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3.04 Acquired coagulation disorders

ACQUIRED HEMOPHILIA A IN LIVER CIRRHOSIS: BALANCING HEMOSTASIS BETWEEN HIGH HEMORRHAGIC AND THROMBOTIC RISKS

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Background. Acquired hemophilia A (AHA) is a rare, life-threatening bleeding disorder caused by autoantibodies against factor VIII (FVIII). Diagnosing AHA alongside pre-existing bleeding etiologies is challenging, as emerging symptoms are easily misattributed to baseline conditions, delaying treatment. Furthermore, liver cirrhosis induces alterations in pro- and anti-coagulant pathways, resulting in an unstable hemostatic equilibrium that maximizes risks for localized hemorrhage and systemic thrombosis.

Case Report. We report a 75-year-old male with Child-Pugh B7 alcoholic-metabolic cirrhosis complicated by portal hypertension (F2 blue esophageal varices, congestive gastropathy, mild thrombocytopenia) with multiple comorbidities (see table 1). Following seven months of recurrent bleeding episodes requiring 25 packed red blood cell (PRBC) units despite normal baseline coagulation times, he was readmitted with severe anemia (Hb 5.0 g/dL). His aPTT ratio was 1.52 upon admission and progressively lengthened; AHA was suspected when it reached 2.76, accompanied by extensive truncal ecchymoses and a large subscapular hematoma requiring near-daily transfusions (totaling 35 PRBC units). Assays confirmed FVIII deficiency (FVIII:C 4%) and a 5.0 Bethesda Unit inhibitor. Hemostasis was initiated with an activated prothrombin complex concentrate (aPCC, 50 U/kg q12h) and rapidly switched to recombinant porcine FVIII (rpFVIII, 50 U/kg q12h). Strict FVIII:C monitoring enabled safe dose tapering to 25 U/kg/day and successful discontinuation after three weeks; tranexamic acid (1 g q12h) was maintained until discharge. Immunosuppression began with prednisone (1 mg/kg/day); due to early steroid resistance, cyclophosphamide (50 mg/day for 5 weeks) was added, successfully eradicating the inhibitor. Transfusion requirements dropped after starting rpFVIII. Clinical improvement of ascites was observed. Treatment-related complications included severe glycemic flare, oral candidiasis, and intercostal Herpes Zoster reactivation. A temporary fibrinogen drop occurred post-hemostatic therapy (nadir 1.06 g/L), returning to baseline (3.10 g/L) at discharge. Long-term octreotide (10 mg/month) was introduced for intestinal angioectasias.

Conclusions. Pre-existing bleeding phenotype can confound AHA diagnosis, especially in liver cirrhosis where prolonged coagulation times can be overlooked. In this case, the sequential use of bypassing agents and monitored rpFVIII achieved safe hemostasis without inducing thrombotic events. In patients with tight thromboembolic and hemorrhagic thresholds, monitorable agents represent a crucial asset to navigate narrow therapeutic windows. Concomitant improvement in ascites may reflect the emerging role of FVIII in vascular permeability regulation.

Table 1. Baseline comorbidities and overlapping clinical risk profiles of the patient

Thrombosis risk	Child-Pugh B7 Cirrhosis	Thrombosis risk	Coronary Sclerosis
Bleeding risk	F2 Blue Esophageal Varices with red signs	Thrombosis risk	Active Smoking
Bleeding risk	Congestive Gastropathy	Thrombosis risk	Dyslipidemia
Bleeding risk	Chronic Thrombocytopenia	Thrombosis risk + Treatment complication risk	Type 2 Diabetes Mellitus
Bleeding risk	Chronic Multifactorial Anemia (with recorded Iron-Deficiency)	Thrombosis risk + Treatment complication risk	Hypertension
Bleeding risk	History of Gastric Ulcer & Hematemesis	Treatment complication risk	Severe Chronic Obstructive Pulmonary Disease
Bleeding risk	*Intestinal Angioectasia**	-	Polyneuropathy
-	*Severe Aortic Stenosis**	-	Subclinical Hypothyroidism
-	Colonic Diverticulosis	-	Mild Monoclonal Gammopathy of Undetermined Significance
-	Benign Prostatic Hyperplasia		

Key Legend: Thrombosis risk; Bleeding risk; Treatment complication risk

** Initial suspect of Heyde syndrome, but von Willebrand Factor levels above normal*