

CASE REPORT

Sickle Cell Disease in Pregnancy – A Rare Condition with Detrimental Outcome: A Case Report

NIK SUMAYYAH NMN¹, KALOK A², NAGANDLA K³, SHAFIEE MN²

¹Department of Obstetrics and Gynecology, Hospital Tuanku Jaafar Seremban, Jalan Rasah, 70300 Seremban, Negeri Sembilan, Malaysia.

²Department of Obstetrics and Gynecology, Faculty of Medicine, Universiti Kebangsaan Malaysia (UKM) Medical Centre, Jalan Yaacob Latif, Bandar Tun Razak, 56000 Cheras, Kuala Lumpur, Malaysia.

³Department of Obstetrics and Gynecology, School of Medicine, International Medical University, Clinical Campus Seremban, Jalan Rasah, 70300 Seremban, Negeri Sembilan, Malaysia.

ABSTRAK

Penyakit 'sickle cell' ketika mengandung bukanlah satu perkara biasa di Malaysia. Kami membentangkan satu kes berkenaan penyakit 'sickle cell' ketika mengandung dengan beberapa komplikasi ibu dan bayi. Pesakit kami telah mengalami sakit badan secara tiba-tiba dan pemecahan darah pada trimester yang ketiga. Walaupun ubat pencegah darah beku telah diberikan, pesakit tersebut telah mendapat darah beku pada kaki. Kandungan itu juga telah mengalami masalah seperti darah tinggi yang bahaya dan pembesaran bayi dalam kandungan yang terbantu sehingga memerlukan pembedahan untuk melahirkan bayi. Bayi berkenaan telah dimasukkan ke unit rawatan rapi bayi kerana pra-matang dan kurang berat badan ketika lahir. Pendekatan dari pelbagai disiplin dalam merawat pesakit 'sickle cell' ini sangat penting bagi mendapatkan proses kehamilan yang lebih baik.

Kata kunci: anemia, sickle cell, kandungan kehamilan

ABSTRACT

Sickle cell disease (SCD) in pregnancy is uncommon in Malaysia. We present a case of sickle cell disease in pregnancy with maternal and fetal complications. The patient presented with acute pain crisis and hemolysis in the third trimester. Despite thromboprophylaxis, she developed deep vein thrombosis. The pregnancy was

Address for correspondence and reprint requests: Mohamad Nasir Shafiee, Department of Obstetrics and Gynecology, Faculty of Medicine, Universiti Kebangsaan Malaysia Medical Centre, Jalan Yaacob Latif, Bandar Tun Razak, 56000 Cheras, Kuala Lumpur, Malaysia. Tel: +603-91456485 Fax: +603-91456672 E-mail: nasirshafiee@hotmail.com

further complicated by severe pre-eclampsia and intrauterine growth restriction which require preterm caesarean section. The baby was admitted to Neonatal Intensive Care Unit due to prematurity and low birth weight. Multidisciplinary approach in managing pregnant patient with SCD is essential in achieving good obstetrics outcome.

Keywords: anemia, sickle cell disease, pregnancy outcomes

INTRODUCTION

Sickle cell disease (SCD) is an autosomal recessive disorder, in which abnormal haemoglobin is formed due to mutation in the β -globin gene. In low oxygen condition, polymerisation of this abnormal haemoglobin results in fragile sickled erythrocytes. The mechanically weak red blood cells break down easily, causing capillary blockage. As a result, patients with SCD often present with haemolysis and acute pain crisis. Pregnancy in women with SCD has been associated with higher rate of maternal and fetal complications including pre-eclampsia, preterm birth, fetal growth restriction and stillbirth (Villers et al. 2008). In this case report, we discuss the complications and management of pregnant woman with sickle cell disease.

CASE REPORT

The 32-year-old woman, of Odisha origin, Gravida 2 Para 1, a known case of sickle cell disease, presented to us at 31-wks gestation with generalised body ache and diarrhoea. She was a late booker and her first antenatal assessment was only performed at 26

wks of gestation. She did not receive any pre-pregnancy counselling as she had defaulted previous haematology follow up. Her hemoglobin (Hb) at booking was 8.6 g/dL.

Her first pregnancy was complicated by acute pain crisis and hemolysis. She delivered a low birth weight baby (2.05kg) at 37 wks via emergency caesarean section due to fetal distress.

She was diagnosed with sickle cell disease at the age of eight. Severe veno-occlusion resulted in auto amputation of her right fingers and toes. She had recurrent acute pain crisis and multiple blood transfusions since childhood. She underwent a cataract surgery at the age of 14. Her younger brother was also diagnosed with SCD. Her husband and first born were both screened negative for the disease.

On assessment, she appeared pale and dehydrated with blood pressure of 120/60 mmHg and pulse rate of 94 beats/minute. Abdominal examination revealed symphysio-fundal height of 31 cm which was appropriate for gestation. There was no evidence of calf swelling or tenderness. Hb was 7.7 g/dL and ultrasound assessment showed a singleton fetus with growth parameters plotted at 10th centile and

estimated fetal weight of 1697 g.

Diagnosis of acute pain crisis secondary to dehydration was made. She was treated in a high dependency ward, with nasal oxygen, intravenous fluid and parenteral morphine. She also developed acute haemolytic episode which was evident by low Hb (6.7g/dL), raised lactate dehydrogenase level (1139 U/l) and reticulocytes counts (14.8%). She was subsequently transfused with two units of packed red cell. Unfortunately, despite receiving low molecular weight heparin (LMWH) as thromboprophylaxis, she developed deep vein thrombosis in her left leg.

One week later, she developed pre-eclampsia with elevated blood pressure of 158/95 mmHg and significant proteinuria (urine protein creatinine ratio was 0.574 g/mmol). Thus she was given oral nifedipine for blood pressure stabilization before delivery. Intramuscular dexamethasone was administered to promote fetal lung maturity. Patient underwent emergency Caesarean section at 33 week of gestation and delivered a baby boy with birthweight of 1690 g and Apgar score of 5 at 1 minute and 8 at 5 minutes of life. The baby was admitted to the Neonatal Intensive Care Unit (NICU) due to prematurity and low birth weight.

Anti-hypertensive drug was discontinued post-delivery as her blood pressure subsequently normalised. Therapeutic dose of LMWH was continued for three months postpartum. Patient had opted for intramuscular Depo Provera as contraception and is currently under

active follow up by the haematology team.

DISCUSSION

Sickle cell disease (SCD) is common amongst individuals of African, Greek, India, Turkish and Middle Eastern descent (Centers for Disease Control and Prevention 2017). This condition is rare in Malaysia with only a few cases observed particularly in Indian patients of Odisha origin. Odisha is one of the 29 states in India with high prevalence of SCD (Balgir 2007).

The number of pregnant women with SCD is increasing despite its higher risk of maternal and fetal morbidities. In a latest retrospective cohort study, women with SCD were found to have higher odds for severe preeclampsia (odds ratio, 3.75; 95% confidence interval, 2.21-6.38), preterm delivery (odds ratio, 2.50; 95% confidence interval, 1.93-3.21), small for gestational age (odds ratio, 1.96; 95% confidence interval, 1.18-3.25), and caesarean delivery (odds ratio, 1.93; 95% confidence interval, 1.40-2.67) (Kuo & Caughey 2016).

Women with SCD should be counselled regarding the maternal and fetal risks in pregnancy. It is imperative that these women receive thorough examination during antenatal booking, blood pressure and urinalysis monitoring, and serial fetal growth scan for early detection of pregnancy complications (Royal College of Obstetricians and Gynecologists 2011). Low dose aspirin should be commenced as early as 12 weeks in these women as pre-eclampsia

prophylaxis. Unfortunately, our patient was not prescribed aspirin due to her late booking.

Physiological changes in pregnancy may precipitate sickle cell crisis. Avoidance of the precipitating factors such as excessive vomiting or stress should be emphasized during pre-conception counselling and antenatal booking. Opioids is the treatment of choice in pregnancy for acute painful crisis whilst non-steroidal anti-inflammatory drugs (NSAIDs) are contraindicated during the first and third trimester due to an increased risk of miscarriage and premature closure of fetal ductus arteriosus (Canadian Haemoglobinopathy Association 2014). Our patient was well managed with intravenous morphine during her acute pain crisis.

The previous case report in 1984, resulted in spontaneous vaginal delivery of a normally grown fetus at term (Rachagan et al. 1984). In contrast, our patient developed severe preeclampsia at 33 weeks which warranted a preterm delivery via caesarean section.

Some studies have shown an increased risk of pre-eclampsia, placenta abruption, peripartum cardiomyopathy and acute sickle cell crisis in near term pregnancy. Thus, delivery for normal growing fetus should be aimed at 38 weeks to prevent late pregnancy and perinatal complications (Villers et al. 2008).

Despite thrombo-prophylaxis, our woman developed deep vein thrombosis. This could be contributed by multiple factors including reduced mobility, increased blood viscosity

by the pregnancy and dehydration, as well as endothelial dysfunction which was worsened by her sickled erythrocytes.

Routine prophylaxis transfusion is not recommended unless the pregnancy is complicated by recurrent acute pain crisis, acute chest syndrome and symptomatic anemia (Canadian Haemoglobinopathy Association 2014; Davis et al. 2017). The decision for blood transfusion should be made judiciously, taking into account the risks involved such as alloimmunisation, transfusion reaction and transmission of infection.

Pre-pregnancy counselling plays an important role in preventing pregnancy complications for women with SCD. Contraception counselling is equally important in preventing unplanned pregnancy in women with uncontrolled disease. An intrauterine copper device (IUCD) is efficient for long term contraception while progesterone-only contraception is safe in sickle cell disease (Canadian Haemoglobinopathy Association 2014).

CONCLUSION

While pregnancy carries significant risks to a woman with sickle cell anemia and her fetus, a favorable outcome is possible with a holistic approach by a multidisciplinary team. Adequate pre-conception counselling, avoidance of sickle cell crisis precipitants, regular antenatal visits with serial growth scans, thrombo-prophylaxis with LMWH and prompt treatment of acute events will improve the outcome of

the pregnancy of a woman with SCD. Contraception is important and should be discussed with every woman with SCD.

ACKNOWLEDGEMENTS

The authors thank Dr. H. Krishna Kumar, Dr. Jameela Sathar, Dr. Joanne Lim Xu Mei and Ir. Abdullah Dinsuhaimi for the assistance received in preparing this case report.

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Received: 8 April 2017

Accepted: 13 October 2017