

# Thrombotic thrombocytopenic purpura in the first trimester of pregnancy

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## Abstract:

Thrombotic thrombocytopenic purpura (TTP) occurs more commonly in women and so can be associated with pregnancy. The time during pregnancy with greatest risk for development of TTP is near term and during the post partum period. TTP occurring in early trimester is uncommon and is also associated with great maternal and fetal mortality. We report a successful outcome of pregnancy in a woman with TTP in early first trimester who was treated with therapeutic plasma exchange.

## Key words:

First trimester, plasmapheresis, pregnancy, thrombotic thrombocytopenic purpura

## Introduction

Thrombotic thrombocytopenic purpura (TTP) is a very rare complication of pregnancy, and occurs in 1 in 25,000 pregnancies. Majority of the cases occur in the late third trimester or during the puerperium. Only about 10% occur in the 1<sup>st</sup> trimester.<sup>[1,2]</sup> Before the advent of plasma infusion and therapeutic plasma exchange (TPE) as the primary treatment, pregnancy-related TTP was associated with a high maternal and fetal mortality which approached 80%.<sup>[3]</sup> Currently plasma infusion and TPE have improved the maternal and fetal survival rates dramatically. There have been several reports on TTP in later trimesters of pregnancy and puerperium but very few in the 1<sup>st</sup> trimester.

We describe the case of a first pregnancy in a woman with severe TTP diagnosed in early 1<sup>st</sup> trimester with maternal survival and delivery of a healthy, term baby.

## Case Report

A 21-year-old primigravida presented at 10 weeks of gestation with pain in abdomen, hematuria, oliguria, and altered sensorium with episodes of generalized tonic clonic seizures. On general examination her blood pressure records were normal. She had pallor, tachycardia and facial puffiness. Laboratory investigations showed anemia with hemoglobin of 4.7 gm% and thrombocytopenia with a platelet count of 29,000/cumm. On peripheral blood film there was anisopoikilocytosis with abundant schistocytes. Renal function tests were deranged with blood urea of 131mg/dl, serum creatinine of 6.2 mg/dl and spot urine albumin was 2+. Total

serum bilirubin was 2.8 mg/dl, indirect bilirubin was 2 mg/dl. Levels of plasma lactate dehydrogenase were very high the value being 1062 IU/L (normal range 70-150 IU/L). Ultrasound of abdomen showed increased echogenicity of the pelvicalyceal system of both kidneys. There was no evidence of renal vein thrombosis on renal Doppler. Her thrombophilia screen was negative. This screen included a normal lupus anticoagulant factor, antinuclear antibody and anticardiolipin antibodies. Her coagulation studies were also normal. Unfortunately the measurement of ADAMTS-13 levels was not carried out due to nonavailability of this investigation in our laboratory.

In view of a deteriorating neurological condition, a lumbar puncture was done and the cerebrospinal fluid analysis was normal. Hemodialysis was started in view of oliguria and azotemia after nephrology consultation. The next day she needed mechanical ventilation in view of poor respiratory efforts. Platelet count further fell down to 12,000/cumm. On the basis of symptoms and laboratory tests, a clinical diagnosis of TTP was made. Treatment with daily TPE which comprised replacement of 30ml/kg plasma in combination with infusion of fresh frozen plasma (FFP 15ml/kg) was started using Cobe Spectra TPE machine. After four cycles of TPE, her platelet count rose to 1.5 lakh/cumm. The platelet count was subsequently monitored initially weekly and then biweekly till term and the values remained normal. Hemoglobin increased to 7.9 gm% and there was decrease in number of schistocytes on peripheral blood film. Plasma lactate dehydrogenase levels decreased to 180 IU/L. Urine output improved. She was extubated within next 48 hrs. Her condition was monitored till day 20 of treatment and there was progressive improvement in clinical and laboratory parameters. She thus did not receive any further

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plasma exchange. Tablet folic acid 5 mg/ day, supplemental iron and calcium were continued. Corticosteroids were not used. Patient was discharged on day 24 of admission in a satisfactory condition. Here pregnancy was monitored regularly in the medicosurgical unit at our institution. At 37 weeks of gestation, her blood pressure was found to be high in the range of 130/90 – 140/100 for which her pregnancy was terminated. She had intrapartum fetal distress for which an emergency Cesarean section was done and she delivered a healthy 2.4-kg baby. She received postpartum prophylaxis for deep vein thrombosis (heparin 5000 IU subcutaneously twice a day). Both the mother and baby were discharged 1 week later in a satisfactory condition.

## Discussion

TTP is a multiorgan system disorder characterized by microangiopathic hemolytic anemia, thrombocytopenia, neurological symptoms and renal involvement. Pregnancy is one of the common precipitating events for acute episodes of TTP. It is because of the association of pregnancy with increasing concentration of procoagulant factors, decreasing fibrinolytic activity, loss of endothelial cell thrombomodulin and decreased activity of ADAMTS-13.<sup>[4,5]</sup> ADAMTS-13 is a protease involved in the cleavage of Von Willebrand factor. All these abnormalities become progressively severe as pregnancy advances, in fact majority of cases of TTP occur at delivery or immediately postpartum. Whatever, be the period of gestation it is associated with a considerable risk to both maternal and fetus in the absence of treatment. Even diagnosis of this condition is not easy during pregnancy because the symptoms mimic those of preeclampsia, HELLP, eclampsia or any acquired coagulopathy. Fetal death is secondary to placental infarction and maternal death occurs due to acute renal failure, disseminated intravascular coagulation or thrombocytopenia.<sup>[6]</sup> Plasma exchange is the mainstay of treatment. When the disease occurs late in pregnancy and the fetus has gained considerable maturity, pregnancy can be terminated for aggressive treatment. However, when it occurs very early in pregnancy as was in our case, therapeutic abortion is considered by many because of the risk of repeated episodes. Prolonged courses of plasma exchange may achieve and maintain a remission of TTP allowing full-term delivery. However, if plasma exchange fails to induce remission, therapeutic abortion may be considered. The response of TTP to termination of pregnancy is unknown. Although, we had a successful experience with this patient and her pregnancy, there was always a concern of risk of subsequent episodes of TTP.

According to the guidelines of British Committee for standards in hematology, pregnant patients with TTP should be treated with plasma exchange as is done for nonpregnant patients.<sup>[7]</sup> Delivery is advised only for those who do not show any response to TPE. Plasma exchange should be continued even after normalization of platelet count and resolution of hemolysis is

documented. McMinn *et al* have documented that the increased plasma volume of pregnancy does not impair the response of TTP to TPE and TPE does not adversely affect the outcome of pregnancy.<sup>[8]</sup> Overt ADAMTS-13 deficiency is not a universal finding, its estimation is not recommended for diagnosing TTP. TTP may respond to TPE in absence of the deficiency of this enzyme. Intravenous immunoglobulins and glucocorticoids like methylprednisolone and oral prednisolone can be included in the treatment, it being an autoimmune disorder. Our patient did not receive any glucocorticoids. We cannot be sure whether this patient had pregnancy induced TTP or that the pregnancy was just coincidental. The risk of recurrence in subsequent pregnancies will remain an important issue.

From our experience, we advocate that such women if managed collectively by a unit of obstetricians, hematologists, nephrologists in institutes where TPE is available a successful outcome can be expected.

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