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Epidemiological, Clinical and Biological Characteristics of Cameroonian Children and Adolescents with Sickle-Cell Anemia

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Abstract

A total of 126 patients were included in the study- 65 girls and 61 boys. The mean age of the subjects was 8 ± 4 years (ranging from three to sixteen years). The most represented age group was that from 3-4 years of age. Most of the participants were from the Central Region (46%). All of the subjects were homozygous for hemoglobin SS. Subjects with yearly frequency of three vaso-occlusive crises represented 42.9% of the sample. 57.9% of the patients had been poly-transfused within the past 3 years (>2 transfusions). Up to 45.2% of the patients had been hospitalized within the past year.

Keywords: Sickle-cell anemia; Hemoglobin; Hereditary; Hemoglobinopathy

Introduction

Sickle-cell anemia (SCA) is a hereditary hemoglobinopathy characterized by the presence of hemoglobin S inside red blood cells [1]. It is very widespread throughout the world with a peculiar geographical distribution [2]. According to the WHO, about 5% of the world's population bears the genes that are characteristic of hemoglobinopathies [3], 2.9% carry mutations for sickle-cell anemia; about 120 million people [4].

SCA manifestations can affect all tissues with slow blood circulation. They involve cranio-facial bones and the mouth where the most described manifestations include gingival hypertrophy, periodontal disease, dental caries, pulp necrosis and tonsillar hypertrophy, and a light-pink coloration of the mucosa [5-7]. These consequences have physical, emotional and social impacts which often compromise the quality of life of these patients, leading to the involvement of several medical specialties in the management of this pathology [2]. Meanwhile, because of genetical and hematological differences, socio-economic specificities and different levels of access to

healthcare, the clinical forms of these manifestations vary according to place and time. In Cameroon, bucco-dental manifestations of SCA are not well-known owing to the paucity of studies carried out on these subjects. This motivated us to carry out this study with the main aim of describing and analyzing the socio-demographic, clinical and biological characteristics of Cameroonian children and adolescents with sickle-cell anemia.

Materials and Methods

We carried out a cross-sectional, descriptive and analytical study. It involved children and adolescents with SCA consulting at the Mother and Child Center of the Chantal Biya Foundation from the 14th November 2014 to April 2015 and those sensitized during monthly meetings of the Cameroonian Sickle-cell Association within the same period. Sampling was consecutive and non-randomized. Included were patients aged 3 to 16 years with no sex distinction whose parents gave consent for participation. Excluded were patients with incomplete medical files and those with other associated pathologies.

Patients were examined and socio-demographic, clinical and biological data collected on data forms. The patients were examined in the sickle-cell anemia ward of the Chantal Biya Mother and Child Center. Eligible patients were required to have done beforehand work-up including a complete blood count (CBC) and reticulocyte count during a stable period, conjugated, unconjugated and total bilirubin and serum LDH.

1. Interview

During the interview, the following data were collected:

Patient identification: name, first name, sex, region of origin

2. Patient files provided information on:

Baseline hemoglobin values dating less than 3 months prior.

Reticulocyte count

Hemolysis work-up

Anthropometric parameters

Type of SCA on the hemoglobin electrophoresis results.

Yearly frequency of hospitalization and number of blood transfusions during the past 3 years.

For each patient, data collected was recorded using Epi Info version 3.5.3, 2011 software. Statistical analysis was then done using the R software.

Results

The study was carried out on 126 homozygous SS patients. The mean age was 8 ± 4 years (range from 3 to 16 years). The most represented age group was the 3-4 years group. The most represented region was the Center Region which made up 46.0% of the study population (N=58) followed by the Western Region with 16.7% (N=21). The least represented region was the South West Region comprising 2.4% of the study population (N=3). All of the patients were homozygous for hemoglobin S. The mean percentage of hemoglobin S was $78.8 \pm 12.2\%$; $15.5 \pm 10.4\%$ for hemoglobin F and $12.8 \pm 8.6\%$ for hemoglobin A2. The patients had regenerative anemia with a low mean baseline hemoglobin (7.4 ± 1.02 g/dl) and elevated mean reticulocyte count (268.1 ± 128.4 G/L). LDH and indirect bilirubin levels were also elevated. The mean weight of the patients was 24 ± 9 kg with a minimum of 12 kg and a maximum of 49 kg. The mean height was 125.6 ± 17 cm with a minimum of 80 cm and a maximum of 160 cm. On average, 42.9% (N=54) of the patients had three vaso-occlusive crises per annum and 38.9% (N=49) had at least four crises per annum. Twenty-eight (28) patients (22.2%) had been transfused at least once during the past three years, most of the patients had been hospitalized at least once during the year (45.2%).

Discussion

The mean age of the sample was 8 ± 4 years (range from 3 to 16 years). The predominant age group was that from 3 to 4 years. A similar mean age was found in the Ivory Coast by Kple-Farget et al. [7] who in a similar group, reported a mean age of 8 ± 3 years. Similarly, Benoist et al. in Senegal in 2006 found a mean age of 9 ± 3 years which is close to our finding [8]. Female sex was predominant with a sex ratio of 0.9 in favor of girls. This was similar to that found by Ramatoulaye in Senegal in 2006 in her doctoral thesis which showed a sex ratio of 0.89 in favor of girls [9]. On the contrary, Kondani et al. found equal sex distribution in their own study [10]. Inversely, Nembe in Cameroon in a doctoral thesis working on a similar sample found male predominance with a sex ratio of 1.2 [11]. The divergence of these results shows that SCA is a hereditary disease of autosomal transmission, which means the anomaly is not sex-related. The most represented region was the Center Region comprising 46% of the participants followed by the Western Region with 16.8%. The South West Region was the least represented (2.4%). This distribution was similar to that found by Mendouga in Cameroon in a group of children with SCA (Center Region: 42.9%; Western Region 17.1%; South West Region 2.9%) [12]. This shows inhomogeneity of SCA throughout the national territory. The very high frequency of participants

from the Center Region could be justified by the fact that the study was carried out in this region.

The sample was completely made up of homozygous SS SCA patients. This concurs with the studies carried out by Ngone [13] and Mendouga in Cameroon [12]. However, it was different from that of Kple-Farget et al. [8] in the Ivory Coast who had 67% of homozygous SS patients, 24.2% of SC heterozygotes and 8.8% of patients with $S\beta$ -Thalassemia. These results show that the major sickle-cell syndromes are made up essentially of homozygous SS form as Bardakdjian et al. have suggested [4]. This study population had a mean hemoglobin S percentage of $78.8 \pm 12.2\%$; $15.5 \pm 10.4\%$ for hemoglobin F and $12.8 \pm 8.6\%$ for hemoglobin A2. These results are close to that described in literature which shows that in homozygous SS individuals, electrophoresis reveals three migratory bands with HbS: 75 to 95%; HbA2: 2 to 4% and HbF: 1 to 15% [14]. The mean baseline hemoglobin was 7.4 ± 1.02 g/dl. This result was like that of Nembe [11] who found mean baseline hemoglobin of 7.44 ± 1.55 g/dl. The low mean baseline hemoglobin in our patients is due to anemia which is pathognomonic for their disease. The mean platelet count was 372.3 ± 126.3 G/L and did not correspond with that of Mendouga [12] who found a mean platelet count of 499.5 G/L. The mean leucocyte count was 16.4 ± 9.8 G/L. Mendouga found a mean of 12.4 G/L. Despite the disparity in these results, it remains that the white cell count was high in our patients. The Mean Corpuscular Volume was averagely 87.7 ± 11.4 fl, and the mean reticulocyte count was 268.1 ± 128.4 G/L. These results were close to that of Ramatoulaye who found a mean of 80.7 ± 16.2 fl for the MCV and mean reticulocyte count of 233.4 ± 120 G/L. High reticulocyte counts signify regenerative anemia in these patients. Mean LDH was 1437 ± 551 IU/L while mean unconjugated bilirubin levels were 61.7 ± 38.4 μ mol/L. Elevated levels of unconjugated bilirubin and LDH are due to intra-tissular hemolysis principally in the spleen and which is a consequence of destruction of falciform red cells [15].

The mean weight of our patients was 24 ± 9 kg and their mean height was 125 ± 17 cm. Our results were similar to that of Ramatoulaye who found a mean weight of 24.7 ± 9.2 kg and a mean height of 131.5 ± 18.9 cm [9]. The weight and height in our study were reported without calculation of the Body Mass Index (BMI).

Yearly frequency of 3 vaso-occlusive crises was found in 42.9% of the patients and no patient was free from vaso-occlusive crises. Our results were different from that Rakoto Alson et al. in Madagascar who found that 23.5% of their subjects had 3 crises a year and 35.3% had no crises [16-19]. This can be justified by the difference in climatic conditions from one place to another as we know that exposure to cold, high altitude and humidity increase the risk of vaso-occlusive crises [15]. In our study, 57% of patients were poly-transfused while only 22.2% had one transfusion each. These findings were similar to that of Kple-Farget et al. in the Ivory Coast who found that 65% of their subjects were poly-transfused [7]. This can be justified by the fact that blood transfusion is considered a therapeutic measure in these patients who suffer from chronic anemia. The frequency of yearly hospitalization in our subjects was 45.2%. Nembe et al.

also found a frequency of hospitalization of 74.1% [11,20-23]. This can be explained by the early management and follow up of the patients by their parents. It could have positive repercussions on the life expectancy of these patients [24].

Conclusion

Sickle-cell anemia is a serious public health problem whose prevalence even in urban zones is yet unknown in our context. We observe that the clinical and biological characteristics of this disease are influenced by the number of hospital admissions and blood transfusions. Diagnosis and early management permit improvement in the quality of life of these patients.

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