ORIGINAL ARTICLE

Clinical profile of Sickle Cell Disease patients coming to a tertiary care hospital from central Gujarat

Keyur Brahme[1], Kedar Mehta[2*], Kalpita Shringarpure[3], Mahendra Parmar[4]

[1]MD (Medicine) Assistant Professor, [4]MD Associate Professor, Department of Medicine, Baroda Medical College, Vadodara, [2]MD (PSM), DMCH, CNM, CIH, Assistant Professor, Dept. of Community Medicine (PSM), GMERS Medical College, Gotri, Vadodara, [3]MD (PSM), DPH, PGCNM, MBA, Tutor, Department. of Community Medicine Baroda Medical College, Vadodara.

ABSTRACT

BACKGROUND AND OBJECTIVES: The situation of the hereditary hemoglobinopathies including sickle cell disease (SCD) is very grim in India as well as in Central Gujarat. So to study the regional diversity in hemoglobin variants prompted us to look at the clinical profile of patients presenting with this condition. **METHODS:** This was a cross sectional study involving a total of 41 patients with sickle cell diseases over a period of two years attending tertiary care government hospital in Vadodara, Central Gujarat. The data was entered in Microsoft excel sheet and analysis was done using Epi Info software. **RESULTS & CONCLUSION:** Among the 41 study participants having SCD, we found that 23 were males and 18 were females. The mean age of patients was 22.4 years. The common presenting symptoms include arthralgia, bodypain, abdominal pain, breathlessness, fatigue and edema. The mean BMI of the patients of SCD was 20.36 kg/m^2 and their average duration of hospital stay was 6.3 days. Almost all (40) patients were discharged and one patient died due to severe complications of vasoocclusive crisis.

Key words: Sickle cell disease, clinical profile, Gujarat

INTRODUCTION

Inherited disorders of the human hemoglobin which are the commonest gene disorders impact a great burden of on the health services, of which the sickle cell disease is the most common. Such hemoglobinopathies affect nearly 4.5% of the world population.[1]Sickle cell diseases are a group of autosomal recessive genetic disorder caused due to point mutation at the sixth position in beta globin chain, valine substituting glutamic acid. In India, Sickle cell disease (SCD) is more common in central, southern and north-eastern parts of the country. [2]Moreover, SCD is the second most common hemoglobinopathy after thalassemia in India. [3]The prevalence of SCD in Gujarat is around 6.5% and the sickle gene has a high prevalence as high as 30-40% in the tribal areas of Gujarat.

Corresponding Author:
Dr. Kedar Mehta,
1st Floor, College Building,
GMERS Medical College,
Gotri, Vadodara
Contact No: 9879036835
Email-Id: kedar_mehta20@yahoo.co.in

[4]Sickle cell diseases include sickle cell trait, sickle cell anemia and sickle beta thalassemia. Nearly half of sickle cell anemia individuals suffer from the first vasoocclusive episode between 6-12 months and majority before the age of 6 years. Dactylitis and acute chest syndrome have the highest incidence in the first year of life. Mortality and morbidity is very high in sickle cell disease patients. Sickle cell trait is nearly 40 times more than sickle cell anemia. Sickle cell trait is rarely associated with clinical or hematological manifestations. [5]Many studies have been done regarding the clinical manifestations of the sickle cell diseases. But at the same time there is no attempt at national level to enlarge the epidemiological database, establishing special screening programmes for population at risk, imparting genetic counselling or establishing special centres for treatment. There is genetic, ethnic and regional diversity in the hemoglobin variants. So the present study was undertaken to understand the clinical spectrum and profile of sickle cell disease patients coming to a tertiary care Government hospital named Shri Sayaji

General hospital in Vadodara, the third largest city of Central Gujarat.

MATERIAL AND METHODS

The current study was a descriptive cross-sectional hospital based study. Shree Sayaji General Hospital (SSGH) is the third largest tertiary care government hospital in Gujarat. In addition to Vadodara city, SSGH also attracts a large section of population from Vadodara district with tribal pockets like Naswadi, Chhotaudepur, Kawant and nearby districts of Panchmahal, Narmada and Bharuch. This study consists of 41 cases of sickle cell diseases coming to this SSGH. The study participants were recruited over a period of 2 years from 2003 to 2005 after taking written informed consent. Data was collected using a pretested semistructured questionnaire proforma. This being a descriptive study the primary objective was to study the clinico-demographic profile of the patients with SCD. A detailed clinical history of each patient including general biodata, chief complaints for their presentation and physical examination like general examination, vital signs, height, weight and systemic examination was recorded in the study proforma. All the patients were treated for any complications if present due to SCD. Data was collected in hard copies of the study proforma which was then entered in the Microsoft excel work sheet. Data was analyzed using Epi Info software. The study reports proportions of the variables under study in percentages.

RESULTS

As shown in Table 1, 56.1% were males and 43.9% were female patients of SCD in this study. Majority (48.8%) of patients were in the age group of 12-20 years followed by 36.5% were in the age group of 21-30 years and 14.7% were in the age group of 31-40 years. Ethnic group distribution suggests that SCD was commonly observed in Bariya, Parmar and Solanki caste. Nearly 65% of the study participants were illiterate and average family income was Rs. 2144 per month. Most of the SCD patients in this study were from Vadodara (43.9%), Panchmahal (21.9%) and Chhotaudepur (19.5%) districts. Table 2 depicts the clinical profile of the patients with SCD. Almost 63.4% of patients presented with fever, 58.5% with abdominal pain and 53.6% were having arthralgia/bodypain classified as musculoskeletal pain. Further analysis of patients presented with fever, indicated that 23% of them had fever due to malaria while in almost 35% of fever cases cause was not known. Whereas among patients with abdominal pain, nearly 54% of them had abdominal pain due to vasoocclusive crisis and in 16% it was due to hepatosplenomegaly. On General examination of the patients, their mean height, weight and BMI was 149.9 cms, 45.7 kg and 20.36 kg/m2 respectively. Pallor was observed in all patients and icterus in 41% of patients. Other signs like tachycardia and tachypnea were present in 58% and 39% of patients respectively. Average duration of hospital stay of these 41 patients of SCD was 6.3 days while unfavourable outcome of death was seen in 1 patient only.

DISCUSSION

The hemoglobinopathies are the commonest human lethal recessive conditions. In developing countries like India, it has become an important public health problem. It demands on increase in the important health resources in developing countries. Age distribution of the SCD patients in this study showed that it is common in the age group of 12-20 years and the mean age is 22.46 years. Similar findings were also observed by study conducted by Ibiddapo in Nigeria and also in a study by S. Diop et al in Senegal.[6-7] The gender distribution in our study showed 23 (56.1%) males and 18 (43.9%) females. Male preponderance was not found in the studies by Archana et al and Sahu et al.[8-9] In India, Hb S has been detected in more than 50 distinct subgroups predominantly tribals. In Gujarat also it is prevalent in tribes like Bariya, Parmar, Solanki, Vasava, Tadvi and others. Such ethnic distribution of patients in our study is clearly shown in Table 1. Majority of the SCD patients in our study were illiterate and their average family income was around Rs. 2144 per

month. Nearly 45% of patients were from Vadodara district. This dominance of Vadodara inspite of it being a non tribal area, could be accounted by the fact that probably patients in and around Vadodara would come to this hospital even for uncomplicated pain crisis, whereas patients from far-away places like Chhota Udepur would prefer this hospital in search of a tertiary level health care, for acute severe complications or chronic organ damage. The clinical features of SCD are variable. The patient suffers from a severe chronic hemolytic anemia with frequent episodes of crisis to a variety of organ system damage and shortened life expectancy. In our study, fever, abdominal pain and musculoskeletal pain in the form of joint pain and body pain was observed in 63.4%, 58.5% and 53.6% respectively. Similar findings of common clinical presentation as fever and pain were also observed by Patel Archana et al and in a study by Ibidapo in Nigeria.[6,8] Vasoocclusive crisis (54.16%) presenting as abdominal pain has been the commonest manifestation in the present study. Yetunde et al and Archana et al also concluded that vasoocclusive crisis is very common among patients of SCD in their studies.[8, 10] Fever was seen in 63.4% of SCD patients overall, of which 23% had malaria, 15% had pneumonia, 11% had acute chest syndrome and 39% had unknown etiology. Ibidapo in Nigeria observed fever in 72% patients, of which 24% had malaria, 17% had infections and 16% had fever of unknown origin.[6] Height less than 155 cm was seen in 31 (75.6%) patients and weight less than 50 kgs were seen in 27 (66%) patients. Gupta et al found that in 80-90% of children of SCD in his study were having weight below 5th percentile. [11]In present study, hemolytic facies was observed in 12.19% patients. While pallor was present in all patients which was in line with the findings by Ibidapo et al. [6]Jaundice is present in 23% of patients while contradictory to this nearly 70% of patients had jaundice in study by Ididapo et al. [6]The mean duration of hospital stay was 6.3 days and 40 patients of SCD were discharged while one patient died due to severe complications of hemolytic crisis. There were some limitations of this study. A long term follow up could not be carried out in this study due to poor patient compliance. A large scale, prospective population and community survey could have been planned for the actual prevalence determination of SCD.

Table1: Demographic profile of the patients with SCD (N=41)

Variables	Frequency (%)
Gender	
Male	23 (56.1)
Female	18 (43.9)
Age group (in years)	
12-20	20 (48.8)
21-30	15 (36.5)
31-40	6 (14.7)
Ethnicity	
Bariya	6 (14.6)
Parmar	5 (12.2)
Solanki	5 (12.2)
Vankar	4 (9.7)
Rathwa	4 (9.7)
Bhil	4 (9.7)
Muslim	3 (7.3)
Vasava	3 (7.3)
Tadvi	3 (7.3)
Others	4 (9.7)
Education	
Illiterate	27 (65.8)
Primary	8 (19.5)
Secondary	6 (14.6)
Family Income (in Rs.)	2144 per month
Residence	
Vadodara	18 (43.9)
Panchmahal	9 (21.9)
Chhotaudepur	8 (19.5)
Rajpipla	2 (4.8)
Others (MP, Maharashtra)	4 (9.7)

Table2: Clinical profile of the study participants

Variables	Frequency (%)
Symptoms (N=41)	
Fever	26 (63.4)
Abdominal pain	24 (58.5)
Musculoskeletal pain	22 (53.6)
Breathlessness	12 (29.2)
Jaundice	11 (26.8)
Pedal oedema	5 (12.2)
Others	4 (9.7)
Abdominal pain analysis (N=24)	
Vasoocclusive crisis	13 (54.1)
Hepatitis	3 (12.5)
Hepatosplenomegaly	4 (16.6)
Hemolytic jaundice	2 (8.3)
Others	4 (16.6)
Fever analysis (N=26)	
Malaria	6 (23.0)
Pneumonia	4 (15.3)
Acute chest syndrome	3 (11.5)
Hepatitis	3 (11.5)
Cholecystitis	1 (3.8)
Unknown	9 (34.6)
General Examination (N=41)	

Mean Height	149.9 cms
Mean Weight	45.7 kg
Mean BMI	20.36 kg/m^2
Tachycardia	24 (58.5)
Tachypnea	16 (39.0)
Pyrexia	13 (31.7)
Pallor	41 (100.0)
Icterus	17 (41.4)
Pedal oedema	6 (14.6)
Hemolytic facies	5 (12.2)
Average duration of hospital stay (N=41)	6.3 days
Outcome (N=41)	
Discharged	40 (97.5)
Death	01 (2.5)

REFERENCES

1. Angastiniotis M, Modell B, Englezos P, Boulyjenkov V. Prevention and control of hemoglobinopathies. Bull World Health Organ. 1995; 73(3): 375-86.
2. Bhatia HM, Rao VR.Genetics Atlas of Indian tribes. Institute of Immunohematology, (ICMR) Bombay, India; 1986.
3. Balgir RS. The genetic burden of hemoglobinopathies with special reference to community health in India and the challenges ahead. Indian Journal of Hematology and Blood Transfusion. 2002; 20(1): 2-7.
4. Patel A, Naik M, Shah N, Sharma N, Parmar P. Prevalence of common hemoglobinopathies in Gujarat: an analysis of a large population screening program. Natl J Community Med 2012:3(1);112-6.
5. John P. Greer et al. Sickle cell anemia and other sickling syndromes. Wintrobe's Clinical Hematology, 12th Edn 2008;Chp37:1039-71.
6. Ibidapo MO, Akinyanju OO. Acute sickle cell syndromes in Nigerian Adults, Clin Lab Hematol 2000; 22 (3): 151-55.
7. Diop S, Mokono SO et al. Homozygous sickle cell diseases in patients above 20 yeasssrs of age in Dakar. East Afr med J 2003; 71 (11):742-4.
8. Patel Archna et al. Clinical profile of children of sickle cell anemia with reference to oral penicillin prophylaxis. Indian Pediatr 2003; 12(11):1017-21.
9. Sahu T, Sahani NC. Sickle cell anemia in tribal children of Gajapati District of Orissa. Indian J Comm Med 2003; 12 (4):180-5.
10. Yetunde A, Anyaegbu CC. Profile of sickle cell anemia patients above 30 years of age in Nigeria. Cent Afr J Med 2001, 47(4):108-11.
11. Gupta RB, Yadav R, et al. Morbidity profile of sickle cell disease in Central India. Jour Ind Council Med Research 2003; 12 (1):23-26.