Acute abdomen in a young girl with factor XIII deficiency: Perianesthetic issues

Sir,

Surgeries in women with coagulation pathway disorders need good teamwork and communication between gynecologist, hematologist and anesthesiologist. Congenital factor XIII deficiency is a rare disorder. We report the case of a 13-year-old girl with known congenital factor XIII deficiency who presented with acute abdomen, diagnosed as an ovarian hematoma and was posted for laparoscopic evacuation of the same.

The patient had a history of umbilical cord bleed in the first week after birth and thereafter multiple bruises and hematoma till one year of age. She was diagnosed as congenital factor XIII deficiency and was enrolled for follow up. Transfusions of
fresh frozen plasma (FFP) (15 ml/kg) monthly were initiated for which she reported irregularly due to which she suffered intracranial hemorrhage, pelvic hematoma, renal subcapsular hematoma and a subdural hemorrhage in the past which were managed conservatively with FFP transfusion.

Preoperatively her hemoglobin was 7.1 gm/dl, platelet count 1,60,000/ml and coagulation tests including clot solubility test were within normal limits. Our team of hematologists, gynecologists and anesthesiologists meticulously discussed and planned her operation. The hematologist advised preoperative coagulation profile and clot solubility test, which were normal. Infusion of two units of FFP and one unit of blood were advised a day prior to the operation. In the operation theatre, two 20G intravenous (IV) cannula could be secured on the dorsum of her wrist. Ultrasound machine was kept standby for guided central cannulation, if needed. Supplementation of analgesia by regional anesthesia was avoided to prevent spinal hematoma. After standard anesthesia induction, size three Proseal laryngeal mask airway was inserted smoothly. Oral intubation was avoided to prevent airway trauma and bleeding. Two liters of blood clot was evacuated from the pelvis for which two units of FFP and two units packed erythrocytes were transfused intraoperatively after securing hemostasis. Lactated Ringer’s solution was used as replacement fluid and starch/gelatin/dextran avoided for preventing further coagulopathy. Tranexamic acid 500 mg IV was injected and advised four times a day for the next two days. Neuromuscular blockade was reversed after gentle suctioning. Postoperatively intramuscular injections of non-steroidal anti-inflammatory drugs were avoided. Oral paracetamol one gram QID along with tramadol injections of non-steroidal anti-inflammatory drugs were avoided. Oral paracetamol one gram QID along with tramadol 50 mg IV for rescue analgesia was advised. Postoperatively, she was transfused another two units of blood. In the recovery ward she complained of severe headache and pain abdomen. However, computerized tomography scan and ultrasound abdomen revealed normal study. Both these subsided on its own in a week. She was discharged after 15 days after prescribing oral contraceptives to prevent ovulation.

Factor XIII deficiency is a rare congenital coagulation abnormality of which only 200 cases were reported till 2002. It could be congenital or acquired. Congenital factor XIII deficiency, originally recognized by Duckert in 1960, is a rare autosomal recessive disease usually associated with a severe bleeding diathesis with M: F of 1:1. The incidence of factor XIII deficiency is about one case per 2.5 million populations. Acquired factor XIII deficiency has been described with hepatic failure, inflammatory bowel disease, and myeloid leukemia.

Factor XIII is a plasma transglutaminase that catalyzes the final step in the coagulation cascade, cross-linking the loose fibrin polymer into a highly organized structure. In addition, factor XIII covalently binds fibronectin, α2-plasmin inhibitor, and other molecules to the fibrin plug; this enhances adherence to the wound site, resistance to fibrinolysis, and wound healing.

Standard hemostatic screening tests are normal and deficiency is diagnosed by clot solubility test.[1] Plasma, cryoprecipitate, and factor XIII concentrates have been used for replacement of factor XIII and the treatment of bleeding. Severe bleeding episodes or preoperatively; a dose of 50-70 U/kg IV is required.

Not many cases have been reported in literature. Most cases were diagnosed during surgery when the patient bled more than expected without any apparent cause and coagulation studies and platelet value were normal.[2-4] A case series from Japan has reported successful pregnancies in 8 women with judicial replacement with FFP and concentrates.[5] During the latter part of pregnancy, patients transfused factor XIII almost every week and kept the level higher than 25-30% during labor. Even if the abnormality is known beforehand, the bleeding is difficult to control sometimes and can result in mortality.[6] Factor XIII also plays a role in stabilization of wounds.

We are reporting successful outcome of our patient with factor XIII deficiency post surgery. This was made possible by careful teamwork and communication between hematologists, gynecologist and anesthesiologists.

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