Abstract: P2273

Title: MANAGEMENT OF ACQUIRED SEVERE HEMOPHILIA ACCORDING TO THE LATEST INTERNATIONAL GUIDELINES AND THE INFECTIOUS AND INFLAMMATORY COMPLICATIONS DERIVED.

Abstract Type: e-Poster Presentation

Topic: Bleeding disorders (congenital and acquired)

Background:

Acquired Hemophilia A is a hemorrhagic disease of immunological origin (it affects one in a million people each year) that occurs when an autoantibody begins to inhibit the activity of coagulation factor VIII. It is an entity with variable etiology (50% idiopathic). The clinical presentation of the disease usually consists of severe hemorrhages in the subcutaneous, muscular, gastrointestinal, genitourinary, and retroperitoneal locations. The new 2020 international guidelines (International recommendations on the diagnosis and treatment of acquired hemophilia A) establish treatment with cyclophosphamide or rituximab plus systemic corticosteroid therapy in those patients who debut with factor XIII levels less than 1% and/or with inhibitor levels. in Bethesda units greater than 20 Bethesda units/mL.

Aims:

This case exemplifies the clinical difficulty in diagnosing Acquired Hemophilia A, highlighting that therapeutic management according to recent international guidelines is effective in resolving the disease but can lead to the appearance of severe complications, both infectious and toxic at other levels, hence the importance of adequate monitoring and management of them.

Methods:

We present the case of a 63-year-old man diagnosed of Severe Acquired Hemophilia A who underwent serious infectious complications due to the treatment.

Results:

We present the case of a 63-year-old man with a relevant history of paucisymptomatic psoriasis who was admitted in September 2023 for spontaneous right thigh hematomas 2 weeks ago with poor pain control, and subsequent appearance of spontaneous hematomas without previous trauma in the upper limbs, without taking any treatment or any other type of exogenous substance. No arthritis, arthralgia, or B symptoms. Coagulation essays revealed Factor VIII: C 0.4% and factor IX 113%, with inhibitor 33.70 Bethesda Units. It is started at this time with the diagnosis of Severe Acquired Hemophilia A, eptacog alfa 5 mg/8 hours, methylprednisolone 100 mg/24 hours and, on October 3, 2023, cyclophosphamide 2 mg/Kg is started. In the absence of improvement, Rituximab (RTX) 830 mg 4 doses was started. He presented infectious complications with CMV infection and JC virus, as well as Fusarium proliferatum infection and Staphylococcus haemolyticus bacteremia. He also presented an increase in acute phase reactants with elevated ferritin and triglycerides, which together with the fever support the suspicion of hemophagocytic syndrome, although he did not meet the criteria for diagnosis. On the other hand, he presented central erythroblastopenia with transfusion requirements that resolved spontaneously. These complications, after ruling out other causes, were put in the context of the treatments received for severe Acquired Hemophilia A.

Summary/Conclusion:

Early diagnosis of acquired hemophilia is of vital importance to provide timely treatment in order to avoid bleeding with serious complications.

The availability of the new guidelines that support the first-line use of corticosteroids and immunosuppressants together with Rituximab when it is a severe case, represents an important advance in terms of the resolution of

the condition, but may condition the appearance of a greater number of infectious complications.

Infectious complications are a cause of important morbidity and mortality in patients with acquired Hemophilia A under immunosuppressive treatment.

It is necessary to create working groups and develop research to collect non-hemostatic complications in patients with severe acquired coagulopathies such as Acquired Hemophilia A to establish predictive models for them and improve their management.

Keywords: Rituximab, Cyclophosphamide, Acquired hemophilia, Infection