A Parturient with TTP Complicated with Severe Pre-eclampsia

Aseem Kumar, M.D.; Yunping Li, M.D.
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Introduction: Thrombotic Thrombocytopenic Purpura (TTP) has been classically defined as a pentad of fever, thrombocytopenia, microangiopathic hemolytic anemia, neurological signs and renal failure. [1] Most cases are caused by either congenital or acquired deficiency of vWF-cleaving protease known as ADAMTS 13. The survival of patients with TTP has improved to 78% with plasma exchange making it the treatment of choice. [2]

Case Report: A 30-year-old woman, G2P1, presented at 24-week gestation for peripartum management of severe pre-eclampsia, anemia and severe thrombocytopenia associated with new onset neurological deficits.

Extensive laboratory workup showed diminished ADAMTS 13 activity and elevated ADAMTS 13 inhibitor level consistent with TTP requiring initiation of daily plasma exchange. Her brain MRI demonstrated multiple chronic infarctions, suggestive of thromboembolic etiology. Fetal management included daily Doppler assessment of umbilical artery blood flows and steroids to facilitate lung maturity.

On hospital day 10, a precipitous drop in platelet count along with reversal of umbilical diastolic blood flow prompted an emergent c-section by the care team.

She underwent rapid sequence induction of general anesthesia with left uterine displacement. A 520-gm neonate was delivered with apgars of 1 and 5 and subsequently transferred to NICU. Surgical microvascular bleeding was marked and required administration of PRBCs, FFP and Platelets.

The patient underwent multiple plasma exchange therapies postoperatively along with corticosteroids. Her platelet count stabilized on postoperative day (POD) 5. She was discharged home on POD 14 in stable condition without any long term sequelae.

Discussion: Our patient was unique in the fact that her TTP was further complicated with severe pre-eclampsia making her management quite challenging.

Plasma exchange was helpful in improving her platelet count by providing replacement of ADAMTS 13, the enzyme deficient in TTP. The challenges for anesthetic management included: severe preeclamptic patient with mental status changes, unknown fluid status immediately after plasma pheresis, and the controversy over platelet transfusion in TTP. Although transfusion of platelet products seems counterintuitive, a systematic review of case reports and series [3] did not document any risks.

References: