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A STUDY OF SICKLE CELL DISEASE IN RELATION TO CLINICAL AND HEMATOLOGICAL PROFILE OF CHILDREN IN A TERTIARY CARE HOSPITAL

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ABSTRACT:

Introduction: In the world, Sickle cell disease (SCD) is the commonest genetic disease. The most common single gene disorders are Haemoglobinopathies. 59th World Health Assembly in 2006, sickle-cell disease (SCD) recognized by WHO as an inherited disorder of hemoglobin as a priority of public health. Sickle hemoglobin (HbS) is abnormal hemoglobin difference in which adenine is substituted by thiamine. Deoxygenating of red blood cells form tactoid aggregates and distorts RBCs into sickle shape in HbS. repeated sickling and desickling RBCs convert into permanent sickle shape which can occur sickle red blood cell anemia, crisis and organ damage. There is extremely variable in Hematological profiles of sickle cell disease. There are very few studies are available on hematological profiles of sickle cell disease. Individuals express the homozygous form (HbSS) manifest the disease whereas with the heterozygous form (HbAS), also known as sickle-cell trait (SCT), are usually asymptomatic carriers. Initially Sickle cell disease is limited to sub-Saharan Africa, Middle-East and some parts of India. However nowadays it is spread to all continents with the migration of populations. In India, sickle cell disease (SCD) is common in Chhattisgarh, Madhya Pradesh, Gujarat, Orissa, Vidarbha, Andhra Pradesh and Tamil Nadu. For sickle cell disease there is more need for hospital care in children than adult patients. Children younger than five years of age suffering from SCD especially with age of 1-3 years are most endangered to mortality.

Aim: The main aim of this study is to study hematological profiles clinical manifestation of children with sickle cell disease.

Material and method: This is cross- section study which is done in the Department of Paediatrics at Surat Municipal Institute of Medical education and research Gujarat (SMIMER) and from 2016 to 2017 during the period of one year. This study was carried out on patients between the age group 6 months to 14 year old which found with diagnosed of sickle cell disease on routine blood investigations and further investigations were carried out to ascertain the cause of sickle cell.

Result: A total of 60 patients with different age group from 6 months to 14 years were admitted during the study period that fulfilled the inclusion criteria. Out of 60 patients 46 were homozygous sickle cell disease and 14 heterozygous sickle cell trait patients. Total 38 (63.3%) male patients and 22(36.7%) female patients were included.

Conclusion: Child presenting for detecting sickle cell with the symptom like abdominal pain, musculoskeletal pain, pallor, icterus, splenomegaly and hepatomegaly should lead to high suspicion of sickle cell where its prevalence is more. in pediatric age group commonest manifestation like hemolytic in nature, hematological parameters were suggestive of hypochromic microcytic anemia. In patients with sickle cell anemia, parental counseling and preventive measures like management of pain with simple analgesics, early treatment of infection with antibiotics and regular folic acid

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supplementation will be more useful for decreasing morbidity and mortality. Hence With comprehensive medical health care life status expectancy can be improved considerably.

Keywords: Sickle cell disease, Pediatric, hematological profiles, Acute painful crisis

INTRODUCTION

In the world, Sickle cell disease (SCD) is the commonest genetic disease. The most common single gene disorders are Haemoglobinopathies[i]. In India from south Indian tribal groups Sickle cell anemia was first described and subsequently in central India[ii, iii]. 59th World Health Assembly in 2006, sickle-cell disease (SCD) recognized by WHO as an inherited disorder of hemoglobin as a priority of public health[iv]. Sickle hemoglobin (HbS) is abnormal hemoglobin difference in which adenine is substituted by thiamine. Deoxygenating of red blood cells form tactoid aggregates and distorts RBCs into sickle shape in HbS. repeated sickling and desickling RBCs convert into permanent sickle shape which can occurs sickle red blood cell anemia, crisis and organ damage[v]. There is extremely variable in Hematological profiles of sickle cell disease. There are very few studies are available on hematological profiles of sickle cell disease[vi]. In early life clinical manifestations of sickle cell anemia (SCA) begin which continue with an increased incidence of adverse matter with the physiologic decline in fetal hemoglobin (HbF)[vii]. In SCA one of the predominant clinical features associated are Vaso-occlusive pain episodes[viii]. Individuals express the homozygous form (HbSS) manifest the disease whereas with the heterozygous form (HbAS), also known as sickle-cell trait (SCT), are usually asymptomatic carriers. Initially Sickle cell disease is limited to sub-Saharan Africa, Middle-East and some parts of India. However nowadays it is spread to all continents with the migration of populations[ix]. Approximately 300,000 children with severe hemoglobin disorders are born every year worldwide[4]. Sickle cell disease is the most common genetic disease that identified through state mandated newborn screening programmed in USA, occurring in 1:2647 births. About 72,000 people, especially whose ancestors come from Africa was affected. About 1 in every 500 African-American births and 1 in every 1000 to 1400 Hispanic-American births affected this disease. 1 in 12 African Americans or About 2 million Americans carry sickle cell allele[x]. In India, sickle cell disease (SCD) is common in Chhattisgarh, Madhya Pradesh, Gujarat, Orissa, Vidarbha, Andhra Pradesh and Tamil Nadu. For sickle cell disease there is more need for hospital care in children than adult patients. Children younger than five years of age suffering from SCD especially with age of 1-3 years are most endangered to mortality[xi]. The main aim of this study is to study hematological profiles clinical manifestation of children with sickle cell disease. In both the groups' pain (61.7%) was the commonest symptom, seen in more than half of the patients. In sickle cell disease patient pain (67.4%) was the most common symptom followed by fever (34.8%) and cough (17.4%). Pallor (47.8%) and splenomegaly (28.3%) was the common sign observed. massive splenomegaly had seen in four sickle cell disease patients. In sickle cell trait patients pain was presenting symptom in 42.85% of cases, fever in 61.7% and cough in 20.0% of cases. Severe anemia was seen in 14.3% and splenomegaly in 25.0% of cases.

MATERIAL AND METHODS:

This is cross- section study which is done in the Department of Paediatrics of at Surat Municipal Institute of Medical education and research Gujarat (SMIMER) during 2016 to 2017 the period of one year. This study was carried out on patients between the age group 6 months to 14 year old which found with diagnosed of sickle cell disease on routine blood investigations and further investigations were carried out to ascertain the cause of sickle cell.

Age, sex, detail history and the current diagnosis for which the cause sickle cell and also noted down detail relevant clinical examination findings. Clinical finding like total and differential counts, hemoglobin, platelet count, MCH, MCHC were done using automated cell counter method. Patients with Sickle cell disease or trait patients who visit pediatrics OPD as well

as hospitalized for any morbidity in pediatrics ward included in the study. This study was carried out after the approval of Institutional Ethics Committee and written informed consent was obtained from all patients' parents in this study.

OBSERVATIONS AND RESULT:

A total of 60 patients with different age group from 6months to 14 years were admitted during the study period that fulfilled the inclusion criteria. Out of 60 patients 46 were homozygous sickle cell disease and 14 heterozygous sickle cell trait patients. Total 38 (63.3%) male patients and 22(36.7%) female patients were included.

In this study more male patients are affected in 9-12 years of age group, while more female patients in 5-8 years of age group. However there was no significant difference in gender in patients with sickle cell disease. Most patients belonged to the age group of 5-12 years comprising 68.1% of total sickle cell disease patients as shown in table below (table no 1.).

Table1: Age and gender wise distribution of patients with sickle cell disease.

Year	Male	Female
0-4	6	0
5-8	7	9
9-12	10	6
13-14	5	3

In the table below showing most patients in both gender belonged to the age group of 5-8 years comprising 50.0% of total sickle cell trait patients. However there was no significant difference in gender in sickle cell trait patients (table no 2).

Table2: Age and gender wise distribution of sickle cell trait patients.

Year	Male	Female
0-4	1	1
5-8	5	2
9-12	3	0
13-14	1	1

In this study nutritional status of patients was also studied. In sickle cell disease patients 63.8% had under nutrition while in sickle cell trait patients only 35.7%.had under nutrition.

Table showing that in both the groups' pain (61.7%) was the commonest symptom, seen in more than half of the patients. In sickle cell disease patient pain (67.4%) was the most common symptom followed by fever (34.8%) and cough (17.4%). Pallor (47.8%) and splenomegaly (28.3%) was the common sign observed. massive splenomegaly had seen in four sickle cell disease patients. In sickle cell trait patients pain was presenting symptom in 42.85% of cases, fever in 61.7% and cough in 20.0% of cases. Severe anemia was seen in 14.3% and splenomegaly in 25.0% of cases (table no 3).

Table3: Clinical profile of patients with sickle cell disease and trait.

Clinical profile	Sickle cell disease (N=46)	Sickle cell trait (N=14)	Total (N=60)
Pain	31(67.4%)	6(42.9%)	37(61.7%)
Cough	8(17.4%)	4(28.6%)	12(20.0%)
Vomiting, Diarrhoea	2(4.3%)	3(21.4%)	5(8.3%)
Fever	16(34.8%)	4(28.6%)	20(33.3%)
Splenomegaly	13(28.3%)	2(14.3%)	15(25.0%)
Heptamogaly	3(6.5%)	1(7.1%)	4(6.7%)
Pallor	22(47.8%)	2(14.3%)	24(40.0%)
Icterus	5(10.9%)	1(7.1%)	6(10.0%)

Table4: Hematological profile of sickle cell disease and trait patients on admission.

Hematological profile	Disease (N=46)	Trait (N=14)	P value
Hemoglobin (gm/dl)	8.8±2.4	10.1±2.4	<0.001
Total leucocyte count (cumm)	14158±7859	6841±2923	<0.001
Platelet (lac/cumm)	2.7±1.2	3.2±0.8	0.154
Hematocrit (%)	23.3±6.4	31.2±2.5	<0.05
MCV (micro liters)	75.9±2.1	82.6±3.1	<0.001
MCH (picograms)	25.2±1.1	25.8±1.2	<0.05
MCHC (gram/dl)	26.3±1.8	25.7±1.2	0.109

In this study musculoskeletal pain in 15 (40.5%), abdominal pain in 14 (37.8%), generalized body ache in 7 (18.9%) and chest pain in 1 (2.7%) of total patients with acute painful crisis. It is observed that hemoglobin level with indices MCH, MCV and hematocrit were low in patients with sickle cell disease as compared to sickle cell trait and statistically significant on laboratory analysis. It was found that high leucocyte count in sickle cell disease which is statistically significant. Platelet count was low in sickle cell disease as compare to sickle cell trait but statistically not significant as shown in above table.

DISCUSSION AND CONCLUSION:

Total 46 diagnosed cases of homozygous sickle cell disease and 14 heterozygous cases were included in this study in which male preponderance was seen which was similar to other studies from central India[xii,xiii]. This may be due the gender-selective use of medical facilities and management of patients. Most patients with the age group of 5-12 years as comprising 68.1% total sickle cell disease patients. In both gender sickle cell trait 50% of patients belonged to the age group of 5-8 years. There was no statistically significant difference in gender in both groups which is similar to the study of Swarnkar K etal[xiv]. Pain was the most common presenting complains seen in sickle cell disease and sickle trait patients followed by Splenomegaly in this study which is also similar to different studies carried out in India as panigrahi S etal[xv] and Mandot S etal[xvi]. In this study shows Hemoglobin, MCH and MCH were low as comparing to other studies done by Rao SS etal[xvii] and Kaur M etal[xviii]. In India according to National Family Health survey (NFHS-3), anemia is common among the children with low socioeconomic status[xix]. In this study there was no mortality of Sickle cell patients however sickle cell patients with crisis had mortality due to splenic sequestration or severe sepsis. Therefore like this study should be community-based cohort study or a birth cohort study.

In this study morbidity events were common in males' patients in 5-12 years of age groups as comparing various studies in other parts of India. Child presenting for detecting sickle cell with the symptom like abdominal pain, musculoskeletal pain, pallor, icterus, splenomegaly and hepatomegaly should lead to high suspicion of sickle cell where its prevalence is more. In pediatric age group commonest manifestation like hemolytic in nature, hematological parameters were suggestive of hypochromic microcytic anemia. In patients with sickle cell anemia, parental counseling and preventive measures like management of pain with simple analgesics, early treatment of infection with antibiotics and regular folic acid supplementation will be more useful for decreasing morbidity and mortality. Hence With comprehensive medical health care life status expectancy can be improved considerably.

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