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Research Article

CLINICAL PROFILE OF SICKLE CELL DISEASE PATIENTS IN SOUTH RAJASTHAN

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ABSTRACT

Background: Sickle cell disease is a major health problem among the tribal population which leads to varied complications and even resulting in premature death. Probable estimate of this condition in Southern Rajasthan is quite high. Authors aim to determine the spectrum of manifestations of the Sickle Cell Disease patients. ***Objective:*** To study the clinical manifestations of Sickle cell disease patients visiting a tertiary care hospital at Udaipur, Rajasthan and to compare between manifestations of homozygous state (HbSS) v/s Sickle beta thalassemia patients. ***Study Design:*** This study was a hospital based observational prospective study. *Participants:* 38 patients with sickle cell disease were included in this study. *Methods:* Present study was conducted at Center of Excellence for Sickle cell Disease, RNT Medical College, Udaipur, Rajasthan. Patients with Sickle Cell Disease visiting our center were thoroughly examined and investigated along with scrutiny of past medical records. ***Results: Conclusion:*** This study shows that sickle cell disease leads to various complications leading to a significant impact on quality of life. There is a need for more extensive studies over the subject in this region.

INTRODUCTION

Sickle cell disease (SCD) is an autosomal recessive group of genetic blood disorders caused by sickle haemoglobin (HbS). It is the result of a single base-pair change, thymine for adenine, at the 6th codon of the β-globin gene. This change encodes Valine instead of Glutamate in the 6th residue in the β-globin molecule. Sickle cell disease refers not only to patients with sickle cell anemia, but also to compound heterozygotes where one β-globin allele includes the sickle cell mutation and the 2nd β-globin allele includes a gene mutation other than the sickle cell mutation, such as HbC, β-thalassemia, HbD, and HbOArab.[1]. This conformational change in β globin chain leads to formation of sickle hemoglobin (HbS) which has propensity to polarise in deoxygenated state leading to various complications.

Sickle cell disease (SCD) is highly prevalent among the tribal population of India with prevalence ranging between 1 to 40 %.[2-8] According to the Census of 2011, 49.71% of the population residing in Udaipur district, 76.4% in Banswara, 70.8% in Dungarpur, 63.42% in Pratapgarh and 8.08% in Sirohi district belongs to Tribal community.[9] With such a huge tribal population, probable estimate of sickle cell disease in this region is quite high. In this study, we intend to determine the characteristics of Sickle cell disease among the SCD patients visiting a tertiary care hospital at Udaipur, Rajasthan and to identify the clinical spectrum of presentation and distribution of vaso-occlusive crisis in Sickle cell disease.

METHODS

This was a hospital based observational prospective study conducted at the Center of Excellence for Sickle Cell Disease, RNT Medical College, Udaipur, Rajasthan on 38 SCD patients. Detailed history, clinical examination and laboratory investigations including CBC with PBF and other relevant investigations according to the signs and symptoms were done. Diagnosis of SCD was confirmed with haemoglobin electrophoresis whenever reqquired. Patients diagnosed as SCD were managed accordingly with supportive treatment, hydroxyurea and blood transfusions whenever reqquired.

STATISTICAL ANALYSIS

All collected data were entered into Microsoft excel worksheet and then managed and analysed with standard software of biostatics (SPSS version 26). Categorical data were analysed using Chi square test and Fischer exact. A p-value <0.05 was considered statistically significant.

RESULTS

In this study, 38 SCD patients were enrolled, out of which 26(69.2%) were males and 12(30.8%) females (Male: Female ratio- 2.2:1). Of these 20 were homozygous (HbSS) and 18 were Sickle Beta thalassemia. **Table 1** shows demographic profile of the patients.

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Table1 Demographic profile of the study patients

characteristic	Sickle beta thalassemia	Sickle cell anemia	Total
AGE			
0-5 years	0	3	3 (7.9)
5-18 years	17	16	33 (86.8)
>18 years	1	1	2 (5.3)
GENDER			
Male	12	14	26 (69.2)
Female	6	6	12 (30.8)

Most common presenting complaints of these patients are easy fatiguability (55.6%), pain/swelling of limbs (50%), abdominal pain (33.3%) and back pain (27.8%). Pallor (68.4%), splenomegaly (55.3%) and heaptomegaly (39.5%) were the common examination findings amongst the study patients.

CONCLUSION

This study provides an account of common clinical presentations of SCD patients. Pain in limbs, abdominal pain, heaptomegaly and splenomegaly are the manifestations that should arise a suspicion of SCD. It also provides an account of comparision between sickle β thalassemia than sickle cell anemia.

LIMITATIONS

- This is a hospital based study of small number of patients. More studies are required to establish the facts.

FUNDING

None

CONFLICTS OF INTERESTS

None

Table 2 Clinical Manifestations of SCD patients

Presenting complaints	**Sickle β Thalassemia (n=18)**	**Sickle cell anemia (N=20)**	**Total**	**p-value**
Easy fatiguability	12 (66.7)	8 (40)	20 (52.6)	0.18
Pain/swelling of limbs	9 (50)	9 (45)	18 (47.3)	1
Abdominal pain/distension	8 (44.4)	4 (20)	12 (31.6)	0.16
Back pain	4 (22.2)	6 (30)	10 (26.3)	0.46
Limping/AVN	2 (11.1)	3 (15)	5 (13.2)	0.72
Headache	1 (5.5)	1 (5)	2 (5.3)	1
Examination:				
Pallor	17 (94.4)	9 (45)	26 (68.4)	**0.002**
Icterus	10 (55.6)	4 (20)	14 (36.8)	**0.04**
Hemolytic facies	9 (50)	5 (25)	14 (36.8)	0.17
Hepatomegaly	9 (50)	6 (30)	15 (39.5)	0.310
Splenomegaly	13 (72.2)	8 (40)	21 (55.3)	**0.046**

Pallor (p 0.002), icterus (p 0.04) and splenomegaly (p 0.046) were significantly more in sickle β- thalassemia patients as compared to sickle cell anemia patients. **(Table 2)**

DISCUSSION

The common presenting complains amongst the SCD patients in this study were easy fatiguability, Pain/ swelling in the limb and abdominal pain. These were similar to observations of Mandot S, Ameta G [8] who found that weakness/fatigability (75%), pain abdomen (33.3%), fever (43%) and bone pain (15%) were common manifestations in sickle cell disease. Common examination findings in our study were pallor (68.4%), splenomegaly (52.6%), heaptomegaly (39.5%), icterus (36.8%). Yadav R et al. [10] also found splenomegaly (67.7%) and icterus (69.0%) as common findings in their study of SCD patients. Panigrahi S et al.[11] have observed in their study of SCD patients at Chhattisgarh that splenomegaly was found in 57% and hepatomegaly in 31%. Their findings were similar to our study. In our study, pallor, icterus and splenomegaly were found to be significantly higher in sickle beta thalassemia patients as compared to SCA patients ($p<0.05$). Yadav R et al. [10] also found in their study that splenomegaly was significantly more common in sickle β thalassemia than sickle cell anemia.

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