ACQUIRED HEMOPHILIA A AND SYSTEMIC CONNECTIVE TISSUE DISEASE: A CASE REPORT

Enisa Zaric*[1]*, *Isidora Rubezic*[1]*, Sanela Muminovic*[2]*, Sofija Marojevic*[1]

[1]*Center for Hematology, Clinical Center of Montenegro, Montenegro*
[2]*Primary health care Podgorica, Montenegro*

Introduction. Acquired hemophilia A is a rare, life-threatening condition that manifests itself in spontaneous bleeding, mostly in soft tissues. The diagnosis of this disease should be considered in unexplained bleeding, especially in elderly patients.
Case presentation. An 83-year-old female patient was initially hospitalized at the vascular surgery of the Clinical Center of Montenegro due to deep venous thrombosis (DVT) of the right leg. After being discharged from vascular surgery, she noticed a bruise on the skin of her abdomen, for which she was hospitalized again. Surgical drainage of the hematoma in the anterior abdominal wall and tamponade, then new tamponade and revision were performed. During the second hospitalization, the laboratory findings showed anemia with prolonged aPTT values (Er 2.74, Hg 76, MCV 86, D dimer 3.73, aPTT 113.7, PV normal). A reduced level of factor F VIII (< 0.4 I.U./dl) is found. In a 50:50 mixing study with normal plasma, aPTT and factor VIII did not normalize. The inhibitor level was high (224 BU/mL). The patient was treated with corticosteroids, and intravenous immunoglobulins, and therapeutic plasma exchange with factor VII recombinants was performed. There is a gradual stabilization of the laboratory findings as well as the resolution of the hematoma, there have been no repeated manifestations of cowering. Screening tests were carried out in terms of malignancy and autoimmune diseases. The findings of immunoserology indicated a possible systemic connective tissue disease, which was also confirmed by a rheumatologist. She was treated with pulse doses of corticosteroids. The aPTT values remained stable. Due to previously verified DVT, treatment with rivaroxaban was started. Conclusion. Acquired Hemophilia A can be a reversible coagulopathy. Early diagnosis and early initiation of treatment can lead to the successful resolution of the disease. An adequate approach includes screening for the etiology of acquired hemophilia.

Key words: acquired hemophilia, inhibitor, etiology, diagnosis, treatment