Morbidity Profile of Sickle Cell Disease in Central India

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Abstract

Patients registered/enrolled in sickle cell clinic at NSCB Medical College Jabalpur were studied to understand the clinical profile and natural history of sickle cell disease (SCD) and also to evolve standard guidelines for management and prevention of the disease. Splenomegaly was reported in 67.7% patients. Non-palpable spleen was observed in 30.3 % patients. The most frequent clinical symptoms observed in SCD patients were Icterus (69.0%), Joint pains (59.7%), bony pains (51.6%), fever (51.3%) and abdominal pain (30.3%), joint swelling (16.4%) and chest pain (10.6%). Combination of symptoms commonly observed were splenomegaly with hepatomegaly (37.7%), joint pain with fever (34.5%) and bony pains with fever (30.3%). About 4.1% of cases had multiple blood transfusions. More than half of the cases (52%) had shown no variation in clinical symptoms in any weather. However 16% of cases were shown variation during rainy season and 6% in summer. The patients were followed up at three months interval. A reduction in severity of the disease was observed in the majority of cases after the intervention. Intervention constitutes a package of medicines, health education and avoidance of risk factors susceptible to disease. Frequency of severe cases (7%) was reduced to half (4%) after intervention.

Introduction

Sickle cell disease is the structural disorder of haemoglobin that results in anemia. In India, it is reported mainly in tribal populations of central and southern parts of India (Bhatia and Rao, 1986). The haplotype of sickle haemoglobin gene prevalent in various tribal populations of Central and Southern India revealed that the gene is identical in nature and has evolved independently (Kozolik et al, 1986; Gupta et al, 1991). It is stated that the disease prevalent in India is of milder type than Africa.

The present study is aimed to understand the clinical profile and the natural history of sickle cell disease of the patients who had attended the sickle cell clinic at NSCB Medical College Jabalpur. An attempt shall also be made to evolve standard guideline for management and prevention of the sickle cell disease.

Material and Methods

2-3 ml of blood was taken in EDTA vials from each patient, referred from various OPDs of NSCB Medical College, Jabalpur to Genetic laboratory of Regional Medical Research Centre for Tribals, Jabalpur. Patients diagnosed as sickle cell disease through appropriate laboratory techniques were registered in the sickle cell clinic for detail clinical assessment and follow up. The clinical history, various investigations are recorded in structured proforma. Total of 310 SCD patients were studied.

Complete Blood Counts were done by automatic blood cell counter. Presence of sickle haemoglobin was identified by sickling test with 2% sodium metabisulphite and confirmed by electrophoresis on cellulose acetate membrane with TEB buffer at pH 8.6 and agar gel electrophoresis at pH 6.0. Estimation of Hb F was done by alkaline denaturation method.

Following intervention package was given to each patient of the present study with an interval of three months for a period of one year.

The patients were examined clinically as per structured proforma. All patients were given folic acid to be taken daily. Patients and/or their parents were advised to avoid disease precipitating factors like exposure to extreme climate, excessive dehydration, excessive physical stress etc. Patients were also told to seek appropriate medical intervention quickly upon any minor ailment. Patients were advised to take enough water/fluids. Patients were given folic acid, B-complex and anti-pyretic/anti-inflammatory tablets. They were given symptomatic treatment as outdoor patients and referred to respective clinics in case of emergency. The severity of the disease was assessed by converting the clinical observations in to the numerical score.

Results and Discussion

About three fourth of patients were males. Nearly one third of patients were Scheduled caste (Jhariya, Mehra, Dehariya etc.), another one third was OBC and remaining 17.4 % were Scheduled tribe mainly 'Gonds'.

Splenomegaly was the most common clinical sign observed among 67.7 % the patients of all age groups. Prevalence of splenomegaly decreased with age from 73.3% (0-5 year's age group) to 50% (20-25 years age group). Non palpable spleen was observed in 30.3 % of patients. Incidence of nonpalpable spleen increases slightly with the advancement of age from 26.7% (<5 years) to 44.4% (20-25 years). Similarly splenectomy increased from 1.4% in age group of 5-10 years to 5.6% in 20-25 years (Table 1).

Table 1: Distribution of age and spleen size (cms) in SCD

Age group (Yrs.)	N	S_0 (NP)	S_1 (1-3)	S_2 (3-6)	S_3 (6-9)	S_3 (9+)	Total Splenomegaly	S_X Splenectomy
0-5	75	26.7	38.7	30.7	2.7	1.3	73.3	0
5-10	73	28.8	28.8	30.1	5.5	5.5	69.9	1.4
10-15	78	28.2	28.2	28.2	5.1	6.4	67.9	3.8
15-20	43	34.9	30.2	23.3	7.0	2.3	62.8	2.3
20-25	18	44.4	33.3	11.1	5.6	0	50..0	5.6
25+	23	34.8	17.4	30.4	13.0	4.3	65.2	0
Total	310	30.3	30.6	27.7	5.5	3.9	67.7	1.9

Pallor was observed in majority of patients (91.0%). The other most frequent clinical symptoms observed in these patients were Icterus (69.0%), Joint pains (59.7%), bony pains (51.6%), fever (51.3%) and abdominal pain (30.3%). About 16% of patients had joint swelling and 10.6% of patients had the complaint of chest pain (Table 2).

Multiple clinical signs and symptoms were frequently observed. As an instance, 37.7% of patients had splenomegaly with hepatomegaly; a joint pain with fever was observed in 34.5% of cases and bony pains with fever in 30.3% of patients (Table 3).

Table 2 : Common symptoms observed in SCD patients (N=310)

Findings	No. of cases
Joint pain	59.7
Fever	51.3
Abdominal pain • Gastric • Splenic	30.3 4.8 25.5
General weakness, fatigue & giddiness	27.4
Joint swelling	16.4
Chest pain	10.6
Body pain	11.0
Bony pain	51.6
Pallor	91.0
Icterus	69.0
Hospitalization	38.1
No complaints	4.2

Table 3 : Combination of signs and symptoms observed frequently in SCD patients

Combination of symptoms	% of patients
Fever + Joint pain	107(34.5)
Fever + Bony pain	94(30.3)
Fever + Joint pain +Spleenomegaly	75(24.2)
Fever + Bony pain + Splenomegaly	64(20.6)
Fever + Abdominal pain	46(14.8)
Fever + Abdominal pain +Splenomegaly	34(11.0)
Hepatomegaly + Splenomegaly	117(37.7)
Hepatomegaly + Icterus	123(39.7)
Gallstones	1(0.3)

About 4.1% of patients had history of multiple blood transfusions i.e. more than three units of transfusion (Table 4).

Table 4 : Frequency of blood transfusion in SCD patients

Age	N	No. of blood transfusion						
		0	1	2	3	4-6	7-10	10+
0-5	75	49.3	40.0	6.7	2.7	1.3	0	0
5-10	73	50.7	35.6	6.8	5.5	1.4	0	0
10-15	78	44.9	33.3	11.5	5.1	2.6	2.6	0
15-20	43	20.9	30.2	18.6	20.9	7.0	0	2.3
20-25	18	72.2	11.1	5.6	5.6	5.6	0	0
25-30	12	16.7	66.7	0	8.3	0	0	8.3
30-35	6	33.3	33.3	16.7	0	16.7	0	0
35-40	2	0	50.0	0	50.0	0	0	0
40+	3	0	100.0	0	0	0	0	0
TOTAL	310	43.6	35.8	9.4	7.1	2.9	0.6	0.6

About 43% of patients had no history of blood transfusion and about 35.8% of patients had received only one unit of transfusion in their life time. It is also observed that frequency of cases who had received multiple transfusions increases with age i.e. it was 1.3% in the age group of 0-5 years and 16.7% in 30-35 years. No variations in clinical symptoms with weather were observed in 52.2% of patients. While in remaining cases, 26% patients had suffered more in rainy season followed by 16% in winter and 6% in summer thus indicating the effect of weather on 48% of SCD patients among the studied group (Fig. 1).

Among all these patients, a marked reduction in severity of the disease was observed after intervention (Figs. 2 and 3). Frequency of severe cases (7%) reduced to half (4%) after the implementation of intervention package while the moderate cases reduced from 53% to 40%. After intervention it was observed that the frequency of severe and moderate cases reduced to mild category.

Conclusion

In this study, 30% of the patients did not have palpable spleen. A large proportion of patients among the age group of ten years and above had autosplenectomy.The joint pain (59.7%) and bony pain(51.6%) found in this study are same as in the patients of UK (Murray and May 1988). Clinically it is characterized by localized tenderness and after swelling of the affected area. Thirty percent multiple sites are commonly affected simultaneously (Serjeant, 1994). Fever in Jamaican study (Serjeant, 1994) reported 40% of cases as compared to present study i.e. 51%.

Fig. 1: Effect of weather on frequency of crisis (N=310)

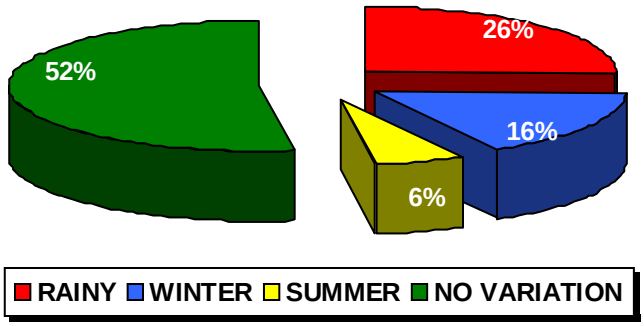

Fig. 2: Severity Index of The Sickle Cell Disease patients before intervention

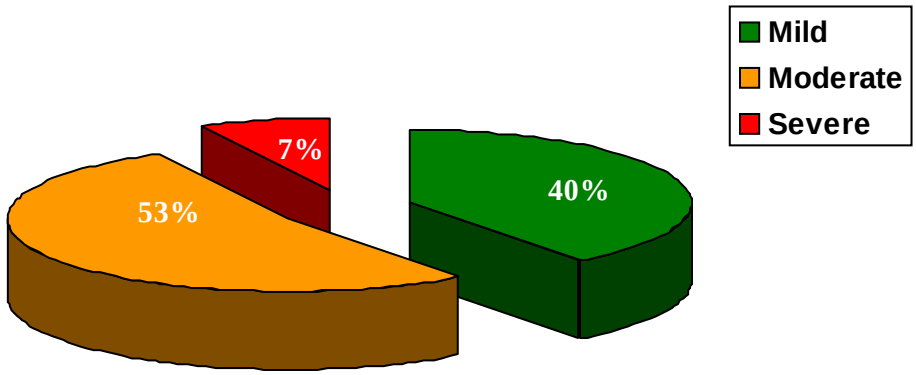

Fig. 3: Severity Index of the Sickle Cell Disease patients after intervention



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