Case Report

Role of plasma exchange in a post-partum case of severe thrombotic thrombocytopenic purpura with acute kidney injury

Jyoti Bharti[1], Tulika Chandra[1], Archana Solanki[1], Ashutosh Singh[1], D. Himanshu Reddy[2], Mallika Agarwal[3]

[1]Department of Transfusion Medicine, King George Medical University, Lucknow, Uttar Pradesh, India, [2]Department of Medicine, King George Medical University, Lucknow, Uttar Pradesh, India, [3]Department of Pathology, D.Y. Patil Medical College, Pune, Maharashtra, India

Abstract

Thrombotic thrombocytopenic purpura (TTP) is a life-threatening disease present with the classic pentad of microangiopathic hemolytic anemia (MAHA), fever, neurologic changes, thrombocytopenia, and renal dysfunction. In a diagnostic dilemma, therapeutic plasma exchange (TPE) is a choice of life-saving intervention. In this, we assess the efficacy of TPE in a suspected case of post-partum TTP. A 27 years old female was admitted in an emergency on day 8 after a lower segment cesarian section (LSCS) with unresponsive behavior for 3 days and with TTP. She was normal 32 days back with her second, 7-month pregnancy. Ultrasonography (USG) showed an umbilical cord around the neck of the baby. On the fifth post-operative day, she was shifted to emergency with fever, generalized anasarca, gastrointestinal tract (GI) bleeding, low platelet count, and low Hb, with a poor Glasgow coma scale (GCS) of 6. On the bases of serum urea and serum creatinine, she presented acute kidney injury with encephalopathy. At emergency, she was unresponsive to mechanical ventilation and supportive treatment; hence, therapeutic plasma exchange was performed. After eight TPE cycles, the patient presented with an improved hematological and renal profile with good GCS. TPE is helpful and life-saving for suspected TTP patients with AKI.

Keywords: Acute kidney injury, fresh frozen plasma, post-partum thrombotic thrombocytopenic purpura, therapeutic plasma exchange, TTP

Introduction

Thrombotic microangiopathy (TMA) syndromes are characterized by MAHA, platelet clumping, and organ failure of variable severity, for example, TTP, hemolytic-uremic syndrome (HUS), elevated liver enzymes, low platelet (HELLP) syndrome, acute fatty liver of pregnancy (AFLP), antiphospholipid syndrome (APS), and systemic lupus erythematosus (SLE).[1] TMA can feature several pregnancy-related disorders such as TTP/HUS, TTP, HELLP syndrome, or AFL. TMA from sepsis with disseminated intra-vascular coagulation (DIC) is associated with prolongation of clotting time, prothrombin time, and activated partial thromboplastin time (PTT, aPTT) due to consumption of clotting factors.[2]

Address for correspondence: Prof. Tulika Chandra, Department of Transfusion Medicine, King George Medical University, Lucknow, Uttar Pradesh, India.
E-mail: drtulikachandra@gmail.com

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Case History

A 27-year female, multigravida with two stillbirths ($G_2P_oL_o$; P_1-male fetus 2 kg normal vaginal delivery, P_2-male 2.5 kg cord around neck

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and low-lying placenta), presented 25 days back in the emergency department with unresponsive behavior since 1 day on day 8 post LSCS. She was normal 32 days back with her second, 7-month pregnancy. The USG at the 7th month of pregnancy revealed cord around the neck of the baby with a low-lying placenta; hence, she was advised for LSCS under anesthesia. On the 5th post-operative day, she was unresponsive and shifted to a tertiary care center. At the time of admission, her heart rate (HR) was 140/minute, blood pressure (BP) was 110/82 mmHg, and SpO_2 was 70%. Respiration was gasping, the temperature was 98.6F, and pallor and anasarca were present. GCS was $E_2V_2M_2$, with normal pupils, B/L coarse crepitation was present, and S1 and S2 present with no murmur and healthy abdomen. At emergency, she was intubated and under mechanical ventilation with sedation.

At CCM, she was unresponsive for last 10 days with reduced urine output for 4 days. Examination revealed the following: pallor (+); anasarca (++); icterus (-); cyanosis (-); clubbing (-); heart rate, 119/minute; BP, 124/80 mmHg; SpO_2, 100%; respiratory rate, 24/minute; and body temperature, 98.4F. Her platelet count fell unexpectedly to 0.30 lac cells/mm^3, with a falling Hb of 6.4 gm/dl. ADAMTS13 test was not done due to financial issues. The results of the investigations are in [Table 1].

Hence, we decided to perform TPE to use 1.5 plasma volume exchange using FFP as a replacement fluid. The patient's informed consent was obtained before each procedure. Complication managed by prophylactic administration of calcium gluconate.[3] We performed all the procedures at 25–30 ml/minute blood flow and ACD (acetate, citrate, dextrose) as an anticoagulant at 1:10 ratio.

She was under sedation at first TPE cycle and exchanged 0.8 volume of plasma by 22 units FFP. After 10 days of the first TPE cycle, her serum LDH level (1142 U/L) increased. By the second and third TPE cycles, 0.8 volume plasma exchanged. At third TPE cycle, the patient was fully oriented with GCS 9. The fourth, fifth, sixth, seventh, and eighth TPE cycles exchanged 1.0 volume plasma [Table 2].

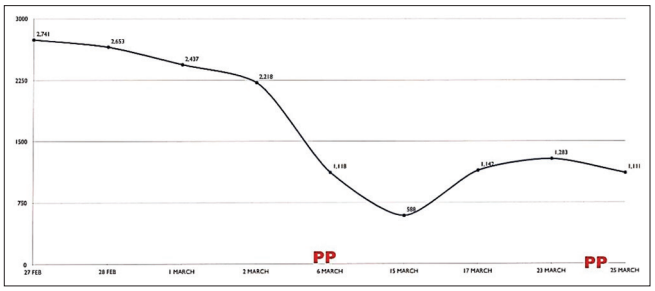
Discussion

TMA is a term used to describe a group of disorders characterized by thrombocytopenia, hemolytic anemia, and widespread thrombosis in the microvasculature, with HELLP syndrome.[4] Diagnosis of this condition is not easy during pregnancy because the symptoms mimic pre-eclampsia, HELLP, eclampsia, or any acquired coagulopathy.[5] This case is based on diagnostic dilemma [Table 3]. N. Gonnade, A. Bajayee, and A. Elhence *et al.*[3] stated that 42% of the patients did not respond to TPE, but 58% of patients gave adequate response. M. Soffer, P. Bendapudi, and D. Roberts *et al.*[6] stated that TPE drastically decreased the rates of maternal mortality resulting from TTP. K. Artinger, G. Hackl, and G. Schilcher *et al.*[7] reported delayed recovery from HELLP syndrome and primary TMA syndromes in the post-partum period. Immediate treatment with PEX and revisiting the first tentative diagnosis decrease mortalities. G. Ve and L. Trombotik[8] stated in their study thrombotic microangiopathies of pregnancy and the post-partum period should be treated by TPE in tertiary care units promptly. M. Wind, A. Gaasbeek, and L. Oosten *et al.*[9] statSed that TPE procedures can be used safely during pregnancy. P. Care, G. Ary, and H. Ipscomb *et al.*[10] said in their study that early plasma exchange is a relatively safe procedure during pregnancy and also used in the treatment of TTP and HUS.

In our case, after every TPE cycle, platelet counts increased and serum LDH was decreased [Table 3]. The patient was weaned off from IV drugs and stabilized on oral medicines. She was able to sit on her bed with support. After the eighth cycle of TPE on post-procedural investigation, serum LDH [graph 1] [Table 3], she was discharged after full recovery.

Conclusion

By this study, we could be aware of the importance of TPE for primary care physicians as early as possible. Basic screening methods in pregnancy like blood pressure monitoring and testing for proteinuria can identify and prevent potentially life-threatening complication for patients. Plasma exchange therapy should be performed without delay in suspected cases of during pregnancy and post-partum cases with a carefully



Graph 1: Serum LDH levels with days

Table 1: Blood investigations with no. of units and blood components transfused

	12th March	13th March	14th March	15th March	16th March	17th March	18th March	19th March	20th March	21st March	22nd March
Hb%	6.7	7.8	6.5	6.7	-	8.6	7.8	6.9	7.2	7.5	9.4
Platelet counts	4.4	3.5	2.6	3.0	-	2.9	2.7	3.0	2.6	2.2	2.1
Blood products	1PRBC	-	1PRBC	-	-	-	-	1PRBC	1PRBC	1PRBC	-
Serum creatinine	3.4	-	1.5	-	-	1.3	1.4	2.4	2.3	1.6	1.6
Serum urea	65	-	40	-	-	27	26	40	43	27	23
Serum LDH	-	-	588	-	-	1142	-	-	-	-	-

Table 2: Pre- and post-procedural laboratory (hematological and renal) profile for all TPE cycles

No. of TPE Cycle	Hematological profile								Renal profile							
	Hb%		Platelet count		PT		aPTT		Serum creatinine		Serum urea		Serum LDH		Serum alkaline phosphate	
	Pre	Post	Pre	Post	Pre	Post	Pre	Post	Pre	Post	Pre	Post	Pre	Post	Pre	Post
1st	6.44	8.4	0.80	0.60	18.1	20	9.8	1.5	2.14	3.01	45.6	96.4	1279.8	1289.9	241.7	
2nd	9.4	9.4	2.1	2.1	20	20	1.5	1.5	1.67	1.67	23.1	23.1	1283	1283	241.7	
3rd	7.5	8.6	2.2	1.2	20	16.1	1.5	1.2	3.45	3.67	29.8	77	1283	1527	241.7	
4th	8.6	6.5	1.5	1.6	16.1	18.1	1.20	1.35	3.89	3.49	77	79.3	1527	1196		
5th	7.29	7.3	0.80	0.60	18.9	78.9	1.34	3.98	3.89		89		1196.4	1063		
6th	6.5	7.6	1.6	1.6	18.1	14.3	1.35	1.06	3.49	3.17	79.3	67.1	1063	757		
7th	7.6	5.3	1.6	0.90	14.3	14.5	1.06	1.09	3.17	2.16	67.1	43.1	757	454		
8th	5.7	7.6	0.90	1.5		15.1		1.11	3.16	2.65	43.1	48.3	416	405		

Table 3: Differential diagnosis of patients

	Points in Favor	Points against
PRE ECLEMPSIA (± HEELP)	Thrombocytopenia, anemia, hemolysis AKI, cerebral disturbance, pulmonary edema	Liver enzymes normal; no recovery after delivery AKI usually mild
THROMBOTIC MICROANGIOPATHY	Thrombocytopenia, hemolytic anemia, cerebral involvement, fever present, PT, INR normal, fibrinogen: normal pregnancy may trigger either TTP or C-TMA, TTP (associated with ADMAST-13 Def) second or third trimester. CMTAP: Usually post-partum, CMTAP: AKI usually sever	CM TMA: TPE may not be effective
RENAL COERTICAL NECROSIS	Abrupt onset of oliguria or anuria	Associated with catastrophic obstetric emergencies such as placental abruption with massive hemorrhage or amniotic fluid embolism, no hypoechoic area in renal cortex
NSAIDs induced AKI, acute pyelonephritis, Obstructive uropathy		
APLAS	Poor obstetric history	APLA: negative
ITP	Thrombocytopenia, hemolysis, Any time pregnancy, PT INR normal, fibrinogen normal	No large platelets on PBS creatinine in raised
DIC/Sepsis	Thrombocytopenia, mucosal oozing	Fibrinogen normal, PT aPTT normal
AFLP	Encephalopathy, hypoglycemia, elevated TLC, elevated ammonia	Post-partum presentation, bilirubin normal, SGOT normal

monitored platelet count and serum LDH level. In conclusion, both maternal and fetal survival depend largely on early diagnosis, early initiation of TPE, and close monitoring. In our case, we cannot be sure about confirmation of patient diagnosis, and after TPE, the condition of the patient was significantly improved. Thus, increased morbidity and mortality burden of pregnancy-induced microangiopathies can be decreased through the early management in pregnancy/post-partum with timely initiation of TPE therapy.

Ethical clearance

The Institutional ethical committee, King George's Medical University, Lucknow, INDIA approved my study protocol and procedures of informed consent before the formal survey Ref. code: III PGTSC-II A Thesis/P21.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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