**International Journal of Medical Science and Clinical Inventions 4(3): 2779-2780, 2017**
**DOI:10.18535/ijmsci/ v4i3.10** **ICV 2015: 52.82**
**e-ISSN: 2348-991X , p-ISSN: 2454-9576**


**Case Report**

# Alloimmunisation in Sickle Cell Disease Presenting As Haemolytic Crisis

***Priyansh Bhayani[1], Sourya Acharya[2], Samarth Shukla[3]***

[1]Resident Internal Medicine JNMC Sawangi (Meghe) Wardha
[2]Professor Department of Internal Medicine JNMC Sawangi (Meghe) Wardha
[3]Professor Department Of Pathology Sawangi (Meghe) Wardha

Datta Meghe Institue Of Medical Sciences(DU), Jawaharlal Nehru Medical College Sawangi (Meghe)Wardha-442004, Maharashtra

**Address for Correspondence:** Priyansh Bhayani
F-1 Raghobaji PG boy's hostel JNMC Sawangi Meghe Wardha

**ABSTRACT:**
Bystander haemolysis is a devastating consequence of delayed haemolytic transfusion reaction (DHTR) in cases of sickle cell disease receiving multiple transfusions. This can be potentially a life-threatening complication, which should be kept in mind while treating patients of sickle cell disease with haemolytic crisis.
We present a case of a 22-year-old male who presented to us with symptoms of severe anaemia; later, diagnosed as DHTR and managed accordingly.

**Key Words**: **DHTR, sickle cell disease, anaemia, transfusion.**

## INTRODUCTION:

Sickle cell disease is a chronic haemolytic anaemia with various compound heterozygote genotypes. The clinical manifestations range between haemolytic and vaso occlusive sub phenotypes. [1] Patients of SCD frequently require RBC transfusions because it is an important treatment modality. Basically, it increases oxygen carrying capacity and improves the physiologic property of blood in these patients [2]

Absolute indications for transfusion are: Aplastic crisis, severe anaemia causing haemodynamic compromise, cardiac failure and splenic sequestration.

Exposure to multiple RBC transfusions, may put the patient for the risk of infections, iron overload, and DHTR because of alloimmunisation. DHTR can be potentially serious and should be identified at an early stage because it can masquerade hyper haemolytic crisis [3]

## CASE REPORT:

A 22-year-old male, known case of sickle cell disease for 15 years, presented to us with chief complaints of increasing fatigue, lassitude, palpitations, pre-syncope and chest pain of 10 days' duration. 1 month back he was admitted for painful crisis in another hospital and was transfused 2 units of packed red cell. There was no history of fever, bone pains, cough, haemoptysis, urinary complaints. Examination revealed pulse of 115/min regular high volume, blood pressure 100/70 mmhg, severe pallor was present, icterus was present. JVP was normal. There was no Petechiae, gum bleeding, oral ulcers, leg ulcers. Per abdomen, CVS, RS were within normal limit.

**INVESTIGATIONS:** Hb-3.2 gm/dl, bilirubin 12 mg/dl unconjugated 8 mg/dl, LDH-4000 U/L, direct and indirect coombs test were positive. Haemoglobin electrophoresis revealed HBA – 0%, HBF-30%, HBA2-3.1% AND HBS-66.9% suggestive of sickle cell disease pattern. Reticulocyte count was 6%.

The discharge document of the hospital where he received blood transfusions, showed initial haemoglobin (pre-treatment was 5 gm/dl and after two transfusions was 8.2 gm/dl) Reticulocyte count (Pre-treatment 25 % and Post treatment was 18%). In view of severe decrease in the haemoglobin level along with reticulocyte count, a probable diagnosis of DHTR was kept and antibody study was done which revealed ANTI JK ANTIBODIES BY AHG GEL CARDS. The patient was treated with corticosteroids, antigen negative blood units by antigen typing. After 5 days, his haemoglobin was stable at 7 gm/dl. Patient was discharged and 2 weeks follow up haemoglobin level was 8.1 gm/dl.

**DISCUSSION:** DHTR is a dangerous complication especially in patients of sickle cell disease. It typically occurs around 1-2 weeks' post transfusion and presents with fever, abdominal and back pain. Severe anaemia deepening icterus and haemoglobinuria [4]. It occurs due to stimulation of alloantibody to a foreign surface antigen on the transfused RBCs. It is sometimes confused with hyper haemolytic crisis which is another manifestation of sickle cell disease patients.

Prevalence and risk factors: Alloimmunisation rate in SCD

has been reported to be around 19-47 % in adult patients [2]. Ironically these antibodies may become undetectable in a substantial number of cases [5].

Table 1 shows risk factors FOR DHTR in SCD patients.

| | |
|---|---|
| 1. | Old age at time of 1st transfusion |
| 2. | Female sex |
| 3. | Racial difference (Afro-American recipients/Caucasian donors) |
| 4. | RBC antigens E , C , Kell , $Fy^a$ , $Fy^b$ AND $JK^b$ [6] |
| 5. | HLA DRB1 1503 is associated with worst prognosis and HLA DRB1 0901 is protective |
| 6. | ? Inflammation. |

Decreased haemoglobin levels with reticulocytopenia indicates DHTR.

PATHOPHYSIOLOGY: Bystander haemolysis mechanism is the cornerstone for DHTR in SCD. Immune haemolysis of RBCs that are negative for the antigen against which the culprit antibody is directed and destroys patients own cells and the transfused red cells by activating complement. Suppression of erythropoiesis that accompanies transfusion also contributes to the anaemia which follows DHTR [7, 8 ,9,10,11]. The situation is further aggravated because patients of SCD already have shortened RBC survival. Some case reports also suggest that rather than suppression of erythropoiesis increased peripheral consumption of RBCs may be another mechanism [12].

DIAGNOSIS: DHTR is diagnosed based on appropriate history and laboratory testing. New onset painful crisis, decreasing haemoglobin levels approximately 1-2 weeks following transfusion of RBC, reticulocytopenia, haemolytic jaundice, positive coombs test (direct and indirect) and increased LDH are the indicators of DHTR. Urine examination may show haemoglobinuria suggestive of intravascular haemolysis. In many cases antibody screening remains negative [13].

TREATMENT: Whenever DHTR is encountered RBC transfusion should be withheld temporarily because it may precipitate acceleration of haemolytic reaction. High dose corticosteroids appear to improve the situation. Resistant cases may benefit from IVIG therapy. Rituximab a monoclonal antibody that targets B cells by binding CD20 allows successful transfusion [14]. Post transfusion haemoglobin levels should be kept below 10 gm/dl because more RBC transfusion increases blood viscosity thereby paradoxically decreasing the oxygen carrying capacity.

CONCLUSION: DHTR is an important complication of transfusion in SCD. A high index of suspicion should be kept while treating SCD patients who deteriorate with severe anaemia and features of haemolysis usually after transfusions. Once DHTR is detected further transfusions should be withheld unless necessary. Treatment is with high dose steroids with or without immunoglobulin.

**References:**


**[1]** Kato GJ, Gladwin MT, Steinberg MH. Deconstructing sickle cell disease: Reappraisal of the role of hemolysis in the development of clinical subphenotypes. Blood Rev 2007;21:37-47.

**[2]** King KE, Ness PM. Treating anemia. Hematol Oncol Clin North Am 1996;10:1305-1320.

**[3]** Wayne AS, Kevy SV, Nathan DG. Transfusion management of sickle cell disease. Blood 1993;81:1109-1123.

**[4]** Talano JA,Hillery CA,Gottschall JL,Baylerian DM, Scott JP .Delayed haemolytic transfusion reaction/hyperhemolysis syndrome in children with sickle cell disease.Pediatrics 2003;111(6 Pt 1):e661-5

**[5]** Vichinsky, E. Transfusion. In: Embury, SH.; Embury, SH.; Hebbel; Mohandas; Steinberg, editors. Sickle Cell Disease: Basic Principles and Clinical Practice. New York: Raven Press; 1994. p. 781 -798.

**[6]** Vichinsky EP, Earles A, Johnson RA, et al. Alloimmunization in sickle cell anemia and transfusion of racially unmatched blood. N Engl J Med 1990;322:1617-1621.

**[7]** King KE, Shirey RS, Lankiewicz MW, et al. Delayed hemolytic transfusion reactions in sickle cell disease: Simultaneous destruction of recipients' red cells. Transfusion 1997;37(4):376—381.

**[8]** Garratty G. Severe reactions associated with transfusion of patients with sickle cell disease. Transfusion 1997;37:357-361.

**[9]** Petz LD, Calhoun L, Shulman IA, et al. The sickle cell hemolytic transfusion reaction syndrome. Transfusion 1997;37:382-392.

**[10]** Diamond WJ, Brown FL Jr, Bitterman P, et al. Delayed hemolytic transfusion reaction presenting as sickle-cell crisis. Ann Intern Med 1980;93:231-234.

**[11]** Cox JV, Steane E, Cunningham G, Frenkel EP. Risk of alloimmunization and delayed hemolytic transfusion reactions in patients with sickle cell disease. Arch Intern Med 1988;148:2485-2489.

**[12]** Win N, Doughty H, Telfer P, Wild BJ, Pearson T. Hyperhemolytic transfusion reaction in sickle cell disease. Transfusion 2001;41:323-328.

**[13]** Rosse WF, Narla M, Petz LD, Steinberg MH. New views of sickle cell disease pathophysiology and treatment. Hematology 2000;2000:2-17.

**[14]** Noizat-Pirenne F, Bachir D, Chadebech P, et al. Rituximab for prevention of delayed hemolytic transfusion reaction in sickle cell disease. Haematologica 2007;92:e 132-e 135.