Refractory Thrombotic Thrombocytopenic Purpura in Pregnancy: A Case Report

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Introduction

Thrombotic thrombocytopenic purpura (TTP) is a rare, life-threatening form of acute thrombotic microangiopathy which can be triggered by pregnancy (1). Untreated, it has a high mortality rate and is associated with placental ischaemia and pregnancy loss(2).

Case Report

A 36 year-old primigravida presented at 27 +2 weeks gestation with a three day history of a frontal headache and oedema. She had a past medical history of asthma, polycystic ovaries and high BMI (38).

She had normal observations and an unremarkable clinical examination. She was thrombocytopenic (platelet count 29 x10^9/L) and anaemic (haemoglobin 81 g/L), with normal liver function tests (ALT 15 AST 28), except for a raised bilirubin at 31. She had a normal CT head. A blood film showed red cell fragmentation. ADAMTS13 level at presentation was <0.2%, with a reticulocyte count of 222 and a diagnosis of TTP was made by haematology.

The patient was started on plasma exchange within 24 hours of admission, and received prednisolone (1mg/kg), folic acid and omeprazole. She also received betamethasone and magnesium in the foetal interest. The obstetric team undertook daily uterine artery dopplers.

The baby was delivered by caesarean section at a gestation of 28 +1 /40. This was due to risk to foetus from placental thrombosis, and to facilitate maternal treatment with agents contraindicated in pregnancy. She had two units of red cells transfused and was cross matched 4 units of red cells and 2 units of platelets. She had cell salvage and a GamCath high flow cannula in situ. The caesarean section was undertaken under a general anaesthetic, and the livebirth of a baby weighing 980g who was admitted to the neonatal intensive care unit (NICU). Estimated blood loss was 800ml.

Day of admission	Haemoglobin (g/L)	Platelet count (x10^9/L)	White cell count (x10^9/L)	LDH (U/L)	ADAMTS 13 activity (IU/dL)
D1	81	29	12	795	<0.2
D3	71	39	21	431	
D5	84	50	16	403	
D6 (delivery)	99	60	23	318	
D7 (D1 postpartum)	94	190	32		
D8 (D2 PP)	84	174	20		
D12 (D6 PP)	103	358	10	270	<0.2
D17 (D11 PP)	101	383	8	259	<0.2

Table 1: Shows the patients blood results through her admission

The patient was started on caplacizumab one day post-partum and rituximab on day 2 post-partum. She received a course of eight plasma exchanges, over seven days. She had no procedure related side effects and no significant post operative bleeding. The patient was discharged after an eighteen day admission.

References

1. Scully M, Thomas M et al. Thrombotic thrombocytopenic purpura and pregnancy: presentation, management and subsequent pregnancy outcomes. *Blood.* 2014; 124 (2): 211-219
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Discussion

TTP is an acute, potentially life-threatening disease characterised by a deficiency on ADAMTS13, a protease which cleaves von Willebrand factor (vWF) (2). Large vWF multimers bind to the endothelium and trigger platelet aggregation (3).

Pregnancy is a risk factor for an episode of TTP (2). TTP is an aggressive presentation and has an untreated mortality rate of 90% (3, 4). It is characterised by a haemolytic anaemia (Hb <80) and severe thrombocytopenia (<30) (1). Fluctuant neurological symptoms such as headache or confusion are present in 70% and can be severe (4). In pregnancy, placental thrombosis is a risk and it is associated with foetal growth restriction, intra uterine death and pre-eclampsia (4).

First line treatment is with therapeutic plasma exchange (TPE), which is associated with a fivefold decrease in mortality (2) Platelet transfusion is avoided whilst in the hyper-aggregatory state as it may result in increased thrombus creation (3).

For autoimmune TTP, immunosuppression with steroids is a first line therapy (1). Delivery is not a therapeutic tool for TTP but is considered for foetal concerns or maternal deterioration (1)

Further immunosuppression with agents such as rituxamb lack safety data for use during pregnancy and should only be used in the post partum period (1). Future pregnancies can be associated with recurrence, however, relapse is less common in women with normal ADAMTS13 levels prior to pregnancy (4).