Sickle Cell Disease Complicated With Multiple Pregnancy: A Case Report

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Abstract

There is very little information available on how best to manage patient with multiple pregnancy and sickle cell disease. A Gravida2 Para1, 30 year-old female known patient with sickle cell disease, complicated with multiple pregnancy, and previous history of caesarean section, presented with anemia for which she was transfused. In the course of treatment she ruptured membranes, in view of this; a repeat caesarean section was performed. Operation was complicated with uterine atony and bleeding hence hysterectomy was done. Post-operative was unremarkable. Anemia is one of the major complications in a patient with sickle cell complicated with multiple gestation, hence prophylactic blood transfusion is recommended.

INTRODUCTION

Sickle cell disease (SCD) and multiple pregnancy are associated with an increased risk of complications during pregnancy. [1,2,3,4]. There are still some areas of controversy with regard to managing sickle cell patient, however with multi-disciplinary approach it help to improve the pregnancy outcome [1]. Ideally all patients with sickle cell disease should have their antenatal care at tertiary hospital [1]. Unfortunately less is known about managing patient with sickle cell complicated with multiple pregnancy.

CASE REPORT

A gravida 2 para 1, 30 year-old female known patient with SCD reported to be doing well until 2 weeks ago prior to admission when she started having gradual swelling of both lower limbs, swelling associated with mild pain on both limbs. She also reports generalized body weakness of one week duration, unrelated to awareness of heart beat, or headache. One day prior to admission she experienced chest pain which was non specific, which radiate to the back, relieved by resting and she did not use analgesic for that.

Her past obstetric history: she had history of cesarean section in 2006 due to IUGR at 36 weeks and failure of induction at our hospital. She is also allergic to penicillin and has been given chloroquine and folic at medical outpatient where she has been attended regularly before pregnancy. She has had several blood transfusions during her childhood due to complication of sickle cell disease

On physical examination, the patient was found to have obvious bossing of skull, slightly jaundiced, pale; she had bilateral non tender pitting edema. Her vital signs were stable. She had uterine fundus equivalent to 40 cm, pfannenstiel incision scar, multiple fetal parts. Fetal heart detected.

Multiple pregnancy with sickle cell anemia was suspected, twins were confirmed by an ultrasound with average gestation of 30 weeks. Fetal weight was estimated to 1567 and 1566 grams for 1st and 2nd respectively. Two separate placenta with adequate liquor volume in each sac.

Initial Hemoglobin was 4.4 g/dl for which she was transfused 3 units of blood within 4 days. Blood slide for malaria parasite was negative. She was prescribed folic acid and Paracetamol.

Three weeks later in the ward, she developed labour like pains and fever, and history of draining fluid per vagina which started overnight. Upon examination she was found to febrile, and per abdomen; fetal heart detected for both fetuses, with palpable contractions. Vaginal examination revealed a 2cm dilated cervix which appeared to be effaced and soft, membranes were ruptured with thick meconium noted. The impression was latent phase of labour with non reassuring fetal status and possible chorioamnionitis.

Emergency cesarean section was performed, under spinal anesthesia: the finding was, gravid uterus with omental adhesions to the anterior fundus. Twin babies were delivered, first in cephalic presentation, female baby of birth weight of 1.250 Kg, second also a female in breech with birth weight of 1.8 Kg in a different sac. Both babies had APGAR score of 8 at 1st minute and 9 at 5th minutes, meconium stained liquor with smell was noted. Two placentas were extracted.

Patient was given ceftriaxone, gentamycin and oxytocin infusion immediately after cord clamping. Uterus was exteriorized, and repaired with intermittent massage of the fundus. Despite oxytocin and massage, uterus remained atonic. Hence the decision was made to perform supracervical hysterectomy. Abdomen was then repaired in layers. Patient received 2 units of blood intra-operatively and adequate infusions. Blood loss was estimated to be 1.5 litre.

She was continued with routine post-operative care in ICU for 24-hours, and then transferred to the normal post-natal ward. Subsequent days, her recovery was unremarkable, sutures were removed on 6th day. She remained nursing her premature twin babies in neonatal unit for 3 weeks after delivery. Upon discharge her full blood picture (FBP) was normal.

COMMENT

This patient presented with severe anemia in pregnancy, this could have been as a result of hemolysis and/or aplastic crisis due to SCD [2, 3], or could have been due to increased demand and hemodilution seen in multiple pregnancy [4]. Routine prophylactic transfusion is no longer recommended in pregnant women with sickle cell disease [4], however in a situation where you have sickle cell complicated with multiple pregnancy some investigators recommend prophylactic Blood transfusion [2,3].

A FBC is a very important investigation; it should be done

frequently in patients with sickle cell disease [2]. A FBC would have helped the caregiver to determine whether to give iron supplement or not. Although Iron supplementation is relatively contra-indicated in sickle cell disease [2,3], it may be very important in patients with multiple pregnancy [4]. The normal range of hemoglobin in patient with sickle cell is between 6.5 and 9 g/dl [2]. One should not be overly alarmed by low haemoglobin levels.

The preferred method of the delivery for a patient with sickle cell disease is vagina delivery [2]. The indication for abdominal delivery in this patient was multiple pregnancy with previous cesarean scar and meconium. Spinal anaesthesia is a better choice during the operation, as there is little incidence of iatrogenic hypoxia [2]. Atonic uterus in this patient was more likely to have been caused by chorioamnionitis and as a consequence of multiple pregnancy [4], and was probably not related to sickle cell disease.

Both sickle cell and multiple pregnancy increases risk of morbidity and mortality to mother and to the fetus as seen in the presented case. We recommended close follow up during antenatal period and still advocate prophylactic blood transfusion in situation like this.

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