Management challenge between bleeding and thrombosis in an elderly woman with acquired hemophilia

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Introduction:

Acquired hemophilia is a rare but serious disease with morbidity and mortality rates that can reach 20%. It is most often underdiagnosed and is associated with the presence of inhibitors against factor VIII, leading to an increased tendency to bleed. Acquired hemophilia can occur in the postpartum period or can be secondary to dysimmune or neoplastic pathologies, which highlights the importance of thorough etiological investigation. In 50% of cases, it is idiopathic, representing a true diagnosis of exclusion. The therapeutic objective is twofold; it aims both to treat the hemorrhage and to eradicate the inhibitor.

Clinical Case:

In this clinical case, I present to you the paradox of developing deep vein thromboses with simultaneous bleeding in a patient diagnosed with acquired hemophilia and the real challenge in therapeutic management?

We report the case of an 84-year-old woman who presented on 05/01/2023 for consultation with generalized bruising on her back and legs, 10 days after the diagnosis of a left popliteal vein thrombosis treated with curative Lovenox at a dose of 0.8 IU/12 hours. The biological tests revealed anemia with a hemoglobin level of 8.5 g/dL with a normal platelet count, a normal prothrombin time, an activated partial thromboplastin time (aPTT) prolonged to 111 sec/30 sec, and an emergency mix aPTT still prolonged to over 100 sec, a subsequently measured factor VIII level returning to less than 1%, and anti-factor VIII antibodies (ACC) at 20 UI, confirming the diagnosis of acquired hemophilia. An etiological investigation was conducted, specifically a contrast-enhanced CT scan of the thorax, abdomen, and pelvis, which returned normal, along with a normal mammogram. The biological assessment found the presence of lupus anticoagulant, but the FAN and anti-DNA and ENA and APL were negative, with a normal thyroid profile and negative anti-ATPO and anti-ATG antibodies. A treatment with bypass agents was started based on Novoseven 90 IU/kg/3 h with an inhibitor eradication scheme using Solupred 1 mg/kg, which was not combined with Endoxan due to the patient's advanced age and product rupture. After 7 days of treatment with bypass agents, there was a regression of the hemorrhagic syndrome. We restarted the anticoagulant treatment on 12/01/2023 with Lovenox 0.8/12 hours subcutaneously, continuing with bypass agents. The patient was discharged on 14/01/2023 after an improvement in her clinical condition. The patient was readmitted on 19/02/2023 for hypovolemic shock due to upper gastrointestinal bleeding type hematemesis, and worsening edema of the left lower limb with a TCA of 59/32'. An esophagogastroduodenoscopy (EGD) revealed hemorrhagic gastritis, and a venous Doppler ultrasound showed recent left femoral vein thrombosis. Treatment with proton pump inhibitors (PPIs) in PSE with FEIBA was administered, resulting in very good clinical and biological improvement. The patient was discharged on 08/03/2023 with Innohep, with corticosteroids stopped and Endoxan started alone to try to eradicate the inhibitor. An ACC check was done on 05/04/2023, finding a TCA of 40/32' and ACC at 5 ui. Since then, the patient is still alive but refuses to undergo further checks.

Conclusion:

This case illustrates the diagnostic and therapeutic challenges posed by acquired hemophilia in elderly patients, particularly when it presents with severe clinical manifestations. Additional studies are necessary to better understand the pathophysiology and treatment of this condition in elderly individuals. The use of FVIII and bypassing agents can promote the propagation of an existing thrombosis through the TF pathway and independently of the TF pathway. unlike cases of venous thrombosis in congenital hemophilia where venous thrombosis often follows the administration of FVIII or aPCC, the occurrence of thrombosis in acquired hemophilia generally precedes the administration of hemostatic agents with no significant exacerbation of the existing thrombus reported.