Sickle Cell Anemia with Malaria: A Rare Case Report

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Abstract Sickle cell disease is the prototype of hereditary hemoglobinopathies, characterized by the production of structurally abnormal hemoglobin. Sickle cell anemia results from a point mutation that leads to substitution of valine for glutamic acid at the sixth position of the β globin chain. We report a young male admitted with fever and weakness for 3 days. Hematological test reveals *Plasmodium falciparum* malaria parasite and sickle cell anemia. Patient was treated and get cured from malaria and discharged.

Keywords Sickle cell disease · Malaria

Introduction

Sickle cell disease is autosomal recessive disorder first described by Harrick in 1910. About 7 % of world population is carrier of some form of haemoglobin disorder. Sickle cell disease is fairly common in our country. In India it was first detected by Lehman and Cutbush [1] in 1952 among the tribals of Nilgiris hills. In India sickle cell gene is mainly restricted to tribal and schedule caste population where carrier frequency ranges between 5 and 40 % [2]. Prevalence of sickle cell disease is found to be 0–18 % in north eastern India, 0–33.5 % in western India, 22.5–44.4 % in central India and 1–40 % in southern India [3]. The protection against falciparum malaria by HbS is

well known. Sickle cell trait confers a high degree of resistance to severe and complicated malaria [4–7]. To some extent it almost certainly related to the peculiar physical or biochemical properties of HbS red blood cells: invasion, growth, and development of *Plasmodium falciparum* parasite are all reduced in such cells under physiological condition in vitro [8, 9] and parasite infected HbS red cells also tend to sickle [8, 10, 11] and process that may result their premature destruction by the spleen [8, 12]. Fifty years ago observation had made that geographical distribution of sickle cell gene is highly correlated with malaria endemicity. During the course of human evolution in regions where malaria is life threatening, naturally occurring genetic defence mechanisms have evolved to resist to malaria infection. The gene for sickle cell among the first as an example of natural selection by the protection afforded to the carrier through a survival advantage against death from severe malaria. This is the reason where malaria is endemic, as many as 30 % black Africans are heterozygous.

Case Report

A 25 year old young male patient was admitted for fever and weakness for 3 days. On examination, general condition—fair, pulse 110/min, blood pressure 110/78, respiratory rate 20/min, febrile having temperature 103.2 F, systemic examination was normal, no hepato or splenomegaly. There was no past history of tuberculosis, syphilis, diabetes or hypertension. Patient was known case of sickle cell disease and he was taking herbal and ayurvedic treatment for it, although he was not having any written document of this treatment. Blood investigation—haemoglobin 9.0 g %, total leucocyte count 8,600/cumm, polymorph 67 %, lymphocyte 30 %, eosinophil 02 %, monocyte 01 %,

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basophile 00 %, erythrocyte count 3.0 million/cumm, platelet count 1.0 lac/cumm, PCV 28, ESR 10 mm after 1 h by Wintrobe method, reticulocyte 3 %. Peripheral smear shows anisocytosis, poikilocytosis, hypochromasia, target cells, normoblast (06 %), occasional Howell Jolly bodies, few sickle cell and malaria parasite (*P. falciparum* with 2 % parasitic index). Malarial antigen test and quantitative Buffy coat were positive. Sickling test was positive by reducing substance sodium metabisulfite. Haemoglobin electrophoresis shows HbS 78.5 %, HbF 15.5 %, HbA 3.5 %, and HbA² 2.5 %. Biochemistry investigation show fasting blood glucose 72 mg %, creatinine 0.99 mg %, total bilirubin 2.10 mg % (conjugated 0.29 mg %, unconjugated 1.81 mg %), SGPT 110 IU, SGOT 65 IU, Alkaline phosphatase 80 IU/dl, Australia antigen negative, HIV 1 and HIV 2 was nonreactive, G6PD was normal. Patient was treated with oral artesunate 200 mg, pyremethamine 75 mg, and sulfadoxine 1,500 mg on day 1, then oral artesunate 200 mg on day 2 and 3. On day 1 patient became afebrile, on day 3 his peripheral smear was negative for malaria parasite. Patient was cured from malaria and discharged.

Discussion

Sickle cell disease is hereditary hemolytic anemia. The overall prevalence in general population ranges from 4.30 to 5.35 %. At present about 109 countries are endemic to malaria [13]. Among the WHO regions, the South East Asia region has a malaria burden second only to Africa. All countries of South East Asian region of WHO are affected except Maldives [14]. India contributes about 70 % of malaria and 28 % of total deaths in South East Asian region of WHO [15]. Malaria is major public health threat in India [16]. There were 1.67 million cases of malaria in India in 2006, out of which 0.75 million cases of *P. falciparum*. There were 1,487 total deaths from malaria in 2006 [17].

Red cells from people with sickle cell disease do not sickle to any significant degree at normal venous oxygen tension. Very low oxygen tension will cause cell to sickle [18]. Red cells in person of sickle cell disease infected with *P. falciparum* deforms, because parasite causes low oxygen tension in red cells. These abnormal cells become vulnerable for phagocytosis [13].

Experiments carried out in vitro with sickle cell, the red cell infected with falciparum sickle more readily than uninfected cell [11]. Selective sickling of infected cells reduces parasite burden. In other experiment *P. falciparum* parasite cultured in red cells of sickle patient, died when the cells were incubated at low oxygen tension [8]. Ultra-structural study show extensive vacuole formation in *P. falciparum* parasite inhabiting sickle cell trait and causes damage of parasite [19]. Other study suggests

oxygen radicle formation in sickle cell disease, retardation of growth and damage of parasite [20]. Homozygous HbS red cells produce hemin [21]. Infected cell might generate enough hemin which damages parasite [22].

People with sickle cell trait are less likely to get malaria. The trait does not completely protect a person from infection, but makes death from malaria less likely.

Conclusion

Concomitant presentation of sickle cell disease with malaria is very rare; but proves that sickle cell disease is not always immune to malaria.

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