Plasma Exchange and Continuous Infusion of Factor VIII for Life-Saving Surgery in Hemophilia A with High-Titer Inhibitor: Two case reports

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Background:

- Control of bleeding in surgery in hemophiliac patients with high-titer inhibitor is challenging.
- By-passing agents are limited in low-resources settings.
- Plasma exchange combined with continuous infusion of factor VIII concentrate is a treatment option.

Method:

- Report two cases of patients with hemophila A and high-titer inhibitor who underwent surgeries.
- They received - plasma exchange with cryo-removed plasma perioperatively.
  - FVIII conc.100 U/kg followed by continuous infusion rate 14 U/kg/hr for 7- 14 days.
  - rFVIIa, APCC and antifibrinolytic agent for breakthrough bleeding.

Results:

Figure 1

1st case: an 11-year-old boy

- no definite history of bleeding disorder and later diagnosed with hemophilia A with inhibitor
- bleeding from surgical wound with osteolytic lesions at the right foot (Figure 1)
- the highest FVIII:Ab titer > 660 BU (Figure 2)
- right below-knee amputation due to infection and uncontrolled bleeding
- 9 times of plasma exchange, 17 days of continuous FVIII infusion and 2doses of rFVIIa
- bleeding was stopped and FVIII: Ab decreased to 5.2 BU

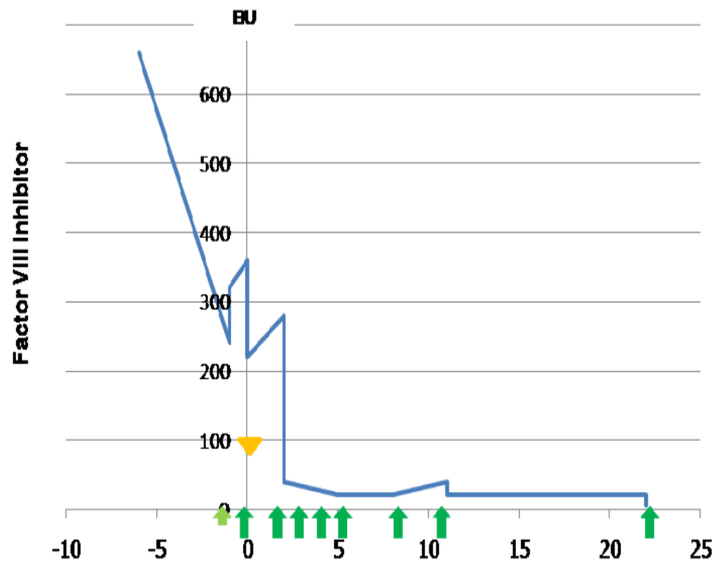

Figure2

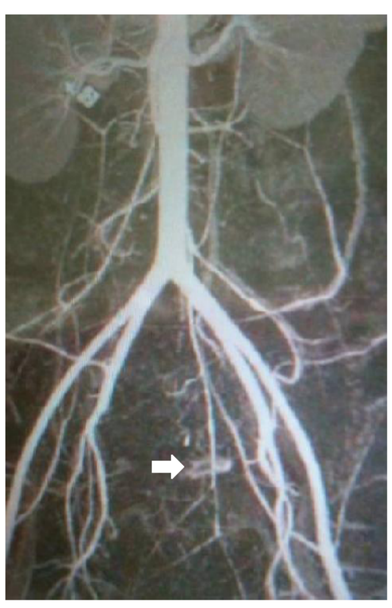

Figure3

2nd case: a 14-year-old boy

- hemophilia A and known high-titer inhibitor
- abdominal distension after a minor fall
- CT angiogram revealed pelvic blood clots and bleeding from sigmoidal artery (Figure 3)
- the highest FVIII: Ab titer was 4,400 BU (Figure 4)
- 1st surgery to relieve abdominal compartment syndrome and 2nd surgery for abdominal closure
- 6 times of plasma exchange, 7 days of continuous FVIII infusion, 5 doses of rFVIIa and 10 doses of APCC
- bleeding was stopped and FVIII: Ab decreased to 3,680 BU

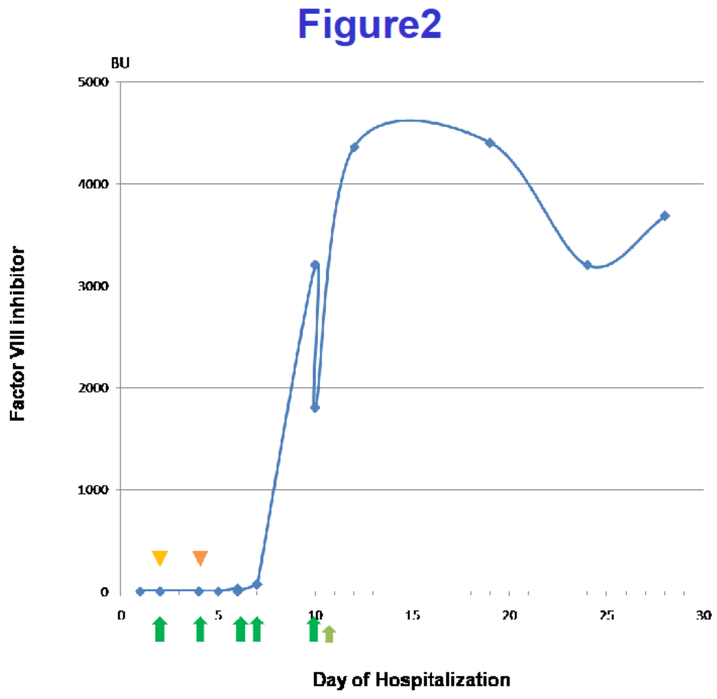

Figure 4

Plasma exchange 1.5-2 time of plasma volume

Plasma exchange 0.75-1 time of plasma volume

1st surgery 2nd surgery

Conclusion:

- Plasma exchange combined with continuous factor VIII infusion is an effective treatment of life-threatening bleeding in hemophiliac patients with high-titer inhibitor.

References:

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