



Malaria With Sickle Cell Disease : The Changing Scenario in Endemic Area

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ABSTRACT

Introduction : It was believed that current prevalence of MALARIA in many tropical populations reflects selection for the carrier form and sickle cell disease(SCD) through a survival advantage. 150 years after this hypothesis, the epidemiological description of relationships between SCD and malaria has changed specially in malaria endemic areas. In this study our aim was to study the occurrence of malaria, its clinical features and complications in sickle cell disease.

Methods : This was a retrospective analysis of the data collected from children age 1 to 18 years admitted to Dhiraj Hospital during January 2010 through December 2010. We studied the clinical features, complications, lab investigations of malarial infections in these children, stratified by SCD status.

Results : we had 44 patients with SCD who had *p.vivax*(26 i.e. 58%), and *p.falciparum* (18 i.e.42%) malaria proven by smear as well as RDT. In these patients 48%(22) had complicated malaria. 75%(34) had thrombocytopenia, 80% (36) had severe anemia (Hb<7gm/dl), all patients had bodyache and limb pain as chief complaints with high grade fever, 10%(i.e. 4) had acute chest pain and 20%(i.e. 8) had severe pain in abdomen, 2-2(5%) had acute renal failure, acute respiratory distress and convulsions as complications. Mean HbF concentration was 18.6 % .

Conclusion : Although hemoglobin S is considered to be protective against Plasmodium, this is not always the case and in these children SCD complicated the diagnosis of malaria because hemolysis was related to the infection rather than to SCD.

KEYWORDS

Malaria, Sickle cell disease

Introduction :

Sickle cell disease is caused by a variant of the β -globin gene called sickle hemoglobin (Hb S). Inherited autosomal recessively, either two copies of Hb S or one copy of Hb S plus another β -globin variant (such as Hb C) are required for disease expression. Hb S carriers are protected from malaria infection. Despite this advantage, individuals with sickle cell disease exhibit significant morbidity and mortality due to malaria.¹

The gene for sickle hemoglobin (HbS) is a prime example of natural selection. It is generally believed that its current prevalence in many tropical populations reflects selection for the carrier form (sickle cell disease through a survival advantage against death from malaria.² Nevertheless, 150 years after this hypothesis was first proposed, the epidemiological description of the relationships between SCD and malaria has changed completely.²

In this study our aim was to study the occurrence of malaria, its clinical features and complications in sickle cell disease.

Material Methods :

We conducted a retrospective analysis of the data collected prospectively from children age 1 to 18 years admitted to Dhiraj Hospital during January 2010 through December 2010. We studied the clinical features, complications, lab investigations of malarial infections in these children, stratified by SCD status. Collected data was classified and statistically analysed to get the results.

Results :

Total 44 patients were selected who had SCD with either *p.vivax*(26 i.e. 58%), or *p.falciparum* (18 i.e.42%) malaria proven by smear as well as RDT. In these 44 patients 48%(22) had complicated malaria clinically and/or by laboratory parameters. Complicated malaria patients included 75%(34) with thrombocytopenia, 80% (36) with severe anemia (Hb<7gm/dl). All studied children were in some form of crisis like : all patients had bodyache and limb pain as chief complaints with high grade fever, 10%(i.e. 4) had acute chest pain and 20%(i.e. 8) had severe pain in abdomen, 2-2(5%) had acute

renal failure, acute respiratory distress and convulsions as complications. Mean HbF concentration was 18.6 % .

Discussion :

The allele that causes sickle cell anemia also imparts partial resistance to malaria. In individuals with two "normal" alleles, the malaria parasite can infect the red blood cells. The bursting of these infected cells can cause kidney and liver failure, anemia, hypoglycemia, or block blood vessels to vital organs, such as the brain (causing cerebral malaria); children under the age of 5 have a high risk of death if this occurs. But the red blood cells of individuals with one sickle cell allele are relatively resistant to malaria; furthermore, these individuals do not get sickle cell anemia but the people with homozygous SCD are now having complicated malaria and sometimes death due to it in endemic areas.³

Though malarial parasite counts are said to be lowest in sickle-cell homozygotes, intermediate in heterozygotes, and highest in normal homozygotes³ suggesting, perhaps, that malaria in sickle-cell anemia patients is not severe.³ But in malaria endemic countries malaria in sickle cell anemia patients can be very severe, and is a serious common precipitating cause of Crisis which sometimes can lead to death. Causative factors of crisis due to malaria in SCD can be : pyrexia causing in vivo sickling, hyperhidrosis, Anorexia, Vomiting, diarrhoea in young children. These factors can lead to serious dehydration with massive intravascular sickling, severe erythrocyte sequestration, and instant death.^{4,5,6}

In a study conducted in Tanzania, it was concluded that, Although malaria was rare among patients with SCA, parasitemia during hospitalization was associated with both severe anemia and death. Effective treatment for malaria during severe illness episodes and further studies to determine the role chemoprophylaxis are required.⁷ Malaria is widely considered a major cause of illness and death in patients living with SCA in sub-Saharan Africa.^{8,9}

Conclusion :

Although hemoglobin S is considered to be protective against

Plasmodium , this is not always the case and in these children SCD complicated the diagnosis of malaria because hemolysis was related to the infection rather than to SCD. Malaria and its complications should be considered as a differential diagnosis in a patient with SCD in malaria endemic area.

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