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Original Research Article

Evolutionary profile of patients with hemoglobin SC disease regularly followed in Côte d'Ivoire

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ABSTRACT

Background: West Africa is recognized as the elective focus of hemoglobin C. The S and C combination in the same patient gives a major sickle cell syndrome. In our country, very few series dealing with the evolutionary features of this SC form have been published contrary to the homozygous SS form. The aim of this study was to describe the evolutionary profile of double heterozygous SC sickle cell patients.
Methods: This was a retrospective and prospective study with descriptive and analytical purpose of 174 SC sickle cell patients.
Results: The median age was 26 years with extremes of 6 years and 57 years. 96% of patients had less than 4 vaso-occlusive seizures per year. The evolutionary complications were mainly ischemic (56.30%) and infectious (39.10%). Among ischemic complications, sickle cell retinopathies and aseptic osteonecrosis are the most common with 59.20% and 31.63% respectively. Infectious complications were dominated by ENT (36.76%) and osteoarticular (35.29%) infections. Only age had an influence on the occurrence of ischemic complications (p = 0.0001). The probability of survival at 5 years was 99.38% and that at 20 years was 91.57%. The overall survival was not influenced by evolutionary complications.
Conclusions: Infectious and ischemic evolutionary complications show the importance of vaccination and an early screening program.

Keywords: Complication, Hemoglobin S disease, Hemoglobin C disease, Hemoglobin SC disease, Mortality, Prevention

INTRODUCTION

Asterion is the confluence of the temporal, occipital and sickle cell disease is the most common hemoglobinopathy in the world. It is characterized by an abnormal structure of hemoglobin due to the replacement of glutamic acid by a valine at the level of the 6th amino acid of the beta chain (β6Glu-Val).[1] In Côte d'Ivoire, hemoglobin S affects 12% of the population, including 2% in its severe forms.[2] Hemoglobin C is a structural variant of normal hemoglobin (hemoglobin A) resulting from a genetic mutation caused by the substitution of an amino acid (lysine replaced by glutamic acid) in position 6 of the beta chain of globin (β6Glu-Lys).[1]

West Africa is recognized as the elective focus of hemoglobin C.[3] The S and C combination in the same patient gives a major sickle cell syndrome. In our

country, very few series dealing with the evolutionary features of this SC form have been published contrary to the major SS form.[4,5]

The reasons are related to clinical signs relatively less expressive than in the SS form. Our work was therefore aimed at determining the evolutionary profile in patients with this double heterozygous SC form regularly followed.

METHODS

This was a retrospective and prospective study with descriptive and analytical purpose that was carried out in the clinical hematology department of Yopougon university hospital from September 2002 to September 2017 that is 15years and which focused on patients carrying hemoglobin SC who are followed. The diagnosis of the hemoglobin type was made by zone capillary electrophoresis with the Capillarys® system (Sebia, France) were included in this study sickle cell SC patients over 5years or with at least 5years of follow-up with a complete medical record. Socio-demographic data included age, gender, and socio-economic level.

Clinical data investigated the annual frequency of vaso-occlusive seizures, infectious complications, and anemic seizures. The paraclinical data were searched for from the hemogram, abdominal ultrasound (looking for vesicular lithiasis, cholecystitis, splenic infarction and appearance of kidneys), cardiac ultrasound (looking for cardiomyopathy, fundus and retinal angiography (looking for an ophthalmologic complication), bone radiography (looking for osteitis, osteomyelitis or osteonecrosis).

Kidneys were investigated from microalbuminuria, urea and serum creatinine. Infectious complications were investigated from an infectious assessment according to the clinical context (hemogram, thick drop, C-reactive protein, blood culture, bacteriological examination of the cerebrospinal fluid, chest X-ray, cytobacteriological examination of the urine, stool culture, swabbing or others as needed). These data made it possible to select 174 patients.

Data were entered using the Epi-info 6.04 FR software and processed using the SPSS 12.0 software. In the context of the analytical study we did some matching to determine the likely risk factors influencing the outcome and survival.

Thus, authors used Mantel-Haenzel's Chi-Square test (x^2), Student's test and Fisher's test. The significance threshold of p was 0.05. The adjusted relative risk (RR) and its confidence interval (CI) at 95% were used to assess the degree of significance of the observed differences. The calculation of survival was done according to the Kaplan-Meir method. The comparison of the survival curves was done by the Log-Rank test.

RESULTS

About 174 patients with hemoglobin SC disease were included in this study. Young adults (aged 20 to 49) were the majority in this study that is 68.9%. Only 5 patients were under 10years old. 10 patients had an age ≥50years. Patients with fewer than 4 vaso-occlusive seizures were 96%. Ischemic and infectious complications were the most found. The death rate was only 2.90% over the study period (Table 1).

Two of the patients died during a road accident, 1 died of decompensation of post-alcoholic cirrhosis of the liver, 1 died of diabetic acido-ketosis and the last one died of a tumor of the cerebellum.

Table 1: General features.

Parameters numbers (%)	
Age group (years)	
< 10	05 (2.90)
10-19	42 (24.14)
20-29	55 (31.61)
30-39	30 (17.20)
40-49	32 (18.40)
≥50	10 (5.75)
Number of vaso-occlusive seizures/year	
00-03	167 (96)
04-06	07 (04)
Type of complications	
Anemic	08 (4.60)
Ischemic	98 (56.30)
Infectious	68 (39.10)
Mortality	
Living	169 (97.10)
Dead	05 (2.90)

Acute anemic complications, although rare (5 patients), were especially acute because of malaria. As for chronic anemia complications, they were found in only 3 patients. The frequency of ischemic complications was 62.07%. These complications were dominated by sickle cell retinopathy (proliferative retinopathies) and osteonecrosis. 12.96% of patients had neurosensory disorders. Infectious complications were dominated by ENT involvement in 36.76% of cases, osteoarticular disorders in 35,29% of cases and pulmonary involvement in 20,58% of cases. Urinary tract infections were rare (1 case only).

Ischemic complications occurred after the age of 10years. The age of the patients influenced the occurrence of ischemic complications (Table 3). The probability of survival at 5years was 99.38%. This probability of survival was 98.64% at 10years. At twenty, it was 91.57%. The survival was thus comparable to that of the general population.

Table 2: Evolutionary complications.

Complications	Numbers (%)
Anemic	
Acute	05 (62.50%)
Chronic (pigmentary lithiasis)	3 (37.50%)
Ischemic	
Osteonecrosis	31 (28.70%)
Retinopathy	58 (53.70%)
Priapism	5 (4.64%)
Neurosensory disorder	l4 (12.96%)
Infectious	
Tonsillitis	4 (5.88%)
Angina	6 (8.82%)
Rhino pharyngitis	15 (22.06%)
Arthritis	6 (8.82%)
Osteomyelitis	18 (26.47%)
Urinary infection	1 (1.47%)
Pulmonary tuberculosis	1 (1.47%)
Bronchitis	5 (7.35%)
Acute lobar pneumonia	2 (2.94%)
Broncho pneumopathy	8 (8.82%)
Viral hepatitis B	2 (2.94%)

Table 3: Distribution of ischemic complications according to the age.

Age group (years) patients with ischemic complications	Ischemic complications
< 10	00 (00%)
10-19	15 (15.30%)
20-29	27 (27.55%)
30-39	22 (22.45%)
40-49	24 (24.49%)
≥50	10 (10.20%)

Khi^2 =25.57, p= 0.0001

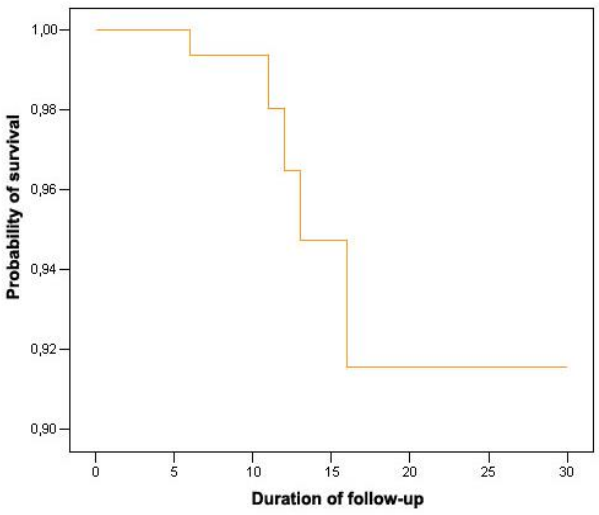

Figure 1: Overall survival curve.

DISCUSSION

Côte d'Ivoire is a West African country populated by 22,671,331 inhabitants with a population growth rate of 2.6%.[6] The health system does not have a primary prevention program for hemoglobinopathies. Consequently, neonatal screening is non-existent. The average age of our patients at diagnosis was 27.4years with a standard deviation of 6. Young subjects in the 20 to 29 age group predominated (Table 1). Late diagnosis is probably due to the low prevalence of severe anemia in these patients and the late onset of symptoms. This fact has also been reported in other studies.[7,8] A neonatal screening program would certainly reduce the age at diagnosis of hemoglobinopathies in general.[9,10] The high frequency of ischemic complications in our study is reported by most authors with retinopathies, hearing disorders and osteonecrosis.[8,11,12] Sickle cell retinopathy (Table 2) is thought to be the prerogative of the double heterozygous SC form.[8,13] This was most commonly proliferative retinopathy. Also, the authors agree that the prevalence of osteonecrosis is higher in the SC form than in the SS form.[8,11,13] Neuro-sensory complications (hearing loss and deafness) of hearing deserve special attention because they are serious and constitute for Gualandro SFM et al, a mortality factor.[14] Pulmonary, bone and ENT Infectious complications are the most commonly contracted by sickle cell SC patients (Table 2). Sangaré A and Latoundji S justified the high frequency of pulmonary and bone infections by the fact that the lungs and bones of sickle cell patients are the site of multiple foci of infarction favorable to the multiplication of germs.[15,16] We noted in our study that age had a statistically significant influence on the occurrence of ischemic complications (P=0.0001). These complications occurred in patients over 20years (Table 3). The other parameters did not influence the occurrence of complications. About the overall survival of sickle cell SC patients, a study carried out by Latoundji S on the morbidity and mortality of sickle cell SC patients in Benin showed no death related to sickle cell disease. The survival was thus comparable to that of the general population (Figure 1).[16] In our study, anemic, ischemic and infectious complications did not statistically have significant influence on survival.

CONCLUSION

It emerges from this study that the evolution of the SC form of sickle cell disease is primarily towards ischemic and infectious complications if it is not diagnosed early and managed correctly. It is therefore important to focus on immunization and the implementation of an early screening program in countries with limited resources such as Côte d'Ivoire.

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