Original Article

Assessment of Iron Status in Patient of Sickle Cell Disease and Trait and its Relationship with the Frequency of Blood Transfusion in Paediatric Patients Attending at B.S. Medical College & Hospital, Bankura, West Bengal, India

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Abstract

Introduction: Sickle cell disease (SCD) is common in Indian subcontinent.Despite the tremendous advances in diagnostic and therapeutic modalities, Children with sickle cell anemia continue to suffer from repetitive crisis and have frequent severe complications. These morbid events as well as mortality can be greatly reduced by specialized medical care like blood transfusion and with or without chelation therapy and that focuses on prevention and active intervention.

Objective: To assess the iron status in children with sickle cell disease (SCD) and sickle cell trait (SCT).

Methods: The study was conducted on 150 consecutive patients of SCD and SCT and complete blood count (CBC) with serum iron, serum feritin were measured.

Results: Patients with SCT were more at risk of having iron deficiency (ID) than SCD. Iron deficiency was present in patients who had not received <5 units of blood transfusion (BT). Elevated level of serum iron was found in all the patients who had received more than 10 units of BT and serum ferritin level had a linear relationship with the same.

Conclusion: Patients with SCT were more in number than that of homozygous SCD (2.6:1).Patients with SCT had more chances to have iron deficiency than homozygous SCD.

Keywords: Sickle cell disease (SCD), Sickle cell trait (SCT), Serum iron, Serum ferritin

INTRODUCTION

Sickle cell disease (SCD) is a type of hemoglobinopathy and is produced by single base pair change at the 6th codon of the β-gene followed by replacement of an amino acid glutamine by valine. Subsequent polymerization of hemoglobin under hypoxia and destruction of red blood cells (RBC) is an outcome. About 50% of world populations of SCD cases are found in India.[1] Estimates indicate that SCT is predominant among the tribal population of eastern India.[2,3] Incidence of SCD is 9.3% in tribal children of Chotonagpur.[4] The predominant population of Bankura, is tribal. Iron status of children in SCD from Bankura district, West Bengal is not studied earlier with large number patients. Our aim of study is to evaluate the iron status in children of SCD/SCT and with blood transfusion.

MATERIALS AND METHODS

This was a prospective, observational and descriptive study. One hundred and fifty (150) children enrolled as SCD and trait, between the ages of 3-18 years attending outpatient department (OPD) and admitted in pediatric

ward of B.S Medical College and Hospital, Bankura, West Bengal, India from January 2011 to February 2013. Patients with double heterozygous conditions like SCD, Sickle β-thalassaemia and others are confirmed by hemoglobin electrophoresis and those on iron chelation therapy were excluded from study. Nutritional status was assessed in all cases by weight for age, height for age and weight for height and comparing with age and sex specific WHO growth charts. Patients with weight for height and height for age less than 2 Standard Deviation (SD) has been considered as 1leucocyte count, differential leucocyte count, total RBC, reticulocyte count, mean corpuscular hemoblobin, mean corpuscular volume,mean corpuscular hemoglobin concentration. Iron profiles of those patients including serum iron (μg/dl) and serum ferritin (mg/ml) were estimated. Stool sample of all children were examined for ova, parasite and presence of occult blood. Cases having hemoglobin S (HbS) >50% of total hemoglobin were defined as SCD, those with HbS < 50% as SCT.[5] Normal serum iron and ferritin level were considered to be 22-184 μg/dl and 7-140 ng/ml respectively.[6]

RESULTS

One hundred fifty (150) consecutive SCD and SCT were enrolled in the study. Out of 150 patients ninety two (61.1%) were boys and fifty eight (38.9%) girls.

In this study tribal children dominated the group (108/150). Among the study population,one hundred eight (72.2%) children were having SCT and forty (27.8%) with SCD.

Serum Iron level in SCT varied from 8.8-226 μg/dL, with mean of 67.37 μg/dL, whereas in SCD the range was from 12-221 μg/dL, with mean of 112.8 μg/Dl. Serum ferritin level in SCT varied from 4.7-450 ng/ml; mean 79.6 ng/ml and in SCD varied from 4.8-380 ng/ml; mean 140.2 ng/ml. Twenty seven (27) patients had low level of serum ferritin and serum iron, fifty seven (57) had normal level and twenty four (24) patients had high level of serum iron and ferritin. Out of twenty seven (27) patients with low level of serum iron and ferritin, twenty four (24) were tribes. Chi-square test have been applied between SCT and homozygous SCD with 1st degree freedom, the observed value was 1.809 ($p < 0.05$). Hence we concluded patients with SCT had more chances to have iron deficiency than homozygous SCD.

Malnutrition was observed in sixty seven (67) patients of SCT (85.9%) and twenty eight patients of SCD (93.33%).

Those who were transfused with more than ten units of blood had serum iron level between 80-226 μg/dL (mean 141.5 μg/dL) and ferritin level 120-450 ng/ml (mean 256.8 ng/ml). A fairly linear relationship was observed between amount of blood transfusion and serum ferritin level. Though these patients had high iron and ferritin level, serum ferritin level was always below 1000 ng/ml.

DISCUSSION

This study was conducted at B.S.Medical College, Bankura, located in the region where SCD and trait is prevalent and 72.2% of our study group was in tribal community. Burn HF et al[2] and Balgir RS et al[3] also observed that SCT is predominant among the tribal population of India (Table 1).

We observed that sixty seven patients of SCT (85.9%) and twenty eight patients of SCD (93.33%) had malnutrition and it is the major risk factor for IDA. Our study is comparable with studies by Prasad R K et al,[7] Radha Raghupathy et al,[8] L.King et al,[9] Rao et al[10] and Vichinsky et al.[11]

Chronic haemolysis, increased absorption of iron from gastrointestinal tract as well as iron provided by blood transfusion would suggest that ID is unlikely in SCD. ID anemia had been described in pediatric population with SCD both due to nutritional status and intravascular haemolysis with urinary iron losses.[12-14]

Study done by Das P K et al[15] in Orissa found malnutrition and worm infestation as the commonest cause behind ID in children of SCD and trait but, in another study Haddy et al[16] found that overt ID in SCD and trait was due to suspected blood loss (Table 2).

Our study as well as study by L.King et al[13] indicated that iron deficiency was more common among SCT than SCD, which is statistically significant (Table 3).

High iron status was observed in 40% of SCD and 15.38% of SCT in our study. Hussain et al[17] observed that 86% of SCD had ferritin level greater than 101 ng/ml. Serjeant et al[18]

Table 1: Demographic profile of patients

Race	Male	Female	<10 yrs	>10 yrs	Number	Percentage
Tribal	68	40	45	63	108	72.2%
Non tribal	28	14	16	26	42	27.8%

Table 2: Distribution of serum iron and serum ferritin level in patients with SCT and SCD

Serum Iron & Feritin level	Sickle cell trait (SCT)	Sickle cell disease (SCD)
Normal level	52 (48.14%)	25 (59.52%)
Low level	34 (31.48%)	5 (11.9%)
High level	22 (20.37%)	12 (28.57%)

Table 3: Distribution of Iron status in different ages

Units of blood	Total no of patients	Normal Iron status(%)	Low Iron status(%)	High Iron status (%)
Nil	21	15 (71.42%)	6 (28.57%)	0
1-4	36	15 (41.66%)	21 (58.33%)	0
5-10	18	18 (100%)	0	0
11-15	15	9 (60%)	0	6 (40%)
16-20	6	0	0	6 (100%)
21-25	6	0	0	6 (100%)
26-30	6	0	0	6 (100%)

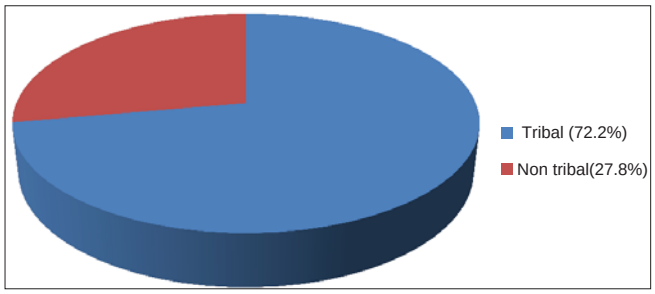

Figure 1: Demographic distribution of patients in tribal and non tribal community

had reported the higher serum iron level in SCD than control. The probable reason is the excessive intravascular haemolysis as well as increased blood transfusion in SCD.

In present study it was found that there were some correlations between blood transfusion (BT) and serum ferritin. High iron status was found only in children who needed frequent BT but, according to study,none of our patients had serum iron more than 1000 ng/ml. Das et al found the same result in his study. In another study on effect of BT on iron status in SCD and trait by Devis et al[19] found that the serum ferritin was lower than normal in patients who were not transfused. Hussain MA et al[17] observed that 6% of SCD had ferritin level greater than 1000 ng/ml.Vichinsky et al described 43 adult patients with SCD who were previously transfused for a mean of 6 years, resulting in elevated mean ferritin levels at 2916 ng/ml[20,21] but, patients under our study never required chelation therapy, as serum ferritin level was always below 1000 ng/ml. Probable reason is that all the patients in our study were of pediatric age group and a significant proportion of our patients had moderate to severe malnutrition.

CONCLUSION

Sickle cell disease as well as sickle cell trait is more common in tribal population of Bankura. Patients with SCT were more than that of homozygous SCD (2.6:1). Patients with SCT had more chances to have iron deficiency than homozygous SCD. Iron deficiency was found in those who were not transfused or transfused with <5 units of blood. All the patients who required transfusion with more than 15 units of blood had high serum iron and ferritin level.

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