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Postpartum thrombotic microangiopathy - A diagnostic dilemma

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Background

During pregnancy, thrombotic microangiopathy is linked with HELLP syndrome, thrombotic thrombocytopenic purpura, and atypical hemolytic uremic syndrome (aHUS). Achieving a conclusive diagnosis is tricky but critical since management strategies and prognosis will differ. While ADAMTS13 test, biopsies and other laboratory tests can assist in diagnosis, they are time intensive and not easily accessible. We present a case in this report that exemplifies the diagnostic and treatment challenges associated with this illnesses.

Objectives

- To be acquainted with the spectrum of TMA during pregnancy and postpartum.
- To report a case of postpartum aHUS
- To be able to identify the early symptoms of TMA and to establish the diagnosis of aHUS.

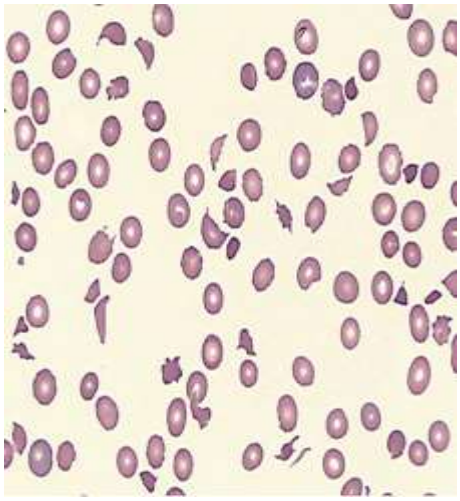
Case History

- A 28 year female G2P1L1 with 28 WOG presented with abdominal pain bleeding per vagina, reduced fetal movements. Patient had tachycardia, elevated BP, tender abdomen and other systemic examination were fairly normal.
- Routine lab investigations at admission showed low Hb while other investigations were normal.
- Patient was diagnosed with PIH with abruption and started on labetalol and emergency LSCS was done and extracted a stillborn
- Patient initially was managed with antibiotics and blood transfusion.
- Despite multiple blood transfusions and patient had severe anemia with thrombocytopenia, raised Creatinine (eGFR 55) mildly deranged LFT, peripheral smear showing MAHA with schistocytes, urine routine showing proteinuria (3+), elevated LDH
- Followed by progressive worsening anemia and thrombocytopenia in the absence of bleeding manifestations.

INVESTIGATION	DAY 1	DAY 2	DAY 3
Hb(g%)	8.5	5.8	7.8
Plt Count(cu/mm)	2.38	45000	1.38
Total Count(cu/mm)	15670	9030	8000
PT/Aptt	Normal	Normal	Normal
LDH	-	1756	497
Blood urea(mg/dl)	20	38	18
Creatinine(mg/dl)	0.7	1.3	0.8
Bilirubin	0.9	1.4	0.8
AST & ALT	39/8	83/32	42/21

Differential Diagnosis

- HELLP
- TTP
- Ahus



Management

We excluded TTP and HELLP syndrome as possible causes of the postpartum microangiopathic hemolytic anemia (MAHA). This decision was based on the history, clinical presentation, and laboratory findings. aHUS was retained as the final diagnosis, and in the absence of anti-complement therapy, the patient underwent three sessions of hemodialysis which improved the blood parameters. Patient was stabilized and discharged.

DISCUSSION

Clinical features	HELLP	TTP	aHUS
Time of onset	3T	2T/3T	Post partum
Hypertension	80 – 100%	-/+	+
AKI	Mild/Moderate	Mild/Moderate	Severe
Renal prognosis	Recovery	Fair	76% ESRD
CNS Findings	+	Dominant	-
Purpura with bleeding	-	++	-
Thrombocytopenia	+	++	++
Coagulopathy	-/+	-	-
Liver transaminases	++	-	-
LDH	+	++	++
ADAMTS13 <10%	-	++	+
Alternate complement pathway	-/+	-	++
Management	Supportive/Delivery	Plasma exchange	Plasma exchange

Key points

- Worsening clinical signs of gestational hypertension pre-eclampsia, aggravation of anemia, thrombocytopenia and renal abnormalities after delivery, points to TTP and aHUS.
- 50-76% of aHUS develop ESRD and overall mortality rate of 25% in acute phase.
- Since the basis of aHUS is genetic, even after complete remission, relapses are always possible.

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