Factor XI Deficiency and Postpartum Hemorrhage After Necessary Precautions

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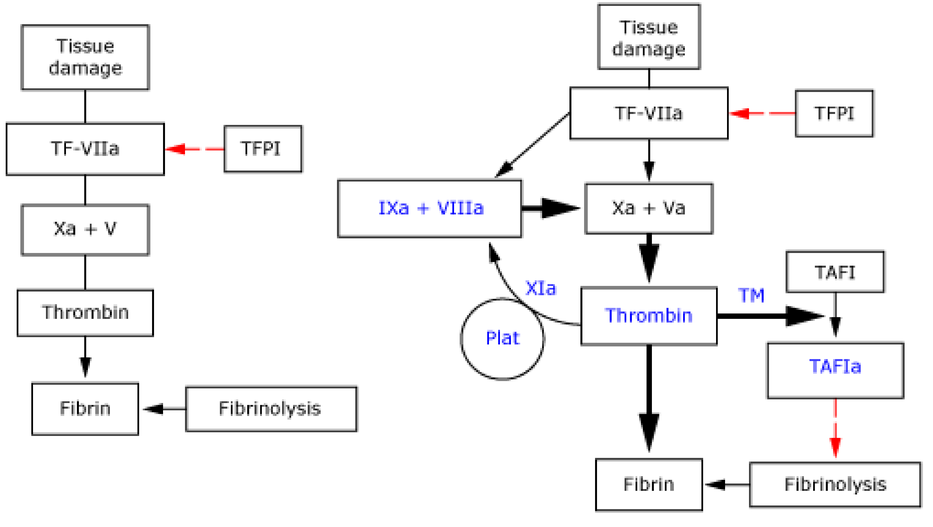
- Background
  - Rosenthal syndrome or Hemophilia C affects 1 in 1 million patients worldwide
  - Pregnant patients with Factor XI deficiency have an increased incidence of bleeding of about 19%
  - Difficult to predict the extent of bleeding, because each patient's presentation is vastly different and sometimes absent
- Case presentation
  - 39-year-old G1P0 38 weeks pregnant dated by Invitro fertilization with known factor XI deficiency(discovered during IVF workup)
  - Presented after a 3 day failed induction from an outside hospital desiring vaginal birth
  - The patient was found to be negative for any bleeding history, but factor XI activity was 28% (normal >50%)
  - Hematology recommended 3 units of fresh frozen plasma,1g IV Tranexamic acid before Neuraxial and continue TXA 1G QID for 3 days after vaginal delivery and 5 days for C-section

- Arrest of labor after 20 Hrs
- Before C/S 7U of FFP and 1 G TXA was given. Single dose spinal was successfully performed for anesthesia.
- The C-section preceded uneventfully with blood loss of 760mL.
- Once on the unit it was discovered there was continued bleeding
- Bedside D&C discovered 2 liters of blood loss and a BP of 80s/50s
- Taken back to the OR and a Bakri balloon was placed under general Anesthesia. Labs drawn at that time demonstrated Factor XI activity of 43% and PLT of 140.
- In total the patient received 12 units of fresh frozen plasma, 3 units of red blood cells, 2 units of platelets, 1 gram of TXA before and after C/S and then continued 4 times a day for 5 days, 3 doses of 30 mg of Pitocin, and 2 doses of methergine.



Initiation of Coagulation **Maintenance of Coagulation**

Reference "Factor XI Deficiency." *UpToDate*, https://www.uptodate.com/contents/factor-xi-eleven-deficiency?search=factor+xi+in+pregancny&source=search_result&selectedTitle=1~82&usage_type=default&display_rank=1.

Day	Factor XI activity	PT	Hgb	PLT	What was done
1	28%	143	12.6	166	7U FFP, 1g TXA pre and post op
C/S					
2 0200	43%		7.1	140	1 FFP, 1 RBC, Methergine, Pitocin
2 1830			6.8	123	2 RBC, 2 FFP
3	23%		8.3	95	2 FFP, 2 PLT
4			7.6	143	

Learning Points

- Our patient may have experienced a catastrophic outcome if it was not for continued vigilance from staff.
- Factory XI concentrates are not FDA approved to be used. (*Hemoleven*)
- At this time we rely on FFP to resuscitate our patients with factor XI deficiency and continue to treat a patient's needs individually.
- What would we change:
  - The correct dose of FFP and TXA was administered before procedure
    - Amount of factor XI in plasma is equivalent to 1 unit/mL. Thus, a dose of Fresh Frozen Plasma (FFP given at a dose of 10 to 20 mL/kg is usually sufficient to raise the factor XI level to 10 to 20 percent above the baseline level.)
  - Should we have gone back to the OR sooner for possibly better tone?
  - If Hemoleven was available would we have used it?