Behavior of Sickle Cell Disease (SCD) and it's Management in Adult Patients

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ABSTRACT

Introduction: Sickle cell disease SCD is an inherited chronic hemolytic anemia. Clinical manifestation arise from the tendency of hemoglobin (Hb s or sickle hemoglobin) to polymerize and deform red blood cells into the characteristic sickle shape. Aim of study: To study the behavior and complications of sickle cell anemia in adult life and to assess the benefit of Hydroxyurea in selected patients. Patients and Methods: All patients who were referred from pediatric hematology department to medical hematology department in Tripoli Medical Center as a case of sickle cell anemia proved by hemoglobulin electrophoresis in the period between Jan 2012-Dec 2017 were included in our study. Results: 34 patients were included in our study. The Median age was 30.5 years. 64.7% (22) were males and 35.3% (12) were females. 91.2% were black. 47% of patients had strong family history of SCD. Mean hemoglobin level 7 g/dl. Jaundice occurred in 88.2%. The causes of admission were infection in 50% and bone pain crisis in 73%. One patient had bone tuberculosis. The mean frequency of admissions was 3 times/year. All our patients were on folic acid tab. 5 mg daily and 20(60%) patients received Hydroxyurea capsules 1g/day. 70% (14/20) of patients who received Hydroxyurea had hemoglobin concentration between 9-11 g/dl and the admission frequency became less than one/year. During follow up of all patients, one patient died because of acute chest syndrome & one patient died because of complication of cerebrovascular accident CVA. Conclusion: Painful crisis, hemolytic crisis and infections were the most common causes of admission in our patients. The frequency of admissions to the hospital were less in adult life compared to childhood indicating the quiescent behavior of SCD. The use of Hydroxyurea increases hemoglobin F concentration with less attacks of painful and hemolytic crisis.

Keywords: Sickle cell disease, Quality of life, Hydroxyurea

1. INTRODUCTION

Sickle cell disease (SCD) is the most common gene disordered worldwide, caused by mutation in both Beta Globin genes, presenting by chronic anemia since childhood due to chronic hemolysis, which leads to frequent hospital admission and blood transfusion, complicated by multi organ disease that leads to premature death. The Hemoglobinopathies are a group of inherited autosomal recessive hemoglobin disorders, resulting in the homozygous state, in chronic severe anemia.
Thalassemia and sickle cell diseases (SCD) constitute the most monogenic hemoglobin disorders worldwide, initially described in the tropical and subtropical regions. They are now common all around the world because of migration. A global epidemiological database for hemoglobin disorders has been established and published recently by the World Health Organization (WHO) [1]. Data collected in 229 countries, clearly indicated that hemoglobinopathies constitute a significant health problem in 71% of those countries which include 89% of all births worldwide.

The prevalence of carriers of abnormal hemoglobin is within the range of 5-7% in the world and the number of new cases of affected infants is estimated at 300,000 per year. More than 70% of total hemoglobin disorders are localized in Africa. African population represents less than 10% of the world population but has the highest crude birth rate (39.0‰) and also the highest rate of affected conceptions (10.78‰) compared to the one registered in the world (2.73‰).

North Africa consists of 5 countries, Mauritania, Morocco, Algeria, Tunisia and Libya with a total population of 87 million of inhabitants and a birth rate ranging between 16.8‰ and 40.9%. The geographical position of this area at the parting of the ways between sub-Saharan African countries and the Mediterranean makes North Africa well concerned by hemoglobinopathies [2].

In Libya, first screening study reported the presence of HbS, HbC and thalassemia genes in Libyans in 1975, but it was found that the incidence of abnormal haemoglobins in the indigenous population of Libya was low. More recent studies confirmed that SCD occurs at a low frequency among Libyan. The disease is associated with several complications and seems to be severe [3].

SCD can vary according to the type of hemoglobin with Hb SS and Hb SC being the most common variants. Although SCD in the Hb SC form presents a more benign clinical evolution than Hb SS. Both may present with similar complications [4].

People in a vulnerable socioeconomic situation are more exposed to the determining social factors of the disease, which can lead to an aggravation of the patient’s general health [5]. The severity of the disease is inversely proportional to the quality of life (QOL) [6].

According to the World Health Organization (WHO), the QOL is defined as “the individuals’ perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns [1].

Presented in this definition are the multiple dimensions incorporated in the individual’s perception of the different aspects of life [7]. The Sickle cell disease has been largely studied in terms of population frequency and clinical variations. In this research that addresses aspects related to hemoglobin analysis by Hemoglobin Alkaline Electrophoresis to assess the quality of life as SCD is an important Public Health problem in Libya and assess the tolerance and effect of Hydroxyurea, which is the only drug approved by FDA for SCD.

2. PATIENTS AND METHODS

Patients

All patients who were referred from pediatric hematology department to medical hematology department in Tripoli Medical Center as a case of sickle cell disease proved by hemoglobin electrophoresis in the period between Jan 2012 - Dec 2017 were included in our study.

The hemoglobin Analysis

The blood was collected in tubes with EDTA anticoagulant, the test was done by Hemoglobin Alkaline Electrophoresis. The blood sample were collected from 34 adult patients, 22 were male and 12 were female. They were diagnosed as sickle cell disease, when the result of HbS more than 80%.

The Quality of life Measurements

The quality of life for those patients was assessed by number of hospital admission and blood transfusions.

Hydroxyurea Administration

Hydroxyurea capsule is the only drug approved by FDA for SCD. All of our patients were on folic acid 5mg daily. 20 patients received hydroxyurea as 500mg capsules P.O. 2x1 at early morning for 6 months, then we checked the response by repeating Hb electrophoresis.

Response was considered when Hb SS was less than 80% of total hemoglobin, with increase on the level of Hb F more than 8%.

This study was approved by medical ethic committee in Tripoli Medical Center.

3. RESULTS

The hematological indices were investigated in a group of adult male and female, diagnosed as SCD.
with Median age was 30.5 years old, 91.2% were black (Negroid). 47% of patients had strong family history of SCD. Mean hemoglobin level range between 7g/dl. Leukocytosis occurred in 76.5%. 38.2% had hepatomegaly, Gall bladder stone occurred in 44%. Jaundice occurred in 88.2% of patients with mean Serum bilirubin level range between 2.5mg/dl. Splenectomy was done in one patients with sickle thalassemia. The causes of admissions in these patients was due to infection in 50% of them, one patient had bone tuberculosis. 73% of admissions were due to bone pain crisis. The mean frequency of admission was three times per year.

By Echocardiographic examination, 15% of patients had Mitral regurgitation and Tricuspid regurgitation. Two patients developed nephrotic syndrome due to glomerulosclerosis which diagnosed by histopathological analysis of kidney biopsy. 3 patients had cerebrovascular accident (CVA) and one patient developed recurrent venous ulcers in right leg. During regular follow up on Hematology clinic, the patient who received Hydroxyurea orally had good tolerance and some good compliance with increase in mean Hb level between 9-11 g/dl. It decreases the frequency of blood transfusion and hospital admission. 10%of them had adverse effect as leucopenia, thrombocytopenia which is common side effect of hydroxyurea. During follow up, one patient died because of acute chest syndrome and another patient died because of complication of CVA.

4. DISCUSSION

The sickle cell disease is a worldwide Hemoglobinopathies. In Africa, SCD represents 70% of total Hemoglobin disorders. The geographical position of North Africa, at the parting of the ways between sub-Saharan African countries and the Mediterranean makes North Africa well concerned by hemoglobinopathies [8]. The incidence of the sickle cell gene in Libya is low, but that of the thalassemia gene appears to be high in the Libyan population which is racially mixed, containing Arab, Mediterranean and Negroid types. Electrophoretic study of haemolysates from five hundred and forty-five 545 subjects from the University of GARYOUNIS, Benghazi, was carried out to

find out the incidence of abnormal hemoglobin in Libya. Abnormal hemoglobin were encountered in 23 subjects of unrelated families, giving an overall incidence of 4.2 per cent. Sixteen of these subjects had Hb-AS trait (3.0%), five subjects had Hb-AC trait (0.9%) and two subjects had Hb-AD trait (0.36%). This study indicates that the incidence of abnormal hemoglobins in the indigenous population of Libya is low [8,9].

In other study an electrophoretic study of hemolysates from 1350 subjects from the indigenous population of Libya was carried out to find the incidence of abnormal hemoglobin’s and thalassemia. Sickle cell disease was detected in five patients (0.37%), sickle cell trait in 61 (4.51%), sickle cell thalassemia in three patients (0.21%). Homozygous beta_ thalassemia in 16 (1.20%), heterozygous delta-beta _thalassemia in 25 (1.85%) and heterozygous beta-thalassemia in 105 (7.77%). The incidence of the sickle cell gene is low, but that of the thalassemia gene appears to be high in the Libyan population because of racial mixture [10]. In this retrospective study among 34 adult patients from Tripoli Medical center, due to lack of previous studies regarding hemoglobin analysis in general population for general survey of SCDs, and a quality of life measurement’s, tolerance of Hydroxyurea and complication of SCDs in Libya , it was impossible to make a comparison. This study found that SCD had high incidence in people with Black races (Negroid) population and they had strong family history of SCDs due to consanguinity.

There was an association between SCDs and QOL, due to high frequency of hospital admissions as manifestation of sickle cell disease. The complication of sickle cell disease as (CVA), Heart disease and ESRDs lead to long term morbidity and high mortality.

The patients who were on hydroxyurea had a good performance in their life and symptoms and had decreased frequency of hospital admission. Their hemoglobin level was between (8 -11g/dl), with some adverse effect as leucopenia and thrombocytopenia in 10%.

The mortality and morbidity were low in those patients. Our findings confirmed the need to conduct a more detailed analysis of quality of life of patients with sickle cell disease and the role of Hydroxyurea in adult SCDs, and also reinforced the need to promote education regarding the manifestation and the
complication of SCD and organize multidisciplinary teams to manage them.

5. CONCLUSION

The hemolytic, painful crisis and infection were the most common causes of morbidity and early death. Use of hydroxyurea as definitive treatment in SCD improve hemoglobin level and QOL.

REFERENCES