OROPHARYNX HEMATOMA IN A PATIENT WITH CONGENITAL AFIBRINOGENEMIA

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ABSTRACT

INTRODUCTION: The congenital afibrinogenemy is an autosomic recessive disease defined by absence of fibrinogen. It causes not only small amounts of bleeding, but also severe hemorrhages. This is a case report of a patient who developed a large hematoma in the oropharynx, caused by great abnormalities at the coagulogram and turning back into normal after cryoprecipitate administration.
OBJECTIVE: To describe a rare case of afibrinogenemy.
CASE REPORT: Patient 12 years old female was admitted to the emergency department of otorhinolaryngology complaining of swelling of the face and right arm for five days. Her mother told us a history of manipulation of the lower left second molar, with local pain associated with edema in left submandibular area for 4 days. Medical antecedents: Afibrinogenemy and hemorrhagic stroke a year ago. Examination evidenced bruising of the mouth floor on the left, friable and bleeding, together with neck edema. The patient was admitted to the ICU, underwent necessary support. She was evaluated by an hematologist, and it was decided to apply 3 bags of cryoprecipitate, prophylactic antibiotics and expectant management. The patient evolved with complete clinical reversal, not requiring surgical drainage, and improvement of laboratory parameters.
CONCLUSION:
In practice in otolaryngology, we must be alert to cases of blood disorders, which can be detected or be part of differential diagnosis in our physical examination, such as gingival hyperplasia, mucosal bruising, bleeding, among others.
KEYWORDS: Afibrinogenemia, bruising, coagulopathy

INTRODUCTION

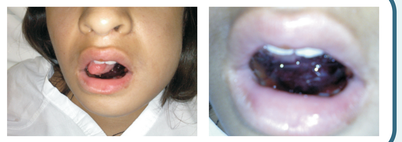
Afibrinogenemia is characterized by congenital absence of fibrinogen plasma coagulation factor (type I). It is considered a rare autosomal recessive disease, with an estimated incidence of 1:500,000 births (1), and only 250 cases reported in the world literature (2).
Fibrinogen is a glycoprotein of hepatic origin involved in the final stage of coagulation as a forerunner of fibrin, necessary for the formation of platelet plug. There are 3 disorders of fibrinogen: afibrinogenemia (absence of fibrinogen), hypofibrinogenemia (low fibrinogen levels) and disfibrinogenemia (malfunction of fibrinogen). Afibrinogenemia is an autosomal recessive disease, located on chromosome 4 (q26-q28), which affect both genders (3).
The clinical symptomatology ranges from mild to severe hemorrhages. Hemorrhagic episodes may appear in the neonatal period, as traumatic hematomas in childbirth, umbilical bleeding, intracranial hemorrhages , mucosal hemorrhages among others.
The hemorrhagic episodes although untimely, are generally pos traumatic, but there are reports of asymptomatic individuals for prolonged periods. The most frequent hemorrhages are abortions, gastrointestinal bleeding and epistaxis.
The diagnosis starts from a detailed clinical history: beginning of bleeding, family history, spontaneous or traumatic bleeding, location of bleeding (skin, mucosa, cavitations); combined with the laboratory evaluation with fibrinogen dosing in blood (undetectable), dosage of prothrombin time (PT), activated partial thromboplastin time (aPTT) and time of prolonged thrombin, absent clotting time (4).
When this coagulopathy is diagnosed, physicians must be prepared to welcome and guide patients and relatives about genetic counseling, discourage contact sports, care in surgical procedures, administer medicinal products when necessary (5).
The treatment of choice may be done with fresh frozen plasma or cryoprecipitate. Due to the importance of hematological disorders in otorhinolaringology practice, we described a case of congenital afibrinogenemia accompanied by the emergency room of otorhinolaringology from Hospital Nossa Senhora de Lourdes São Paulo Brazil.

CASE REPORT

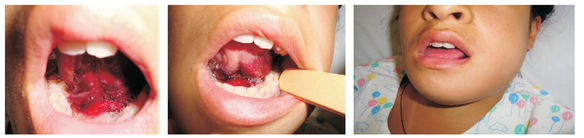
Patient C. F. R. M 12 years, female, natural from São Paulo, was admitted at an otorhinolaringology ER from Hospital Nossa Senhora de Lourdes São Paulo Brazil, accompanied by her mother, referring a 5 day edema on left face. The mother reported an history of dental procedure. The girl presented with local pain with edema in left submandibular region, progressive, salivary swallowing difficulty without other symptoms. Examination revealed afebrile, regular general state, left submandibular bulging with cervical extension , voluminous hematoma in mouth floor. Evolved with fall of the tongue due to the oral hematoma. Antecedent of afibrinogenemia and hemorrhagic stroke 1 year ago.
She wasn't using any medication daily.
Familiar antecedent of parents cousins first degree and a brother of 2 year old with the same hematological disorder.
Coagulogram in ER: PT > 10 seg; prothrombin activity < 8%; INR >10; aPTT > 180 seg; fibrinogen < 50; platelets 280,000 ;
Hemogram HB 14; HT 40; VCM 78; leuco 13,800 ; sticks 5%; segmented 56%; neutrofilos 61%; eosinofilos 2%; lymphocytes 36%.

Cervical CT: left hypoattenuation formation in masseter muscle region ,that may correspond to hematoma, presence of cervical lymph node enlargement on left submandibular region.
Patient evolved with airway obstruction due to the fall of tongue, being then admitted in the ICU. Evaluated by hematologist that chose treatment with cryoprecipitate (3 bags) and prophylactic antibiotics. At the end of the treatment, patient presented total resolution of the hematoma and coagulogram normalization.

Oral hematoma



Oral hematoma after cryoprecipitate treatment



CONCLUSION

Congenital afibrinigenemia is a rare hematological disorder , with high mortality rate due its severe complications.
It can be diagnosed since birth, normally pos traumatic, and has relationship with consanguineous marriage antecedents. In the case reported the child presented an history of stroke, family antecedent of consanguineous parents, a brother with the same disorder; and laboratory test that confirmed the pathology.

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