

Hyperhaemolysis Syndrome in a Patient with Delta-Beta Thalassaemia

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Hyperhaemolysis syndrome is a life threatening delayed haemolytic transfusion reaction which leads to destruction of transfused and autologous erythrocytes characterized by reticulocytopenia and post transfusion haemoglobin is lower than pre transfusion haemoglobin. It is common in sickle cell disease but rare in other haemoglobinopathies. A 25-year-old female with delta-beta thalassaemia managed as infectious mononucleosis was found to have haemoglobin of 6.9g/dl and one pint of blood transfusion was given. A week after transfusion, patient presented with dark coloured urine and symptomatic anaemia and was found to have haemoglobin of 4.71g/dl, indirect hyperbilirubinaemia and reticulocytopenia. Initial direct Coombs test was negative and repeat one was positive with C3d. Twenty one units of red cells were given but the patient continued to show evidence of haemolysis. She was successfully managed with life-saving red cell transfusion under IVIG cover and further red cell transfusions were avoided and managed with prednisolone, cyclosporine, IV rituximab and subcutaneous erythropoietin therapy. In conclusion, early identification of hyperhaemolysis syndrome, avoidance of blood transfusions and prompt start of immunosuppressive therapy is life saving. Blood transfusion should be done under IVIG or steroid cover in life threatening anaemia.

Keywords: Hyperhaemolysis syndrome, Delta-beta thalassaemia, Immunosuppressant