

Patient Posted for Laparoscopic Appendectomy With von Willebrands Disease.

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Summary

This is a case of acute appendicitis that was diagnosed in a patient of pediatric age group who was also a known case of Von willebrand disease since birth. The patient was posted for lap appendectomy with packed red blood cells and fresh frozen plasma on standby. Intraoperative events were uneventful and postoperatively she was transferred to the pediatric intensive care unit for further follow up. The patient was given von willebrand factors to address the continuous post op bleeding.

Background -

The following case report helps in dealing with patients belonging to the pediatric age group with inherited bleeding disorders who are posted for surgery and their management intraoperatively and post operatively and the role of an anaesthetist in the same.

Case Presentation –

The patient was diagnosed with VD at the age of 5 years following history of recurrent epistaxis. and easy bruisability since 3 years of age. She was treated at the time with Inj Von Willebrand factor 230mg IV BD, plasma products and 2 units PRBC. She attained menarche at 12 years of age causing severe menorrhagia with bleeding that lasted for 15 days with the use of 4-5 pads per day. She was started on T. Tranexemic acid and oral contraceptive pills.

She presented with complaints of abdominal pain for the past 1 day, ultrasound abdomen was done which showed evidence of acute appendicitis. The patient was planned to be posted for laparoscopic appendectomy.

Investigations -

All routine blood investigations done - Hemoglobin – 8.3, Platelet – 3.76 lakhs, Total count – 7000, Sodium – 139. Potassium – 3.3 , Chloride – 107, Bicarbonate – 23.8, PT – 12.6, INR – 1.09. APTT – 66.1 .PRBC and FFP reserved prior to surgery.

Treatment -

Inside the OR ,after administration of Desmopressin nasal spray 10mcg in each nostril. Routine monitors were connected, 18G venflon was secured on right upper limb. Basal heart rate - 70bpm, blood pressure - 120/70 mmHg. Patient in supine position, premedicated with Inj. Glycopyrolate 0.2mg, Inj. Midazolam 1mg, induced with Inj. Propofol 120mg, Inj. Fentanyl 100mcg and Inj. Atracurium 30mg. Preoxygenated for 3 mins. Patient was intubated using size 3 McIntosh Laryngoscope with 6.5 size cuffed endotracheal tube. Intra operatively bleeding of 80ml was seen. 50ml of platelets transfused and Inj. Tranexemic acid 100mg in 100ml NS started. Inj. Dexamethasone 8mg IV along with Inj. Paracetamol 1g IV given. Duration of surgery - 1 hour 30 mins, following which patient was extubated after adequate reversal. Patient was shifted to PICU. Patient was found to have significant drain collection post-operatively for which she was transfused everyday. Inj. Von Willebrand Factor was started. POD 3 – No collection in drain. Patient shifted to ward and discharged.

Discussion –

If the VWD is suspected diagnostic tests should include an APTT, BT, factor VIII: C Ristocetin cofactor and vWF antigen. Additional testing of ristocetin induced platelet adhesion (RIPA) the multimeric structure and collagen binding test and genanalysis allow diagnosing the different types of Von Willebrand Disease.

VWD should always be considered in the assessment of children suspected of non-accidental injury, the risk for increased bleeding should be kept in mind when elective and medical procedures are undertaken in this hemostasis disorder.