	5-20 mg/dl	Increased
Creatinine	0.78 mg/L	0.6-1-1 mg/L	Normal
PTT	35.70 seconds	25-35 seconds	Upper limit
PT	15.30 seconds	11-13.5 seconds	Prolonged
INR	1.08	0.9-1.3	Normal

PTT: *Partial thromboplastin time (population mean 28 seconds).*

PT: *Prothrombin time.*

INR: *International normalized ratio.*

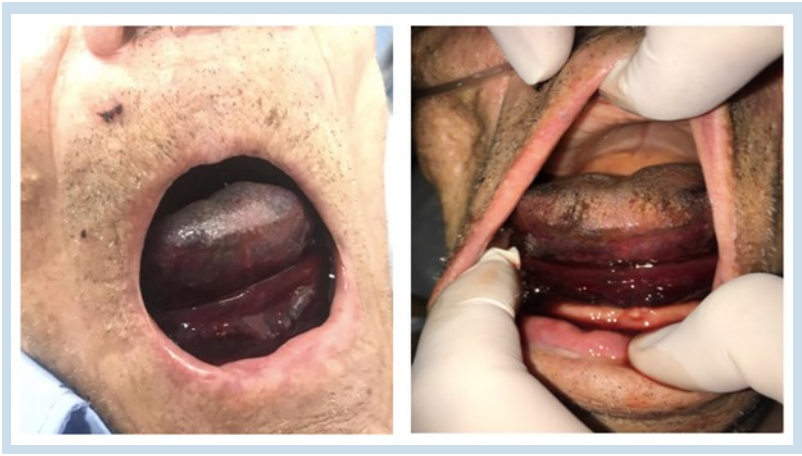

Figure 1. Initial photo of the patient at the time of admission

An emergency videofibrolaryngoscopy was performed. The hematoma was observed up to the base of the tongue. The pyriform sinuses were without masses, foreign bodies, or additional alterations. Due to the risk of collapse of the airway, an emergency tracheostomy was performed and an angiotomography of the face and neck was indicated (Figure 2). The tomography shows extensive inflammatory changes with the presence of hematic remains that extend through the pharyngeal mucous space and the glottis with obliteration of the airway at the level of the hyoid bone. Obliteration of the nasopharynx was also identified with a suggestive collection image in the soft palate with diameters of 19x8 mm and extensive inflammatory changes in the floor of the mouth, as well as in the submandibular spaces with a predominance on the left side. Emphysema of the retropharyngeal space and right carotid space is also observed.

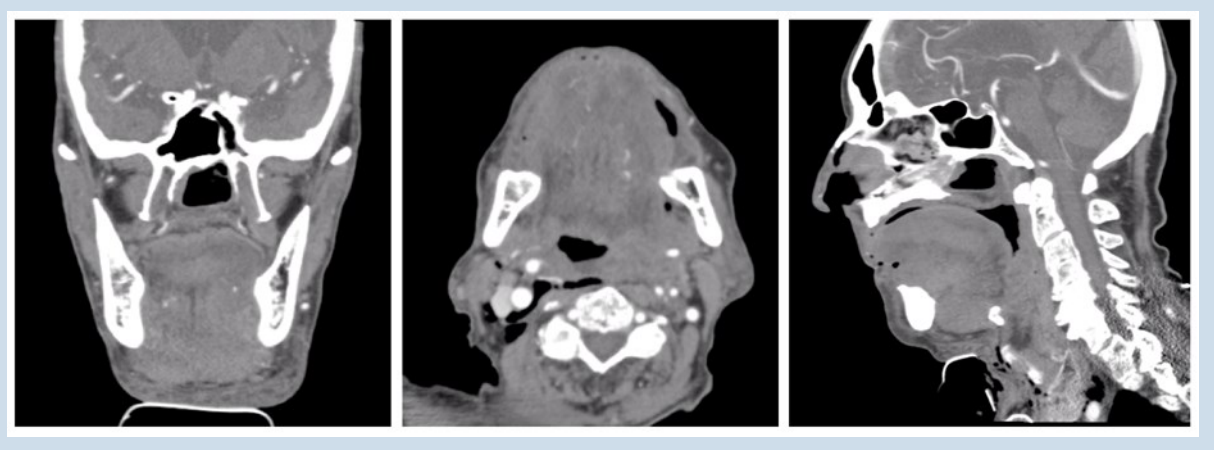

Figure 2. Angiotomography showing the hematoma. The tomography shows extensive inflammatory changes with the presence of hematic remains that extend through the pharyngeal mucous space and the glottis. The day after the patient was admitted, there was no evidence of improvement, bleeding started at the tracheostomy site and new laboratory tests were indicated. (Table 2) and the patient was transferred to the intensive care unit. Laboratory tests are repeated in the afternoon due to previous results and they report similar results. Transfusion of two packaged red blood cell units is performed without complications and tranexamic acid is administered 1-gram, single dose. The patient continued without ventilatory support with a tracheostomy, without vasoactive support, without pulmonary aggregates, with stable blood pressure values, mean values within goals, and good urinary output.

Table 2. Laboratory results taken on the second day of the patient's admission to hospitalization.

Laboratory tests of second day	Results	Reference value	Interpretation
Leukocytes	7.54 x 10^9/L	5.0-10.0 x 10^9/L	Normal
Neutrophils	62,39 %	50-65%	Increased
Lymphocytes	13%	20-45%	Low
Hemoglobin	7.2 g/dl	16 ± 2. g/dl	Low
Hematocrit	25.5 %	37-47%	Low
Platelets	149 x 10^9/L	150-400 x 10^9/L	Low
PTT	Greater than 400 seconds	25-35 seconds	Prolonged
PT	17.10 seconds	11-13.5 seconds	Prolonged
INR	1.22	0.9-1.3	Normal
Fibrinogen	555 mg/dl	200-400 mg/dl	Prolonged
PaFi	277 mm Hg	> 300 mm Hg	Normal
pH	7.35	Between 7.35 and 7.45	Normal
HCO3	20 mEq/L	20-24 mEq/L	Normal
Bood Lactate	2.7 mmol/L	< 2 mmol/L	Normal
Base Excess	-5.1 mEq/L	−2 to +2 mEq/L	Increased

PTT: *Partial thromboplastin time (population mean 28 seconds).*

PT: *Prothrombin time.*

INR: *International normalized ratio.*

PaFi: *PaO2/FiO2*

pH: *Is a scale used to specify the acidity or basicity of an aqueous solution.*

HCO3: *Bicarbonate*

It was decided to continue monitoring the ventilatory pattern: the analgesia was adjusted, the acid-base balance was maintained, and a crossed partial thromboplastin time was indicated, which gave a positive result, so the administration of Vitamin K 10 mg endovenous every 12 hours was started. Factor VIII, VIII, IX, X, XI is requested for documentation of deficit and for suspected acquired hemophilia. The patient became adynamic, asthenic, hemodynamically unstable, hypotensive, tachycardic, with desaturation. On chest radiograph, right basal atelectasis with pleural effusion was observed. The general condition of the patient deteriorated until he presented cardiac arrest with a type of asystole rhythm and died. The results of the requested blood factors report that the patient had a deficiency of coagulation factor VIII, so the diagnosis of acquired hemophilia was confirmed.

Table 3. A summary of all cases of spontaneous upper airway hemorrhage in acquired hemophilia reported.

Study (Year)	Gender	Patient age	Site of spontaneous airway hemorrhage	Airway management
Joynt et al. [12] (1996)	Female	34	Tongue, floor of mouth.	Nasotracheal intubation
Wendell et al.[13] (2004)	Female	74	Tongue.	Nil
Weise et al. [14] (2005)	Male	67	Floor of mouth, tongue, hypopharynx.	Tracheostomy
Harper et al. [15] (2007)	Female	75	Retropharyngeal space.	Orotracheal intubation followed by tracheotomy
Al-Hilou et al.[16] (2011)	Female	65	Oral floor, Sublingual.	Orotracheal intubation
Kelesidis et al.[17] (2011)	Male	94	Dorsal surface of the tongue.	Not intubated
Yanagi et al.[18] (2016)	Male	79	Oral floor, pharynx, and larynx.	Nil
See et al.[19] (2016)	Male	61	Submandibular, submental, retropharyngeal and parapharyngeal spaces.	Orotracheal intubation followed by emergency tracheotomy
Spindler et al.[20] (2017)	Male	73	Sublingual, Oral floor.	Orotracheal intubation
Kaur et al.[21] (2018)	Female	78	Tongue, floor of mouth.	Nil
Seo et al.[22] (2019)	Male	64	Posterior pharyngeal wall and oral mucosa.	Nil
Howden et al.[23] (2020)	Male	80	Supraglottic area.	Nasal intubation.
Endermann et al.[24] (2020)	Male	60	Retropharyngeal space.	Nasal intubation.

Discussion

The diagnosis of acquired hemophilia A is based in a finding of an isolated prolongation of the activated partial thromboplastin time (APTT), that does not correct by incubating the patient's plasma with equal volumes of normal plasma (mixing study), also related to reduced values of coagulation factor VIII levels, and evidence of a factor VIII inhibitor in a patient with no previous personal or family history of bleeding. [5] The diagnosis is not always made on the first appointment or the first day of patient evaluation; different initial studies are generally carried out in order to reach a final diagnosis. In general, these patients urgently come with a complication and deny a previous pathological history, which makes the authors think of different causes instead of the presence of acquired hemophilia A.

In case of the patient in the present case, he presented with an unusual spontaneous hematoma on the floor of the mouth that first caused the professionals to consider local causes such as aneurysms or trauma. Furthermore, the first-day coagulation tests had no relevant alterations, and it was on the second day that an exaggerated increase in coagulation times was observed. This led them to explore other more local causes.

Hematomas or hemorrhage of the oral cavity can be fatal to the patient. Spontaneous bleeding into the sublingual and submaxillary spaces can create a "pseudo-Ludwig's phenomenon" that can obstruct the airway and cause the death of the patient. Hematoma in the oral cavity has been described as a complication of high blood pressure [6,7], spontaneous fracture of mandibular genial tubercle [8], the use of warfarin [9], vascular malformations [10], dental procedures [11], among other unusual causes but the review of the literature 1945 to 2020 revealed only 13 reported cases of upper airway bleeding in patients with diagnosed acquired hemophilia. Of the 13 cases (Table 3), 8 corresponded to bleeding in the oral cavity. Males were the most reported in the literature, with 8 cases and the average age of 69.53 years old. Although studies and cases of bleeding in the head and neck area have been reported in relation to congenital hemophilia, there are few reports of specific clinical cases for acquired hemophilia. [19,25]

The treatment depends on the cause. Identifying that the cause is acquired hemophilia is not straightforward. Generally, patients who come for a spontaneous hematoma consultation require an initial contrasted topography to evaluate soft and bony structures, although a computed tomography angiography can be useful in some cases. Hematological studies should also be performed to confirm the diagnosis and guide the treatment plan. Once diagnosed, the treatment aims to prevent the production of autoantibodies against coagulation factor VIII and normalize hemostasis. This should start as soon as the diagnosis is confirmed, since the risk of serious bleeding will persist. The most widely used method of treatment is the use of steroids, either alone or in combination with low-dose cyclophosphamide, chemotherapy, and rituximab as the alternative of choice if first-line treatment fails.[26,27]

In the patient of this case report, it was not possible to make a timely confirmatory diagnosis, and therefore, the adequate treatment could not be administered. Knowledge of these types of cases is important as they are unusual and can occur in daily practice. Acquired hemophilia is rare and the presentation of a hematoma on the floor of the mouth is very unusual. It requires differential diagnoses, and images and laboratories performed fast so as to establish a timely diagnosis and swift act.

Conflict of Interest

The authors declare that they have no conflict of interest.

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