



Original Research Article

Comparative analysis of some haematological parameters in sickle cell patients in steady and crisis state at Michael Okpara University of Agriculture, Umudike, Abia state, Nigeria

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ABSTRACT

Keywords

Haematological parameters, Sickles cell patients, Steady state and crisis.

Sickle cell anaemia is excruciating and life threatening disease. It affects the whole system of the patients. It is important to study the changes that occur in them to find out how to take proper management of those affected. The study was carried out in the University Health services department of Michael Okpara University of Agriculture, Umudike, Abia State, Nigeria. The study was carried out on 30 confirmed sickle cell patients, 7 were in crisis and 23 were in steady state. The age ranges from 4-34 years. The study started from October 2012-January 2014. The study showed significant increase ($P < 0.05$) in mean values of TWBC, Neutrophil, significant decrease ($P < 0.05$) in PCV, Lymphocytes and no significant change ($P > 0.05$) in mean values of Monocytes when these values were compared to the sickle cell anaemia patients in crisis relative to those in steady state.

Introduction

Sickle cell disease is an inherited multisystem disorder. Its cardinal features, chronic haemolytic anaemia and recurrent painful episodes, relates to the presence of mutant sickle cell haemoglobin S within the red blood cell. The illness affects most organ systems and sickle cell has reduced solubility, sickling and occlusion by sickle

red cells to the microvasculature (Hoffbrand *et al*, 2001).

In addition to its abnormal electrophoretic mobility and solubility, HbS is unstable. Consequence of HbS instability are the increased generation of methaemoglobin and release of haem, processes that contribute to the increased generation of

oxidative radicals by sickle red cells. In addition, the normal physiological process of haemoglobin oxygenation and deoxygenation generate methaemoglobins and oxidative radicals continuously. These processes perpetuate an oxidative stress within sickle red cells that was major pathophysiologic importance to metabolism, membrane lipids, membrane proteins, and the integrity of the HbS molecule itself (Bender *et al*, 1973; Adachi *et al*, 1980; Asakura *et al*, 1973; Macdonald and Charache, 1982).

Sickle cell disease is a genetic abnormality involving the haemoglobin. Patients present with a wide spectrum of disorders because of single-point mutation in which thymine substitute for adenine, thereby encoding valine instead of glutamine in the sixth position of the beta-globin chain. Haemoglobin S resulting from the substitution causes polymerisation of Haemoglobin and red cell sickling on exposure to low oxygen tension and unsickle on oxygenation. The repeated sickling and unsickling damages the red cell membrane leading to irreversibly sickled red cell even when the oxygen pressure is increased thus reducing red cell life span as a result of membrane damage inducing anaemia. The white blood cells and platelets are also affected by the mutation (Akinbami *et al*, 2012).

Quantitative and qualitative changes in red blood cells have been reported. Haemolysis consequent to the damaged red cell membrane could be intravascular or extravascular. The former results from the lysis of complement-sensitive red cells (Test *et al*, 1991) and the haemoglobin lost during sickling induced membrane damage (Allan *et al*, 1982; Platt, 1982). The induced membrane damage occurs by phagocytosis of red cells that have

undergone sickling (Galili *et al*, 1986; Grenn and Kalra, 1988) and physical entrapment of rheologically compromised red cells (Kaul *et al*, 1986). Degree of haemolysis is inversely related to haemoglobin concentration and packed cell volume in sickle cell anaemia patient (Serjeant *et al*, 1969).

Sickle cell anaemia is a major cause of morbidity and mortality in Africa where there is no readily effective treatment (Omoti, 2005). The disease amounts for over 60% of the world's major haemoglobinopathies with an estimated 2-3 million Nigerians affected by S gene (Olatunji, 2002). The extent of the problems of sickle cell disease in Nigeria cannot therefore be overemphasized because of the S gene said to be between 25-30% (Omoti, 2005). The majority of the patients born to rural dwellers do not usually survive childhood (Ukpong, 1992). Furthermore, there is no widely acceptable and readily available cure for patients with sickle cell anaemia at present. Curable methods such as gene therapy and bone marrow transplantation, which may be associated with several complications, are not readily available in developing nations (Omoti, 2005).

Many patients with sickle cell anaemia are in reasonably good health most of time and achieving steady state level of fitness. The importance of early recognition and subsequent clinical and haematological assessment of the disease are greatly facilitated by familiarity with the patient's steady state. A patient with sickle cell anaemia is said to be in steady state when there is absence of infection, acute complicating factors or acute clinical symptoms or crisis for at least three months (Bookchin and Law, 1996).

Crisis refers to episodes of acute illness attributable to the sickling phenomenon in which there is a sudden deviation for the worse or a sudden exacerbation of symptoms and signs of patients with sickle cell anaemia who had hitherto been in stable condition (Gustave *et al*, 2013). The occurrence of the crisis makes the disease incapacitating to the patient and frustrating to parents and physicians (Omoti, 2005). Although sickle cell disease is primarily a disease of the red blood cells, leucocytes, because of their sizes obstruct blood vessels more effectively than red blood cells when attached to the endothelium.

Many complications of sickle cell disease are associated with leucocytosis. It is a risk factor for early sickle cell disease-related death (Platt *et al*, 1994). It is implicated in clinically overt stroke (Ohene-frempong *et al*, 1998; Powers, 2000). Pathogenesis of silent infarction (Kinney *et al*, 1999) and acute chest syndrome (Castro *et al*, 1994) have been associated with leucocytosis.

It will be necessary to find out some changes in haematological parameters among these sickle cell patients for proper management by the parents, the clinicians and all health workers to ensure improvement in their health status to prolong their life span. Many works have been documented on steady state but not much has been done in crisis state and the comparison.

Materials and Methods

Study Area

The study was done in University Health Services Department of Michael Okpara University of Agriculture, Umudike, Abia State, Nigeria.

Subjects

30 Confirmed sickle cell patients, 23 subjects were in steady state and 7 subjects in crisis state were used for the study.

Samples

Venous blood samples were collected from the subjects into EDTA anticoagulated containers for the haematological parameters analysis.

Ethics

Oral consents were made to the patients before the samples were collected.

Statistical Analysis

The data were analysed using t-test with the level of significance set at $P < 0.05$.

Results and Discussion

The study showed significant increase ($P < 0.05$) in the mean values of WBC and Neutrophil and significant decrease ($P < 0.05$) in the mean values of PCV, Lymphocytes but no significant change ($P > 0.05$) in the mean values of Monocytes when these parameters were compared among the sickle cell patients in crisis relative to those in steady state. This is in agreement with report of Omoti (2005) except the monocyte which had significant change in his study but no significant change in this study. These changes occur because of the crisis which results in sudden exacerbation of symptoms and signs of patients with sickle cell anaemia who had been in stable condition (Gustave *et al*, 2003). The leucocytosis observed in this study is associated with many complications in sickle cell disease. It is a risk factor for early death sickle cell disease-related death (Platt *et al*, 1994).

Table.1 Some haematological parameters in sickle cell anaemia patients in steady state and crisis

| Parameters | HbSS steady state (n=23) | HbSS Crisis State (n=7) | P-Value |
|--------------------------|--------------------------|-------------------------|---------|
| WBC(*10 ⁹ /L) | 11.5+/-2.3 | 20.5+/-3.5 | P<0.05 |
| PCV(%) | 20.0+/-1.5 | 15.0+/-1.3 | P<0.05 |
| Neutrophil(%) | 70.0+/-7.3 | 76.0+/-4.2 | P<0.05 |
| Lymphocyte(%) | 29.0+/-4.0 | 23.0+/-3.7 | P<0.05 |
| Monocyte(%) | 1.0+/-0.3 | 1.0+/-0.5 | P>0.05 |

It is implicated in in clinically overt stroke (Ohene-frempong *et al*, 1998; Poweres, 2000), pathogenesis of silent infarction; Kinney *et al*, 1999) and acute chest syndrome (Castro *et al*, 1994) have been associated with leucocytosis. Leucocytosis is a common feature of sickle cell disease and was thought to be due to redistribution of granulocytes from marginal pool to the circulating pool (Ahmed *et al*, 2006).

This is expected because of an increase concentration of neutrophils in venous blood of sickle cell anaemia patients which include demargination of intravascular neutrophils, accelerated release from the bone marrow and reduction in the rate at which neutrophils leave the blood. Those involved with the management of these patient should take proper care of them especially during crisis to avoid los of lives. There haematological parameters should be monitored regularly to maintain sound health for them.

There are significant changes in the haematological parametres studied. Those changes are important clinically. There should be adequate health education given to the patients and the parents to avoid exposing them to factors that can trigger crisis such as dehydration, cold, infection, etc.

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