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HEMOGLOBINURIA FOLLOWING BLOOD TRANSFUSION IN SICKLE CELL HOMOZYGOUS PATIENT: DELAYED TRANSFUSION REACTION; A CASE REPORT

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Abstract

The sickle hemoglobinopathies are hereditary disorders in which red cells contain Hb-S. Hb S (a2b2 6 Glu-Val) polymerizes reversibly. Alloimmunization with sickle cell disease (SCD) has A very low incidence report.

Keywords:
hemoglobinuria, sickle cell anemia, direct coombs test, transfusion reaction.

Introduction

The sickle hemoglobinopathies are hereditary disorders in which red cells contain Hb-S. Hb S (a2b2 6 Glu-Val) polymerizes reversibly when deoxygenated to form a gelatinous network of fibrous polymers that stiffen the RBC membrane. 1

Alloimmunization with sickle cell disease (SCD) has A very low incidence report. One complication of alloimmunization is delayed haemolytic transfusion reaction/ Hyperhemolysis (DHTR/ H) syndrome2. In patients with SCD, clinical finding in DHTR/H syndrome occurs approximately one week after RBC transfusion and includes onset of increased hemolysis associated with pain and profound anaemia3. Hb levels often drops below the pre-transfusion values. In many reported adult cases, Direct Antiglobulin Test (DAT) remains negative and no new alloantibody is detected as cause for these transfusion reactions. Here we report a case of DAT remains positive following treatment with 3 units of blood transfusion in sickle cell disease.

Case report

A 17 year old female presented with fever, yellowish discoloration of urine and conjunctiva, pain abdomen for last seven days. Fever was low grade with no diurnal variation, not associated with chills or rigor, dysuria ,joint pain, rashes, headache, vomiting . Patient had history of multiple episodes of blood transfusion (approx 12 units)in the past and is a diagnosed case of sickle cell anemia 7 years back.

On examination the lady was found to vitally stable with severe pallor. Icterus was present. Per abdominal examination revealed non tender hepatomegaly with liver span being 12 cm, splenomegaly of 3 cm. Other systems being normal.

Investigation revealed Hb: 5.3gm%, TLC: 26560/mm3 with N44.2, E2.8, L50, TPC: 2.8Lakh/mm3. . Renal function test revealed sr urea 41 and creatinine as 1.9. liver function tests revealed SGOT 729, SGPT 137, ALP 308, Total Bilirubin 2.9, Direct Bilirubin 0.6.sr protein was found to be 6.2g/dl

Patient was given 2 units of blood transfusion. After seven days, colour of the urine became cola coloured. Repeat investigations revealed Hb 2.5gm%, TLC 23540/mm3, N 40.6, L 56.2, E 0.4, M2.8 and Nucleated RBCs 12.7%. urine occult blood test was positive. Usg report revealed hepatomegaly and splenomegaly with inhomogenous echotexture with no evidence of any gall stones. Direct Coombs test was positive. The patient was started on low dose oral steroids for 3 days following which the urine colour normalised so also the haematological parameters. The patient was discharged with haematinics.

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Discussion

Alloimmunization in SCD has a reported incidence of 5% to 36% 4. Delayed haemolytic transfusion reaction/ Hyperhemolysis (DHTR/H) which has incidence of 11%, approximately occurs after 1 week of blood transfusion5.haemoglobin often drops below the pre-transfusion level. In many cases, DAT remains negative and no new alloantibody is detected as cause for tissue transfusion reaction 6.

Conclusion

This report clearly demonstrates that hemoglobinuria develops after Anti-S antibody development, following one week of blood transfusion. Many such cases of transfusion reaction goes undiagnosed and unreported and patient comes repeatedly for blood transfusion thinking it to be due to the disease per se.

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