http://jmscr.igmpublication.org/home/
ISSN (e)-2347-176x ISSN (p) 2455-0450
crossref DOI: https://dx.doi.org/10.18535/jmscr/v8i10.24

IGM Publication
Journal Of Medical Science And Clinical Research
An Official Publication Of IGM Publication

Case Report

Rare Case Report of TTP Associated with Pregnancy

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Abstract
Thrombotic thrombocytopenic purpura (TTP) is thrombotic microangiopathies, is rare medical condition that occur in pregnancy but if neglected then it will lead to serious maternal and fetal morbidity. TTP clinical findings are similar with preeclampsia, especially the HELLP (Hemolysis, Elevated Liver tests, Low Platelets) syndrome. In this case report patient is known case of TTP in pregnancy, therefore regular ANC visits and timely transfusions of fresh frozen plasma at the interval of 15 days and followed by routine CBC, BLOOD SMEAR, LFT, RFT, LDH done .
TTP in pregnancy is reversible, but life-threatening medical emergency there is need of strict mother and fetal surveillance for the good result, which was done in this case, from 24 wks her first visit till the discharge of the patient as discussed in case report.
Using the regular pentad solely for diagnosis of TTP will lead to underdiagnosis of many cases and should be avoided.
Keywords: *TTP, pregnancy.*

Introduction

TTP (thrombotic thrombocytopenic purpura) is one of the thrombotic microangiopathies, its incidence is 2 to 6 per million persons per year (Miller 2004).

Its resembles to HELLP SYNDROME allude to their obstetrical ramifications (George, 2014). The possible etiopathogenesis in most of cases to be caused by antibodies to or a plasma deficiency of ADAMTS13 (Ganeshan2011; Sadler, 2010).

Therefore the endothelium- derived protease cleaves vWF (Von Willebrand factor) to decrease its activity.

Also intravascular platelet aggregation which stimulates the cascade of events leading to end organ failure. There is endothelial activation and damage, but it is unclear whether it is a consequence or cause and the aggregates of multimer of vWF and microthrombi of hyaline material consisting of platelets and small amounts of fibrin in arterioles & capillaries cause ischemia and infarct.

In pregnancy, the activity of ADAMTS13 enzyme decreases by 50 % and it will drop further if it is associated with preeclampsia syndrome. It is seen that TTP is more commonly seen during pregnancy.

The PARKLAND HOSPITAL experiences was described by the DASHE and coworkers (1998), who identified 11 pregnancies complicates by

these syndromes among nearly 275,000 obstetrical patients -a frequency of 1 in 25,000.

Case Report

A 25 -year-old G2P1L0 woman was admitted in our hospital at 36 weeks of gestation k/c/o TTP induced in pregnancy with no fresh complains planned for elective lscs as ultrasonography of her 34 weeks had s/o stage 1 foetal growth restriction. she was compliant patient and had regular visits in our hospital since 24 weeks of second (this pregnancy) pregnancy for transfusion of fresh frozen plasma at the interval of 15 days and periodic CBC,CBC with PBF, LDH,LFT ,RFT investigations done afterwards .she was diagnosed with TTP in her last pregnancy after an episode of hemiparesis of left side of her body and raised bp, then to rule out the cause of paresis the various investigations done like USG ,MRI the case discussed with the haematologists, as there was continuous fall of platelet count even after 1 month of her deliver , thrombosis induced due to TTP was considered as root cause of paresis and the persistent fall of platelet count till she was not diagnosed, she delivered through normal vaginal delivery in last pregnancy 3 years back and the baby was still birth after that episode.

Therefore this time she was assumed to be high risk pregnancy and routine investigations were done after every transfusions & her vitals were normal, no signs of the petechiae on skin of the body in all her ANC visits, in our hospital with negative family history but history of depression for few days hence she was counselled & referred to psychiatrist, had h/o road traffic accident and head injury on head at the age of 8 yrs (the possible cause of thrombosis was ruled out for hemiparesis).

The covid-19- testing also done prior to elective LSCS in the pandemic era, her report was positive and she was admitted in COVID maternity wing, planned for LSCS at the completion of 37 weeks with the consultation of anaesthetist and her treating haematologists, also after the corticosteroid dose coverage and repeat covid-19 testing became negative.

Investigations

Before the LSCS her Hb =14.1, Plt count=155000 Wbc=10600, LDH=272, normal LFT, RFT, PT & APTT, bp=124/78mmhg.

Postoperative Period

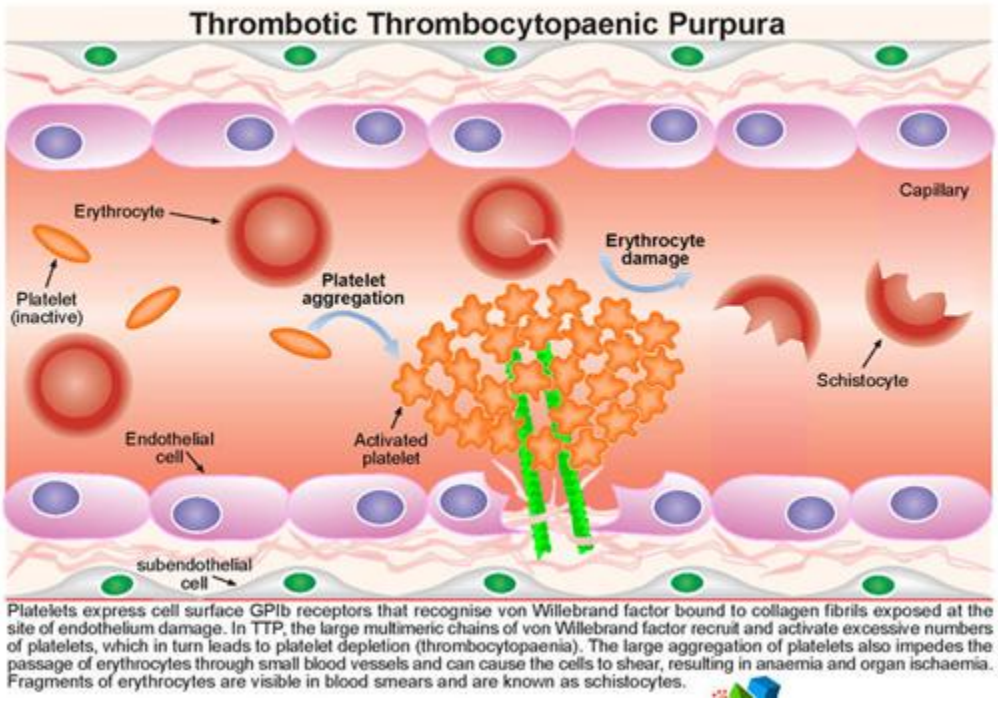
Her LSCS was done on 12/09/20 at 10.12 am she delivered female child of 2.14 kg and the procedure was uneventful and her intraoperative & preoperative vitals were normal.

But after around ½ hours of post op period she complained of severe headache and excessive nausea and vomiting and her bp was 160/110 mmhg, pr=92/min, urine output=500ml, uterus w/c and bleeding per vagina was average, at that time.

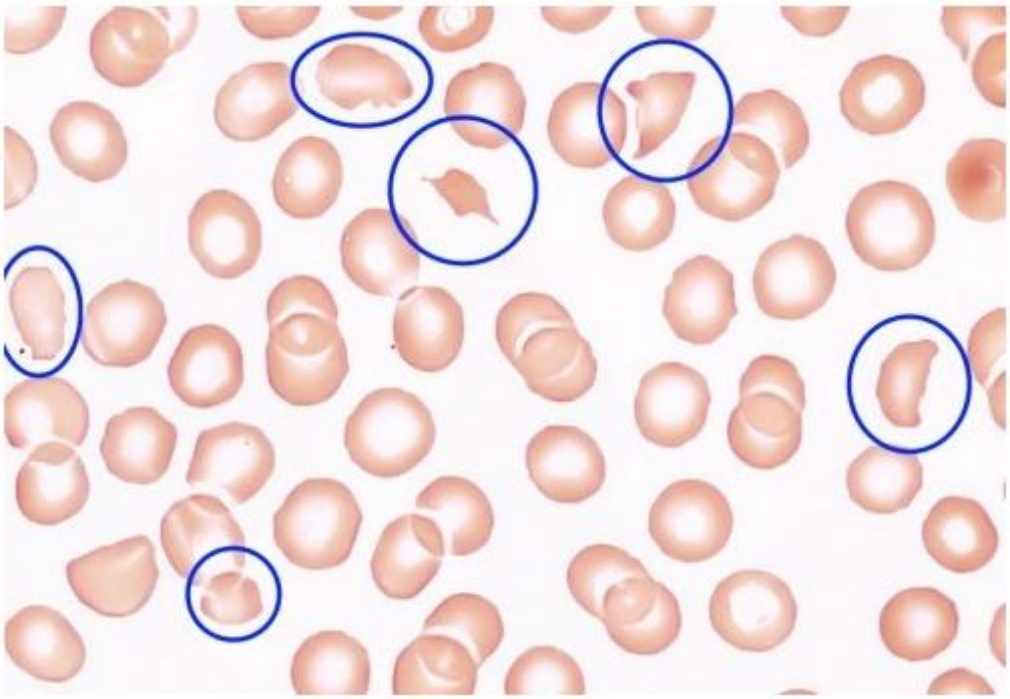
Urgent physician, anaesthetist, neurology and haematology reference done.

Simultaneously symptomatic treatment was given to decrease the bp, Injection labetalol 20 mg in 100 ml of normal saline was given and c/w/o vitals were done, neurologist advised the NCCT of head on an urgent basis to see the thrombosis, and shifted to neurology side for further management with keep in touch with obstetrics and paediatric department for her and baby need.

On her history she had similar episodes of events in her last delivery followed by recovery afterwards and again developed thrombocytopenia in this pregnancy only without any variations of platelet count in her 3 yrs gap after her last delivery. Her NCCT scan and her MRI was normal and her bp reduced with days onwards after giving the magnesium sulphate and amlodipine. She was well discharged on day8 of LSCS.



Platelets express cell surface GPIb receptors that recognise von Willebrand factor bound to collagen fibrils exposed at the site of endothelium damage. In TTP, the large multimeric chains of von Willebrand factor recruit and activate excessive numbers of platelets, which in turn leads to platelet depletion (thrombocytopaenia). The large aggregation of platelets also impedes the passage of erythrocytes through small blood vessels and can cause the cells to shear, resulting in anaemia and organ ischaemia. Fragments of erythrocytes are visible in blood smears and are known as schistocytes.



Discussion

>Thrombotic thrombocytopenic purpura (TTP) is a potentially reversible, life-threatening medical emergency. In its full-blown form, it is characterised by the pentad of microangiopathic haemolytic anaemia (MAHA), thrombocytopenia, neurological abnormalities, fever and renal dysfunction. Nonetheless, in actuality, only 20%–30% of patients present with the classic pentad

>TTP-related maternal mortality can be reduced from 90%, without treatment, to less than 10% with prompt initiation of plasma exchange.[(1)]

>We present this case to discuss a rare disease entity and the potential role for obstetrical monitoring and surveillance to improve fetal outcomes. Thrombotic thrombocytopenic purpura in pregnancy is a rare but potentially fatal condition for both the patient and her fetus if left untreated.

> The disease is a known mimicker of preeclampsia, and often the two entities can coexist within the same patient thus requiring a low index of suspicion to initiate further workup[(2)]

> TTP and HELLP syndrome in pregnancy is very confusing, therefore the differentiating feature between two is that deposition of hyaline and microthrombi within the liver with the thrombotic microangiopathy hepatocellular necrosis with elevated serum hepatic aminotransferase levels characteristics of preeclampsia is not a common feature of TTP, whereas HELLP syndrome is improved by delivery but not in case of TTP.

>The investigations advised in these patients to see CBC with blood smear which shows erythrocyte fragmentation, schizocytes, reticulocytes and increased nucleated red blood cells are increased.

Also there will be increased in LDH (lactate dehydrogenase), haptoglobin.and deranged LFT and RFT.

>The cornerstone treatment of TTP is plasmapheresis with fresh frozen plasma

>As the available literature is limited, it is not known whether treatment with TPE (therapeutic plasma exchange) or plasma infusions during the pregnancy will lead to improved foetal outcomes. As described, there is a high incidence of stillbirth in patients with TTP, most pronounced if the diagnosis is made in the late second trimester.

>Further confounding the discussion regarding fetal outcomes with TTP is the known association with comorbid preeclampsia[3], also experienced by our patient though in the postpartum period.

>It is known that preeclampsia is associated with Foetal growth restriction, placental abruption, and poor fetal outcomes, however, it is not known to what degree comorbid TTP may contribute to these outcomes. Therefore, decisions regarding delivery in the case of significant maternal disease must be made judiciously.

Conclusion

This is the diagnosed case of TTP with pregnancy and the regular follow up made to reduce both maternal and fetal morbidity.

This case suggests that pregnancies affected by maternal TTP require ongoing fetal surveillance to determine the timing of delivery, potential role for antenatal steroid administration, and assure optimal neonatal outcomes

Using the regular pentad solely for diagnosis of TTP will lead to underdiagnosis of many cases and should be avoided.

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