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Major Sickle Cell Syndromes in Maradi, Niger: Epidemiological, Clinical, Biological and Therapeutic Aspects

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Abstract

Introduction: Sickle cell disease is the most widespread genetic disorder in the world. This study aimed to investigate the epidemiological, clinical, biological, and therapeutic aspects of sickle cell anemia in Maradi, Niger. **Methodology:** This was a retrospective descriptive study carried out at the Centre Hospitalier Régional (CHR) of Maradi over 12 months, from September 2021 to August 2022. **Results:** This study included 246 patients with sickle cell anemia. The average age was 7.9 years, ranging from 0 to 16 years, and a sex ratio of 1.2. We observed kinship in 54.9% of the parents of sickle cell patients. Among the fathers, 62.6% were primarily blue-collar workers, while 87.4% of the mothers were housewives. Clinically, we observed pallor in 78.5% of the patients, jaundice in 43.5%, splenomegaly in 12.6%, and hepatomegaly in 11.8%. The most common vaso-occlusive crises involved osteoarticular pain (34.6%), followed by hand-foot syndrome (26.4%) and abdominal pain (20.3%). The SS form predominated biologically, representing 93.5% of cases, while double heterozygotes SC represented 6.5%. Most patients (56.5%) presented with severe anemia, with moderate anemia observed in 38.6% of cases. All patients received folic acid. Level I and II analgesics were used in 82.6% and 46.7% of patients, respectively, and patients received non-steroidal anti-inflammatory drugs in 72.3% of cases. A small proportion of patients (7.8%) were on background treatment with Hydrea. **Conclusion:** The results of our study are similar to those often described in sickle cell anemia.

Keywords

Major Sickle Cell Syndromes, Epidemiological, Clinical, Biological, Therapeutic Aspects Maradi/Niger

1. Introduction

Sickle cell disease is an autosomal recessive genetic disorder characterized by the substitution of valine (Val) for glutamic acid (Glu) at position six on the β-chain of globin (chromosome 11) [1]. It is the most common genetic disease in the world, particularly affecting black populations. Almost 5% of the world's population currently carries a gene responsible for a hemoglobin abnormality [2]. It was recognized as a public health priority by UNESCO in 2005, the African Union in 2005, the World Health Organization (WHO) in 2006, and the United Nations in 2008 [3]. According to the WHO, every year, more than 330,000 children are born with hemoglobinopathy, 83% of them with sickle cell disease [4]. Niger is one of the countries most affected by this condition, with almost one Nigerien in five, or around 20% of the population, carrying the sickle cell trait [5]. In Senegal, prevalence is estimated at 10%, including 1% homozygous forms [6]; 12% in Mali [7]; 12%, including 2% primary forms, in Côte d'Ivoire [8]; and around 25% of the population is thought to carry the gene in Gabon. [9]. This study aimed to investigate the epidemiological, clinical, biological, and therapeutic aspects of major sickle cell syndromes in Maradi, Niger.

2. Methodology

This study was a retrospective descriptive study conducted at the Centre Hospitalier Régional (CHR) of Maradi in Niger from September 2021 to August 2022, which is 12 months. The study included all sickle cell patients aged 0 to 16 years, of all sexes and ages, confirmed by hemoglobin electrophoresis (done on cellulose acetone strips) seen in pediatric consultations at the CHR de Maradi. The variables studied included sociodemographic, clinical, biological, and therapeutic parameters. Data was collected using a pre-established survey form, and consultation registers, patient files, and patient medical follow-up notebooks were used for data collection. Microsoft Office 2013 and SPSS version 20 were used for data entry and analysis.

3. Results

3.1. Socio-Demographic Aspects

During the study period, we consulted 9733 patients. The study included 246 patients with sickle cell disease, a frequency of 2.52%. The sociodemographic characteristics are provided in **Table 1**.

Table 1. Distribution of patients according to socio-demographic parameters.

Parameters	Number	Percentage
Age (years)		
Average age	7.9 ± 2.6 years (0 to16 years)	
Age range		
0 - 5 years	139	56.5
6 - 10 years	55	22.4
11 - 16 years	52	21.1
Sexe:		
Sex ratio	1.2 (M = 136; F = 110)	
Kinship		
Yes	135	55
No	111	45
Parents' professions		
Fathers		
Workers	154	62.6
Shopkeepers	72	29.3
Civil servants	20	8.1
Mothers		
Housewives	215	87.4
Civil servants	22	8.9
Student	9	3.7

3.2. Clinical Features

Pallor was observed in 78.5% of patients, jaundice in 43.5%, splenomegaly in 12.6%, and hepatomegaly in 11.78%. Vaso-occlusive crises were characterized by osteoarticular pain in 34.6% of cases. Detailed clinical parameters and complications are provided in Table 2.

Table 2. Distribution of patients by clinical parameters and complications.

Paramètres	Number	Percentage
Age at diagnosis (years)		
0 - 5	190	77.2
6 - 10	48	19.5
11 - 16	8	3.3
Clinical signs		
Pallor	193	78.5
Jaundice	107	43.5

Continued

Splenomegaly	31	12.6
Hepatomegaly	29	11.8
Osteoarticular pain	85	34.6
Abdominal pain	50	20.3
Hand-Foot Syndrome	65	26.4
Complications		
Infection	125	50.8
Osteomyelitis	3	1.2
Acute thoracic syndrome	9	14.5
Cardiac	6	9.6
Renal	1	1.6
Hepatobiliary	30	48.4
Osteoarticular	16	25.8

3.3. Biological Aspects

SS phenotype predominated (93.5%) with 6.5% SC phenotype. The majority of patients (56.5%) had severe anemia. Table 3 shows the distribution of patients according to biological parameters.

Table 3. Distribution of patients according to biological parameters.

Parameters	Number	Percentage
Electrophoretic profile		
SS	230	93.5
SC	16	6.5
Hemoglobin level (g/dl)		
<6	139	56.5
6 - 9	95	38.6
>9	12	4.9
Rhesus Blood group		
A^+	57	23.2
A^-	2	8
B^+	57	23.2
B^-	3	1.2
AB^+	10	4.1
O^+	111	45.1
O^-	6	2.4

3.4. Therapeutic Aspects

Follow-up was regular in 61.8%, and all patients regularly took folic acid. Level I and II analgesics were used in 82.6% and 46.7% of patients respectively, and non-steroidal anti-inflammatory drugs in 72.3%. Very few patients (7.8%) were on background treatment with Hydrea. Table 4 shows the distribution of patients according to therapeutic parameters.

Table 4. Distribution of patients according to therapeutic parameters.

Parameter	Number	Percentage
Follow-up		
Regular	153	61.8
Irregular	73	29.7
Lost to follow-up	21	8.5
Therapeutic education	146	100
Vaccination status		
Up to date		73
Not up to date		27
Number of hospitalizations		
1 - 7	194	85.9
8 - 14	32	14.1
Number of transfusions		
0	36	14.94
1 - 5	189	78.42
6 - 10	18	6.64

4. Discussion

We conducted a study of 246 sickle-cell patients at the Centre Hospitalier Régional of Maradi in Niger. Epidemiologically, the mean age was 7.9 ± 2.6 years (0 - 16 years), with a predominance of the 0 - 5 age group (56.5%), and the sex ratio was 1.2. Similar results were reported in Niger (at the National Hospital of Niamey) in 2014 by Malam-Abdou B. and coll; the mean age was nine years (1 to 65 years), with a predominance of the 1-5 age group and a sex ratio of 1.2 [10]. Lamine T and colleagues in Senegal at Hospital of Peace in Ziguinchor in 2017 also reported similar results; the mean age was eight years (11 months - 21 years), with a predominance of the 0 - 5 age group and a sex ratio of 1.3 [11]. This was also the case in studies conducted by Alain F and coll. at Mahajanga Teaching Hospital of Madagascar in 2022 and Boiro D and coll., at Abass Ndao Hospital in Dakar, Senegal in 2016 who found a mean age of 6 and 8.26 years and a sex ratio of 1.3 and 1.4 [4] [12]. Clinically, pallor, jaundice, and splenomegaly were found in 78.5%, 43.5%, and 12.6% of patients. Lamine T and coll. reported pallor (95.6%), jaundice

(36.9%) and splenomegaly (21.7%) [11]; Camara E and coll., at Donka National Hospital in Conakry in 2017 found 77% pallor, 64% jaundice and 19% splenomegaly [13] and Cissouma A and coll. also reported pallor (62.5%), jaundice (29.1%) and splenomegaly (13.8%) [7]. These results are close to those of this study. Vaso-occlusive crises were dominated by osteoarticular pain (34.6%), followed by hand-foot syndrome (26.4%) and abdominal pain (20.3%) in this series; Alain F and coll reported this predominance of osteoarticular pain with 52.8%, followed by abdominal pain (23.7%) and hand-foot syndrome (8.2%) [4]. Mekone Nkwele I and coll. in Cameroon (at the Chantal Biya Foundation center for Mothers and children in Yaoundé) in 2022 also reported similar results, with osteoarticular pain at 85.9%, followed by abdominal pain (31.7%) [14]. Biologically, the SS phenotype predominated, with 93.5% and 6.5% of double heterozygotes SC in this series. Igala M and coll. In Gabon at the Teaching Hospital of Libreville in 2019 found the same predominance with 95.5% SS and 2.3% double heterozygous SC [15]. Most patients in this study (56.5%) presented with severe anemia, followed by moderate anemia (38.6%). Alain F and coll. and Apollinaire KS and coll., at the Graben University Clinique in the DRC in 2020 reported this predominance of severe anemia, representing 47.2% and 42.9% of patients in their series [4] [16]. In contrast, Doupa D and coll. in Senegal (at the national center for blood transfusion in Dakar) found a predominance of moderate anemia (48%), followed by mild (34%) and severe (18%) anemia [6]; Igala M and coll. also reported a predominance of moderate anemia (73.9%), followed by severe (13.6%) and mild (12.5%) anemia [15]. These differences may be explained by the fact that the studies carried out by the latter authors were based on sickle cell patients in the stationary phase. Regarding treatment, all patients in this study were on folic acid. Level I and II analgesics were used in 82.6% and 46.7% of patients, respectively, and non-steroidal anti-inflammatory drugs in 72.35%. Roger D and coll. (at Hubert Koutoukou Maga National University Hospital center in Benin) reported the use of level I (73.5%), level II (59.8%), and level III (23%) analgesics and non-steroidal anti-inflammatory drugs (40.2%) [17]. In our study, only 7.8% of patients were on Hydrea. Several authors had reported rates significantly higher than ours, this was the case in the Apollinaire KS and Coll, Mashako M. and Coll at the North-Kivu Province Hospital in the DRC in 2019 and Harrak A and Coll. in Morocco (at Ibn Rochd Casa teaching hospital) studies, with 51.4%, 11.6%, and 10% of cases, respectively [16] [18] [19]; this could be explained by demanding access to Hydrea due to its high cost.

5. Conclusion

The results of this study are similar to those often described in sickle cell anemia. The majority of patients presented with severe anemia, and vaso-occlusive crises were the main acute complication. Adherence to background treatment with Hydrea remains very limited due to its inaccessibility and high cost.

Declaration of Interest

The authors declare that they have no conflicts of interest in relation to this article.

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