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Anaesthetic Management in a Parturient with Thrombotic Thrombocytopenic Purpura (TTP) for Emergency Caesarean Section

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Abstract: Introduction: TTP is a rare and life threatening condition characterized by microangiopathic hemolyticanemia, severe thrombocytopenia and organ ischemia. A 25 - year old pregnant patient at 30 weeks of gestation was posted for emergency lscs in view of non reassuring intrapartum monitoring. Patient was a known case of congenital TTP. She was being treated with Inj. LMWH and 4 units of FFP transfusion 4 times a week since the start of pregnancy. She was also diagnosed with preeclampsia in the current pregnancy and was on tab. labetalol. Patient's starvation status was 3 hours at the time of being taken in for surgery. Laboratory investigations revealed thrombocytopenia. Informed consent was taken. General anaesthesia was administered to the patient. A male neonate was delivered. Result: The surgery was completed in 1.10 hours. Blood loss was approximately 700ml. Patient was extubated successfully and recovery was uneventful. Conclusion: As this patient was prone for thrombocytopenia and was on LMWH but had an inadequate starvation, weighing the risk benefit ratio of general anaesthesia over regional anaesthesia, general anaesthesia is preferred over regional anaesthesia.

Keywords: TTP (thrombotic thrombocytopenic purpura), LSCS (lower segment caesarean section), LMWH (low molecular weight heparin), FFP (fresh frozen plasma)

1. Introduction

TTP is characterized by formation of platelet rich thrombi in both arterial and capillary microvasculature, resulting in thrombocytopenia and microangiopathic hemolytic anemia. The cause of TTP is a deficiency of ADAMTS13, a protease that disrupts Vwf (von willebrand factor) multimers into smaller pieces. Pregnancy can trigger TTP, although diagnosis can be difficult since clinical and laboratory findings can be very similar to those of preeclampsia and HELLP syndrome.

2. Literature Survey

Eli Moschowitz was the first to describe TTP, when performing the autopsy of a 16 - year - old girl whom he had treated. TTP has a mortality rate of 90% without treatment ⁽¹⁾. TTP is classified according to its etiology, although TTP can be congenital, as a result of a mutation of the gene responsible for desynthesizing ADAMTS13 ⁽²⁾. The most common form of TTP is idiopathic or autoimmune, where antibodies directed against ADAMTS13 are created ⁽³⁾.

Marie Scully et al in 2014, conducted a prospective study of TTP cases from the United Kingdom Thrombotic Thrombocytopenic Purpura (UK TTP) registry and found that congenital TTP presents more frequently than acquired TTP during pregnancy and must be differentiated by ADAMTS 13 analysis. They concluded that careful diagnosis, monitoring and treatment in congenital and acquired TTP resulted in excellent pregnancy outcomes ⁽⁴⁾.

3. Case Report

A 25 year old pregnant patient (G4A2IUFD1) at 30 weeks of gestation, diagnosed with congenital TTP and preeclampsia, was posted for emergency lower segment caesarean section in view of non reassuring intrapartum monitoring. She was diagnosed with congenital TTP in 2019 (during previous pregnancy, after a miscarriage at 6th month of gestation) and subsequently treated with plasma exchanges, prednisolone, cyclophoshamide, and rituximab. In the current pregnancy she was started on Inj. lmwh 0.4mg sc once daily. She was also receiving FFP transfusions at thrice weekly intervals in the initial gestational weeks of pregnancy, which was later increased to 4 times/week as platelet counts were in decreasing trend. Besides this, she was also started on Tab. Labetalol (100 mg), twice daily, for preeclampsia.

This patient presented to us in the operation theatre for an emergency caesarean section with a starvation status of 3 hours. Her lab investigations were of previous day which revealed a hemoglobin of 11.6 g/dl, platelet counts 80, 000 mm³ and an INR of 1.01. She had received Inj. LMWH and 4 units of FFP transfusion on the day prior to the surgery and had taken Tab. Labetalol on the morning of the surgery. Patient was evaluated and an informed consent was taken. Standard ASA monitors were attached and two wide bore iv lines were secured. She was premedicated with Inj. Ondansetron (4mg) and Inj. Metoclopramide (10mg) for aspiration prophylaxis. Patient was preoxygenated with 100% oxygen and rapid sequence induction was done with 2mg/kg propofol along with 1mg/kg succinylcholine and endotracheal intubation was performed. An orogastric tube

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was also inserted. Maintenance of anaesthesia was done with sevoflurane in a 50% O2/N2O mixture, post baby delivery. A male neonate was delivered with an APGAR score of 8. After the delivery of the baby, infusion of oxytocin 20 IU in 500 ml 0.9% NS was started. Hemodynamic parameters were stable and urine output was monitored throughout the procedure. Total blood loss was approximately 700 ml and she received 1500 ml of crystalloids and had a urine output of 150ml. Patient was extubated uneventfully after ensuring complete neuromuscular reversal at the end of operation.

4. Discussion

TTP can be described as a complex of signs including thrombocytopenia, microangiopathic hemolytic anemia (MAHA), fever, renal failure and neurologic manifestations. Not all patients with TTP have all 5 signs. Prompt diagnosis and effective treatment can improve both maternal and fetal outcome.

These patients have deficiency of ADAMTS 13 which increases platelet microthrombi leading to MAHA, so they are given FFP transfusions to replace ADAMTS and heparin to prevent coagulation.

One of the main concerns in these pregnancies is the decision of performance of regional anaesthesia, since thrombocytopenia in pregnancy is considered a relative contraindication. The lowest threshold of platelet count for a safe performance of neuraxial block is controversial in obstetric anaesthesia. Along with the platelet count, the platelet function may be affected when thrombocytopenia in pregnancy is present. Another significant challenge is the use of anticoagulant and antithrombotic drugs that can increase the risk of formation of spinal hematoma.

5. Conclusion

Anaesthetic management of patients suffering from TTP is quite challenging in view of increased risk for perioperative complications because of their thrombotic as well as thrombocytopenic state. Besides platelet count, other laboratory findings should be paired with TEG and other point of care testing and a decision should be rendered after considering patient specific risks and benefits regarding the use of general or regional anaesthesia. In order to avoid the risk of neurological complications associated with regional anaesthesia, we considered general anaesthesia would be more appropriate in our patient as her platelet counts were in decreasing trends and she was also on LMWH. Obstetricians, Anaesthesiologists and Hematologists should work as a multidisciplinary team to optimise the outcome of these clinical cases.

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