“Swimming in Dangerous Waters”

The Case Of A Paturient With Platelet Storage Pool Disorder

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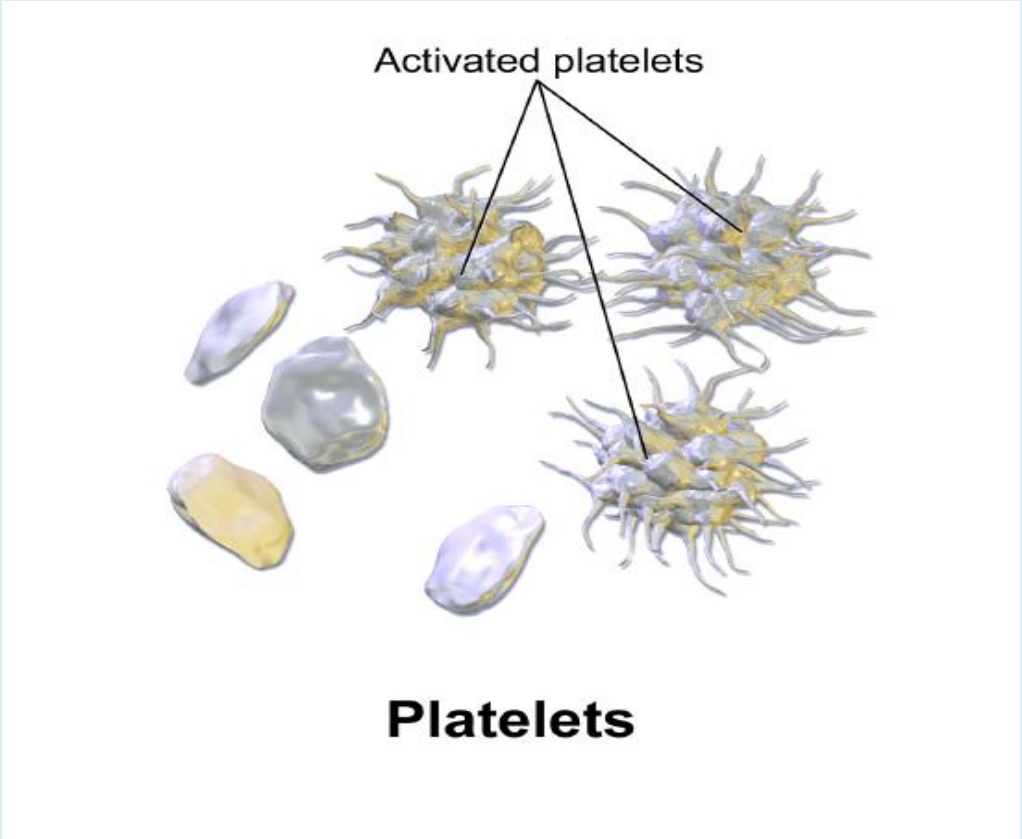
INTRODUCTION

- Platelet Storage Pool Disorders (PSPDs) are a rare group of bleeding disorders that result from either a qualitative or quantitative deficiency in platelet granules (alpha, dense or both)
- They can be inherited (by a variety of patterns) or acquired through disease processes such as systemic lupus erythematosus and cardiopulmonary bypass (1).
- The abnormality in the platelet granules results in a deficiency in platelet aggregation
- Cases of PSPD in parturients are scarce in the literature
- Despite the significant implications to mother and child, there remains no consensus on management of these patients



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CASE REPORT

- 26 yo female G3P0 at 35w5d presented to the MetroHealth Medical Center due to a 1 week history of intractable headaches, associated vision changes, elevated blood pressures and decreased fetal movement
- IOL planned due to diagnosis of preeclampsia with severe features
- History significant for platelet storage pool disorder, diagnosed at age 9 after sibling was diagnosed
- Additional history of gestational diabetes, asthma and hypothyroidism
- Planning included interdisciplinary team discussions including anesthesiology, obstetrics and hematology
- Management included:
  - Baseline rotational thromboelastogram (ROTEM), which was normal
  - 1g tranexamic acid IV 1hr prior to IOL followed by 650mg po q8h x 5 days
  - DDAVP 0.3mcg/kg IV 30min prior to interventions (epidural, stage 2 of labor)
  - 1 unit platelets cross matched
  - 2 units packed red blood cells cross matched
  - IV magnesium sulphate for prevention of eclampsia
- Neuraxial ultrasound done and an experienced member of team performed epidural placement (after DDAVP)
- Successful vaginal delivery (after DDAVP)
- Estimated blood loss 200ml
- 600mg Misoprostol PR given prophylactically

DISCUSSION

- Management of parturients with PSPD should include multidisciplinary approaches including hematology, anesthesiology and obstetrics and ideally a clear plan before arrival to hospital (2,5)
- Hematology specific recommendations in literature include thromboelastograms (TEG), HLA-matched platelets to avoid alloimmunization, adjuncts including fibrinolytics, DDAVP and optimization of coagulation factors (3,4,5). In this case, TEG was normal but case reports have shown TEG used to guide the need for platelet transfusion (2)
- Management should also include reduction of risk of bleeding and trauma to mother and fetus (since these conditions are often hereditary) – avoidance of prolonged labor, intrauterine catheters, fetal scalp electrodes, vacuum, forceps and external cephalic version, as well as reduction of chorioamnionitis risk are a few examples. Case reports have documented successful, uncomplicated vaginal and cesarean deliveries.
- Different forms of PSPD are associated with different inheritance patterns so not all patients will have a family history (2). It should be a differential for any patient presenting with unexplained post partum hemorrhage, menorrhagia, mucosal bleeding. One paper estimates incidence of inherited platelet disorders as high as 30% in those referred to hematology for menstrual bleeding (5)
- Preeclampsia management was a nuance in this case. DDAVP use in those with preeclampsia is controversial due to its antidiuretic effect and risk of elevating blood pressures. Our patient received it safely after careful consideration

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TEACHING POINTS

- Platelet storage pool disorders (PSPDs) can be effectively managed in parturients
- PSPDs are characterized by a deficiency in platelet aggregation which increases bleeding risk
- Management in parturients should focus on a multidisciplinary approach including
  - Anesthesiology
  - Hematology
  - Obstetrics
- We postulate that appropriate management of these patients also includes:
  - Thromboelastography to guide need for platelet transfusion
  - Having available cross matched blood products
  - Use of antifibrinolytic agents
  - Use of desmopressin prior to procedural interventions
  - Neuraxial ultrasound to reduce trauma associated with epidural placement
  - Avoidance of trauma, where possible, to mother and baby
- With appropriate care, mothers can be safely delivered vaginally or via cesarean section
- PSPD can be managed safely even in parturients with preeclampsia

Image Credit: Pregnancy Maternity Motherhood– Free photo on Pixabay

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