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SCIENTIFIC LETTER

Sickle Cell Intrahepatic Cholestasis with Acute Liver Failure and Acute Kidney Injury: Favourable Outcome with Exchange Transfusion

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To the Editor: An 11-y-old girl presented with 2 wk of jaundice, right upper abdominal pain and previous history of two episodes of jaundice. Examination revealed markedly deep icterus and hepatosplenomgaly. Laboratory investigation showed evidence of hemolysis (hemoglobin 9.1 g/dl, reticulocytosis 19%, fragmented RBCs, Lactate dehydrogenase 1674 U/L, negative direct and indirect Coombs test), liver dysfunction (total serum bilirubin 68 mg/dl, direct 58 mg/dl, Asparate aminotransferase 100 IU/L, Alanine aminotransferase 200 IU/L, Alkaline phosphatase 660 IU/L, prothrombin time - International normalised ratio 1.6) and Acute kidney injury (AKI) (urea/creatinine 52/1.5 mg/dl, microscopic hematuria). Serology for Dengue, Chikungunya, Hepatitis viruses, Malaria and Leptospira was negative. Within 24 h she progressed to encephalopathy (Grade II), coagulatopathy (INR 2.4), further hemolysis (Hb 5 g/dl), anuria and metabolic acidosis. Considering Acute Wilsonian crises which presents with this constellation of findings (Acute liver failure, Coomb's negative hemolysis, AKI), plasmapheresis (2 cycles) and hemodialysis was initiated. Meanwhile, investigations for Wilsons disease were normal and repeat blood smear demonstrated multiple sickle shaped cells. Acute sickle cell intrahepatic cholestasis (SCIC) was considered. Single volume exchange transfusion (EBT) was done. Post exchange HPLC showed HbS 6.1% (both parents had sickle cell trait). Hydroxyurea was started. She gradually improved over next 45 d and renal and liver function normalized.

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Acute SCIC is a rare and usually fatal complication of sickle cell disease (SCD) and should be suspected when a patient presents with very high level of conjugated bilirubin and right upper quadrant pain. The progression to AKI, coagulopathy and liver failure is rapid unless timely interventions are instituted and hence the index of suspicion should be high. The pathophysiology is, extensive adhesion of sickled RBCs to endothelium causing sluggish flow, sinusoidal congestion, ischemia, infarction and hepatic dysfunction [1–3]. Exchange blood transfusion has been shown to be beneficial by lowering the fraction of HbS [4]. However, even after aggressive EBT and liver transplantation, outcomes may be poor [5]. In light of the published poor outcome in SCIC with encephalopathy, liver failure and renal dysfunction, the case is unique with successful outcome with exchange transfusion, Hydroxyurea and hemodialysis.

Compliance with Ethical Standards

Conflict of Interest None.

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