

Platelet storage pool disease case discussions

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Case I

- 29 yo WF presented for evaluation of possible platelet storage pool disease, given her long standing bruising and her 4-yo son was diagnosed with platelet storage pool disease
- ISTH BAT score was 8
 - Bruises >1 cm
 - Minor wounds take >10 min to stop
 - Gum bleeding daily
 - Prolonged bleeding after dental procedure which did not require intervention
 - C-section x2, no intervention prior or post, but thinks she “bled more than expected”
 - Bled > 6 weeks post partum
 - Tonsillectomy as a child, could not remember if there was bleeding
 - Menorrhagia: monthly menstrual periods, each lasting >7 days, all days were heavy and needed to change hourly. Had to leave work occasionally because of it

Case I

- Son presented with significant bleeding symptoms (covered in blood from head to toe) after a small scalp cut from trauma
- heel surgery prior to diagnosis, had excessive bleeding within the cast. No intervention was done and bleeding stopped.
- used to be "a walking bruise" when he started to crawl
- Son was treated with stimate prn and "some medications" but not transfusion prior to recent tendon transfer surgery without bleeding

Labs

- Normal CBC with plt count of 312K
- INR 1.0, PT 12.7, PTT 27
- PFA-100 normal
- vWF Ag 107%, activity 107%, factor VIII 153%
- Plt agg:
 - Normal response to 10 uM ADP. Adequate primary and secondary aggregation followed by mild disaggregation to 2.5 uM ADP. Diminished primary aggregation and absent secondary aggregation to 1.0 uM ADP. The abnormal findings are overall mild and nonspecific and could represent normal variations. However, the possibility of an underlying platelet storage pool disorder/release defect cannot be excluded. Clinical correlation is recommended.
- Plt EM:
 - Platelets (plt) acquired from peripheral blood have an average of 2.70 delta granules (dg) per platelet, which is consistent with a delta granule storage pool deficiency. (Normal 4-6 dg/plt)

Case I continue...

- Shortly after diagnosis, she became pregnant
- She is now followed by high risk OB, 24 weeks gestation and is doing well other than daily gum bleeding managed conservatively
- She has had two C-sections previously and is planning on having another C-section
- Peri-partum management
 - Spinal anesthesia or general?
 - Platelet transfusion?
 - DDAVP?
 - Anti-fibrinolytics?

Case

- 57 yo WF c a prior diagnosis of VWD unknown type, presented for pre-op evaluation.
- Pt has severe aortic stenosis, increasing symptoms of chest pain and syncope, plans for an aortic valve replacement
- Since diagnosis of VWD in 2000, she has used DDAVP 3 sprays each nostril 30-60 min prior to the following procedures

Case II

- ISTH BAT score of 12
 - Frequent nose bleeding requiring hospitalization and cauterization previously
 - Large ecchymosis with minimal trauma, >5cm
 - Minor trauma led to bleeding > 10 min
 - At age 18. wisdom teeth removal lead to prolonged bleeding, not requiring other intervention
- Surgery:
 - 1972 R wrist cyst removal- no bleeding
 - 1980 appendix and GB surgery - no bleeding
 - 1990 laparoscopy for endometriosis- no bleeding
 - 2000, diagnosed with vWD, given stimate prior to the following procedures
 - 2002 L overy removal - **bleeding** requiring one night of hospital stay. No transfusion
 - 2004 L Knee scope- no bleeding
 - 2004, 2006 L heart cath- no bleeding
 - 2006 R Foot resection for osteomyelitis - no bleeding
 - Jan 2014 L heart cath- **retroperitoneal bleeding** needing 3 units pRRBC. No use of Humate P

Labs

- Normal CBC with plt count of 162K
- INR 1.0, PT 12.8, PTT 32
- PFA-100 significantly prolonged with epi >300 (ref 73-172), ADP 180(ref 53-111)
- vWF Ag 131%, activity 111%, factor VIII 150%
- Plt agg:
 - This study demonstrates a decreased secondary wave of aggregation to 1.0 and 2.5 micromolar ADP. This abnormality is overcome by the 10 micromolar dosage of ADP with complete aggregation. There is delayed but ultimately complete aggregation with epinephrine stimulation. Collagen and arachidonic acid agonism demonstrates adequate aggregation. The results are not pathognomonic for a particular platelet defect. The differential diagnosis includes abnormalities of the ADP receptor P2Y₁₂, signal transduction disorders, and storage pool disorders. Notably, the platelet morphology appears unremarkable on peripheral blood smear review. Specialized testing may be necessary for further classification, and correlation with clinical findings, including history of current prescription drugs, herbal therapies, and nutritional supplements is suggested.
- Plt EM:
 - Platelets (plt) acquired from peripheral blood have an average of 1.13 delta granules (dg) per platelet, which is consistent with a delta granule storage pool deficiency. (Normal 4-6 dg/plt)

Case II continued...

- Tried DDAVP challenge, but pt was admitted with chest pain 24 hours later, no trop elevation. Chest pain was attributed to her aortic stenosis.
- DDAVP was deemed unsafe for her and any further use was not recommended
- She was given 2 units of platelet transfusion pre/intra-op
- Two units of platelet on POD #1 and 2 each (POD#2 transfusion was given due to drop in Hgb by 2 grams)
- No further transfusion, no clinical bleeding H/H was stable.

Things to consider

- Patients who were diagnosed with platelet storage pool disease present with a variety of bleeding symptoms
- Should recommendations for peri-op planning based on their prior bleeding symptoms
- Are we over diagnosing or overtreating this disease?
- Do patients with platelet storage pool disease need sport precaution?