Acquired hemophilia

A rare, spontaneous, and potentially deadly condition1

- Only 1 to 1.5 per million people are affected yearly2
- Characterized by an inhibitory antibody to FVIII that appears spontaneously3

Associated with severe and life-threatening bleeding4

- Up to 2% mortality rate

Often involving a delayed diagnosis

- 30% of patients go undiagnosed for more than 7 days5

Who "acquires" acquired hemophilia?

- Underlying conditions include autoimmune disorders, malignancies, dermatologic disorders, and pregnancy; however, in approximately 90% to 95% of patients, the cause is unknown6

Signs include:

- Isolated prolonged aPTT7
- Fever and soft tissue hemorrhage8
- Gastrointestinal, urological, retroperitoneal, or postpartum bleeding9
- Prolonged bleeding following surgery10

Patients usually present to physicians who are not specialists in the field and have not previously managed a case...

Lack of familiarity with the disorder may lead to delayed diagnosis and suboptimal treatment...

—Collins et al. J. Clin. Pathol. 2010

Spot it. Stop it.

Stop the bleed with the only bypassing agent FDA approved for acquired hemophilia11

Important Safety Information (cont’d)

Adverse Reactions

- The most common and serious adverse reactions in clinical trials are thrombotic events...

Warnings and Precautions

- Exercise caution when administering NovoSeven® RT in patients with an increased risk of thromboembolic complications, such as those with disseminated intravascular coagulation...

Important Safety Information

Model is used for illustrative purposes only.

Indications and Usage

NovoSeven® RT (Coagulation Factor VIIa [Recombinant]) is a coagulation factor indicated for:

- Treatment of bleeding episodes and peri-operative management in adults with acquired hemophilia A
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Please see additional Important Safety Information, including Boxed Warning, throughout.

References:

7.ALLERGIES TO BLOOD PRODUCTS: Patients who have a known allergy to human blood products should not be given NovoSeven® RT. NovoSeven® RT is a recombinant protein. Only patients allergic to bovine or hamster proteins should not be given NovoSeven® RT...
An acquired hemophilia case in which delayed diagnosis put a patient at risk

**CASE STUDY**

**The patient**

A 78-year-old woman with a swollen leg

**Initial presentation**

• Patient presented to her PCP with painful lower extremity swelling in one leg and shortness of breath.
• Echocardiogram was negative for DVT.
• Anticoagulation therapy with enoxaparin sodium injection was continued in light of the diagnosis of suspected DVT.
• Enoxaparin sodium injection was started to treat the patient for suspected DVT.
• Doppler ultrasound was negative for DVT.
• Anticoagulation therapy with enoxaparin sodium injection was continued in light of the diagnosis of suspected DVT.
• Patient developed bluish lower extremity swelling in one leg.

**Initial evaluation**

• Coagulation studies (PT/aPTT) were not performed during initial presentation.
• Coagulation test (PT/aPTT) were drawn after enoxaparin sodium injections were initiated.

**The delay**

• Prothrombin time was prolonged on initial presentation.
• Prolonged aPTT was noted when bleeding and prolonged aPTT did not improve with vitamin K.
• Enoxaparin sodium injection was stopped and the patient was given vitamin K.

**The diagnosis**

• Patient was transferred to a tertiary care hospital affiliated with the HTC.
• Repeat aPTT was 75 seconds, and the 1:1 aPTT mixing study showed an aPTT of 35 seconds with immediate mix and 85 seconds after a 2-hour incubation.
• The diagnosis of acquired hemophilia was considered.

**Management**

• Patient was diagnosed with acquired hemophilia.
• FVIII levels were <1% and inhibitor titer was 90 BUs.
• Repeat aPTT was 75 seconds, and the 1:1 aPTT mixing study showed an aPTT of 35 seconds with immediate mix and 85 seconds after a 2-hour incubation.
• Patient was transferred to a tertiary care hospital affiliated with the HTC.
• A PCP's lack of knowledge of the effects of enoxaparin sodium injection on PT/aPTT led to further misdiagnosis and improper treatment with vitamin K and heparin.
• Failure to properly diagnose acquired hemophilia in this case led to prolonged hospitalization and contraindicated anticoagulation treatment.

**For hematology/oncology specialists**

• Failure to properly diagnose acquired hemophilia in this case led to prolonged hospitalization and contraindicated anticoagulation treatment.
• The immediate consult from a hematologist/oncologist to a benign hematologist at the HTC supported the eventual diagnosis of acquired hemophilia.

**For nonspecialist HCPs**

• Failure to properly diagnose acquired hemophilia in this case led to prolonged hospitalization and contraindicated anticoagulation treatment.
• The immediate consult from a hematologist/oncologist to a benign hematologist at the HTC supported the eventual diagnosis of acquired hemophilia.

**For pharmacists**

• Use of multiple units of FFP could have served as a red flag to consider the diagnosis of acquired hemophilia.
• Communication between the pharmacist and health care team could support earlier diagnosis of acquired hemophilia.
• Failure to properly diagnose acquired hemophilia in this case led to prolonged hospitalization and contraindicated anticoagulation treatment.

**Key takeaway**

For hemostasis in acquired hemophilia:

- Patient started treatment with NovoSeven® RT (Coagulation Factor VIIa [Recombinant]) 90 mcg/kg every 2 hours for GI bleeding until hemostasis was achieved.
- Patient then began immunosuppressive treatment with rituximab 900 mg/m² over 4 hours, repeated every 2 months for 6 doses.
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