Perioperative Management of Plasminogen Activator Inhibitor 1 Deficiency in the Parturient – A Case Report

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Background

- Plasminogen Activator Inhibitor 1 (PAI-1) deficiency: a rare, autosomal-recessive disorder characterized by mild-to-moderate bleeding associated with injuries, trauma or surgeries due to the unopposed fibrinolysis.[1]
- Females are diagnosed with this condition more frequently than males, primarily because they experience bleeding episodes earlier in life (e.g. menorrhagia or postpartum hemorrhage).
- Management of PAI-1 deficiency in the parturient patient is currently anecdotal.[2] This case report details pre- and intra-operative management of a parturient patient with a known PAI-1 deficiency who underwent cesarean section under general anesthesia.

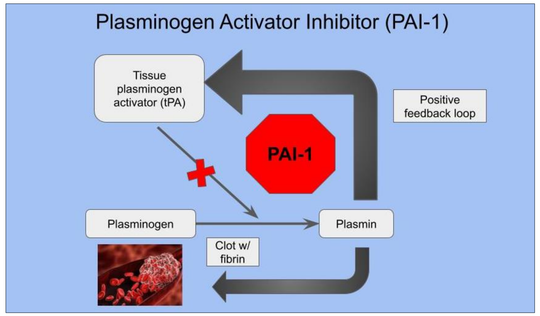

Figure 1. Role of PA-1 in Fibrinolytic System

Methods

This case was reviewed retrospectively and the evaluation of current pertinent literature was completed.

The patient consented to the creation and presentation of this case report. Given that the report does not include identifiable patient information, it is exempt from IRB review in accordance with Hartford Healthcare policy.

Case Presentation

- 33 year-old woman (G1P0) with planned cesarean section delivery with known PAI-1 deficiency.
- Past Medical Hx: obstructive sleep apnea, menorrhagia, and iron deficiency anemia
- Surgical Hx: Tonsillectomy (14yo) and Wisdom teeth extraction (16yo)
- PAI Deficiency Diagnosis: 14 years old following tonsillectomy with significant bleeding
- Family history: PAI-1 deficiency in sister (hx of significant blood loss following cesarean delivery)
- Patient presented to labor and delivery for a planned cesarean section with a mostly uncomplicated pregnancy with the only intervention being occasional iron infusions for low ferritin levels and mild iron deficiency anemia.

Case Management

At Admission:	• 2 units fresh frozen plasma • 4 units packed red blood cells made available for use as necessary
Labs:	• Hematocrit 38.4, platelets 242, sodium 135, potassium 3.7, BUN 6, Cr 0.48, Prothrombin time 10.3, INR 0.9, Partial Thromboplastin Time 29, Fibrinogen 527, CMP and thrombo-elastography.
Patient Education:	• Communicated neuraxial anesthesia risks and contraindications due to high bleeding risk and possible epidural hematoma; advised toward general anesthesia.
Intraoperative Management:	• Rapid sequence induction (fentanyl, propofol, succinylcholine) then indirect/video laryngoscopy and intubation; anesthesia maintenance with sevoflurane. • Delivery of viable baby (APGAR 8 & 9). After cord clamping, patient received 1g tranexamic acid (TXA) over 20 minutes followed by continuous infusion of 1g over 8 hours. Total intraoperative TXA administered was 1120mg. • After placental delivery, 10 units of oxytocin was administered over 10 minutes, then 5 units/hr for rest of procedure. Intrauterine oxytocin was administered during uterine closure to improve uterine contraction. • Before extubation, ultrasound-guided bilateral rectus sheath, ilioinguinal, and iliohypgastric single injection nerve blocks were administered for pain relief using 100mL of 0.2% bupivacaine with 0.3 mL of epinephrine 1:400,000, and dexamethasone 5mg. • Total of 750mL crystalloid (lactated ringers) was given, quantitative blood loss was 615mL as expected for C-section, and urine output was 150mL.
Postpartum:	• Administered PO TXA 1300 mg TID over 10 days; no significant bleeding or hemorrhage throughout hospital stay and patient was discharged on day 4 after surgery.

Discussion

The fibrinolytic system balances activation of plasminogen and fibrin degradation, both are controlled by various factors. PAI-1 plays an important role in regulating fibrinolysis by inhibiting the release of free tissue-type plasminogen activator (tPA), hindering the breakdown of fibrin (**Figure 1**).[3]

PAI-1 deficiency is a potentially life-threatening congenital disorder often first identified during surgery. In our case, the use of prophylactic TXA combined with FFP led to effective hemostasis, resulting in procedural blood loss within the anticipated range

Early detection of PA-1 deficiency and subsequent comprehensive perioperative management are crucial for ensuring the safety of an expectant mother undergoing a cesarean section.

References

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