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Major sickle cell syndromes in children in Kenitra, Morocco

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ABSTRACT

Objective: To highlight the epidemiological characteristics and plot the current mapping of the sickle cell syndromes in children under 15 years old.
Methods: A descriptive study was conducted on children with sickle cell disease over a period of 4 years (from January 2011 to December 2015) at the Pediatric Department at El Idrissi Regional Hospital Center in Kenitra, Morocco.
Results: The mean age of patients was (8.56 ± 3.97) years and the age group 6–15 years was the most affected. The male gender was the most dominant with 60.94% of cases versus 30.06% for females. The homozygous form SS was the most frequently identified (81.25% of cases) while the heterozygous form SC was rarely detected (2.08%).
Conclusions: Sickle cell anemia remains a reality in Morocco and may not be perfectly understood yet by health professionals. A screening policy and a sustainable management program can prevent hemoglobinopathies in the studied region. An action plan must be implemented at national level to improve the quality of management of main sickle cell syndromes.

1. Introduction

Sickle-cell anemia is a blood related disorder that affects the hemoglobin molecule, and causes the entire blood cell to change shape under stressed conditions. It is a codominant autosomal disease due to the presence of abnormal hemoglobin Hb S. Sickle cell disease (SCD) has a worldwide distribution and Piel *et al*.[1] estimated that more than 300 000 babies were born with sickle cell anemia in 2010 with approximately 80% of these births occurring in low- or middle-income countries. Authors also reported large regional disparities in the occurrence of SCD sickle cell trait among newborns by race/ethnicity and place of birth[2].

The most common forms of SCD, include the homozygous form SS, the composite heterozygous form SC and the S beta thalassemia form with a significant prevalence of major forms SS[3]. In Morocco, information regarding the occurrence and epidemiology of hemoglobinopathies is very scarce and according to World Health Organization the rate of carriers in Morocco is about 6.5%, suggesting the existence of 30 000 cases of major forms[4].

The north-west of Morocco was reported to be a zone of predilection of hemoglobinopathies and the region of Gharb Chrarda Bni Hssen (GCBH), especially the province of Kenitra, seems to be one of the most affected areas and may be considered as a focus for the major form SCD[5].

The present study is designed to report the current mapping

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of sickle cell syndromes and to determine their epidemiological characteristics which may assist to elaborate appropriate approaches for the disease treatment and management.

2. Materiels and methods

The present retrospective study involved a cohort of 192 children aged from 1–15 years hospitalized during a five-year period (2011–2015) at the Pediatric Department at the El Idrissi Regional Hospital Center in Kenitra, Morocco. A report containing all the details regarding each patient with sickle syndromes was established and completed. The epidemiological parameters regarding the children with hemoglobin SS, SC, or S beta thalassemia were studied and the investigated factors included age, gender, ethnicity, hemoglobin electrophoresis and hemograms data. The recorded data analysis was done using the Epi-Info 2011 software (version 7.0.3.8).

3. Results

We have identified 192 patients with major sickle cell syndromes (MSCS) admitted in the Pediatric Department of the El Idrissi Regional Hospital Center. The data regarding the geographic distribution and SCD phenotypes specified in the medical folders of the studied children are summarized in Table 1.

Table 1
Distribution of MSCS patients by province and locality of origin and SCD phenotype [*n* (%)].

Province	Locality	Phenotypes			
		SS	S beta thalassemia	SC	Total
Kenitra	Benmansour	42 (21.88%)	5 (2.60%)	1 (0.52%)	48 (25.00%)
	Mnasra	21 (10.94%)	6 (3.13%)	1 (0.52%)	28 (14.58%)
	Sidi Mohamed Ben Mansour	12 (6.25%)	4 (2.08%)	1 (0.52%)	17 (8.85%)
	Kenitra	11 (5.73%)	3 (1.56%)	0 (0.00%)	14 (7.29%)
	Mograne	10 (5.21%)	0 (0.00%)	0 (0.00%)	10 (5.21%)
	Sidi Med Lahmar	10 (5.21%)	2 (1.04%)	0 (0.00%)	12 (6.25%)
	Moulay Bousse Iham	9 (4.69%)	3 (1.56%)	0 (0.00%)	12 (6.25%)
	Bhara Oulad Ayad	8 (4.17%)	1 (0.52%)	0 (0.00%)	9 (4.69%)
	Sidi Alal Tazi	3 (1.56%)	0 (0.00%)	0 (0.00%)	3 (1.56%)
	Souk EI Arbaa du Gharb	3 (1.56%)	1 (0.52%)	0 (0.00%)	4 (2.08%)
	Total	129 (67.19%)	25 (13.02%)	3 (1.56%)	157 (81.77%)
Sidi Slimane	Sidi Yahia Du Gharb	12 (6.25%)	3 (1.56%)	0 (0.00%)	15 (7.81%)
	Sidi Slimane	9 (4.69%)	2 (1.04%)	0 (0.00%)	11 (5.73%)
	Total	21 (10.94%)	5 (2.60%)	0 (0.00%)	26 (13.54%)
Sidi Kacem	Sidi Kacem	6 (3.13%)	2 (1.04%)	1 (0.52%)	9 (4.69%)
Total (Region)		156 (81.25%)	32 (16.67%)	4 (2.08%)	192 (100.00%)

In the province of Kenitra, three localities (Benmansour, Sidi Mohamed Ben Mansour and Mnasra) accounted for 59.2% (93 cases) of the MSCS patients identified in this province (157 cases) corresponding to 48.44% of the total cases recorded in the studied area (192 cases). The proportion of MSCS cases varied according to provinces. The highest proportion was detected in Kenitra (Gharbaoui ethnic group) reaching 81.77% of patients, while lower proportions were recorded in the provinces of Sidi Slimane (Hasnaoui ethnic group) and Sidi Kacem (Charadi ethnic group) with respective frequencies of 13.54% (26 cases) and 4.69% (9 cases).

With regard to the prevalence MSCS in 0–15 years aged children, it was significantly higher in the province of Kenitra with a level of 49.8/100 000 versus 26.8/100 000 for the province of Sidi Slimane and 5.6/100 000 for the province of Sidi Kacem (Table 2).

Concerning the SCD phenotypes, most of patients had homozygous sickle cell disease SS form (81.25%), and S beta thalassemia was identified 16.67% of cases while the lowest recorded was SC form with a level of 2.08%.

Table 2
Prevalence of MSCS in children population (0–15 years old) according to provinces and ethnic groups.

Provinces and ethnic groups	No. of cases	Population (0–15 years)	Prevalene (per 100 000)
Kenitra (Gharbaoui ethnic group)	157	315 246	49.8
Sidi Slimane (Hasnaoui ethmic group)	26	96 671	26.8
Sidi Kacem (Charadi ethmic group)	9	158 770	5.6

For the patients' age, it ranged from 1 to 15 years with a mean age of (8.56 ± 3.97) years and the age group 6–15 years was the most affected (69.27%). With respect to sex factor, a preponderance of males was recorded with a frequency of 60.94% against 39.06% for females with a male to female sex ratio of 1.56 (Table 3).

Table 3
Distribution of MSCS cases by age group and sex factor.

Parameter		MSCS cases	Percentage
Age group	1–5 years	59	30.73%
	6–15 years	133	69.27%
Sex	Male	117	60.94%
	Female	75	39.06%

The severity of the disease is assessed through the number of vaso-occlusive crises which was recorded in 20.1% of the study population, while similar rates of 13% were found clinical anemia and conjunctival pallor. Splenomegaly represented 14% and bacterial infections, including pulmonary, bone and digestive infections, were identified in 46.2% of cases.

The mean hemoglobin level was (6.53 ± 0.90) g/dL with an average of 6.45 g/dL for the hemoglobin form SS, 6.7 g/dL for hemoglobin SC and 6.46 g/dL for hemoglobin S beta thalassemia.

4. Discussion

MSCS are considered the most frequent hemoglobinopathies in Morocco. The diagnosis of sickle cell forms in the study population was carried out in the range age of 1–15 years. The age of 0–5 years corresponding to the phase of onset of clinical signs represented 30.73% of patients while the age group of 6–15 years was predominant reaching 69.27% of cases. These findings showed that the disease diagnosis was made within a late stage. This may have severe repercussions on the clinical state of children including delayed growth and bone damage.

The mean age of the investigated children population was 8.56 years and most of cases were 6–15 years old. This may be due to the

fact that patients with major homozygous sickle-cell anemia may survive beyond the age of ten years. These results are in accordance with those reported by previous studies of hospital records carried out in Morocco[6-9]. In the conducted study a male predominance was recorded with a rate of 60.94% and a sex ratio of 1.56. Comparative findings were reported by Hafiani *et al.*[6], Bouzaid[8] and Mahmouh[9] who quoted respective male to female sex ratios of 1.2, 1.2 and 1.6; while a female predominance was revealed by Dokekias[10] and Ouakasse[11] with respective sex ratios of 0.8 and 0.7. Elsewhere, and contrary to these reports, Dreux[12] reported an equal distribution of MSCS, according to the gender factor. In fact, as sickle cell anemia has an autosomal transmission, the recorded differences in the distribution of SCD cases by gender may be due to recruitment or management bias[12].

With respect to the distribution of cases by ethnic groups, a significant predominance of the ethnic group Gharbaoui was recorded with a percentage of 81.77% versus 13.54% for the ethnic group Hasnaoui and the lowest proportion (4.69% of cases) belonged to the ethnic group Charadi. The predominance of the Gharbaoui ethnic group in the investigated series may be a result of cultural and environmental factors[13].

The main clinical signs found in the studied series (conjunctival pallor, splenomegaly, clinical anemia…) are similar to those classically described in the literature. The splenic sequestration crises are very specific to children under 6 years of age. This syndrome affects the hemoglobin level which is reduced by at least 2 g compared to people without sickle cell anemia[14]. The found mean of Hb level was 6.45 g/dL for the SS form, 6.7 g/dL for the SC form and 6.46 g/dL for S beta thalassemia hemoglobin. Comparative hemoglobin levels of 6.83 g/dL in SS form and 9.54 g/dL in SC patients were reported by Tiendrebeogo[15].

The current study results revealed the occurrence of sickle cell syndromes as a pathology in Morocco. Due to the non-exhaustive character of the conducted study, the actual distribution and prevalences may be underestimated. Signs and symptoms of SCD usually begin in early childhood. Early diagnosis is essential in providing proper preventive treatment for some of the devastating complications of the disease. Screen newborn babies for sickle cell anemia is recommended to people belonging to populations at risk and an action plan must be implemented at national level to improve the quality of management of sickle cell syndromes on the basis of experimented programs and strategies deployed in developed countries[16].

Conflict of interest statement

We declare that we have no conflict of interest.

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