

TRAUMA MANAGEMENT IN SEVERE HEMOPHILIA A: A CASE REPORT

J. Di Paolo, R. Malizia, M. Luongo, P. Massaro, M. Di Mascio, S. Verna

Immunohematology Service, Asl 02 Abruzzo, Chieti, Italy

Background. Hemophilia A is an X-linked hereditary bleeding disorder caused by mutations in the factor VIII (FVIII) gene, resulting in deficiency or absence of FVIII, a key component of the intrinsic coagulation pathway. The prevalence of hemophilia A is estimated at approximately 1 in 5,000 male live births. According to residual FVIII activity, hemophilia A is classified as severe (<1%), moderate (1-5%), or mild (>5% to <40%). The bleeding phenotype generally correlates with disease severity. Trauma in patients with hemophilia represents a time-dependent hemostatic emergency, characterized by a high risk of major bleeding even in the presence of apparently moderate injuries. The World Federation of Hemophilia (WFH) guidelines recommend immediate administration of replacement therapy before completion of the diagnostic workup, particularly in cases of major trauma or suspected musculoskeletal or intracranial involvement. Management is especially challenging in patients with a history of inhibitors.

Case Report. We describe the case of a patient with severe hemophilia A (FVIII <1%) with a previous history of high-responder inhibitors, a target joint involving the right knee, and a current inhibitor titer of 2 BU, admitted to the Emer-

gency Department following a road traffic accident. The patient sustained right shoulder trauma with subluxation and multiple lacerations secondary to windshield glass injury. Given the high bleeding risk, replacement therapy with octocog alfa 8000 IU (calculated according to body weight, neutralizing dose, and incremental dose strategy), was promptly initiated, together with serial clinical, laboratory, and radiological monitoring. Imaging studies confirmed right shoulder subluxation without evidence of major bleeding or intracranial complications. Replacement treatment was continued for three consecutive days. The clinical course was favorable, with no bleeding progression or need for surgical intervention.

Conclusions. This case highlights the importance of early hemostatic treatment and a multidisciplinary approach in the management of trauma in patients with hemophilia, even in the presence of injuries initially considered moderate. A history of inhibitors represents an additional management challenge and requires close clinical and laboratory monitoring. Prompt intervention, in accordance with international recommendations, may help prevent severe hemorrhagic complications and improve clinical outcomes.