

CASE REPORT

Successful Outcome in a Rare Condition in Pregnancy - Thrombotic Thrombocytopenic Purpura-Hemolytic Uremic Syndrome: A Case Report*Babita Vaswani^{1*}, R. P. Patange¹**¹Department of Obstetrics and Gynaecology, Krishna Institute of Medical Sciences and Krishna Hospital, Karad-415110 (Maharashtra) India***Abstract:**

Thrombotic Thrombocytopenic Purpura-Hemolytic Uremic Syndrome (TTP-HUS) is a rare occurrence in pregnancy. It is similar in its clinical profile to other more common pregnancy related conditions like preeclampsia, eclampsia, and Hemolysis, Elevated Liver Enzymes, Low Platelets (HELLP) syndrome. This makes diagnosis a challenge thus hampering maternal and fetal outcome if the disease is not diagnosed in time and managed appropriately in the early course of the disease. Here, we report a case of successful management of this rare and potentially hazardous disorder during pregnancy in due course of time and thereby achieving a healthy mother and a healthy baby.

Keywords: Pregnancy, Thrombotic Thrombocytopenic Purpura, Hemolytic Uremic Syndrome

Introduction:

Thrombotic Thrombocytopenic Purpura-Hemolytic Uremic Syndrome (TTP- HUS) is a rare occurrence in pregnancy [1]. The greatest risk for development of TTP-HUS is near term and during the postpartum period [2]. This is also the time of greatest risk for thrombotic events and for the occurrence of other pregnancy-related syndromes: preeclampsia, eclampsia, and Hemolysis, Elevated Liver Enzymes, Low Platelets (HELLP) syndrome. These other syndromes may also be associated with thrombocytopenia, microangiopathic hemolytic anemia, neurologic symptoms, and renal insufficiency. This poses a diagnostic challenge

for obstetricians making it difficult or impossible to differentiate TTP-HUS from pre eclampsia and HELLP syndrome. TTP-HUS has an unfavourable maternal and fetal outcome if it is not recognised in time and treated appropriately early in the course of the disease. Here, we report a rare case of TTP-HUS in pregnancy with prompt management and a successful outcome.

Case Report:

A 26 year old, second gravida with 32 weeks of gestation with previous Lower Segment Cesarean Section (LSCS) was admitted to our hospital with complaints of pain in abdomen since 5-6 days, vomiting and loose stools since 2 days and decreased urine output since 1 day. Her pulse was 102/min and blood pressure was 110/70 mmHg. She was pale with no icterus or edema. Her cardiovascular and respiratory systems were normal. On per abdominal examination, uterus was 30-32 weeks size, cephalic presentation, relaxed and FHS were normal. There was a scar of previous LSCS. Her laboratory reports revealed Hb of 11.2g/dl with mild leucocytosis and thrombocytopenia (platelets - 47,000/mm³). Her urea was 74 g/dl and creatinine was 4.2 g/dl. (INR) was 1.14. Urine showed numerous WBC's. She was started on IV fluids and urine output was strictly monitored. Injection meropenem was started. Blood tests were repeated the following day and it was found that Hb reduced to 10.7 g/dl

and platelets reduced to 45000/mm³. Creatinine increased to 5.5 g/dl. The patient's PTT was within normal limits and urine culture was sterile. USG showed features suggestive of acute renal parenchymal changes. Haemodialysis was done for the patient. Following this her Hb further reduced to 8.7g/dl and creatinine increased to 6 g/dl. A raised Lactate Dehydrogenase (LDH) level and reticulocytosis confirmed haemolysis as well as platelet destruction. A diagnosis of TTP-HUS was made. A second haemodialysis was done. Plasma transfusions (four pints) were given. Her urea then reduced to 55 g/dl and creatinine to 3.7 g/dl. Urine output gradually increased from 100 ml to 500 ml to 1200 ml over 4-5 days. Unfortunately, the patient started complaining of leaking per vaginal and she was found to have Preterm Premature Rupture of Membranes (PPROM). LSCS was done on maternal request and she delivered a female child of 1.5 kg weight. Post operatively also the patient required haemodialysis until creatinine was reduced to 2 g/dl and urine output became normal. LDH and Hb levels also returned to normal. The patient recovered completely and was discharged on the 10th post operative day in an excellent condition.

Discussion:

TTP and HUS are believed to be different clinical manifestations of the same underlying disease. TTP- HUS is now used as one term owing to an inability to distinguish TTP and HUS on clinical or pathological grounds [3]. The central pathologic feature of both disorders is the formation of platelet thrombi in the microvasculature which may produce tissue ischemia and infarction. Immunohistochemically, the platelet thrombi contain large amounts of Von Willebrand Factor (VWF). These thrombi cause shearing stress on the red blood cells passing

through the capillaries causing fragmentation (thrombotic MAHA) [1]. It is a rare disease and 10% of TTP cases have been reported to occur in pregnancy [4]. Pregnancy is identified as a possible trigger for TTP-HUS, with a higher frequency in women having suffered from the disease pre pregnancy [1].

Diagnosis is often difficult due to the resemblance of this condition to a similar spectrum of presentation in pre-eclampsia, eclampsia and HELLP syndrome. This causes delayed diagnosis and treatment thus causing high morbidity and mortality. The onset of renal failure, thrombocytopenia, MAHA, normal PT, PTT and relatively mild elevated LFTs supports the diagnosis of HUS of pregnancy. To a large extent, management of TTP- HUS continues to be empirical and guided by clinical experience. At present there is no absolute consensus on management, but survival rates in TTP-HUS have improved with early management from 10% to between 75 and 92% [3]. This emphasizes the need for early diagnosis and urgent initiation of treatment measures. Clinical suspicion of TTP-HUS necessitates urgent plasma exchange/ plasma transfusions. With plasmapheresis, now the mainstay of treatment, most patients (80-90%) will survive an acute episode of HUS. Plasmapheresis is accomplished by removing the patient's plasma and replacing it with normal plasma [5]. Treatment should begin as early as possible. Plasma infusion may be appropriate till plasma exchange is available [6].

Daily treatments should continue until the patient improves clinically and the thrombocytopenia and the thrombotic MAHA resolves (evidenced by normalization of LDH). Other supportive therapies include red cell replacement and hemodialysis. Steroids and anti-platelet therapy

are often used as adjuvant therapy with plasmapheresis. Immunoabsorption, splenectomy, intravenous gamma globulin, and vincristine should be considered as second line therapies, and used when a patient does not respond to pheresis. It is important to avoid platelet transfusions, as they will add to the extent and severity of the microvascular thrombi. The prognosis for HUS in pregnancy is poor, with a high maternal mortality and associated fetal loss.

Conclusion:

This paper highlights a well managed case of a very rare disorder in pregnancy with a successful maternal and fetal outcome. This emphasizes the need for early recognition and appropriate consultation and referral of cases of TTP-HUS in pregnancy. Early and accurate detection and diagnosis can significantly improve the prognosis and optimize maternal outcomes.

References

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