Hemofilia A adquirida

Acquired hemophilia A

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ABSTRACT

The authors present the case of a man evaluated by Internal Medicine because of a spontaneous lesion of the upper limb, with active hemorrhage difficult to control. The patient was diagnosed with Acquired Hemophilia, a rare disease caused by the presence of antibodies against factor VIII.

Keywords: Acquired hemophilia A, Bleeding, Factor VIII

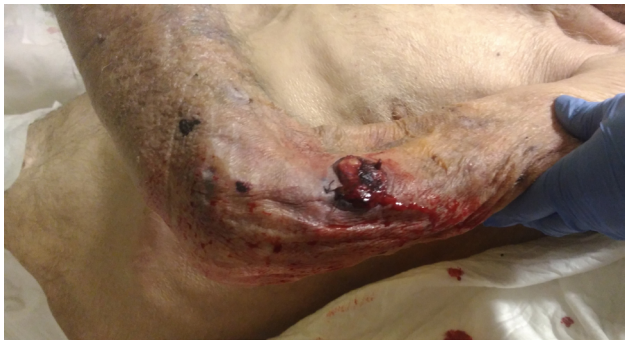
CASE REPORT

Acquired hemophilia A is a rare disease, with an incidence of 1.5 cases per 1,000,000 / year[1], due to the presence of antibodies that inhibit the action of factor VIII (FVIII)[2].

The authors present the case of an 83-year-old man with a history of ischemic stroke about 5 months prior, and in this context diagnosed with paroxysmal atrial fibrillation. He was discharged from hospital on anticoagulation, which he willingly suspended after 1 month.

After multiple visits to the Emergency Room, was requested for an Internal Medicine evaluation, due to the appearance of a non-traumatic lesion in the left upper limb, with active hemorrhage that was difficult to control (Figure 1).

Figure 1. Left upper limb lesion, with suture dehiscence and active hemorrhage.



The initial laboratory evaluation revealed: hemoglobin 7.3 g / dl, platelets 288,000/uL, normal prothrombin time, activated thromboplastin time 106.8 sec and direct negative Coombs test.

The evaluation of the activity of coagulation factors revealed reduced levels of factor VIII 0.5% (N: 50-150%) and an increase in von Willebrand factor (369%), with the presence of factor VIII inhibitor being identified, confirming the diagnosis of acquired hemophilia A.

In the initial approach, transfusion support was performed with red blood cell concentrate and fresh frozen plasma, with progressive worsening. Subsequently, he was transferred to a reference unit to receive FEIBA, after which he started immunosuppressive therapy with prednisone (1mg / kg) and cyclophosphamide (1mg / kg), with a great response.

Acquired hemophilia A is a severe coagulopathy, with a high potential for morbidity and mortality[4], so a timely diagnosis is essential. Physicians should be suspicious when they are facing a patient who shows signs of active bleeding, with abnormal prolongation of isolated aPTT[3].

CONFLICT OF INTEREST

The authors declare that there is no conflict of interest in this work.

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This research had no funding sources.

ETHICAL ASPECTS

All participants submitted a consent form to be included in this study.

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