A Prospective Study on Haemotological Profile of Sickle Disease

Abstract: Background: Sickle cell disease (SCD) refers to a category of inherited hemoglobin (Hb) disorders. The pathophysiology of sickle Hb (HbS) variant emerges from the polymerization of the ensuing HbS variant, which triggers a chain of erythrocyte alterations and the disorder is known to be caused by a single-nucleotide substitution at position 6 of the globin gene. Both acute and chronic sequelae cause significant morbidity in people with sickle cell anemia (SCA). Methods: This prospective cross-sectional study was conducted in the Department of Pediatrics and Sickle Cell Anemia Laboratory, Department of Pathology, Prathima institute of medical sciences, Karimnagar, India, between June 2019-Feb 2020. Most patients were from tribal background and lower socioeconomic status. Before conducting the study, permission was obtained from the Ethical Clearance Committee. Results: A total of 37 children’s homozygous (SS) variant patients were studied. The mean age of male was 9.4 (3.29) years whereas SD age of female was 9.59 (3.59) years. Haematological values of study are mentioned below. Conclusion: We conclude that moderate to severe anemia with low MCV and high HbF dominate the hematological profile.

Keywords: Hb, HCT, RBC, MCV, MCH, MCHC, HbF, WBC

INTRODUCTION:

Sickle cell disease (SCD) refers to a category of inherited hemoglobin (Hb) disorders. The pathophysiology of sickle Hb (HbS) variant emerges from the polymerization of the ensuing HbS variant, which triggers a chain of erythrocyte alterations and the disorder is known to be caused by a single-nucleotide substitution at position 6 of the globin gene. Both acute and chronic sequelae cause significant morbidity in people with sickle cell anemia (SCA). The most serious cases can be devastating within the first few years of life if they are not treated effectively.

The principal pathophysiology is focused on deoxygenation polymerization and the development of long fibers within red blood cells (RBCs), resulting in a twisted sickle shape, which contributes to intensified hemolysis and sickle red cell vaso-occlusion. The clinical appearance of SCD patients, on the other hand, is incredibly complicated, and many events can lead to vaso-occlusion. Recent research has demonstrated the significance of red cell dehydration, irregular RBC attachment to the vascular endothelium, inflammatory events, stimulation of all cells in the vessel, and nitric oxide metabolism disorders in the pathophysiology of this multi-organ disease. Hb E-thalassemia and sickle-cell anemia are two other common Hb disorders that have been confirmed to be widespread in India.

In India, SCD is common among all ethnic groups, with a particularly high prevalence among the tribal population. Odisha has the highest incidence of SCD in India, followed by Assam, Madhya Pradesh, Uttar Pradesh, Tamil Nadu, and Gujarat. In India, the average frequency of SCD is 4.3%, and in Odisha, it is 9.1%. Since neither SCA nor sickle beta-thalassemia (SBT) is uncommon in the eastern part of India; hence, a large-scale community-based study is needed to measure the exact incidence in the population. A hospital-based prospective study was conducted to evaluate the epidemiology and clinical presentation of this disease in children under 15 years of age.
**RESULTS**

A total of 37 children’s homozygous (SS) (mean age 11.6±3.4 years) for sickle cell anemia was studied for their haematological parameters. Out of the total SS subjects, 26 were males and 11 were females. Males to female’s sex ratio: 2.3: 6 years to 14 years of children were taken. Mean (SD) age of male was 9.4 (3.29) years whereas SD age of female was 9.59 (3.59) years. Haematological value of study are mentioned below

<table>
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<tr>
<th>Table 1: Haematological comparison between male and female sickle cell patients (n=37)</th>
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<tr>
<td>Hb (g/dL)</td>
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<tr>
<td>HCT (gm/dl)</td>
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<td>RBC (million/mm³)</td>
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<td>MCV (IL)</td>
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<td>MCH (pg)</td>
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<td>MCHC (g/dl)</td>
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<td>HbF (%)</td>
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<td>WBC (+103 µL)</td>
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| Hindi, hemoglobin; HCT, hematocrit; RBC, red cell count; MCV, mean cell volume; MCH, mean cell hemoglobin; MCHC, mean cell hemoglobin concentration; HbF, foetal hemoglobin; WBC, white blood cells. |

**DISCUSSION**

In the present study, maximum hospitalizations were seen during the winter season (November to February). The mechanism of cold-induced painful crises is postulated to result from cold-induced diuresis, cold agglutinins or cutaneous vasoconstriction with shunting of blood to deeper vascular bed as per a study done in Jamaica. [8] However, the studies from other countries have shown rainy season, low temperature or high wind speed and low humidity as precipitating factors for vaso-occlusive crisis in sickle cell anemia patients. [9]

Hemoglobin, MCH and MCHC were low in our study which is comparable to other studies. [10] It is said that MCV is high in sickle cell disease patients because of the increasing need of erythropoiesis due to chronic hemolysis leading to macrocytosis. It is also related to a folic acid deficiency. However, MCV was low in our study similar to some other studies from different parts of our country. [11] Low MCV in these studies may be due to co-existing iron deficiency anemia. According to National Family Health survey (NFHS-3), anemia is common in India among the schedule caste and tribes and among the children with low socioeconomic status. [12]

Current study shows that, acute painful crisis (59.01%) was the most common cause for hospitalization, followed by severe anemia (39.34%) and infections (36.06%). In a study by Akar NA, [13] Vaso-occlusive crisis was the most common cause of hospitalization in SCD children. Another study from central India has reported severe anemia requiring blood transfusion as the most common cause of hospitalization in SCD children. [14] Study by Sinde S showing respiratory infections in 37%, gastrointestinal infection in 9%, urinary tract infection in 2% and malaria in 9% cases.19 The present study shows similar results. In this study malaria was seen in 7.14% of cases. Previous studies have shown that SCT protects against severe forms and mild malaria infections, although the precise mechanism remains poorly understood. [15]

There was no mortality of sickle cell patients admitted with painful crisis in our study however sickle cell patients with crisis had mortality due to splenic sequestration or severe sepsis. This study was a hospital based study and hence, does not represent the true rate of events for SCD children in the general population. The ideal study should be a community-based cohort study or a birth cohort study. A second limitation of this study is the absence of long-term follow-up to estimate the outcomes of children and the proportion and causes of death mainly among those living with SCD. [15]

**CONCLUSION**

In hospitalized children with sickle cell anemia, acute painful crisis was the most the common morbidity events followed by severe anemia and acute febrile events. Morbid events most commonly occurred during the winter season. It was also observed that vaso-occlusive crisis is the commonest manifestation in pediatric age group and that despite being hemolytic in nature;
hematological parameters were suggestive of hypochromic microcytic anemia which may be due to associated iron deficiency in these patients.

REFERENCES