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SUCCESSFUL MANAGEMENT OF SICKLE CELL ANEMIA WITH RECURRENT VASO OCCLUSIVE CRISIS IN PREGNANCY: A CASE REPORT

Gynaecology

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ABSTRACT

Sickle cell disease is a devastating abnormality of RBC's that results in circulatory impairment in tissue damage, infarctions, severe anaemia and life threatening infections (Yawn et al., 2014). During pregnancy maternal problems can arise from pre-existing chronic underlying organ dysfunction such as renal disease or pulmonary hypertension, from acute complications of SCD such as vaso occlusive crisis and acute chest syndrome. Here we are discussing a case of 19yr old primigravida, a known case of Sickle cell disease with beta- Thalassemia trait having multiple vaso occlusive crises throughout pregnancy that required multiple blood transfusions and ICU admissions. However pregnancy was continued till term with successful outcome due to joint efforts of obstetrician and physician.

KEYWORDS:

Sickle cell anemia, Thalessemia, Vaso-occlusive crisis

Introduction

Sickle cell anemia represents the homozygous state of the Hb S gene inherited from each parent who has either a sickle cell trait or disease. The first report in the literature of pregnancy in women with SCD was published in 1924 by Sydenstricker who briefly described two cases of pregnant women with sickle cell disease. A point mutation replaces the hydrophilic glutamic acid with hydrophobic valine in the sixth position of the amino acid chain of globin chain. This mutation yields an irregular structure causing recurrent damage to the RBC membrane and is responsible for the "sickle" shape of the erythrocytes in this disease (Huntsman, 1976; Oteng-Ntim, Cottee, Bewley, & Anionwu, 2006). These inflexible irregularly shaped cells lack capacity to travel through the microvasculature leading to decreased perfusion and ultimately vaso occlusive crisis. The HbS molecule is prone to auto oxidation and consequent oxidant damage to the cell membrane resulting in alteration in lipid bilayer dynamics which increases the clinical severity of vaso occlusive crisis (Kobak, Stein, & Daro, 1941; LEHMANN & CUTBUSH, 1952). About 20% of sickle cell patients have frequent, severe crisis that require parenteral narcotics and hospitalization, 40% suffer a single painful crisis and remaining 40% rarely experience an episode of painful crisis (Rahimy et al., 2000; TUCK, STUDD, & WHITE, 1983). Thalassemia is of 2 types - alpha and beta - of which alpha can cause anemia and beta - Thalassemia makes it difficult to become pregnant without fertility treatment (Cao & Galanello, 2010).

Case Report

19yr old Primigravida, known case of Sickle cell anemia (HbS 80.6%) and Beta thalassemia double heterozygous trait, detected at the age of 14yrs. At the age of 6 yrs patient had complaints of multiple joint pain, fever and was diagnosed as worm infestation - 2units whole blood given and deworming done. At the age of 8yrs patient had similar complaints with generalised edema – 2 units whole blood given. Patient was asymptomatic from 8-14 yrs of age. At 14 yrs of age patient again complained of multiple joint pain and fever and was diagnosed as Sickle cell anaemia with Thalessemia trait with Hb electrophoresis suggestive of HbS - 80.6% , Hba2 – 5.1%, HbF - 5.7%, HbA0 - 8.6%, Synovial fluid ADA (Adenosine deaminase) - 131, USG s/o Hepatosplenomegaly. Anti malarials were given and Cap Hydroxyurea BD was started. Thereafter patient had multiple hospital admissions for sickle cell crisis and received 12 blood transfusions till she got married (non consanginous marriage) on 3/4/2016. Husband's Hb electrophoresis was normal. She conceived immediately.

M/H: LMP 20/7/2016, EDD 27/4/2017

Since 1st trimester, patient had multiple episodes of vaso-occlusive crisis for which she had multiple admissions under medicine (2 times CCU admission) with total 16 units of blood transfusion. ANC records are as mentioned in Table 1

Since 34 weeks of gestation, she was kept in antenatal ward for close maternal and fetal monitoring. Investigations were done regularly including Complete blood count, Liver function test, Renal function test, Urine Routine Microscopy, Urine culture sensitivity, Blood culture was sent weekly, strict Intake/Output charting was kept. Higher IV antibiotics were given along with Inj Multivitamin. As fetus had Inta Uterine Growth Retardation, daily Fetal heart sound monitoring, biweekly NST and weekly Doppler was done. Her investigations were as follows: Hb 8, WBC 6500, Plt 3 lakhs, Reticulocyte count 10.2, Sr Ferritin TIBC Transferrin LFT RFT URM PT INR WNL, USG s/o SLIUG of 37weeks with cephalic presentation of Wt 1.8kg 2

Table 1 ***ANC Records.***

AN C	Complaints	Ix	BT	T/T
5wks	Fever	Hb 6.4 WBC 22000 Plt 3lakhs	2 P CV	Antimalarials,Antibiotics, Folic acid supple- mentation C.Hydroxyurea stopped
11wks	Joint pain, body ache	Hb 6.8 WBC 8800 Plt 3.2lakhs	1PCV	Folic acid supplementation, Deworming
14wks	Joint pain	Hb 6.4WBC 8900 Plt 2.2lakhs	3PCV	Antibiotics, Folic acid supplementation
29wks	Joint pain, body ache	Hb 6.1WBC 22000 Plt 4lakhs	4PCV	ICU admission - Antibiotics, Folic acid supplementation
32wks	Joint pain, Fever	Hb 7 WBC 15000 Plt 3lakhs	4PCV	ICU admission - Antibiotics, Folic acid supplementation
34wks	Joint pain	Hb 6.9	2BT	Antibiotics, Folic acid supplementation

with Normal Doppler study. Patient went into labor spontaneously at 38 weeks. She was kept on IV fluids for hydration along with oxygen supplementation and strict FHS monitoring. Patient delivered a female baby of 1.9kg on 12/4/17 at 11.09pm vaginally uneventfully. Baby was kept in NICU for investigations and observation for 3 days. Post-delivery Hb was 7. Two units PCV was given and she was discharged with baby after Hb was build up to 9.

Discussion

This patient presented with severe anemia in pregnancy, this could have been as a result of hemolysis and/or aplastic crisis due to SCD or could have been due to increased demand and hemodilution due to pregnancy itself. Routine prophylactic transfusion is no longer recommended in pregnant patient with SCD (LEHMANN & CUTBUSH, 1952; Oteng-Ntim et al., 2006). Hydroxyurea is strongly recom-mended in patients having more than three vaso occlusive crisis per year, however it is generally not prescribed to preg-nant women and advised to discontinue if patient is already taking the drug (Cunningham & Williams, 2005; Huntsman, 1976). In this case, Hydroxyurea was stopped at 5 weeks. The preferred method of delivery for such patients is vaginal delivery with proper hydration and oxygenation along with close intra partum monitoring which was done

for this patient to prevent sickling crisis. Studies from multiple cohorts, including large population studies, have documented the increased risk of IUGR (asymmetric), Preterm delivery and stillbirth. In our case, it was a symmetric term IUGR (Firkin & De Gruchy, 1989; Huntsman, 1976; Rahimy et al., 2000). With multi-disciplinary approach in a tertiary care center such critical patients can be managed successfully.

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