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Therapeutic Plasma Exchange in Postpartum Hemolytic Uremic Syndrome: A Case Report

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ABSTRACT

BACKGROUND AND AIM: Adult, non-infective, haemolytic-uremic syndrome (HUS) although a rare disease in itself, has a high likelihood of occurrence in pregnancy and immediate post partum period. It is an important differential diagnosis in the evaluation of thrombotic microangiopathies. Patients with post-partum HUS display a classical triad of microangiopathic haemolytic anaemia, acute nephropathy and thrombocytopenia.

I hereby present a case of post partum HUS treated with therapeutic plasma exchange (TPE)

MATERIAL AND METHODS: A total of six sessions of TPE were performed daily, three sessions for consecutive days and remaining three sessions were performed on alternate days. All the procedures were carried out with Haemonetics MCS+ exchanging one plasma volume using fresh frozen plasma and saline as replacement fluid. Haemodialysis was started and four sessions were carried out on alternate days.

RESULT: A 37 year old, 85 kg female, G2 P1, underwent emergency LSCS because of foetal distress at 38 weeks of pregnancy. Post surgery she developed decreasing urine output, anuria ensued. Emergency therapeutic plasma exchange was carried out within 24 hours of diagnosis. It could be found that with TPE, patient had improvement in renal function, decrease in LDH levels and increase in platelet count. Patient had sustained remission and discontinuation of haemodialysis.

CONCLUSION: HUS is a disorder with high mortality and long term morbidity, if prompt treatment is not instituted. The decision to intervene with plasma exchange should be based upon the severity of thrombocytopenia, microangiopathic haemolytic anaemia and neurological abnormalities, even if the diagnosis and nomenclature is uncertain. Improved survival after this disorder has been attributed to aggressive treatment with plasma exchange therapy.

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INTRODUCTION

Hemolytic Uremic Syndrome [HUS] of pregnancy most commonly occurs in primigravida, but also has been reported in multiparous women, after an otherwise uncomplicated gestation and delivery [1]. It is a rare disorder with an estimated incidence of approximately 1 in 25,000 pregnancies; however, it is associated with a significant maternal morbidity and mortality [2]. Patients with post partum HUS display a classical triad of acute renal failure, thrombocytopenia and thrombotic microangiopathic haemolytic anemia [MAHA]. Here I report a case of post- partum HUS, occurring on the second post operative day, that was successfully treated with therapeutic plasma exchange [TPE].

CASE REPORT:

A 38 year old [gravida 2, para 1, one living issue] presented at 37 weeks gestation with loss of foetal movements since 3 days for which she underwent emergency caesarean section. Antenatal course was uneventful and there was no history of hypertension, oedema or proteinuria.Patients's vitals were stable throughout the surgery and the baby delivered was healthy. Her Haemoglobin was 11.2 g/dl and platelet count was 67,000/uL.

TABLE I LAB PARAMETERS DURING HOSPITAL COURSE [POST OPERATIVE and POST TPE]

Lab Parameters	Post Op Day 2	Post Op Day 3	Post Op Day 6	Post TPE Day 8	Post TPE Day 10	Post TPE Day 12	At discharge Day 18
Hb (g/dl)	9.8	8.5	8.1	9.8	10.1	10.4	10.8
TLC(*10^3/ul)	19.3	47.1	20.4	13.9	12.6	7.63	5.07
Platelets(*10^3/ul)	22	20	38	78	95	190	196
Urea (mg/dl)	53	145	238	109	95	88	46
Creatinine (mg/dl)	3.26	6.46	5.81	4.76	4.12	2.68	1.63
LDH (U/l)	2558	8540	2703	1185	719	248	189
Haptoglobin (mg/dl)	8.4	6.3	23	45	48	57	59
AST (U/l)	657	740	232		27		24
ALT (U/l)	230	252	125		15		11
Total Bilirubin (mg/dl)	4.2	4.37	1.6		0.65		0.45
Direct.Bilirubin (mg/dl)	2.35	2.71	1.35		0.45		0.35

Hb-Haemoglobin, TLC-Total Leukocyte count, LDH- Lactate Dehydrogenase, AST- Aspartate Aminotransferase, ALT-Alanine Aminotransferase

On the second post operative day, the patient developed oliguria with a urine output of 280 ml, despite adequate hydration and diuretics [Table I], low Haptoglobin and marked elevation of liver enzymes. The blood urea nitrogen level was 53 mg/dl and serum creatinine level was 3.26 mg/dl. Serum bilirubin was elevated [Table I].Fragmented red blood cells [schistocytes and helmet forms] and poikilocytosis seen in peripheral blood film which were significant>5 percent. Urine analysis showed microscopic haematuria.Marked hypertension of 190/105 mm Hg was recorded and was controlled with medications.

On the third post operative day, her laboratory parameters further deteriorated and her 24 hour urine output was 45 ml. Hypertension of 210/103 was noted. The presumptive diagnosis at this stage was HUS with a possibility of HELLP syndrome and acute kidney injury since ADAMTS13 levels were normal. Haemoglobin and platelet count remained low even though no abnormal bleeding was noted and two units of

packed red cells and four platelet concentrates were transfused to correct the parameters.

Keeping in view the clinical deterioration with decreasing haemoglobin, low platelet count, high LDH and renal dysfunction, the possibility of post partum HUS was considered and emergency TPE was carried out on the third post operative day. Haemodialysis was also performed and two sessions were carried out on alternate days.

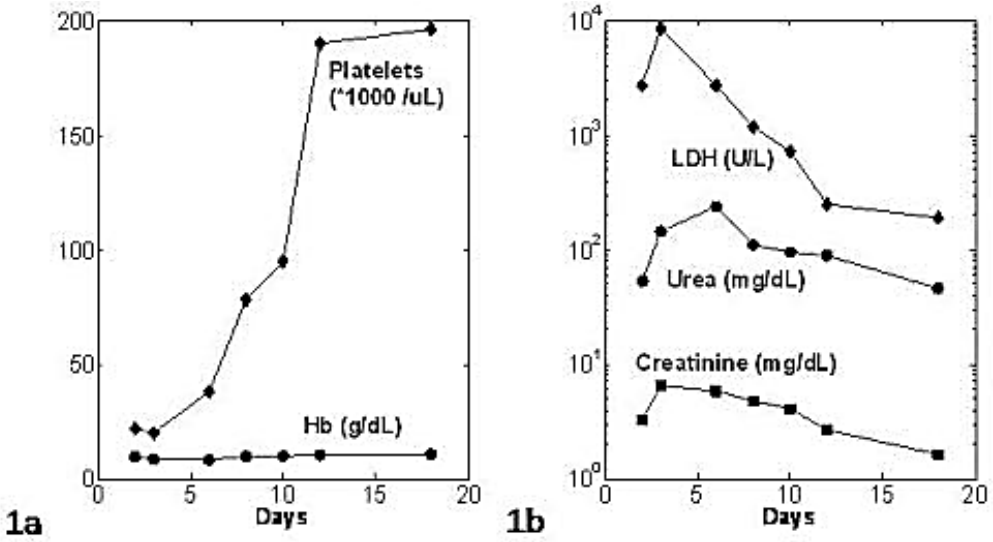

FIGURE 1a: Haematology parameters Haemoglobin [Hb] and Platelets during the Hospital course
FIGURE 1b: Changes in LDH, Urea and Creatinine during the hospital course

A total of six sessions of Therapeutic Plasma Exchanges were performed, three sessions for consecutive days and remaining three sessions were performed on alternate days.All the procedures were carried out with Haemonetics MCS+ exchanging one plasma volume using AB positive Fresh frozen plasma [FFP] and saline as replacement fluid in the ratio of 80:20. [3]

In the 1st session 4 litres of plasma was replaced with 3200 ml of freshly thawed plasma and 800 ml of normal saline. Keeping in view the deranged coagulation profile, freshly thawed Plasma was used in this case instead of Albumin. Ionized calcium prior to the procedure was 1.08 mmol/l.

Daily sessions of Plasma exchange were executed consecutively for 3 days for the patient followed by alternate days for the next 3 days. The replacement fluids used for plasma exchanges were AB positive plasma and 0.9 percent Normal saline with a replacement volume of 3 to 4 litres per session.

Marked decrease in LDH and blood urea was noted. [Figure1]. After two sessions, the patient's general condition improved, 24 hour urine output was 1420 ml with increase in haemoglobin and platelet count and fall in LDH levels.

On the day 18th, patient was discharged from hospital with a haemoglobin of 10.8 g/dl, platelets 1,96,000/cu mm, Creatinine of 1.63 mg/dl and LDH levels of 189 U/L and she was normotensive on follow up after one month.

Dramatic improvement in renal function and haemogram were noted after plasma exchange. [Figure1]. The patient's general condition improved, 24 hour urine output was 1420 ml with increase in haemoglobin and platelet count and fall in LDH levels.

On the day 18th, patient was discharged from hospital with a haemoglobin of 10.8 g/dl, platelets 1,96,000/cu mm, Creatinine of 1.63 mg/dl and LDH levels of 189 U/L and she was normotensive on follow up after one month.

Discussion:

A high index of suspicion is needed to distinguish HUS from Thrombotic Thrombocytopenic Purpura [TTP] and Pre-eclampsia, specifically the variant called HELLP syndrome in pregnant women with thrombocytopenia [4]. HUS and TTP have similar clinical features of hypertension, hemolysis, elevated liver enzymes, low platelet count and renal failure. However TTP ususally occurs in first trimester, while HUS is seen in peripartum or postpartum period [4]. Unlike TTP, HUS has fewer neurological deficits but usually shows severe renal involvement [4].

In 2013, the American Society for Apheresis [ASFA] graded HUS as a level II Grade 2C disease indication for performing TPE that is disorders for which apheresis is accepted as second line therapy, either as a standalone treatment or in conjunction with other modes of treatment [7]

TPE induced a better remission and survival rate at six months compared with plasma infusion, and a higher mortality rate is reported in non-responders [8]. TPE may be initiated even if the etiology of TTP-HUS is not yet confirmed [4]. Authors recommend one daily exchange with FFP until normalization of platelet count and serum lactate dehydrogenase [LDH] concentration. One to two weeks are generally required to induce remission, but the variability between patients is large and unpredictable.

Conclusion:

The early identification of HUS patients along with initiation and maintenance of proper treatment are essential for a favourable outcome in this life-threatening, rare condition. Once the diagnosis is made, prompt and aggressive treatment with Therapeutic Plasma exchange is a successful treatment modality and it can reduce the associated morbidity and mortality of HUS by as much as 90% [6].

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