"Atypical Hemolytic Syndrome in the Context of Pregnancy"

Atypical HUS is often associated with hypertension.

Differentiated by presenting postpartum without improvement following delivery.

>50% end up with ESRD or dialysis within 6 months.

TMA (thrombotic microangiopathic anemia) is the formation of thrombi in the microvasculature leading to hemolysis and, eventually, organ failure.
--Ex: HELLP, PreE, TTP, Shigatoxin-HUS, atypical HUS (aHUS)

Pregnancy is a complement-stimulating condition.

10 - 12% (possibly much higher) of patients with HELLP syndrome and 8% of patients with preE have compliment genetic abnormalities.

aHUS definition: thrombocytopenia (<150k) with microangiopathic hemolysis (schistocytes, elevated LDH, decreased haptoglobin, and/or anemia), AND end organ damage.

PreE/HELLP vs aHUS
--aHUS with variable HTN, but can be severe
--aHUS with LDH > 1.5 x upper normal limit, usually > 1000
--Send ADAMS TS13 if LDH high and PLT < 70k to investigate for TTP
--aHUS is marked by persistent signs of TMA postpartum

LDH is sensitive for hemolysis, but not specific. Though, still of value given its indication of some pathology. Low haptoglobin as well indicates RBC hemolysis.

Complement panels should be sent after diagnosis, but are expensive ($3700).

Recurrent HELLP is suspicious for genetic mutation in the complement pathway.

Incidence of aHUS 1-2/10,000 in the postpartum population. (MUCH more common than incidence in general adult population)

Treatment of aHUS is compliment blocker Eculizumab (Soliris), although it does cause immunosuppression by its mechanism of action.