A Rare Case Presentation of Haemolytic Uremic Syndrome in Post Caesarean Section in Adult Female

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ABSRACT

Hemolytic Uremic Syndrome is characterized by microangiopathic hemolytic anaemia, thrombocytopenia, renal failure, fever and altered sensorium. A 28 years old primigravida patient was admitted on 18-6-2014 with history of 9 MOA and AML of 1 yr with lower abdominal pain since 10-12 hrs & leaking per vaginum. On per abdominal examination, uterus was full-term, cephalic with longitudinal lie. Pt was operated for LSCS for oligohydraminos with all normal investigations. Post-operatively, patient had decreased urine output. HUS was suspected on basis of unexplained thrombocytopenia with normal other coagulation profile, presence of fragmented RBCs, increased serum LDH, unexplained altered renal function, fever in absence of any foci of infection and altered sensorium. Patient was treated with Plasmapheresis, hemodialysis, noninvasive ventilation, other supportive medical treatment and Inj. FFP 8 units, Inj. PCV 3 units.

Keywords: Hemolytic Uremic Syndrome, ADAMTS-13 (A disintegrin and metalloprotease with a thrombospondin type 1 motif, member 13), Plasmapheresis, serum LDH, Hemodialysis.

INTRODUCTION

Hemolytic Uremic Syndrome is characterized by microangiopathic hemolytic anaemia, thrombocytopenia, renal failure, fever and altered sensorium. It is precipitated by infection - acute gastroenteritis & is diagnosis of exclusion with above five features supported with fragmented RBCs, increased LDH, with normal coagulation profile in absence of fever.

CASE REPORT

A 28 years old primigravida patient was admitted on 18-6-2014 with history of 9 MOA and AML of 1 yr with lower abdominal pain since 10-12 hrs & leaking per vaginum. On per abdominal examination, uterus was full-term, cephalic with longitudinal lie, head engaged, with adequate contractions with FHS +/R/144 bpm. On per vaginal examination, dilatation was 1-2 cm, 20-30% effaced, absent membrane. Pt was operated for LSCS for oligohydraminos with all normal investigations. Post-operatively, patient had decreased urine output. It was 300 ml since 12 hrs. Inj. Lasix 40 mg was given, but urine output did not increase. She developed hypotension & tachycardia with normal SpO2. Postpartum patient developed thrombocytopenia and increased creatinine. Then Inj. Noradrenaline was started. She maintained urine output 30 ml/hr and BP 80/40 mm Hg.

Then patient developed breathlessness, tachycardia & tachypnea. Then patient was shifted to ICU and was kept on noninvasive ventilation. HUS was suspected on basis of unexplained thrombocyto-penia with normal other coagulation profile, presence of fragmented RBCs, increased serum LDH, unexplained altered renal function, fever in absence of any foci of infection and altered sensorium. USG Abdo-Pelvis showed renal parenchymal disease, mild ascites, bilateral pleural effusion with consolidation. On blood C/S & urine C/S, no growth seen.

Patient was treated with Plasmapheresis, hemodialysis, noninvasive ventilation, other supportive medical treatment and Inj. FFP 8 units, Inj. PCV 3 units. Patient improved and was discharged vitally stable.

DISCUSSION

Hemolytic Uremic Syndrome (HUS) is a clinical syndrome characterized by progressive renal failure that is associated with microangiopathic (non-immune, Coombs-negative) hemolytic anaemia and thrombocytopenia, fever and altered sensorium.

HUS is the most common cause of acute renal failure in children and is increasingly recognized in adults.[1,2] Childhood HUS and adult HUS have different causes and demographics but have many common features, especially in adults, which include similar pathologic changes such as microangiopathic hemolytic anemia, thrombocytope-nia and neurological or renal abnormalities. Initial therapy in adults is similar for these conditions and includes plasma exchange.

Gasser et al first described HUS in 1955. In 1988, Wardle described HUS and TTP as distinct entities, but in 1987, Remuzzi suggested that these 2 conditions are various expressions of the same entity. With the discovery of von Willebrand factor (vWF)–cleaving metalloprotease ADAMTS-13 (A

disintegrin and metalloprotease with a thrombospondin type 1 motif, member 13), HUS and TTP are clearly different diseases, despite their clinical similarities[3,4,5,6]. Renal circulation in HUS and cerebral circulation in TTP are commonly involved in these thrombotic microangiopathies, likely because of the anatomic restriction of CD36, the thrombospondin receptor. CD36 is found on the human microvascular endothelial cells in a different proportion than on the endothelial cells of large vessels, and this may help to bind ADAMTS-13 to the endothelial cell or the platelet surface, thereby regulating the cleavage of vWF.[7]

Pathophysiology: Damage to endothelial cells is the primary event in the pathogenesis of hemolytic uremic syndrome (HUS). The cardinal lesion is composed of arteriolar and capillary microthrombi (thrombotic microangiopathy [TMA]) and red blood cell (RBC) fragmentation.

Classification: Hemolytic Uremic Syndrome (HUS) is classified into 2 main categories, depending on whether it is associated with Shiga-like toxin or not. Typical HUS (Shiga-like toxin [Stx]–associated HUS [Stx-HUS]) is the classic, primary or epidemic form of Hemolytic Uremic Syndrome (HUS). Stx-HUS is largely a disease of children younger than 2-3 years and often results in diarrhea (denoted D+HUS). One fourth of patients present without diarrhea (denoted D-HUS). Acute renal failure occurs in 55-70% of patients, but they have a favourable prognosis, and as many as 70-85% of patients recover renal function.

Atypical HUS (Non–Stx-associated HUS [non–Stx-HUS]) can be sporadic or familial. As the name implies, infection by Stx-producing bacteria is not the cause, and disease may occur all-round-the-year without a gastrointestinal prodrome (D-HUS). Overall, patients with non–Stx-HUS have a poor outcome, and as many as 50% may progress to end-stage renal disease (ESRD) or irreversible brain damage. Up to 25% of patients die during the acute phase. The familial form is associated with genetic abnormalities of the complement regulatory proteins. Hemolytic-uremic syndrome (HUS) occurs mainly in young children; however, adolescents and adults are not exempt. In young children, spontaneous recovery is common. In adults, the probability of recovery is low when hemolytic-uremic syndrome (HUS) is associated with severe hypertension.

Clinical findings shows prodromal gastroenteritis(83%) - prodrome of fever (56%), bloody diarrhea (50%) for 2-7 days before the onset of renal failure, irritability, lethargy, seizures (20%), acute renal failure (97%), anuria (55%), hypertension (47%), edema and fluid overload (69%).

Hemolytic-uremic syndrome (HUS) predominantly occurs in infants and children after prodromal diarrhea. In summer epidemics, the disease may be related to infectious causes. Causes of the secondary or sporadic form may include pregnancy and pueperium, Quinine - most common cause of drug-induced TTP-[8] oral contraceptives, cancers (chiefly mucin-producing adenocarcinomas), chemotherapeutic agents (Mitomycin-C, Cisplatin, Bleomycin), immune-therapeutic agents (Cyclosporine, Tacrolimus, mTOR inhibitors [eg, Sirolimus], OKT3, interferon [IFN]), antiplatelet agents (Ticlopidine, Clopidogrel), malignant hypertension, collagen-vascular disorder (eg, SLE, antiphospholipid antibody syndrome) - possible to have both true HUS and a lupus anticoagulant, but in most patients, the thrombocytopenia, microangiopathic hemolytic anaemia and renal disease are due to antiphospholipid antibodies rather than true HUS, primary glomerulopathies, transplantation (eg, of kidney, bone marrow, an immunodeficiency-related cause includes thymic dysplasia.

Pregnancy-associated Hemolytic Uremic Syndrome (HUS) occasionally develops as a complication of preeclampsia. Patients may have dysfunction of ADAMST-13 which may precipitate as HUS in post partum period due to stress of delivery. It may progress to full-blown hemolysis, elevated liver enzymes, and low platelets (HELLP syndrome). Postpartum HUS usually occurs within 3 months of delivery. The prognosis is poor, with a 50-60% mortality rate and residual renal dysfunction and hypertension occur in most patients.

Idiopathic Hemolytic Uremic Syndrome (HUS) accounts for 50% of all cases of sporadic non–Stx-HUS. Laboratory study includes CBC, RFT, peripheral blood smear, urine analysis and hemoglobin determination. Severe anemia may be present. Perform peripheral smear for schistocytes (count >1% or 2 or more schistocytes in a 100X magnification field strongly suggests microangiopathic hemolysis).[9] The degree of thrombocytopenia is not correlated with the severity or the length of Hemolytic Uremic Syndrome (HUS) illness. The platelet count usually returns to normal within 2 weeks. Determine activated partial thromboplastin time (aPTT), fibrinogen degradation product (FDP) and D-dimer values. Hemolytic workup: Bilirubin, Lactic Dehydrogenase (LDH) levels may be elevated. Haptoglobin levels may be decreased. Stool culture: Obtain a sample for stool culture, evaluate especially for *E coli* 0157:H7 and *Shigella* bacteria. ADAMTS-13 activity, perform renal ultrasonography in patients with renal failure to rule out obstruction. Biopsy findings clinically establish the diagnosis of hemolytic-uremic syndrome (HUS). However, kidney biopsy is not required in children. In adults, kidney biopsy is rarely required.

Treatment includes plasmapheresis, hemodialysis, antibiotics, supportive treatments like

blood products, iv fluids, cardiac supports, ventilatory supports.

SUMMARY

One may suspect Hemolytic Uremic Syndrome in postpartum period if the patient develops unexplained thrombocytopenia with normal other coagulation profile, fragmented RBCs, increased Serum LDH, unexplained altered RFT, fever in absence of any foci of infection and altered sensorium. Managed by plasmapheresis mainly and other supportive treatment.

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