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SPECTRUM OF CLINICAL PRESENTATION IN SICKLE CELL DISEASE PATIENTS OF CENTRAL INDIA BELONGING TO PEDIATRIC AND ADULT AGE GROUP

Community Medicine

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ABSTRACT

A Cross-sectional study was conducted with objective to determine the spectrum of clinical presentation in patients with Sickle cell disease (SCD). Total 187 patients of either gender who were diagnosed cases of SCD were recruited. Patients of all age groups were included in the study. The diagnosis of SCD was established by solubility testing, hemoglobin electrophoresis and HPLC. Diagnosis of different clinical presentations was established by clinical examination and other necessary investigations where needed. 123 patients were of pediatric age group,64 were adults. 110 patients were male and 77patients were females. In pediatric population, fever, pallor, respiratory tract infections, Jaundice, diarrhea, seizures, typhoid and cellulitis form the first line symptoms. In adults, painful crises, leg ulcer and diabetic keto acidosis form the first line symptoms. We recommend a comprehensive SCD center, which should cater these patients by emphasizing the importance of prevention of disease through health education, genetic counselling, effective manifestations management and regular follow-ups.

KEYWORDS:

Introduction:

Sickle cell disease is a hereditary hemolytic anemia resulting from substitution of amino acid valine residue for glutamic acid at position 6 in the beta – subunit of haemoglobin [(1)] The gene for sickle cell hemoglobin is located on short arm of chromosome 11 and has an autosomal recessive inheritance. Hence it manifest in two forms viz. heterozygous (sickle cell trait) or homozygous (sickle cell anemia) [(1)] . In deoxygenated state HbS tends to polymerize non covalently into long strands that deform the erythrocyte, giving the characteristic —Sickle cell morphology[(1)] .

According to CDC statistics data , It is estimated that SCD affects approximately 100,000 Americans. It occurs among about 1 out of every 365 Black or African-American births and 1 out of every 16,300 Hispanic-American births. About 1 in 13 Black or African-American babies is born with sickle cell trait (SCT) [(2)] . Sickle hemoglobin is highly prevalent among the tribals of central, southern, and western India with variable frequency ranging from 10 to 23%, though increased prevalence is also reported in the non tribal communities[(3)]. The prevalence of sickle cell carriers among different tribal groups varies from 1 to 40 per cent. In Maharashtra, the sickle gene is widespread in all the eastern districts, also known as the Vidarbha region, in the Satpura ranges in the north and in some parts of Marathawada[(4)] .The main focus of the study is to present the variations in the clinical profile of symptomatic homozygous patients of sickle cell disease in pediatric and adult age group.

Methodology:

After obtaining institutional ethical committee approval, the present cross sectional analytical study was carried out in Regional Hemoglobinopathy Diagnosis and Management Centre (Dept of Pathology) and Sickle cell OPD from August 2010- October 2012.The study population included 187 homozygous patients of sickle cell anaemia from indoor and outdoor departments irrespective of the age, caste, origin and ethnic background. ANC patients of homozygous sickle cell disease were excluded from the study.

After taking informed and written consent; diagnosis being based on detailed history and complete clinical examination, solubility testing, hemoglobin electrophoresis, High Performance Liquid Chromato graphy and radiological investigations.

Results & Discussion:

Age group	Number of patients	Percentage N=187
Pediatric(0 to 12 years)	123	65.77
Adult (more than 12 years)	64	34.23
Total	187	100

Table 1: Age wise distribution of study participants

Table 1 shows age wise distribution of SCD patients. Out of 187, 123(65.77%) patients were of pediatric age group (0-12 yrs) and 64(34.23%) patients were more than 12 years of age.

Yadav , Gupta et al (Jabalpur)[(5)], studied 310 patients of SCD and reported that majority of patients were in the age group of 10-15 years(25.16%) followed by 0-5 years(24.19%).[(5)] Sadarangani M et al(Kenya)[(6)] studied 130 SS patients and reported maximum patients between 6-8 years(20.76%)[(6)].In the present study, highest percentage of patients were in the pediatric age group.This could be attributed to more parental awareness in patients who have a previous child of sickle cell disease and also because sickle cell disease is the commonest hereditary disorder in this area. This leads to a tendency that more patients in pediatric age group seek hospital facility .

Gender	Total	Percentage N=187
Male	110	58.82%
Female	77	41.17%
Total	187	100

Table 2: Genderwise distribution of study participants

Table 2 shows genderwise distribution of study participants, Out of 187 patients , 110(58.82%) were male and 77(41.17%) patients were female. In M. N. Khaja et al study the incidence of sickle cell anemia was found to be high in males as compared to females in both populations. Sickle cell trait remained more or less equal in both sexes.[(7)] Shrikhande AV et al (2005), reported male to female ratio of 1.18:1[(8)].In the present study , males outnumbered females in the ratio of 1.42:1.This increase in the composition of male population could be attributed to the negative correlations between Hb A2 and Hb F values between males and females. Marginally high Hb F values in females may attribute to the late onset of symptoms in SCD [(8)] .The social structure in the country is such that there is more availability and accessibility of male population to health care facilities.

First presenting complaint	Pediatric(0 to 12 years)	Adult (Above 12 years)
Pain- 67	25(37.32%)	42(62.68%)
Fever- 42	38(90.47%)	04(9.53%)
Pallor- 25	20(80%)	05(20%)
Respiratory tract infections- 12	10(83.33%)	02(16.17%)
Jaundice – 12	07(58.33%)	05(41.67%)
Diarrhoea- 4	04(100%)	00
Leg ulcer - 1	-	01(100%)

Diabetic ketoacidosis- 1	-	01(100%)
Seizures - 1	01(100%)	-
Typhoid - 1	01(100%)	-
Cellulitis - 1	01(100%)	-
Diminished night vision- 1	-	01(100%)
Asymptomatic-17	15(88.23%)	02(11.77%)
Mortality – 3	01(33.33%)	02(66.67%)

Table3 :Age groupwise distribution of study participants according to first presenting complaint

67 patients presented with pain as the first presenting symptom. Out of which 42 were adults and 25 were pediatric age..42 patients presented with fever as the first presenting symptom. Out of which, 38 patients belong to pediatric age group and 4 were adults. 25 patients presented with pallor as the first presenting symptom. Out of which 20 were pediatric and 5 were adults.12 patients presented for the first time with respiratory tract infections. 10 were pediatric and 2 were adults 12 patients presented for the first time with jaundice.Out of them, 7 were pediatric and 5 were adults.4 patients of pediatric age presented with diarrhea as the first presenting complaint. 1 patient presented with leg ulcer and one with diabetic ketoacidosis as the first symptom , both were adults. Seizures , typhoid, cellulitis were the first symptom in 1 patient each. All were pediatric age group. Diminished night vision was the first presenting complaint in 1 adult patient. Mortality was reported in 3 patients, 1 pediatric and 2 adults. 17 patients were asymptomatic, 15 pediatric and 2 were adults.

Gupta, Yadav et al (India) reported that joint pain (59.7%), bone pains(51.6%),fever(51.3%) form the first line symptoms in SS patients [5] . Splenomegaly (67.7%) and jaundice(69%) are the commonest clinical signs in symptomatic SS patients. Anaemia is seen in 91% patients [5]. Albert Komba et alv (Kenya)(6) reported that Splenomegaly(33%) followed by hepatomegaly(25%) were commoner in homozygous sickle cell patients than joint pains(4%) and fever(12%) [6].This Splenomegaly in African population is attributed to high incidence of malaria and high concentration of circulating immunoglobulins in the African population [6]. A Chijoke,P M Kolo et al Nigeria reported that jaundice(56%),pallor (47.6%), splenomegaly(20%) form the first line symptoms [9] .The Nigerian study illustrates that SS patients have higher levels of Hb F, alpha Thalassemia and malarial infections as compared to Asian population. These three factors combined lead to higher percentages of Splenomegaly [9]..Richard Perrine etal Saudi Arabia[10] reported fever as the first line symptom amounting to 33.33%. Causes of fever included gastroenteritis, respiratory tract infections, septicaemias, osteom yelitis. Brozowick et al [11] quoted vaso occlusive crises and acute chest syndrome as the first line symptoms accounting for 92% and 80% admissions respectively followed by splenomegaly (14%)[11].The British study enlists acute chest syndrome as the first line symptom in addition to vaso occlusive crises[11].Frances, Gill et al reported that painful event(27.63%),acute chest syndrome (26.96%),fever(4.49%), splenic sequestration(3.63%), pallor(4.3%) and stroke(1.15%) as the first line symptoms[12].

In the present study, the first line symptoms included pain(35.82%), fever(22.45%), pallor(13.36%).Whereas ,respiratory tract infections(6.41%),jaundice (6.41%),diarrhea(2.13%) form the minor chunk of first presenting symptoms..Few patients also presented with aprototypical first line symptoms including leg ulcer(0.53%),diabetic ketoacidosis(0.53%), seizures(0.53%), a diagnosed case of typhoid(0.53%),cellulitis(0.53%) and diminished night vision (0.53%).

Conclusion:

Spectrum of clinical presentations varies widely among SCD patients belonging to adult and pediatric age group.We recommend a comprehensive SCD center, which should cater these patients by emphasizing the importance of prevention of disease through health education, genetic counselling,effective manifestations management and regular follow-ups. By adopting a comprehensive care approach, life expectancy and general well being of sickle cell disease patients can be significantly improved and morbidity and mortality from complications of the disease can be significantly reduced.

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