www.jmscr.igmpublication.org
Impact Factor 5.84
Index Copernicus Value: 83.27
ISSN (e)-2347-176x ISSN (p) 2455-0450
crossref DOI: https://dx.doi.org/10.18535/jmscr/v5i6.89

IGM Publication
Journal Of Medical Science And Clinical Research
An Official Publication Of IGM Publication

Clinical Profile and Complications in Sickle Cell Disease and Its Variants in Central India

Authors
Ashwin D K, T N Dubey, R S Saxena, Harsha H B, Gopal Krishna

Abstract

Objective: *Sickle cell anemia is a hereditary anemia, with nearly 50% of the world population affected by sickle cell disease reside in India. Complications of sickle cell disease increases morbidity. This study was designed to asess the clinical profile and complications of sickle cell anemia and its variants.*

Method: *Hospital based cross-sectional observational study was undertaken among adolescent and adult patients conducted at Hamidia Hospital Bhopal. A total of 50 patients were studied and analysed. Clinical features and complications were noted and compared with previous studies.*

Results: *Males are found to be predominantly affected by sickle cell disease with male to female ratio of nearly 3:2.Most common symptoms of presentation are Bone pain and Abdominal pain. Most common signs are Pallor, Fever and Splenomegaly. Splenomegaly was found more common in the adolescent age group. Vaso-occlusive crisis is the most common crisis of presentation.*

Conclusion: *Sickle cell disease is one among the common presentations of anemias in central India. Prevalent in females more than males with morbidity and mortality affecting adolescent and early adult population. Bone pain and pallor are common symptoms and signs of the disease with significant presence of splenomegaly, particularly in sickle-thalassemia patients.*

Introduction

Sickle cell anemia is a hereditary anemia, predominantly seen amongst various tribal populations of India. Sickle cell disease is prevalent in many parts of India, where the prevalence has ranged from 9.4 to 22.2% in endemic areas.[1]It is characterized by chronic hemolytic anemia and vaso-occlusive crisis,which can lead to widespread vascular occlusion by sickled red blood cells leading to multiple organ infarctions. In this respect, SCD can be considered as a multisystem disease.[2]

Haemoglobin S is the result of a single base pair change, thymine for adenine, at the sixth codon of Beta-globin gene. This change encodes valine instead of glutamine in the sixth position in the Beta-globin molecule.[3]

Sickle cell anemia is a disease that is responsible for considerable amount of morbidity and mortality in India and particularly southern regions of Madhya Pradesh. It not only has an adverse effect on the health status of people affe-cted but significantly affects the quality of life.

The complex nature of disease with its variability in clinical features from region to region, along with various risk factors for hospitalization need to be thoroughly investigated.

It is believed that this study will motivate research worker to work for further improvement of the disease, clinical presentation, diagnostic measures

and management in already existing health care delivery system in a tertiary care centre in the disease affected areas and which may lead to even better survival and quality of life for these patients.

Hence the study is taken up to find the clinical profile of Sickle cell disease in adolescent and adult population.

Material and Methods

This prospective observational study was carried out in department of medicine, Hamidia Hospital Bhopal for a period of one year.This study carried out on 50 sickle cell anemia patients and its variants attending the Medicine OPD and IPD, whose age is 13yrs and above and who willing to participate in the study. Detailed history and careful clinical examination performed on each patients. The hemoglobin and hematocrit along with battery of investigations (given below) to detect crisis and organ failure were carried out during study period. The patients with SS pattern on electrophoresis were labelled as 'disease' and patient with AS pattern were labeled as 'trait'. The study was approved by hospital ethics committee and informed consent was obtained from each patients.

Observation

Table No.1: Number of Patients in Various Sickle Cell Diseases

TYPE OF SICKLE CELL DISEASE	NO. PATIENTS	PERCENTAGE
Sickle Cell Anemia	33	66.00%
Sickle Cell Trait	8	16.00%
Sickle Thalassemia Syndrome	9	18.00%
Total	50	100.00%

Table No.2: Type of Sickle Cell Disease and Gender Distribution

	SEX DISTRIBUTION		TOTAL
TYPE OF SICKLE CELL DISEASE	**MALE**	**FEMALE**	
Sickle Cell Anemia	19	14	33
Sickle Cell Trait	5	3	8
Sickle Thalassemia Syndrome	5	4	9
Total	29	21	50

Table No.3: Type of Sickle Cell Disease and Age Group

	SICKLE CELL ANEMIA	SICKLE CELL TRAIT	SICKLE THALASSEMIA SYNDROME	TOTAL
AGE GROUP(YRS)				
13-20	15	2	8	25
21-30	11	6	0	17
31-40	4	0	1	5
>40	2	1	0	3

Table No.4: Symptoms and Signs in Various Sickle Cell diseases

SYMPTOMS	SCA	SCT	STS	p VALUE
Easy fatigability	20	4	3	0.33
Bone pain	24	3	3	**0.03**
Fever	15	3	4	0.92
Pain Abdomen	14	4	8	**0.046**
Breathlessness	7	1	1	0.71
Cough	8	0	0	0.086
Splenomegaly	9	4	8	**0.0036**
Hepatomegaly	5	2	3	0.44
Icterus	12	3	3	0.98
Dehydration	25	2	6	0.11
Febrile	15	3	4	0.12

Table No.6: Type of Sickle Cell Disease and Various Crisis Types

TYPE OF SCD / CRISIS	SICKLE CELL ANEMIA	SICKLE CELL TRAIT	SICKLE THALASSEMIA SYNDROME	TOTAL
Vaso-occlusive crisis	16	1	7	24
Acute chest Syndrome	7	0	0	7
Haemolytic crisis	4	0	2	6
Sequestration Crisis	1	0	4	5
Aplastic Crisis	1	0	0	1
Total	29	1	13	43

Table No.7: Type of Sickle Cell Disease and Systemic Complications

TYPE OF SCD	AFI	Cholecystitis	Epilepsy	Hepatic congest	Normal	Others	Nephrological	Sepsis	Total
SCT	4	1	0	1	3	0	0	0	8
STS	0	1	1	0	5	3	2	2	9
SCA	4	2	1	1	16	3	2	0	33
TOTAL	8	4	2	2	24	4	4	2	50

A total 50 sickel cell anamia and its variant patients were studied and analysed. Among 50 patients majority were sickle cell anemia 33(66%) (table1).

As shown in table number2, 29 patients (58%) are males and 21(42%) patients are females out of total 50 cases observed. Among Sickle cell anemia patients, 19(57.57%) are males and 14(42.42%) are females suggestive of more male predilection. Similarly, sickle thalassemia and sickle cell trait types have 5 male patients each accounting to 60.2% and 50.5% respectively.

As shown in table number 3, more number of sickle cell anaemic(15 out of 25) and sickle thalassemia (8 out of 25) patients are found in 13-20 years age group followed by 21-30 years age group i.e;11 out of 17 for Sickle cell anemia cases. But, more no. of Sickle cell trait patients are in the Age group of 13-20years.

As shown in table 4, Symptoms in various Sickle cell diseases are analysed and found that Bone pain and Pain abdomen are statistically significant presentations. As shown in table 5, Signs in various Sickle cell diseases are analysed and found that Splenomegaly as statistically significant sign of presentation.

As shown in table 6, most common type of crisis in Sickle cell anaemic patients are Vaso-occlusive crisis(16 out of 29) followed by Acute chest syndrome (7 out of 29) and Haemolytic crisis(4 out of 29).In Sickle thalassemic patients vaso-occlusive crisis(7 out of 13) is commoner than Sequestrational crisis(4 out of 13) and haemolysis. But Sequestrational crisis is more common in Sickle thalassemia patients than other Sickle cell syndromes. As shown table 7, most common type of systemic illness in Sickle cell disease and its variant are acute febrile illness (8 out of 50), cholecytitis (4 out of 50), renal impairment (4 out of 50), epilepsy (2 out of 50) and Hepatic impairment (2 out of 50).

Discussion

The sickle cell anemia is a very common genetic disorder, 50% of the world population affected by sickle cell anemia reside in India. Prevalence SCD in India is 5.7% as per study by Kamble[4] et al over Epidemiology of Sickle cell disease in central India and Dangi[5] et al indicating high prevalence rate (7%), in district Bhopal having mixed population of diverse ethnic groups.

In present study, total 50 patients presenting to Hamidia Hospital were included. Of all the 50 cases studied 25 cases(50%) were of ≤20 years age and 47 cases(94%) are within 40 years of age where as only 3 cases(6%) are of above 40 years age which is similar to the earlier study by Vasundhara M[8] et alin which 86% of Sickle cell

disease patients are <40 years of age. The reason for recording few cases in this age group might be that most of the patients did not survive beyond this age.

Considering the distribution of cases in both the sexes male to female ratio is 1.38:1 showing male sex preponderance. Males are more prone to be exposed to known precipitating factors as compared to females as stated by studies of Kar[6,7] et all and Diggs[5] et al. There is also a gender bias in the community as males are given better care and females are neglected. Both these reasons may have contributed to the preponderance of males over females in the present study. But this observation is contrary to the study by Vasun-dhara[8] M et al which found equal distribution of the disease among boththe sexes. In the age group of below 20 yrs, males (19) outnumbered females (11), where as in the age group 20-40yrsthe reverse was seen with ratio of 1.57:1(11:7) with female preponderance, which is because females of 20 to 40 years age group are more prone for crisis as they have risk factors like menstruation, parturition and nutritional deprivation.

Amongst the symptoms, most frequently noted was Painful crisis of the Bones and Joints (30 cases, p value<0.0372) with statistical significance. This was seen in almost 2/3rd of the patients, followed by easy fatigability, abdominal symptoms (pain abdomen with p value<0.0466), fever, chestsymptoms cough and breathlessness), renal and neurological symptoms which is similar to the varied symptomatic presentation in study by Vasundhara[8] et albut is quite different to study by Swarnkar[9] et alconducted among children,which had Recurrent fever (51.14%) as the most common symptom followed by abdominal pain, joint pain and fatigue. The risk of painful crisis and acute chest syndrome begins in the first year of life and increases steadily. Out of all symptoms, Bone pain was present in 24 out of 33 Sickle cell anemia patients and Pain abdomen in 8 out of 9 Sickle thalassemia patients which were more common than easy fatigability, fever, breathlessness and cough. Bone pain and Pain Abdomen were of statistical significance among the groups of SCD in our study.

On evaluation of Signs, most common presentation was Pallor, followed by fever others were splenomegaly, icterus, dehydration, hepatomegaly and edema, pattern of which is not similar to the study by Swarnkar[9] et alin which splenomegaly (44.27%) was the most common sign followed by hepatomegaly, pallor, icterus and lymphadenopathy.Conversely,

splenomegaly(42%) was present in 21 patients out of 50 total patients among which 8 out of 9 Sickle thalassemia patients , with p value of less than 0.0036 is of statistical significance and rest of the signs were of no statistical significance, which is similar to findings of Swarnkar[9] et al study. Splenomegaly was found in 16 patients between the age of 13-20 years, suggesting prevalence of splenomegaly in adolescent sickle cell patients, which is also statistically significant.

Sickle cell disease patients in the study are presented with different crisis, most common was vaso-occlusive crisis with 24 out of 50 patients had this feature followed by Acute chest syndrome (7 out of 50), Haemolytic crisis(6 out of 50), Sequestration crisis (5 out of 50), Aplastic crisis. The findings of present study in correlation with Swarnkar[9] et all and Vasundhara[8] et all studies. Among Vaso-occlusive symptoms Bone Pain followed by Joint pain were common.

Other Systemic complications are also noted in Sickle cell disease patients with most common was Acute Febrile Illness (8 out of 50 patients) followed by Cholecystitis, Renal complications, Epilepsy, Hepatic Complications (Congestion, Chronic liver disease), Antenatal status, Sepsis and Splenic Abscess.

The number of these systemic complications are very low to have a statistical significance. Most of the patients had Acute febrile illness as their complication suggesting frequent infections may be due to altered splenic functioning with increased susceptibility to infections.

Conclusion

Among 50 patients, 33 are of sickle cell anemia, 9 patients of sickle cell trait and 8 patients are of sickle thalassemia syndrome. Most of the patients are present in age group of 13-20 years. Males are found to be predominantly affected by sickle cell disease with male to female ratio of nearly 3:2.Most common symptoms of presentation are Bone pain and Abdominal pain. Most common signs are Pallor, Fever and Splenomegaly. Splenomegaly was found more common in the adolescent age group, especially in Sickle-thalassemia patients. Vaso-occlusive crisis is the most common crisis of presentation. Females of reproductive age group are vulnerable to complications, which even can be fatal.

References

1. Weatherall DJ, Clegg JB(2001).”inherited haemoglobin disorders:an increasing global health problem”. Bull. World Health Organ.79(8):704 12.PMC2566499. PMID11545326.//www.ncbi.nlm.nih.gov/ pmc/articles/.PMC2566499.
2. Meshikhes AW,al-Faraj AA.Sickle cell disease and the general surgeon.J R Coll Surg Edinb 1998;43(2):73-79.
3. DeBaun MR, Jones MF, Vichinsky E.Sickle Cell Disease in Nelson text book of Pediatrics 19th edition. Behrman RE, Kleigman RM, Jenson HB, Saunders Phildelphia,2011;456:1663-1670
4. Kamble M, Chatruvedi P. Epidemiology of sickle cell disease in a rural hospital of central India. Ind Pediatr 2000; 37: 391-396.
5. Dangi C, Sajid M, Sawke G K, Ambhore J. Sickle cell hemoglobinopathies in district Bhopal. Indian J Hum Genet 2010;16:100-2.
6. Kar BC, Satyabhama Devi : Clinical profile of Sickle Cell disease in Orissa.
7. Indian J Pediatrics 1997;64:73-77
8. Kar BC; Sickle cell disease in India, JAPI 1991; Volume 39 (12) ;954-960
9. Vasundhara M, Ramdas R,Bhagya Lakshmi A, Satish Kumar S, Subash R.Study of sickle cell anemia with clinical and hematological correlation. Int J Res Med Sci2016;4:246-51.
10. K Swarnkar, A Kale, B Lakhkar. A Clinico-Epidemiological and Hematolo-gical Profile of Sickle cell Anemia with special reference to Pencillin prophy-laxis in a rural Hospital of central India. Internet Journal of Epidemiology.2010 Volume 9 Number 2.