ORIGINAL ARTICLE

Pregnancy Outcomes In Patients With Sickle Cell Disease In Enugu, Nigeria

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Abstract

Background: *Pregnancy in patients with sickle cell disease(SCD) is associated with increased maternal and foetal morbidity and mortality. The objective of this study was to review the pregnancy outcomes in patients with SCD as seen at the University of Nigeria Teaching Hospital(UNTH), Enugu, South-eastern Nigeria.*

Methods: *This is a retrospective study. The data extracted from the patients' case files include the age, parity, gestational age at booking and complications of disease and pregnancy during the antenatal period, labour and puerperium. Fetal outcomes were also reviewed.*

Results: *During the 30-year period under study(1975-2004), only 10 pregnant women with sickle cell disease were documented to have been attended to. Pregnancies were characterized by high maternal and fetal morbidity and mortality. Maternal complications identified were maternal mortality, lobar pneumonia, HIV and recurrent malaria infections, candidiasis, bone pain crises, haemolytic crises, pseudotoxaemia and pre-eclampsia. Fetal complications included intra-uterine fetal deaths, still births, low birth weights, and breech presentation.*

Conclusion: *From this study, it seems that female SCD patients present more rarely with pregnancy in South-eastern compared to South-western Nigeria. However, the spectrum of complications seen is similar to that recorded in other studies.*

Key words: *Sickle cell disease, pregnancy, outcome, Enugu, Nigeria.*

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INTRODUCTION

The term sickle cell disease (SCD) refers to a life-long disorder arising from the inheritance of two abnormal haemoglobin genes, each from a parent, with at least one of the two abnormal genes being a haemoglobin S. Examples of SCD are HbSS, HbSC and HbSâthal. The S gene results from the replacement of the normal codon GAG at position â[6] by GTG. As a result of this change, valine is inserted at this position instead of glutamic acid usually in that position in HbA individuals. In the deoxygenated state, the HbS molecule forms polymers which progress to the sickled red blood cell. Although SCD has a high prevalence throughout equatorial Africa, the gene is now known to be widespread in parts of Sicily and Southern Italy, Northern Italy, Northern Greece, Southern Turkey, the Middle East, Saudi Arabia and much of Central India[1] as a result of the survival advantage which the heterozygous genotype (HbAS) has against malaria infection. Due to increased education, awareness and improved nutrition, many of the individuals with SCD now attain adulthood and are now encountered in obstetric practice. Since the earliest description of pregnancy in patients with sickle cell anaemia by Koback[2], other authors have documented various complications including increased maternal and foetal morbidity and mortality[3-6].

The various pregnancy complications which have been noted include maternal complications such as bacterial infections, eclampsia, placenta praevia, premature rupture of fetal membranes (PROM), premature labour, and maternal mortality. Fetal complications include spontaneous abortions, still births, small for gestational age, intrauterine growth retardation and perinatal mortality. Pregnancy is also associated with the various crises seen in non-pregnant patients with sickle cell disease. The objective of this study was to review the pregnancy outcomes in patients with sickle cell disease as seen at the University of Nigeria Teaching Hospital, Enugu, South-eastern Nigeria.

MATERIALS AND METHODS

This is a retrospective study of women with SCD who presented with pregnancy at the University of Nigeria Teaching Hospital (UNTH), Enugu, Nigeria between January 1975 and December 2004. A thorough scrutiny of the antenatal, labour and delivery records of the Obstetric unit as well as the records of the Medical Records Department of the hospital was done to identify these patients. Their case files were subsequently retrieved and studied. The data abstracted from the

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patients' case files include: age, parity, gestational age at booking, complications of the disease and pregnancy during the antenatal period, labour and puerperium. Fetal outcomes as well as blood counts were also extracted from the case files. Others include the number of times each woman was transfused with blood in a given pregnancy, mode of delivery and birth weight.

RESULTS

During the 30 year period under review, 10 women with SCD presenting in pregnancy were treated in the hospital. During the same period, there were 50,561 deliveries giving a prevalence of one pregnant SCD patient in 8427 pregnancies. Out of the 10 women with SCD, the case files of only three of them were seen. The three women had a total of 6 pregnancies. One of the patients had 4 pregnancies while the remaining two had one pregnancy each. The mean age of the patients was 28.8 ± 0.8 (range 28 32 years). Two of the patients were primigravidae while the third patient who eventually had 4 deliveries at UNTH Enugu during the study period first presented to the Obstetric unit during her second pregnancy. She had her second to 5th deliveries at the UNTH Enugu. Although she actually had a total of 5 pregnancies/deliveries, the first was not in UNTH Enugu. Therefore, only the records of her last 4 deliveries were available to be studied. The mean gestational age at booking during the 6 pregnancies was 21.3 ± 2.7 (range: 9 29 weeks). With regard to the haemogram during pregnancy and delivery, the mean PCV(packed cell volume) at booking was 23.0 ±3.0%(range 16-26%). The steady state PCV was not recorded for any of the patients. White blood cell counts(WBC) were done in only two of the pregnancies, once in one pregnancy and thrice in the second pregnancy. In one pregnancy, the total count was 9.1 x 10^9/L with a differential count of neutrophils 38%, eosinophils 6%, lymphocytes 52% and monocytes 4%. In the other pregnancy, the WBCs of 12.7 x 10^9/L and 18 x 10^9/L. The differential counts in both results were within the reference ranges. Review of the peripheral blood films showed nucleated red blood cells. No platelet count was done in any of the patients.

Maternal complications

Bone pain crises occurred in 5 of the pregnancies. It occurred once during the 38th week of gestation in one of the primigravid patients. All the 4 pregnancies of the multiparous patient were complicated with bone pain crises. In her first and fourth pregnancies in UNTH Enugu, she had one episode each of bone pain crisis. She had 4 episodes of bone pain crises during her second pregnancy in her 21st, 32nd, 36th and 39th weeks of gestation. In her third pregnancy, she had two episodes of vaso-occlusive crises during her 23rd and 34th weeks of gestation. Four of the six pregnancies required blood transfusion. The multigravid patient received two units of blood each in her first, third and fourth pregnancies while one of the primigravidae that died had one unit of packed red blood cells.

With respect to malaria and other infections, malaria parasitaemia was confirmed in only one of the pregnancies and was treated with chloroquine phosphate. However, therapeutic anti-malarial drugs were also given in some of the other pregnancies even when malaria parasite screenings were not done especially when the patients had bone pain crisis and fever. In one of the pregnancies, HIV I and II infections were confirmed. Candidiasis was confirmed in one of the pregnancies. Lobar pneumonia was diagnosed in three of the pregnancies. These were in the 2nd, 3rd, and 4th pregnancies of the multigravid patient who was not HIV positive. Two of the pregnancies were associated with diarrhoeal illnesses. Pseudotoxaemia occurred in one pregnancy. Pre-eclampsia occurred in 4 of the pregnancies. There was no case of eclampsia.

During delivery, the mean blood loss was 390.3 ± 80.7 mls with a range of 50 1200 mls. Blood loss in 4 pregnancies was less than 500mls each. In the fifth pregnancy, the blood loss was 1200mls. This patient had 2 units of packed red blood cells post-partum. In one of the 6 pregnancies, the woman had an elective caesarean section for breech presentation. For the remaining 4 cases, labour was spontaneous in 3 cases and induced in 1 case. The four cases ended in vaginal delivery. One of the two primigravidae died during the antenatal period at 39 weeks gestational age (GA). She had anaemic heart failure, malaria parasitaemia and severe bone pain crises during her antenatal period. She was also confirmed to have human immunodeficiency virus (HIV) I and II infections as well as an intra-uterine fetal death. In one of the 6 pregnancies, the woman had an elective caesarean section for breech presentation. For the remaining 4 cases, labour was spontaneous in 3 cases and induced in 1 case. The four cases ended in vaginal delivery.

Fetal complications

The mean duration of pregnancy was 38.5 ± 2.5 (range: 29-42) weeks. Three pregnancies terminated with intrauterine foetal deaths (IUFDs) at 39 weeks, 29 weeks and 40 weeks respectively giving a perinatal mortality rate of 500 per 1000. There was one fresh still

birth and one macerated still- birth. One of the cases of IUFD was not delivered because soon after diagnosis of IUFD, the mother died. She was therefore buried undelivered. The birth weights of the 5 babies who were delivered ranged from 1.8 kg to 3.1kg with a mean of 2.48 ± 0.6 kg. Two fetuses weighed below 2.5kg (1.8g and 2 kg) while three weighed above 2.5kg (2.7, 2.8 and 3.1 kg). Table I shows a summary of the pregnancy outcomes.

Table I Pregnancy outcomes in sickle cell disease patients at the University of Nigeria Teaching Hospital Enugu, Nigeria (1975 2004)

Patient's Identity	Gestational age at delivery	Maternal outcome	Mode of delivery	Birth weight	Sex	Foetal status
1	42 wks	Alive	SVD*	2.7kg	F	Live
2	30 wks	Alive	SVD	1.8kg	F	IUFD***/MSB+
2	40 wks	Alive	SVD	2.8kg	M	IUFD/FSB++
2	41 wks	Alive	C/S**	3.1kg	M	Live
2	40 wks	Alive	SVD	2kg	F	Live
3	39 wks	Died & buried undelivered		-		IUTD

* SVD = Spontaneous vertex delivery
** C/S = Caesarean section
*** IUFD = Intra-uterine foetal death
+ MSB = Macerated still birth
++ FSB = Fresh still birth

Discussion

It is rather surprising that only 10 women with sickle cell disease presented with pregnancy to the Obstetric Unit of this institution over the 30 year period covered in this study. The number is rather small when compared to results from other Nigerian centres. In three separate studies from the Lagos University Teaching Hospital (LUTH) covering the periods September 1966 to December 1970[7], January 1980 to December 1984 (5 year period)[8] and 1995 to 1997 (3 year-period)[9], 53, 34 and 60 patients were seen respectively. At the University of Benin Teaching Hospital, Nigeria 20 pregnant women with sickle cell anaemia were seen in 7 years (November 1973 to October 1980)[10]. The low incidence in our centre may be because of a lower incidence of SCD in Eastern compared to Western Nigeria in which the other studies were carried out or it may mean that many of the affected children die before reaching the child bearing age. The latter reason has also been observed in a similar study from Benin Republic[11]. A third possibility may be because of relative infertility in SCD patients compared to AA or AS individuals as has been documented in other studies.[12-14] One of our patients however had a total of 5 pregnancies, delivering one outside and the remaining 4 at UNTH Enugu, a direct contradiction of the widely held believe that there is relative infertility in patients with sickle cell disease.

From the results above, it is obvious that the spectrum of complications seen in these patients during pregnancy and delivery reflect those documented in other studies[15-17]. Maternal complications identified were one maternal death, lobar pneumonia, HIV infections, malaria infections, candidiasis, bone pain crises, haemolytic crises with anaemic heart failure, pseudotoxaemia and pre-eclampsia. Fetal complications included intra-uterine fetal deaths, still births, low birth weights, and breech presentation. The HIV infection recorded in one of the patients may have resulted from frequent blood transfusions, although we do not know her sexual and social lifestyles. With the increasing prevalence of HIV infection, there is a need for HIV antigen-based screening of blood for transfusing sickle cell patients since they are likely to receive multiple blood transfusions.

Conclusion

Although available data are not enough to make a general conclusion, this study supports previous findings that pregnancy associated with SCD carries high maternal and foetal morbidity and mortality. This calls for a closer and more detailed monitoring of this group of patients during antenatal care, labour and puerperium by the Obstetricians, Haematologists and all the other teams involved in their care.

Limitations of this study

The greatest limitation of this study is that of record keeping. It reveals the need for improved record keeping systems in our hospitals.

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