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Case Report

Therapeutic plasma exchange for early aggressive management of postpartum hemolytic uremic syndrome: a tertiary care centre experience

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ABSTRACT

Postpartum hemolytic uremic syndrome is an unusual condition of obscure origin that manifests with hemolytic anemia, thrombocytopenia and acute renal failure after delivery. We describe a case of 28 year old woman referred to our hospital in view of severe renal failure, 24 hours after the delivery by caesarean section for scar rupture and placental abruption. She was in a delirious state and had anuria, severe anemia and moderate thrombocytopenia. After many diagnostic dilemmas, a final diagnosis of hemolytic uremic syndrome was made. Aggressive treatment with plasma exchange in conjunction with hemodialysis was started. Fresh frozen plasma was used for replacement and four consecutive plasmapheresis sessions were instituted. Simultaneously steroids and anti-hypertensive drugs were given. Two weeks later, quick clinical and laboratory response was noted. There was significant improvement in renal functions along with resolution of signs of active hemolysis. This case collaborates with the ideal scenario involving prompt diagnosis and early aggressive treatment with plasma exchange in a postpartum hemolytic uremic syndrome patient.

Keywords: Hemodialysis, Plasmapheresis, Postpartum hemolytic uremic syndrome

INTRODUCTION

Postpartum renal failure as a part of the clinical spectrum of HUS was initially described in 1955.¹ First case of postpartum HUS was reported by Robson in 1968.² Although it is an idiopathic condition, the inciting cause may involve a pregnancy related hormonal change.¹ History of pre-eclampsia is often associated with the disease.³

Hemolytic uremic syndrome is characterized by the classical triad of acute renal failure, microangiopathic hemolytic anemia and thrombocytopenia. Postpartum hemolytic uremic syndrome is a rare clinical entity and is associated with an apparent poor maternal and perinatal outcome. Diagnosis of postpartum HUS illustrates many clinical dilemmas.

Therefore awareness, prompt diagnosis and aggressive treatment using plasma exchange play a life-saving role in these cases.

CASE REPORT

A 28 year old (gravida 4, para 3) was referred to our hospital in view of severe renal failure, 24 hours after the delivery by caesarean section for scar rupture and placental abruption. She was in a delirious state and had anuria (nil output for 24 hours), severe anemia (Hb 6.3g/dl) and moderate thrombocytopenia (platelet count 20,000/cmm). Her blood pressure was 170/110 mmHg. Previous pregnancy records showed a history of pre-eclampsia in all her pregnancies. Thorough workup was done. Her cardiovascular and respiratory systems and fundus examination were normal. Laboratory parameters showed progressive haemolysis (Hb 5.7g/dl, serum

bilirubin 2.6 mg/dl), low platelets (platelet count 20,000/cmm), renal damage (serum creatinine 2.9mg/dl) and raised LDH (2416 U/l) which showed no improvement despite haemodialysis and blood component therapy. Liver enzymes were within normal limits. After many diagnostic dilemmas, a provisional diagnosis of hemolytic uremic syndrome was made in consultation with nephrologist.

Aggressive treatment with plasma exchange in conjunction with haemodialysis was started. Fresh frozen plasma was used for replacement and four consecutive plasmapheresis sessions were instituted. Simultaneously steroids and anti-hypertensive drugs were also given. Stitch removal was done on 8th post-operative day. Two weeks later, quick clinical and laboratory response was noted. There was significant improvement in renal functions (urine output 1500/ml, serum creatinine 1.1 mg/dl) along with resolution of signs of active haemolysis (Hb 9.8 g/dl, low platelet count 1, 10, 000/cmm, LDH 467 U/l). Thereafter patient was discharged in a satisfactory condition.

DISCUSSION

Postpartum hemolytic uremic syndrome is a relatively uncommon clinical entity characterized by the classical triad of acute renal failure, microangiopathic hemolytic anemia and thrombocytopenia, occurring a few days to approximately ten weeks after an apparently normal pregnancy and delivery.

Various authors in different studies have designated postpartum HUS as irreversible postpartum renal failure, postpartum malignant nephrosclerosis, postpartum renal failure, accelerated postpartum nephrosclerosis, late postpartum intravascular coagulation.^{2,4-6}

The diagnosis of postpartum HUS is difficult due to complicated clinical manifestations, which results in misdiagnosis rates as high as 80% following the initial work up.⁷ The syndrome is known to be associated with a global poor prognosis.⁸ In early reports, most women died, survived with severely impaired renal function or progressed to end stage renal failure; complete recovery of renal function was noted in less than 10% cases.^{2,9} However, few cases of good outcome have been reported in later studies.^{10,11}

This case is interesting as despite diagnostic dilemmas, the patient showed quick clinical and laboratory response after aggressive treatment with plasma exchange. The case also reflects good outcome of postpartum HUS patient with complete remission, return of renal function towards normal along with resolution of signs of active haemolysis.

In recent years, the apparent improvement in survival rates are attributed to earlier diagnosis, better

management of hypertension, timely dialysis and early institution of therapeutic plasma exchange.¹²⁻¹⁴

CONCLUSION

Postpartum hemolytic uremic syndrome is a rare clinical entity and is usually associated with a poor maternal and perinatal survival rates. However, a thorough workup, prompt diagnosis and early institution of aggressive treatment using plasma exchange can be of life saving importance.

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