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Clinical profile of sickle cell disease patients attending pediatric department of a tertiary care hospital and Sickle-Thal center: A cross-sectional study

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Abstract:

BACKGROUND: Sickle cell disease (SCD) is a major public health problem in certain states of India associated with morbidity and mortality in children. The characteristics and clinical features of SCD are different in India as compared to those in developed countries with considerable clinical diversity.

AIMS: This study aimed to describe clinical features in patients with SCD at the time of diagnosis and first visit.

SETTING AND DESIGN: Department of Pediatrics at tertiary care hospital or Sickle-Thal Society Amravati,India (Both Collectively called as Unit). Design: Cross sectional Study.

MATERIALS AND METHODS: All SCD patients 0–18 years, attending unit from October 1, 2019, to March 31, 2021, were included in the study. Data collected were based on history, clinical examination, and review of old records. The sample size was 153.

STATISTICAL ANALYSIS: Data was analyzed by using software statistical packages in social sciences (SPSS) version 16. Mean, standard deviation (SD), proportions were calculated and Chi -square test of significance was used.

RESULTS: Of 157 SCD patients (145 families), 90 (57.32%) were males and 67 (42.68%) females. The mean age of diagnosis and unit visit was 53.59 and 115.74 months, respectively. At diagnosis, paleness (68%), pain (54%), and fever (45%) were common complaints with other nonspecific complaints. Pallor (97.45%) and palpable spleen (53.90%) were common signs at the first unit visit. The spleen was palpable in 65 (42.48%) even after 5 years.

CONCLUSION: SCD patients had paleness, pain, and fever as the most common complaints. Palpable spleen was seen even after 5 years of age. Early suspicion is key for diagnosis.

Keywords:

Fever, pain, pallor, splenomegaly, stroke

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Introduction

Sickle cell disease (SCD) is a major public health problem in certain states of India such as Madhya Pradesh, Orissa, Chhattisgarh, Jharkhand, Gujarat, and Maharashtra, with an estimated 5200 live births each year.[1,2] Rather than severe African haplotype, a unique milder "Arabian- Indian'' haplotype is seen in India.[3] The characteristics and clinical features of SCD are different in India as compared to those in developed countries with considerable clinical diversity.[4] Despite the number of patients born with SCD in resource-limited countries such as

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India, only a few detailed studies have described the clinical course and complications of SCD. In the cohort study conducted at Nagpur, 12% of SCD babies died by 2 years indicating early fatality of SCD.[5] With this background, the present study was conducted to find out the different clinical presentations of SCD patients at diagnosis and during Hospital/Sickle-Thal Society visits, which will help pediatricians to suspect the disease early leading to early diagnosis.

Materials and Methods

A cross-sectional study was conducted among all SCD patients up to 18 years at a tertiary care center running pediatric outdoor and indoor services (OPD/IPD) and Sickle-Thal Society Amravati which is a private nongovernmental organization working for sickle cell anemia (SCA) and thalassemia patients (collectively labeled as unit). Consent was taken from parents and in children more than 7 year and assent was also obtained before the study. The study was conducted from October 1, 2019, to March 31, 2021.

Any child with hemoglobin "S" (Hb S) more than 50% or 'SS' or 'AFS' pattern in quantitative Hemoglobin Electrophoresis showing only 'SS' or 'AFS' band respectively while the patients denying consent, age more than 18 years, HbS <50, and only sickling/solubility positive were not included in the study. The sample size of 153 was calculated, using OpenEpi Software version 3 at an 80% confidence interval and 20% error, considering fever (63.4%) as the most common clinical presentation from a previous study.[6]

Data were collected by a pediatrician, detailed history including sociodemographic characteristics of study participants, clinical examination, and review of old records. The complaints at, before, and after diagnosis till unit visit were assessed by detailed history and were correlated with old records wherever available. Clinical features at the time of unit visit were directly seen and noted by a pediatrician. The term recurrent was used in SCD patients if they had more than two episodes of the same complaints. The standard definitions of different crises for SCD were used. Wasting and stunting were said to be present when weight and height were less than the 3rd percentile.

Statistical analysis

Data was analyzed by SSPS software version 16 (IBM, Armonk, New York 10504-1722 United States) and mean, standard deviation (SD), proportions were calculated and Chi -square test of significance was used.

Ethical committee approval was taken before starting the study and anonymity and confidentiality were maintained.

Results

There were 145 families with 157 SCD patients consisting of 90 (57.32%) males and 67 (42.68%) females. Out of 157 patients, 39 (24.8%) were diagnosed at our unit during their visit. Table 1 depicts the different ages of presentation at diagnosis and first visit at unit. The mean age of diagnosis was 53.59 ± 43.84 months (range of 4–204 months). The mean age of unit visit was 115.74 (SD: 55.29) months (range from 9 to 214 months). The mean age of diagnosis in males and females was 49.12 (SD: 38.17) and 59.59 (SD: 50.14) months, respectively.

Out of 157 patients, only 18 (11.46%) had a history of consanguinity, 29 (18.47%) had either positive family history (F/H) suggestive of some disease, 33 (21.01%) had a family history of SCA, and 53 (33.76%) had either of any disease.

Figure 1 [Supplementary Table 1] shows the different complaints described at the time of diagnosis, with paleness, pain, and fever being more common. At diagnosis, 42 (26.75%) had symptoms for the first time. Other features at diagnosis were as follows: vomiting (4), loose motion (9), not gaining weight (1), urticarial rash (1), abdominal distension (3), edema (3), altered behavior with posterior reversible encephalopathy syndrome (PRES) (1), hepatic encephalopathy (1), cerebral malaria (1), decreased activity (7), nonhealing ulcer (1), nasal bleed (1), abdominal mass felt by mother (1), and suspected lymphoma in abdominal sonography (1), and only five were diagnosed during community screening.

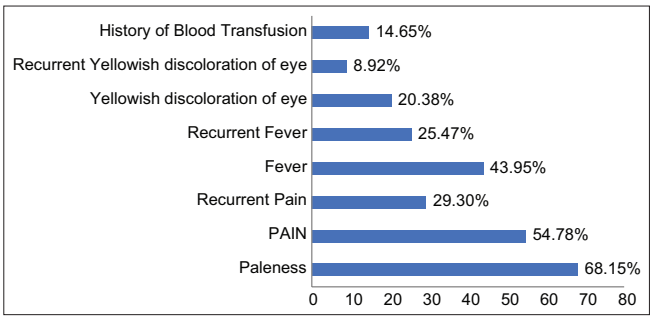

Figure 1: Different complaints at diagnosis (*n* = 157)

Table 1: Age-wise distribution of study participants as per the age of diagnosis and age of first unit visit (*n*=157)

Age	At diagnosis, *n* (%)	At the first unit visit, *n* (%)
0–6 months	2 (1.27)	0
>6 months–1 year	13 (8.23)	1 (0.64)
>1 year–5 years	88 (56.05)	28 (17.83)
>5 years–10 years	39 (24.84)	59 (37.58)
>10 years–18 years	15 (9.55)	69 (43.95)
Total	157 (100)	157 (100)

Total 85 patients had a painful crisis with involvement of upper and lower extremities in 82% and 86%, respectively. Abdomen, back, and chest pain were seen in 34%, 13%, and 8%, respectively. Pain at two sites was present in 53% of cases compared to only one site and three or more sites in 35% and 12%, respectively.

The mean weight of SCD children during unit visit was 24.71 (SD: 1.21) kg and the mean height was 125.94 (SD: 23.26) cm. Wasting and stunting were seen in 29.3% and 21.7%, respectively.

At unit visit, pallor was present in 153 (97.45%), icterus in 54 (34.39%), hemolytic facies in 16 (10.19%), and palpable liver in 72 (45.86%). The palpable spleen was found in total 83 patients, of which 17 were up to 5 years, whereas 34 and 32 were between 5–10 and 10–18 years, respectively. This difference was not found to be statistically significant ($P = 0.352$).

About 98 (62.42%) patients already received blood transfusion, out of which 51 (52.04%), 17 (17.34%), and 30 (30.61%) received 1, 2, or 3 and more transfusions, respectively. There was a history of stroke in 5 (3.18%) patients, and all were <10 years of age with male predominance (4:1). The hepatic encephalopathy-like presentation was seen in two patients, whereas two girls had multifocal osteomyelitis, and in both of these cases, girls were more than 10 years of age.

Discussion

Categorization of SCD patients was done depending on clinical presentation at diagnosis and at the time of presentation to our unit. Pallor, pain, and fever were common clinical symptoms at diagnosis along with other nonspecific symptoms. Pallor, palpable liver, and palpable spleen were the most common signs in SCD patients during unit visits. Compared to height, weight was affected in more SCD patients.

Family history of SCA was seen only in 21% of patients in our study population which was less as compared to other studies, so negative family history cannot be considered absence of SCD.[7,8]

The mean age of diagnosis was 53.59 (SD: 43.84) months with 56% diagnosed between 1 and 5 years of age which was higher than the study by Faruk *et al*. but lower than the study by Adewoyin *et al*.[9,10] Newborn screening for SCD should be strongly recommended for early diagnosis and early intervention. The mean age of presentation to unit was 115.74 (SD: 55.29) months which corresponds with the study by van den Tweel *et al*. but was much higher as compared to another study by Faruk *et al*.[9,11]

Similar to our study, pain, fever, and paleness were common complaints in most of the studies, as shown in Table 2.[4,6,12,13]

Pain is the hallmark of SCD. In our study, 54.78% of patients presented with pain as presenting complaint similar to different studies showing pain as presenting complaint in 40%–70% of sickle cell patients.[6,8,12,14] Our study showed pain in extremities as the most common presentation similar to the study by Salman and Hassan, whereas in the study by Patel *et al*., abdominal pain was more common.[12,15]

In our study, it was evident that at the time of diagnosis, complaint related to pallor was seen in 68% as compared to recognition of pallor by the attending pediatrician (97%) during unit visit, thus signifying that pallor as a symptom is underrecognized by parents and thus emphasizes the importance of educating the community about early recognition of pallor by examining palms or face. During visit to unit, pallor, palpable spleen, and palpable liver were common signs similar to other studies, as shown in Table 2.[8,12,13] Although books mention autosplenectomy is present in most of SCD patients after 5 years, our study and other Indian studies showed splenomegaly after 5 years in 42% of cases, indicating its presence should not be considered a reason for exclusion of SCD diagnosis after 5 years.[8,12]

In our study, stroke was more common in boys as compared to girls (4:1) and all were <10 years of age. This finding was comparable with the study by Lagunju *et al*.[16] Larger studies are required to verify this finding and reasons behind it.

Our study showed that weight was more affected than height similar to the study by Fawzia *et al*., whereas the study by Fadhil *et al*. in case-control study showed stunting in 25% and wasting in 12.5%.[13,17] The larger sample size and inclusion of complete pediatric age group upto 18 years in main strengths of our study. While as genotyping is not done (not knowing exact percentage of SCA and sickle beta thalassemia) and some data is dependent on history given by patient which may lead to recall bias.

Conclusion

Though pallor, pain and fever being most common complaints in SCD patients some uncommon complaints at diagnosis also given by patients. Compared to height, weight was affected in more SCD patients. Palpable spleen was seen in many patients even after 5 years of age. Early suspicion is key for diagnosis.

Table 2: Studies on sickle cell disease: Methodology and results

Study site	Type of study	Number of study participants/duration/age	Results	Comparison with our study
Hospitalized children, GMCH Nagpur, India[4]	Prospective observational study	Participants: 85 Duration: July 2007–June 2009 Age: <5 years	Acute febrile event (MC in 31%), severe anemia (30%), acute painful events (20%), and stoke (0.6%)	Paleness and pain were most common symptoms followed by fever
Central India (Nagpur) and Jamaica Cohorts[5]	CO study	CO×1 Participants: 78 Duration: May 2008–May 12 Age: 4 years CO×2 Participants: 311 Duration: June 73–December 81	Fever, bone pain, and anemia 3 MC diagnosis on admission, BT in 33%	Paleness and pain were most common symptoms followed by fever. BT required in 15% at diagnosis
Tertiary Care Hospital, Central Gujarat India[6]	Cross-sectional	Participants: 41 Duration: 2003–2005 Age: 12–40 years	Fever 63.4%, abdominal pain 58.5%, musculoskeletal pain 53.6%, jaundice 26.8%, pallor 100%, and icterus 41%	Yellowish discoloration in 20%, icterus in 34%
Hematology Unit, Tertiary Care Hospital Yemen[8]	Prospective	Participants: 241 (participants: 150 <16 years) Duration: February 2002–11 Age: 9 months–adulthood	Percentage for<16 years: Pain 63.3%, jaundice 57.33%, splenomegaly 12.7%, and stoke 4.7%	Splenomegaly in 53%, stroke in 3.18%
Medical College, Valsad, Gujarat, India[12]	Prospective Observational trial	Participants: 47 Duration: November 2015–October 2016 Age: 6 months–14 years	Pain (MC 66%), fever 36%, pallor 47%, icterus 11%, splenomegaly 28%, and 64% undernutrition	Paleness and pain were 1st 2 symptoms, fever 3rd Splenomegaly in 53%
Children's Hospital in Benghazi Libya[13]	Retrospective and prospective	Participants: 78 Age: 6 months–26 years	Mean age: 10.84 Anemia 100%, VOC 20%, infection 25%, BT 96%, hepatomegaly 100%, splenomegaly 30%, icterus 100%, and stroke 9.5%	
Sickle cell clinic, Tertiary Care Center, Amsterdam, The Netherlands[14]	Cross-sectional	Participants: 88 Duration: Unscreened population before 2007 Age: 7 months–16 years	Painful crisis 43% and anemia 26%, jaundice 14% 78% specific, and 22% nonspecific symptoms	Paleness in 68%, pain in 55%, nonspecific symptoms present
Hospitalization Events among Children and Adolescents with Sickle Cell Disease in Basra, Iraq[15]	Descriptive study	Participants: 160 Duration: January 2012–July 2012	Acute painful crisis 73.48%, infection 9.28%, and stroke 0.63%	No icterus in their study

GMCH=Government Medical College and Hospital, MC=Most common, BT=Blood transfusion, VOC=Vaso-occlusive crisis, CO: Cohort

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Conflicts of interest
There are no conflicts of interest.

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