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Original Research Article

Study of sickle cell pregnancies at tertiary care hospital

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Abstract

Aim: To study the clinical presentation and complications in pregnancies in sickle cell patients
Methods: The study reported findings in 6 pregnant women with sickle cell disease admitted during the period of May2015-June2016. The incidence of obstetric complications, non-obstetric complications linked to sickle cell disease and complications in the new born were analyzed.
Results: All the patients followed in the age group of 21-31 years, most were first primi, all were in third trimester of pregnancy, and all are referred cases. Patients presented with variety of complaints such as anemia, joint pain, etc. Hepatomegaly was found in all patients. Intra Uterine Fetal Demise was found in one case. All patients presented with moderate to severe anemia. Cesarean section was done in most of the cases.
Conclusions: These study shows that pregnancy is still associated with many clinical and obstetric complications in patients with SCD and there is need to providing genetic counselling and educating SCD women about their disease and followed by a multidisciplinary team in a tertiary hospital.
Keywords: Sickle cell anemia, pregnancy, clinical and obstetric complications.

1. Introduction

Sickle cell disease (SCD) comprises a group of diseases characterized by the presence of sickle hemoglobin (Hb S). In situations of low oxygen tension, Hb S solubility decreases, resulting in the polymerization of these molecules. The intracellular formation of Hb S polymers affects the red cell structure, changing it into a sickle-shaped, thereby damaging the cell membrane, making it more rigid and exposing a greater number of adhesion molecules on the cell surface, thus increasing the adherence of red cells to the vascular endothelium.[1] This phenomenon, named sickling, is responsible for the premature destruction of red cells by the reticuloendothelial system, causing a chronic hemolytic anemia. Sickle cell disease is the most common inherited disorder worldwide with varying clinical severity and potentially serious complications. [2]

Chronic hemolytic anemia and frequent vaso-occlusive crises cause damage to various organs and impair both the survival and the quality of life of patients with SCD.[3]

Pregnancy in sickle cell disease is at very high risk. Many reports have documented a considerable maternal risk of morbidity and mortality and high perinatal adverse outcomes.[4-7] Nowadays, with newborn screening and preventive measures such as vaccination and antibiotic prophylaxis since birth, patient survival has improved.[8] However, despite all the medical advances in recent decades, pregnancy in sickle cell patients is still associated with many clinical and obstetric complications compared to the general population.[9-10] This paper reports 6 cases of sickle cell anemia in pregnancy observed at SAMC & PGI with summary of current concepts of diagnosis and treatment.

2. Material & methods

The retrospective study was carried out at department of Obstetrics and Gynecology, Sri Aurobindo

Medical College and PG Institute, Indore (MP). The subjects were patients with sickle cell pregnancies admitted to the department from May 2015- June 2016. The inclusion criterion was a pregnant patient with sickle cell diseases (HbSS, HbS-beta, or HbSC diagnosed by hemoglobin electrophoresis) Permission was sought from the Ethical Committee to carry out the study. Consent was elicited from the respondents before collection of data.

Clinical data was obtained through a review of medical records from the hospital with the confidentiality of information being preserved. The results of laboratory tests were obtained through the online hospital system.

3. Result

Table 1: Characteristics & presentation in cases

Sr No	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6
Age	26	24	22	23	31	21
gravidity	1 primi	1 primi	2 primi	1 primi	1 primi	1 primi
PoG(WKS)	39	34	35	36	34	39
Presentation & complaints	Generalised body ache, Vomiting, loose stools	Yellowish discolouration, anemia	Swelling over lower limbs	Multiple joint pain right high, yellowish discolouration, severe anemia, fever	High grade fever, yellowish discolouration, severe anemia	High grade fever, Swelling over lower limbs
Diagnosed	11 months back	At 16 yrs	At 14 yrs	At 10 yrs	At 12 yrs	At 8yrs

All patients' falls in the age group of 21-31 years, out of six patients 5 were primiparous and all were in third trimester of pregnancy, all are referred cases. First case was diagnosed 11 months back, just before conception, rest patients were diagnosed either in the child hood or adolescence for sickle cell diseases. Patients presented with variety of complaints such as anemia, joint pain, etc.

Table 2: Clinical parameters, systemic and obstetrics presentation in cases

Sr. No	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6
GC	poor	fair	fair	avg	poor	Avg
temp	Afeb	Afeb	Afeb	Afeb	Feb-99 F	Feb-100F
Vitals (P/BP)	94/m 136/90	70/m 130/86	86/m 150/90	110/m 140/90	92/m 130/90	110/m 150/100
Systemic exam	Hepatosplenomegaly	Hepatosplenomegaly	Hepatomegaly	Hepatomegaly	Hepatomegaly	Hepatomegaly
Obst exam	34wks/V/ FHS=+ RELAXED	36wks/ CEPHALIC/ FHS=+ RELAXED	32wks/ CEPHALIC/ FHS=+ RELAXED	36wks/ BREECH/ FHS=+ IRRITABLE	30wks/ CEPHALIC/FHS ABSENT	34wks/ CEPHALIC/ FHS=+ IRRITABLE

Hepatomegaly was found in all patients associated with splenomegaly - 2 patients. All patients had pre-eclampsia Case no 5 admitted with Intra Uterine Fetal Demise.

. **Table 3: Diagnostic parameters in cases**

Sr. No	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6
Blood GR	AB+	O+	O+	A+	A+	A+
HB	8.6	7.5	8.9	6.2	4.7	8.1
PCV	26.7	24.3		18		26.6
WBC	7,500	11,800		23000	27100	8300
PLT	2.09	2.7	2.45	6.3	1.3	3.35
HIV/HBsAg	NR	NR	NR	HEV+	NR	NR
Billurubin						
Total	3.5	2.8	2.5	1.9	5.2	3.5
Direct	0.8	1.4	0.9	1.3	2.3	0.6
Indirect	2.7	1.4	1.6	0.6	2.9	2.9
Retic count		9	3	10	18	2
Sickling	+	+	+	+	+	+
Hb Electrophoresis	HbSB	HbSB	HbSS	HbSS	HbSS	HbSS
USG Observation	SLF,VX, 3,4WK,2.2KG	SLF,VX, 34WK,2.2KG	SLF,VX, 34WK,2.4KG	34WK/1.9KG/ Breech/AFI-adq	IUFD/1.8/ CEPHALIC/CA-Absent	32wks/AFI 5-6/Pl-post/1.9 kg
MCA D	N	N	N	N	Not done	N

All patients presented with moderate to severe anemia, lowest Hb -4.6gm/dl, highest Hb- 8.7 gm/dl. Case fourth-HEV positive. First two cases associated B-thalassemia. Last four cases-HbSS.

Table 4: Characteristics of new born & mode of delivery in cases

Sr. No	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6
Mode of delivery	Induction-LSCS	Induction-LSCS	Induction-LSCS	LSCS	PTVD Still birth	PTVD
MSL	+	+	+	-	+	-
IUFD	-	-	-	-	+	-
BT	2	1	2	5	6	1
LMWH	+	+	+	+	+	+
Baby wt	2.4/F	2.5/M	2/M	2.3/M	1.6/F/IUD	1.8/F
APGAR	7/9	8/9	7/9	8/9	-	8/9
NICU	+	+	+	+	-	+

Induction required in three patients, case four was taken for elective LSCS (AVN of femoral head, breech, pre-eclampsia, HEV POSITIVE) Case sixth -spontaneous PTVD. All patients received DVT prophylaxis and low molecular weight heparin in post-partum period. 4 out 6 patients were kept in CCU. All patients received multi-disciplinary approach. In our study there was no maternal mortality and out of six. Only one patient had IUFD (referred from outside).

4. Discussion

Pregnancy in a sickle cell woman is at very high risk, especially in patients with more severe sickle cell disease. Indeed, the risk of maternal and fetal complications is higher than in the general population [11]. It is known that pregnancy induces a number of physiologic changes that affect the hematologic indices, and patients with SCD may experience worsening of the anemia and other sickle cell complications. **[**12] Oxygen demand during pregnancy increases to support the metabolic requirements of the placenta and foetus. As the maternal oxygen reserve may be compromised during pregnancy due to the increased oxygen consumption and decreased functional residual capacity, patients may be predisposed to hypoxemia, with exacerbation of sickling and its complications.[13] These changes during pregnancy highlight the need for a multidisciplinary team of experts to monitor pregnant sickle cell women in a tertiary hospital.

The high rate of complications in pregnant patients with SCD has already been reported in previous studies. Complications include an increased number of cesarean deliveries, preterm births, restricted intrauterine growth, and low weight babies, especially in pregnant women with the homozygous form of the disease (Hb SS). Many already published studies were observational ones and confirms high complication rate. Our study also reported obstetric complication (pre-eclampsia and cesarean section) which were similar to other studies [14]

Parity > 1 was also identified as a higher risk (two times) for near miss/death in pregnant women with SCD. The reason for this remains unclear. A hypothesis is that pregnancy-driven physiologic changes could pose more risk to the pregnant women with SCD, a risk that is proportionally higher as the number of gestations and accumulated complications deriving from the disease increases.

Our results also shows fetal characteristics such as, weight lower than 2500 g, at birth 5-min Apgar score less than 7, cesarean section for fetal distress, and perinatal death which was concurrent with other studies Koshy *et al* [15,16] Chronic fetal hypoxia associated with decreased placental circulatory flow level seems the most plausible explanation for this high incidence of perinatal complications [17].

The major follow-up recommendations for SCD pregnant women consist of carefully monitoring hematologic, obstetrical and fetal complications, and recognizing SCD complications as early as possible.[18] Blood transfusions are indicated for acute and severe episodes during pregnancy complications that may be enhanced by SCD.[19] Critical care should be delivered by a concerted team composed by hematologists, obstetricians, general practitioners and intensivists.[20]

5. Conclusions

These study shows that pregnancy is still associated with many clinical and obstetric complications in patients with SCD and there is need to providing genetic counselling and educating SCD women about their disease and followed by a multidisciplinary team in a tertiary hospital.

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