BLOODLESS MANAGEMENT OF BREAKTHROUGH HAEMOLYSIS IN A PATIENT WITH PAROXYSMAL NOCTURNAL HAEMOGLOBINURIA

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Introduction: Paroxysmal nocturnal haemoglobinuria (PNH) is an acquired clonal haematopoietic stem cell disorder characterized by the triad of intravascular haemolysis, thrombosis and bone marrow failure. Patients with PNH can present with severe anaemia secondary to acute haemolytic episodes, necessitating hospitalization and frequently receive packed cell transfusions. These patients are also iron deficient due to chronic haemoglobinuria. Frequent blood transfusions carry many risks including infections, iron overload and transfusion reactions.

Case Summary: We present the case of a young lady with underlying PNH who presented with a severe haemolytic episode and was successfully treated with intravenous iron and supportive therapy, without blood transfusion. This 26-year-old lady presented with symptomatic anaemia (lethargy, palpitations, presyncope, chest discomfort) and tea coloured urine for 2 days. On admission, her haemoglobin (Hb) was 3.6 g/dL, with indirect hyperbilirubinaemia and markedly elevated lactate dehydrogenase (LDH) of 4100 U/L. She was treated with intravenous iron sucrose, folate, hydration, supplemental oxygen and antibiotics to cover for occult infection. She did not receive any blood transfusion throughout her 12-day hospitalization. Her haemoglobin and other haemolytic parameters gradually improved during her stay. Her haemoglobin upon discharge was 8.7 g/dL, with normalization of indirect bilirubin and reduction in LDH to 1812 U/L.

Conclusion: This case demonstrates the successful use of intravenous iron and avoidance of blood transfusion in a PNH patient presenting with acute haemolysis and provides an excellent example of how principles of bloodless medicine can be utilized in everyday practice.

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