

ABSTRACT

We present a 16 year newly diagnosed delta beta Thalassaemia patient who developed dropping haemoglobin to less than pre transfusion value with evidence increased haemolysis and haemoglobinuria eight days after red cell transfusion. She was ill, severely pale, icteric and had moderate hepatosplenomegaly. She had severe anaemia , hyperbilirubinaemia and evidence of haemolysis in the blood picture. Her direct anti globulin test was positive with C3d specificity, but red cell alloantibodies were negative. Despite the ongoing haemolysis she had a reticulocytopenia. She was diagnosed to be having hyperhaemolysis syndrome and was managed with steroids, rituximab and erythropoietin to which she responded.