Case Report

Interesting case of G6PD deficiency anemia with severe hemolysis

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Abstract:

Severe hemolysis was observed in a critically ill patient with G6Pd deficiency where the causative trigger could not be identified. We describe one young patient with severe hemolysis treated with two cycles of plasmapheresis which proved to be an effective tool in the treatment. The patient presented with diffuse pain abdomen, vomiting, yellowish discoloration of sclera and skin and acute breathlessness. Hemoglobin 5.4 mg/dl and total (T) serum bilirubin 17.08 mg/dl: Direct (D) 4.10 mg/dl and Indirect (I) 12.98 mg/dl. Subsequently patient started passing black color urine. As the patient developed severe hemolysis and the trigger agent of hemolysis was unknown, two cycles of plasmapheresis were performed with the aim to remove unknown causative agent. Consequently no trace of hemolysis was found and patient stabilized. Plasmapheresis can be used to treat G6PD deficient patients with severe hemolysis due to unidentified trigger agent.

Key words: Acute hemolytic anemia, Glucose-6-phosphate dehydrogenase deficiency, plasmapheresis, therapeutic plasma exchange

Introduction

Glucose-6-phosphate dehydrogenase (G6PD) is a house keeping enzyme critical in the redox metabolism of all aerobic cells. G6PD deficiency is the most common human enzyme defect. Though majority remains clinically asymptomatic the risk of developing AHA still remains. The three known trigger factors being (i) favism beans (ii) infections (iii) drugs like antimalarial, sulphonamides/ sulphones, antibacterial/ antibiotics/antipyretic/ analgesics etc.[1] We describe the case of young male patient with G6PD deficiency, critically ill with severe hemolysis treated with plasmapheresis, demonstrated sufficient recovery.

Case Report

A 23 year old male patient presented with diffused pain abdomen and vomiting for two days, yellowish discoloration of urine, sclera and skin for one day, acute breathlessness for four hours, fever (100.4°F), pallor, moderate icterus, high pulse rate(124/minute), respiratory rate of 28/minute, cyanosis and reduced oxygen saturation, Hemoglobin 9.6 gm/dl, TLC 38,840/µl with neutrophilia, reticulocyte count 5%, serum bilirubin (T) 24.95 mg/dl, serum bilirubin (D) 3.43 mg/dl, serum bilirubin (I) 21.52 mg/dl , SGPT 61.39 IU/L, SGOT122.48 IU/L,GGT 39.37 IU/L, blood urea 68.62 mg/dl, S.Creatinine 0.62 mg/dl, G6PD (Qualitative)- discoloration in more than 60 minutes, urine – red/ brown, negative screening for malarial parasite and dengue.

In Systemic Examination: Chest: Air entry diminished over bases, CVS: Normal, per Abdomen: mild distension (+) and liver was just palpable which was confirmed by ultrasonography.

Provisional diagnosis: Sepsis with hemolytic jaundice (G6PD def)

Intra venous Meropenam, Teicoplanin administered along with BIPAP support and O2 inhalation.

On day two patient's condition deteriorated. Hemoglobin 5.4 mg/dl and total bilirubin 17.08 mg/dl, s.bilirubin (D) 4.10, s.bilirubin (I) 12.98 mg/dl. Treated with four units of leukocyte reduced packed RBCs. Hemoglobin increased to 8.0 gm/dl but patient started passing black color urine.

Repeat blood grouping, auto control, direct and indirect coombs test were negative.

Final diagnosis: G6PD deficiency with hemolysis.

Results

Patient was administered two cycles of TPE (carried out on Hemonetics Corporation (USA)-MCS+). Total volume processed 9023 ml, total plasma volume extracted 5392 ml, total volume substituted 5393 ml. Color of the extracted product was Black [Figure 1]. Improvement in general condition and parameters noted. Hemoglobin improved to 10.3 gm/dl and Total and Indirect Bilirubin 2.63 gm/dl and 1.53 gm/dl respectively. No trace of hemolysis found. Patient found to be stable and discharged subsequently.

Discussion

As the patient developed severe hemolysis and the trigger agent of hemolysis was unknown, two

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Figure 1: Extracted Plasma

cycles of TPE were performed with the aim to remove unknown causative agent of hemolysis.

Plasmapheresis can be used to treat patients with severe hemolysis in the presence of unidentified trigger agent. Though there is no mention of TPE in the treatment of severe hemolysis due to G6PD deficiency in guidelines,[2] plasmapheresis can be used as an effective tool as demonstrated.

References

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