HELLP! Is it really preeclampsia? A Case Report of Thrombotic Thrombocytopenic Purpura in Pregnancy.

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BACKGROUND

Thrombotic Thrombocytopenic Purpura (TTP) is a rare but serious cause of Thrombotic Micro-Angiopathy (TMA) in pregnancy and the puerperium. It presents with thrombocytopenia and Micro-Angiopathic Haemolytic Anaemia (MAHA) so is often mistaken for the more common preeclampsia and HELLP syndrome (Haemolysis, Elevated Liver enzymes and Low Platelets). Without urgent treatment it is associated with high rates of foetal and maternal mortality.

CASE REPORT

We discuss the case of a previously well 28 year old primigravida at 34 weeks and 6 days who presented with reduced foetal movements for two days and peripheral oedema. Prior to this presentation she had a low risk antenatal course. The patient was normotensive and initial investigations showed proteinuria, acute haemolytic anaemia, significant thrombocytopenia and impaired renal function. Her cardiotocography showed an unprovoked variable deceleration. She was initially diagnosed with severe HELLP and proceeded to an uncomplicated emergency caesarean section of a growth restricted baby born live in good condition. The investigation abnormalities were persistent and on day three postpartum she developed tachypnoea and was transferred to the intensive care unit.

She was diagnosed with TTP with multiorgan dysfunction and commenced therapeutic plasma exchange. She had a very low ADAMTS-13 level which confirmed the diagnosis. During the course of her admission she also received methylprednisolone and monoclonal antibody medications to treat the TTP.

The patient had a protracted intensive care admission with several complications including gastric erosion and upper gastrointestinal bleeding, necrotising pancreatitis and pseudocyst development, sepsis, acute kidney injury requiring renal replacement therapy and acute pulmonary oedema requiring intubation. She was in hospital for over seven months.

DISCUSSION

While uncommon TTP is an important differential for thrombocytopenia and MAHA in pregnancy as it can cause significant morbidity and mortality. Left untreated TTP has a reported maternal mortality rate of 75-90%.(1, 2)

TTP is caused by a deficiency, either acquired or congenital, of ADAMTS-13 which is a plasma protease responsible for the cleavage of Von Willebrand Factor. When the ADAMTS-13 level is low it results in large VWF which interacts with platelets and causes microvascular thrombosis. This results in thrombocytopenia due to consumption of platelets and MAHA. When this occurs it causes multi-organ dysfunction due to reduced blood flow.

In this case report the patient was initially diagnosed with and managed as HELLP syndrome. When she did not follow the expected course of this condition a broad differential was considered and other specialty teams became involved in her care. This demonstrates the importance of considering other causes of thrombocytopenia and MAHA in obstetrics so that appropriate treatment can be commenced to reduced the risk of maternal and foetal mortality.

1. Martin Jr. JN, Tucker JM. Maternal morbidity and mortality in pregnant/postpartum women with suspected HELLP syndrome identifiable as probable thrombotic thrombocytopenic purpura or atypical hemolytic uremic syndrome by high LDH to AST ratio. International Journal of Gynecology & Obstetrics. 2022.

2. Osborn JD, Rodgers GM. Update on thrombotic thrombocytopenic purpura. Clinical advances in hematology & oncology : Hematology & Oncology. 2011;9(7):531-6.

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