Acquired Hemophilia A: Presentation of 5 cases

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Introduction and objectives

Acquired hemophilia A can be defined as a bleeding disorder caused by generation of auto-antibodies towards factor VIII. In untreated patients it often leads to a life-threatening hemorrhage.

Material and Methods

This report presents 5 cases with acquired hemophilia A treated at the University Clinic of Hematology, Skopje from 2006 to 2012. The men-women ratio was 1-4 respectively. The average age of patients was 56 years (24 – 80 years). The risk factors for development of auto-antibodies included: puerperal period, lung cancer and urolithiais. However, in two of the patients the etiology was not defined.

Results

Clinical examination demonstrated extensive hematomas throughout the body and limbs in four of the patients and hematuria in two of them. Laboratory evaluation showed initial elevation of APTT (activated partial thromboplastin time) in all patients; average 64.8sec with control 22.0 sec.

The qualitative test with mixture of normal and patient-derived plasma was positive in all patients. Initially, the average value for factor VIII and the inhibitors' titer were 1.2% and 19.7 B.E. respectively out of which, three of the patients were low responders, with inhibitor titer below 5 B.E. and two of them were high responders, with inhibitor titer over 5 B.U.

The acute hemorrhage was treated with high doses of factor VIII concentrate (Coate) in two patients, fresh-frozen plasma (FFP) in one patient and activated recombinant factor VII (Novoseven) in two patients. In addition, the patients were treated with immunosuppressive therapy including corticosteroids only or, corticosteroids in combination with Cyclophosphamide and/or Rituximab. The hemorrhage was stopped and clinical remission was achieved in all patients for approximately 60 days (18 to 180).

Conclusion

- Acquired Hemophilia A is a very rare disease.

Nevertheless, it may lead to a life-threatening hemorrhage. However, early diagnosis and suitable treatment can prevent fatal outcome.

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