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Letter to Editor

All That Glitters is Not Gold: A Pregnant Woman with a Rare Inherited Bleeding Disorder

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Dear Editor

Inherited bleeding disorders can lead to a lifelong bleeding tendency. The prevalence of these disorders varies among different populations and is higher in areas that consanguinity or endogamy are common. The most common and potentially severe inherited bleeding disorder in humans is hemophilia. The main cause of bleeding in this X-linked disorder is deficiency of coagulant factor VIII or IX. Management of this disorder is basically prevention of bleedings by avoiding high-risk activities, receiving factor prophylaxis in defined situations and treatment of bleeding events with appropriate factors immediately following trauma or surgery.[1, 2]

Parahemophilia or Owren's disease or hereditary deficiency of factor V is a very rare disease. It is an autosomal recessive disorder and is estimated about 10 times more frequently in eastern countries such as Iran where consanguineous marriages are more common.[3] The estimated incidence of this disorder is less than 1 in 1 million.[3, 4]

A 34-year-old primiparous woman who had developed preeclampsia at 39th week of gestational age was admitted for cesarean section due to arrest of normal vaginal delivery. She mentioned that previously she was told to have hemophilia based on coagulation screening tests despite having any history of bleeding. However, initial evaluation of the patient before operation showed abnormal coagulation tests with prolonged PT and PTT. We asked to have the patient's medical ID card and accordingly, she was found to be a case of severe factor V deficiency. Finally, after infusion of 15 ml/Kg of Fresh Frozen Plasma (FFP), anesthesia was induced and operation was performed uneventfully.

Parahemophilia is a clinically well-known disorder that any fault in diagnosis or management could result in a serious complication.[5] The risk of peripartum bleeding is high in a pregnant patient with congenital factor V deficiency during anesthesia. The least levels of factor V necessary to avoid pregnancy loss and postpartum bleeding is reported to be ≥15 and 20 IU/dL, respectively.[6] Beside the primary interventions such as placement of a secure intravenous access, treatment is based on infusion of FFP or plasma-derived factor V concentrate.[7, 8] Recombinant factor VIIa can be substituted as an alternative; however, cryoprecipitate has no place in prevention or treatment of bleeding episodes in patients with parahemophilia.[9]

Remembering "all that glitters is not gold" and being fully informed about the exact type of the disorder in patients with abnormal bleeding tendency is of paramount importance in management of bleeding disorders.

Conflict of Interest: None declared.

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