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Transfusion Medicine

FIRST EXPERIENCE OF AUTOMATED RED CELL EXCHANGE IN SICKLE CELL CRISIS : A CASE REPORT FROM TERTIARY CARE HOSPITAL

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ABSTRACT **Background:** Sickle cell disease is an inherited autosomal recessive blood disorder. Sickle cell anaemia (SCA), is a chronic disorder having qualitative defect in globin chain. It is caused by a single mutation & substitution of valine with glutamic acid at sixth position in beta globin gene resulting in abnormal haemoglobin Hb –S. (1) Red Cell Exchange(RCE) is removal of a patient's red blood cells while replacing with donor red blood cells either manually or using automated systems.(2) In SCD Automated Red Cell Exchange is an evolving technique which prevents new vaso - occlusive events by removing HBSS & HBSβ Cells and provides added oxygen carrying capacity without increasing viscosity of blood.(3) **Case Report:** We report our first experience of automated red cell exchange in 20 year old female diagnosed case of sickle cell disease presented to us with complaints of chest pain, breathlessness at rest, pain in both upper limbs, vomiting, diarrhoea. Red Cell Exchange was planned to tide over the acute sickle cell crisis and provide symptomatic improvement.

KEYWORDS : Sickle cell crisis, Apheresis, Automated Red Cell Exchange.

INTRODUCTION:

Sickle cell disease is an inherited autosomal recessive blood disorder**.** Sickle cell anemia (SCA), is a chronic disorder having qualitative defect in globin chain. It is caused by a single mutation & substitution of valine with glutamic acid at sixth position in beta globin gene resulting in abnormal haemoglobin HbS.[1]

Red cells containing HbS
↓
Passage through microcirculation of spleen
↓
Low oxygen tension
↓
Deoxygenation of HbS
↓
Sickling occurs
↓
Cells pass through circulation with good oxygen tension (other organs)
↓
Desickling
↓
Various cycles of sickling &desickling
↓
Cell membrane affected & change in membrane permeability
↓
Irreversible sickling
↓
SICKLED RBCs[2]

These sickle shaped RBCs get sequestered in the spleen leading to hemolysis & congestion followed by infarction of the organ known as hyposplenism and phenomena known as AUTOPHAGY.[1] Avascular Necrosis (AVN) of femoral head in homozygous sickle cell disease is seen frequently in Indian patients. It results from recurrent episodes of vaso- occlusion & ischemia. Crisis in sickling syndromes are: 1) vaso-occlusion crisis, 2) Hemolytic crisis, 3) Aplastic crisis and 4) Sequestration crisis.[2]

Main stay of treatment for sickle cell anaemia (SCA) was transfusion therapy to keep haemoglobin threshold above 8 g/dl. But repeated transfusions put the patient at risk of iron overload. [3] Red Cell Exchange (RCE) is removal of a patient's red blood cells while replacing with donor red blood cells either manually or using automated systems.[4] In SCD, Automated Red Cell Exchange is an evolving technique which prevents new vaso-occlusive events by removing HBSS & HBSβ Cells and provides added oxygen carrying capacity without increasing viscosity of blood. RCE can also be used as a long term therapy to maintain a low level of HbS in primary & secondary stroke prevention in patients with homozygous SCD. RCE is also employed in patients infested with malaria & Babesiosis, where parasitic index remains high despite medical management.[3] Here we are reporting a case of sickle cell disease successfully managed with automated red cell exchange.

Case Report

A 20 year old female patient, known case of sickle cell disease since last 10 years, Resident of Vadodara –Gujarat, presented with complaints of chest pain, breathlessness at rest and pain in both upper limbs. She also complained of pain in abdomen associated with vomiting &diarrhea. Patient had similar episodes in the past with history of easy fatigability and upper respiratory infections. Patient had history of left total hip replacement (THR) surgery in year 2019 and right THR in year 2021 due to avascular necrosis of femoral head. She needs average blood transfusion of 3-4 times/year and the last one was 6 months ago.

On general examination she was well oriented & conscious to time, place and person. She had pulse rate of 86 beats/minute, Sp02 of 98% on room air and blood pressure of 110/70 mmHg. On auscultation, bilateral equal air entry were present, crepitations & rhonchi were absent. S1S2 heart sounds were regular & murmur was absent. On per abdominal examination, abdomen was soft and non tender.

Laboratory findings as in Table 1 showed severe anaemia with sickle cells on peripheral smear. Two red cell concentrate transfusions were done on 2nd day of admission to correct anaemia. High performance liquid chromatography (HPLC) showed Hb-S =82.90 % , Hb-F =12.1 %, Hb-A=1.8 %, Hb-A2 =2.7 %. Patient was second order female child with no similar complains in other family members. Her father and mother were screened for sickle cell disease by HPLC and were found to be sickle cell trait.

In a view of persisting symptoms and discussion with clinician, Automated red cell exchange was planned. On 10th day of admission, an internal jugular line was secured and red cell exchange was started on automated cell separator. Before the start of procedure, routine blood bank reports were done which suggested blood group = B positive, Direct and indirect Coombs test = negative. The blood volume was replaced with 1356 ml (5 units of packed red cell) of Hb-S negative (tested by Sickle Solubility Test), leucoreduced Rh matched and cross match compatible packed red cell having an average haematocrit of 53%. During the procedure, patient received 233 ml of acid citrate dextrose as anti-coagulant. Whole procedure was completed in 110 minutes and patient was comfortable and did not show any signs of further deterioration.

Complete blood count , liver function test, renal function test and HPLC were repeated after 24 hour as shown in Table 1, showing

improvement in laboratory values. Patient was relieved of chest pain and bone pain immediately after the procedure. On 5th day of post procedure, patient was discharged on hydroxyurea twice daily, folic acid, multivitamin supplements. On discharge she was asked for monthly follow up for next 3 months with Hb-S load.

TABLE-1. Comparison of laboratory parameters at the time of admission, before and after red cell exchange.

PARAMETERS	AT THE TIME OF ADMISSION	BEFORE RED CELL EXCHANGE	AFTER RED CELL EXCHANGE
Hb (g/dl)	7.20	10.19	11.38
TLC (cumm)	7500	7700	7900
DLC (cell%) N	70	67	74
L	28	30	24
E	01	02	01
B	00	00	00
M	01	01	01
PLATELET (cumm)	1,75000	1,65000	1,26000
PCV (%)	30.27	33.96	28.88
TOTAL RBC (million cells/µl)	4.0	4.48	3.44
MCV (fl)	75.62	75.88	83.97
MCH (pg)	22.79	22.79	27.28
MCHC (g/dl)	30.14	29.99	32.49
RETICULOCYTE COUNT (%)	4	3.9	0.5
UREA (mg/dl)	12	18	16
CREATININE (mg/dl)	0.69	0.81	0.82
TOTAL BILIRUBIN (mg/dl)	0.90	0.80	0.75
TOTAL PROTEIN	7.20	7.90	7.70
DIRECT BILIRUBIN (mg/dl)	0.40	0.30	0.28
AST(u/l)	58.0	35	32
ALT (u/l)	9.0	10	10
ALP (u/l)	194	201	139
Na+ (meq/l)	139	137	138
K+ (meq/l)	4.10	4.20	3.50
LDH (u/l)	1039	992	505
Hb-S (%)	82.90	-	23.10
Hb-F (%)	12.10	-	3.70
Hb-A (%)	1.80	-	60.90
Hb-A2 (%)	2.70	-	2.70

DISCUSSION:

Red cell exchange (RCE) (erythrocytapheresis) is an effective important component of the management of sickle-related complications. RCE is superior to simple transfusion in that it reduces the percentage of circulating sickle cells while increasing the red cell oxygen carrying capacity, without causing a corresponding increase in blood viscosity and body iron. Increased blood viscosity alters blood rheology and predisposes to ischemic adverse events such as vaso-occlusive crisis and stroke while iron overload causes marked organ tissue damage, with an increase in disease morbidity and mortality.[7]

American society for apheresis advises RCE to be used as a first line or adjunctive second line therapy for cerebrovascular accidents and acute chest syndrome (ACS).[6] Documented effects of RCE in the setting of ACS include improved tissue oxygenation, changes in Hb oxygen affinity and blood oxygen pressure, increase in transcutaneous oxygen saturation (SpO2), increased HbA content with reduced HbS fraction and inflammatory markers.[7]

RCE is relatively safe procedure, yet the patient is at risk for transfusion-associated adverse events in addition to apheresis risks.[2]

CONCLUSION :

Automated red cell exchange done in our patient proved to be a lifesaving modality with rapid reversal of symptoms and near normalization of laboratory parameters.

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