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EPIDEMEOLOGICAL PROFILE OF SICKLE CELL DISEASE PREVALENT IN CHHATTISGARH, CENTRAL INDIA

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ABSTRACT

330 clinically suspected sickle cell disease (SCD) patients admitted to Dept. of Pediatrics, Medical College & Hospital, Raipur (Chhattisgarh) between 9 months to 14 years of age within the time period of May 2000 to September 2007 were chosen as subjects for this study. The diagnosis of SCD was confirmed by Sodium Metabisulphite method. On the basis of the results of electrophoresis the subjects were divided into two groups' i.e. Sickle Cell Anaemia (SCA) consisting of 195 patients and Sickle cell Trait (SCT) consisting of 135 patients. Detailed clinical examination, complete blood count, peripheral blood smear, bilirubin estimation, USG whole abdomen & digital radiograms of skull &limbs were done for every subject. Weakness is the commonest symptom followed by bone pain, abdominal pain, jaundice & fever respectively in decreasing order of prevalence. Pallor is the commonest sign followed by splenomegaly, lymphadenopathy, hepatomegaly & icterus respectively in the study population. When compared with incidences of clinical features &complications of SCD obtained from the studies on people of other parts of Indian sickle cell belt central Indian SCA are having worst features.

KEY WORDS: Sickle cell disease (SCD), Sickle cell anemia (SCA), Sickle cell trait (SCT)

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INTRODUCTION

Sickle cell disease (SCD) is an autosomal dominant haemoglobinopathy where 6th amino acid Glutamate (polar) of β chain of Globin (HbA) is replaced by non-polar amino acid Valine (HbS).In hypoxia, two or more HbS molecules form rod shaped precipitating crystals to impart a sickle shape to RBCs which are easily destroyed by narrow splenic sinusoids, complement cascade & macrophages. Homozygous Sickle cell anaemia [(SCA or HbSS) is clinically severe whereas heterozygous sickle cell trait (SCT or HbAS) rarely have clinical significance**.** Prevalence of SCD is seen in decreasing order in blacks of Tropical African ancestry, Mediterranean basin, Saudi Arabia, Kuwait, Iran and India [(1,2,3)].] DNA polymorphism studies have shown that three independent mutations have given birth to β^S gene responsible for SCD in blacks and Mediterranean, whereas a new fourth mutation resulted to β^S gene in Saudi & Indian people [(1)]. The sickle cell gene was first described in India in a tribal population in the south [(4)]. It is widespread in the state of Orissa and spreads throughout Hindu society, being more common in scheduled castes and upper castes than in tribal groups [(2,3)]. As per Das et al [(5)] wide range of variation (0.00-0.14) in the frequencies of the HbS allele has been observed among 16 ethnic groups of Orissa, Madhya Pradesh, and Maharashtra. A significant excess of SS individuals over that expected under Hardy - Weinberg equilibrium was observed among 6 of 16 populations which have heterogeneous anthropological origins. An increase in HbS allele frequency from east to west is apparent which can largely be explained by differential migration. Population genetic survey data[(6)] indicates that the prevalence of sickle cell disorder is very high amongst the Oktar, Bhil, Powara, Madia & Pradhan tribal population groups from Nandurbar and Gadchiroli districts of Maharastra & neighboring Gujarat, Madhya Pradesh and Andhra Pradesh. The overall prevalence among tribal populations is about 10% for the carrier state and 0.5% for sufferers. Clinically Indian sickle cell anemia is a moderate to severe anemia with high HbF level that seems to be milder than SCA in Africa and Jamaica & similar to SCA in Central Asia[(7)]. Mostly homozygous sickle cell anemia patients in India seek treatment for vaso-occlusive crises, which have greatest incidence during the rainy season, followed by winter [(8)]. The aim of present study is to obtain a detailed epidemiological profile of SCD prevalent in hospital admitted children of Chattisgarh. **.** In a study [(9)] involving in whole of Madhya Pradesh it was observed that the average prevalence of sickle haemoglobin in Gond tribe varies from 15% to 25%. In Bharia and Korku tribes is 20% and 15 % respectively.SCD among Kol tribe is low i.e. about 5% but in Gond, Bhumia and Baiga tribes it is15 to 20 percent. SCD in Bhil group of tribes, varies from 18% to 33%. The present study is aimed at observing different epidemiological parameters of SCD like distribution across age & sex, among different aboriginal ethnic groups in hospitalized children belonging to pediatric ward of Pt. JNM Medical College Hospital, Raipur (CG). This medical college hospital was the only properly functioning apex hospital in the state of Chattisgarh & thus effectively drains the majority of critically ill patients across different parts of the state.For this reason it was chosen as study place.Another important objective is to observe the incidences of important clinical features of sickle cell disease found in the study population & comparing them with clinical profiles of SCD prevalent in other endemic regions across India as observed in different studies.

MATERIALS & METHODS

330 clinically suspected SCD patients admitted to Dept. of Pediatrics, Medical College & Hospital, Raipur (Chhattisgarh) between 9 months to 14 years of age within the time period of May 2000 to September 2007 were selected as subjects for this study. The diagnosis of sickle cell disease was confirmed by Dolland and Castles' Sodium Metabisulphite method. At first the presence of

hemolytic anaemia was confirmed by complete blood count and hemoglobin estimation by electronic cell counter, serum bilirubin (total & direct) by diazo method using fully automated auto analyzers as well as demonstration of microscopic evidence of haemolysis such as target cell, fragmented RBC through examination of peripheral blood smear. Subsequently hemoglobin electrophoresis by agar gel method and absorption spectrometry was performed. On the basis of the presence of electrophoretic bands of HbS & HbA the subjects were divided into two groups i.e. Sickle Cell Anaemia (SCA) and Sickle cell Trait (SCT). Subjects in each group were then clinically examined after examining certificate issued by appropriate state govt. authorities like domiciliary certificates & caste certificates for the population to which he/she belongs .Detailed history of present illness, past illness , were obtained. Ultra sonogram of whole abdomen and Digital Roentgenograms of skull, both hands & feet were obtained for subjects with clinically suspected hepatospelenomegaly and having clinical evidences of bone & joint diseases respectively.

OBSERVATION & RESULTS

TABLE 1
Distribution of Sickle Cell Disease across age & sex

Parameters	Groups	SCA (total no. of cases=195)	SCT (total no. of cases=135)
Sex	Male	111(56.9%)	72(53.3%)
	Female	84(43.1%)	63(46.7%)
Age	9 months to < 5 years	72(36.9%)	15(11.1%)
	5 years to <10 years	99(50.8%)	48(35.6%)
	10 years to <14 years	24(12.3%)	72(53.3%)

Chi-square = 70.46, df = 2, P < 0.001(age)
Chi-square = 0.42, df = 2, P = 0.51(sex)

TABLE 2
Distribution of Sickle Cell Disease in different castes

% of expected Body weight	SCA	SCT
< 50%	09(4.6%)	06(4.4%)
50 to <60%	54(27.7%)	24(17.7%)
60 to<70%	102(52.3%)	48(35.6%)
70 to <80%	18(9.2%)	42(31.1%)
More than 80%	12(6.1%)	15(11.1%)

Caste	SCA	SCT
Sahu	57(29.2%)	36(26.7%)
Kurmi	39(20%)	21(15.6%)
Panka	33(16.9%)	24(17.8%)
Mahar	21(10.8%)	15(11.1%)
Satnami	12(6.1%)	15(11.1%)
Gond	03(1.5%)	06(4.4%)
Kamar	06(3.0%)	0(0.0%)
Kolta	03(1.5%)	03(2.2%)
Brahmin	03(1.5%)	03(2.2%)
Sindhi	0(0.0%)	03(2.2%)
Oriya	03(1.5%)	06(4.4%)
Muslim	03(1.5%)	06(4.4%)
Cristian	03(1.5%)	0(0.0%)

TABLE 3
Bodyweight & Height distribution in Sickle Cell Disease

% of expected height	SCA	SCT
<80%	54(27.7%)	24(17.7%)
80 to <85%	78(40%)	36(26.6%)
85% to < 90%	57(20%)	45(33.3%)
90% to <95%	06(3.1%)	21(15.5%)
> 95%	0(0.0%	9(6.6%)

The symbol% indicates the relative proportions of height & weight of different study subjects as compared to the Harvard standards of their corresponding age.

TABLE 4
Clinical features of Sickle Cell Disease

Parameters	Sign/symptoms	SCA	SCT
Past episodes	Bone pain	126(64.6%)	30(22.2%)
	Abdominal pain	105(53.8%)	24(17.8%)
	Jaundice	66(49.2)%	21(19%)
	Hand Foot syndrome	24(12.3%)	0(0.0%)
	Acute chest pain	06(3.1%)	0(0.0%)
Presenting symptoms	Generalized weakness	171(87.7%)	81(60%)
	Bone/joint pain/swelling	147(75.4%)	42(31.1%)
	Abdominal pain	120(61.5%)	36(26.7%)
	Fever	66(33.8%)	15(11.1%)
	Cough	21(10.8%)	06(4.4%)
	Chest pain/Breathlessness	12(6.2%)	06(4.4%)
	Convulsions	06(3.1%)	0(0.0%)
Clinical signs	Pallor	174(89.2%)	93(68.9%)
	Icterus	87(44.6%)	12(8.9%)
	Bone/joint swelling/tenderness	78(40%)	09(6.7%)
	Hemolytic facies	10(5%)	06(4.4%)
	Lymphadenopathy	99(56.7%)	36(26.6%)
	Purpuric spot	03(1.5%)	0(0.0%)
	Hepatomegaly	9956.7%)	12(8.9%)
	Splenomegaly	138(70.8%)	33(24.4%)
	Cardiac murmur	72(37%)	09(6.7%)
	Consolidation	06(3.0%)	0(0.0%)
	Dactylytis	06(3.1)%	0(0.0%)
	Vasoocclusive	204(61.8%)	
Crises	Hemolytic	66(20%)	
	Sequestration	18(5.45%)	
	Aplastic	03(.09)%	
	Megaloblastic	00(0.0%)	

SCD has a significant age predisposition (Chi-square = 70.46, df = 2, P < 0.001). It is commonest in between 5 to10 years of age (50.8%), followed by 9months to 5years of age. The disease is relatively rare above 10years of age (12.3%). There is no significant sex predisposition. Moderate growth retardation is overt as observed by finding of < 70% of expected body weight in 84.6% of SCA & 55.7% of SCT patients; however decrease in height is not appreciable. Generalized weakness is the commonest presenting symptom f SCD in the study population; however it is more common in SCA (87.7%) than in SCT (60%). Bone pain (75.4%) & abdominal pain (61.5%) are the next two common presenting symptoms of SCA. Fever (33.8%) is also a common presenting symptom of SCA. Pallor is the commonest sign seen both in SCA (89.2%) & SCT (68.9%). Splenomegaly (70.8%), lymphadenopathy (56.7%), hepatomegaly (56.7%) & icterus (44.6%) are the next common signs in SCA.

DISCUSSION

As per Balgir et al the highest prevalence of SCD in India has been recorded in the state of Orissa (1-44.4%), followed by Madhya Pradesh including Chhattisgarh (1-40.0%)[10]. In another study done by him [11] the frequency of sickle cell hemoglobinopathy (3.2-22.5%) is very high among the tribes of Chhattisgarh. In a study in the present Madhya Pradesh done by Yadav et al[13] involving nearly 1/3 rd Scheduled caste, 1/3rd backward caste and remaining Scheduled tribe the incidences of common clinical features were splenomegaly (70%), joint pain (59.7%) ,bony pain(51.6%) & fever(51%.) respectively in decreasing order. Kamble et al [12] showed that incidence was maximum in the Mahar community (70%) followed by Kunbi (8 %) and Teli (6%) whereas Patel et al [14] found that incidence was very high in the Mahar community. In the present study in hospitalized SCD patients of Chhattisgarh however there is no such strong caste predisposition. Sahu (29.2% of SCA & 26.7% of SCT) is the commonest caste affected followed by Kurmi (20% of SCA & 15.6% of SCT) & Panka (16.9% of SCA & 17.8% of SCT). Mahar (10.8% of SCA & 11.1% of SCT) stands at a distant 4th position which stands against findings of Kamble et al & Patel et al. Contrary to the data from Maharastra [6] the incidence of SCD in Gonds (1.5% of SCA & 4.4% of SCT) which is the only local tribal group inhabiting Chhattisgarh is very low. Kamble et al showed among hospitalized patients of SCD 61.6% had homozygous sickle cell disease (HbSS) whereas 38.4% (n=38) had heterozygous state (HbAS).Similar is the finding in the present study i.e. SCA 59.1% & SCT 40.9%. Of these, 63% were below five years of age in Kamble's study but only 26.36%(36.9% of SCA & 15.1% of SCT) in present study instead 5 to 10 yrs age group is the commonest affected (44.55% in total of which 50.8% of SCA & 35.6% of SCT). Male: Female ratio was 1.65:1 in HbSS cases vs 1.32:1 in SCA cases in present study and 1.71:1 in HbAS cases vs 1.5:1 in SCT cases in present study. As per Shrikhande et al [7] mean age for male is 14.55 yrs vs 7.60 yrs in present study & for female is 18.13 yrs vs 9.22 yrs in present study, mean Hb% for male is 7.11 gm% vs 6.8gm% in present study & for female is 6.85 gm% vs 6.2gm% in present study. As per Kar et al[2] moderate to severe anaemia, vaso-occlusive attacks (86.5-89.36%), splenic sequestration (8.43%-12.76%), crippling avascular bone necrosis (5.7%-35.08%), osteomyelitis (5/700), and epistaxis (28.92%-35.08%) were the key clinical features. In present study generalized weakness was the most commonly observed symptom in the study population (87.7% in SCA & 60% in SCT) followed by bone pain (75.4% in SCA & 31.1% in SCT), abdominal pain (61.5% in SCA & 26.7% in SCT), jaundice (36.9% in SCA & 6.1% in SCT) and fever (33.8% in SCA & 11.1% in SCT) respectively. Pallor was the most commonly observed sign in the study population (89.2% in SCA & 68.9% in SCT) followed by splenomegaly(70.8% in SCA & 24.4% in SCT), lymphadenopathy(56.7% in SCA & 26.6% in SCT),hepatomegaly(50.8% in SCA & 8.9% in SCT), icterus(44.9% in SCA & 8.9% in SCT), bony tenderness(40.0% in SCA & 6.7% in SCT), haemic murmur(37% in SCA & 6.7% in SCT) and hemolytic facies(33.8% in SCA & 11.1% in SCT) respectively. Vasoocclusive crisis is the commonest type of crisis encountered in study population (61.8%) followed by hemolytic crisis (20%).

CONCLUSION

Clinical picture of SCD in different parts of India is variable in spite of mildness in general The overall clinical picture of SCA observed in this study seems to be milder than the same observed by Kamble et al but severer than Kar et al & Shrikhande et al. This may be attributed to extremely varying ethnicity of the populations involved in these studies. However as a whole central Indian SCA are having worse features than rest of Indian sickle cell belt. Further studies are needed to confirm this finding.

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