The Official Journal of the Italian Society for the Study of Hemostasis and Thrombosis
SISET
Società Italiana per lo Studio dell'Emostasi e della Trombosi
Corso Nazionale SISET
Bergamo | 23-25 ottobre 2025

29TH NATIONAL CONFERENCE OF THE ITALIAN SOCIETY FOR THE STUDY OF HEMOSTASIS AND THROMBOSIS, 2025

ABSTRACT N. PO67

PANCREATIC CANCER IN A 71-YEAR-OLD SEVERE HEMOPHILIA A PATIENT WITH INHIBITORS AND SUSTAINED ZERO BLEEDING DURING EMICIZUMAB PROPHYLAXIS.

M. Biglietto, A.L. Faccini, S. Sorella, E. Crisanti, M. Antonacci, A. Taglietti, M. Gherardini, A. Tirnetta, S. Ligia, R. Mormile, R. Ciciani, E. Baldacci, C. Santoro.

Hematology, Department of Translational and Precision Medicine Sapienza University of Roma. .

Background

The HAVEN 1 trial demonstrated the efficacy and safety of emicizumab prophylaxis in severe hemophilia A with inhibitors. However, the management of bleeding events and high-risk surgical procedures remains a clinical challenge.

Case report

In 2020, a 67-year-old man with severe hemophilia A (baseline FVIII activity 0.8%) and chronic HCV infection was referred to our center. He had developed a high-titer inhibitor during on-demand pdFVIII therapy; he never underwent immune tolerance induction (ITI) and therefore was managed with bypassing agents (e.g. rFVIIa). He also developed severe hemophilic arthropathy affecting all index joints, particularly the right knee, for which he underwent two arthroplasties in 2000–2001, complicated by chronic osteomyelitis and fistulization, without benefits in terms of joint mobility.

He was on-demand treatment with rFVIIa, mostly for recurrent fistula bleedings. In November 2016, due to a severe elbow hemarthrosis refractory to rFVIIa therapy, it was necessary to administer pdFVIII with a subsequent transient rise in the inhibitor level (57.2 BU).

In March 2020, during the pandemic, the patient was referred to our Center because of a vitreous hemorrhage occurred after a cataract surgery with rFVIIa prophylaxis; in that critical period it was impossible to perform a posterior vitrectomy and that lead to blindness in one eye. In March 2021, a severe bleeding episode from the fistula required FVIII replacement therapy, resulting in a significant and sustained rise in inhibitor levels (peak 422 BU), and the presence of multiple hemorrhages with an important decrease of hemoglobin levels.

Therefore, emicizumab prophylaxis was started (maintenance regimen with 3 mg/Kg every 2 weeks), resulting in a sustained zero-bleeding state for 3 years and 3 months. During this period the patient underwent 2 dental extractions with no need for any additional hemostatic therapy.

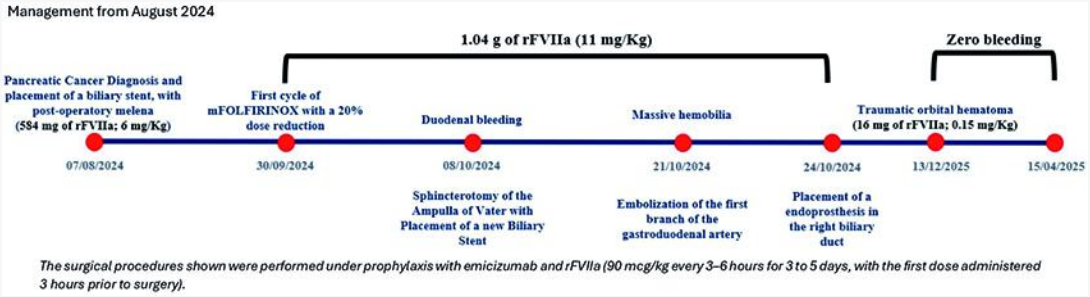
In August 2024, after developing jaundice, the patient was diagnosed with stage III pancreatic cancer. A diagnostic biopsy and a biliary stent placement were performed under rFVIIa prophylaxis (total dose: 64 mg). Nevertheless, he experienced postoperative melena, successfully managed with additional 520 mg of rFVIIa. In September 2024, a modified FOLFIRINOX regimen was started. After a week he developed duodenal bleeding and massive hemobilia, requiring multiple procedures: endoscopic sphincterotomy with common bile duct stenting, embolization of the gastroduodenal artery's first branch, and right biliary tract stenting. A total of 1.04 g of rFVIIa (11 mg/kg) was administered to control bleeding (Figure 1).

Therefore, systemic chemotherapy was discontinued in favor of palliative stereotactic radiotherapy. Then, no further cancer-related bleeding events occurred.

Conclusions

Emicizumab prophylaxis greatly improved the patient's quality of life, maintaining a zero-bleeding state for over three years. However, cancer-related bleeding remained a major challenge and we had to use a lot of rFVIIa to control bleeding on top of emicizumab prophylaxis. In conclusion, inhibitor patients still remain a difficult-to-treat subgroup especially concomitantly to comorbidities and surgeries. This concept is important not to exclude the chance of ITI in the life of our inhibitor's subjects.

Email: *m.biglietto97@gmail.com*


Management from August 2024
1.04 g of rFVIIa (11 mg/Kg)
Zero bleeding
Pancreatic Cancer Diagnosis and placement of a biliary stent, with post-operatory melena (584 mg of rFVIIa; 6 mg/Kg)
First cycle of mFOLFIRINOX with a 20% dose reduction
Duodenal bleeding
Massive hemobilia
Traumatic orbital hematoma (16 mg of rFVIIa; 0.15 mg/Kg)
07/08/2024
30/09/2024
08/10/2024
21/10/2024
24/10/2024
13/12/2025
15/04/2025
Sphincterotomy of the Ampulla of Vater with Placement of a new Biliary Stent
Embolization of the first branch of the gastroduodenal artery
Placement of a endoprosthesis in the right biliary duct
The surgical procedures shown were performed under prophylaxis with emicizumab and rFVIIa (90 mcg/kg every 3–6 hours for 3 to 5 days, with the first dose administered 3 hours prior to surgery).