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Case Report

Sickle Cell Anemia Diagnosed in Pregnancy

Dr. Pravallika Vellanki, Dr. Meenakshi Devarmani*

M.B.B.S, PG Resident in M.S. Obstetrics & Gynecology, Mahadevappa Rampure Medical College, Kalaburagi- 585105

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***Corresponding author:** Dr. Meenakshi Devarmani

Abstract

Introduction: Sickle cell disease (SCD) results from the substitution of a valine residue for glutamic acid at position 6 in the beta-subunit of hemoglobin. It is characterized by chronic hemolytic anemia, vaso-occlusive process and multiple organ infarction resulting from wide spread vascular occlusion. Sickle cell disease is the most common inherited disorder worldwide. Pregnancy in sickle cell disease is at very high risk. Many reports have documented a considerable maternal risk of morbidity and mortality and high peri-natal adverse outcomes. ***Case report***: 26 year old G3P1D1A1 presented with 39 weeks 4 days gestation with gestational hypertension with mild tricuspid regurgitation with pulmonary hypertension. She was diagnosed with sickle cell disease. ***Results***: Patient is taken for emergency LSCS and transfused with 1 pint packed cells along with Thrombo prophylaxis. Both mother and baby are doing well till date. ***Conclusion:*** This case report highlights the fact that anemia in pregnancy should be evaluated systematically and early diagnosis and effective multidisciplinary approach will yield a better maternal and neonatal outcome.

Keywords: Sickle cell disease, Pregnancy, Anemia, Maternal mortality.

INTRODUCTION

Sickle cell disease (SCD) is one of the most common genetic disorders globally, originating in sub-Saharan Africa and the Middle East. It is a group of inherited auto-somal recessive hemoglobinopathies characterized by hemolytic anemia and vaso-occlusive complications. It affects 10000-15000 people in the UK. The mortality rate for women with SCD is 72 deaths per 100 000 deliveries [1].

Sickle cell disease (SCD) results from the substitution of a valine residue for glutamic acid at position 6 in the beta-subunit of hemoglobin. It is characterized by chronic hemolytic anemia, vaso-occlusive process and multiple organ infarction resulting from wide spread vascular occlusion. Sickle cell disease is the most common inherited disorder worldwide. Pregnancy in sickle cell disease is at very high risk. Many reports have documented a considerable maternal risk of morbidity and mortality and high peri-natal adverse outcomes.

CASE REPORT

A 26 year old G3P1D1A1 with 39 weeks 4 days gestation was admitted in view of labor pains with complaints of breathlessness, and generalized weakness occasionally on exertion. Patient is a known case of gestational hypertension from 1 month on tab labetalol 100mg BD. Patient was admitted in hospital in view of lower respiratory tract infection during her fifth month of gestation [2,3]. She received three pints of packed cell transfusion in this pregnancy in the sixth month of gestation.

Patient was admitted in ICU for five days for breathlessness and diagnosed with tricuspid regurgitation with pulmonary hypertension, also osteo necrosis of Right tibia in seventh month. Previous pregnancy: A1: spontaneous abortion at two and half months of amenorrhea, Dilatation and Curettage done with a history of two pints of packed cell transfusion. P1D1: history of four pints of packed cell transfusion and diagnosed with sickle cell trait. She delivered a healthy live male baby, died at 1½ yr age in view of fever. Family members have been screened for sickle

cell disease and were negative. History of menstrual cycles – regular periods with moderate flow, no clots.

On examination, Pulse was 92 beats per minute, regular with normal volume, Blood pressure-150/92mmHg, On auscultation of cardiovascular system, murmur heard in pulmonary area, Respiratory system is clear. Patient is febrile with a body temperature of 100 F on presentation.

Per Abdomen examination revealed uterus of term size, acting at 2 contractions for 20 seconds, with longitudinal lie, cephalic presentation, head $3/5^{th}$ palpable, Fetal heart sounds heard at a rate of 136 beats per minute in Left Occiput Anterior position(LOA)[4].

Per vaginal examination revealed that cervix is soft posterior which is 2 cm dilated and 50% effaced. Membranes are present and Artificial Rupture of Membranes (ARM) done (Non Reassuring Fetal Heart Rate (NRFHR) noted in admission test), thick meconium noted and vertex at -2 station.

Blood picture showed Anemia with Hemoglobin of 8.1g/dl with reticulocyte count of 3%. Total counts of 23,900 cells/cumm with differential counts of 81% neutrophils, 16% of lymphocytes were noted. Erythrocyte sedimentation rate of 70 was noted. Platelet counts were 6 lakh 40 thousand. RBC counts were 10-12. Liver and renal function tests and coagulation profile were within normal limits [5,6].

Hb Electrophoresis: HB F- 9.7, HB A1- 35.6, HB A2-4.90, HB SICKLE- 45.

Peripheral smear showed normocytic normochromic anemia with leucocytosis with thrombocytosis. LDH: 598 U/L, TSH: 6.8 mIU/L and Fever profile showed WEIL FELIX- 1:160.

RESULTS

Based on the patient's condition, Emergency Lower segment Caesarian section (LSCS) was done and extracted a healthy female baby of 2.25kg birth weight. Patient was shifted to ICU for supportive postoperative management where she was on O2 inhalation till 48 hours post LSCS, 3 pints of packed cells were transfused and Inj. Enoxaparin 40mg was started on first post-operative day. Patient improved well and was discharged on ninth post-operative day. Both mother and baby are doing well till date.

DISCUSSION

Pregnancy in Sickle cell disease is associated with increased risk of obstetric complications like pre-eclampsia and eclampsia.

Acute pain crisis is the most common complication requiring frequent hospital admissions. Hydroxyurea is known to decrease morbidity in sickle cell disease. However studies have shown that the use of high dose Hydroxyurea may be unsafe in pregnancy and further research is needed. Prophylactic transfusions in sickle cell disease in pregnancy are controversial. The decision for simple or exchange transfusion ideally should depend on the severity of the situation. The aim is to keep Hemoglobin $\geq$ 9 g/dL and < 35- 40% HB S. Early pregnancy blood transfusions are indicated in high risk patients with chronic organ dysfunction or significant history of acute chest syndrome or acute pain crisis.

CONCLUSION

Careful analysis of pregnant patients with anemia has to done with high index of suspicion to rule out rare conditions like sickle cell disease without which would lead to increased maternal and fetal mortality.

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