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Epidemiology of Sickle cell disease among children in Gadarif Hospital, eastern Sudan

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Abstract

Background and Objective: Sickle cell disease is a common and severe genetic disease affecting children. The aim of this study was to investigate the epidemiological factors of sickle cell disease among children in Gadarif hospital, eastern Sudan.

Methods: A descriptive cross sectional, facility-based study conducted from 1st January to 31st December 2016. A total of 83 children (<18 year old) attended the hospital with confirmed diagnosis of sickle cell disease was selected through non probability- convenient sampling method.

Results: Among the total 83 investigated children, 43 (51.8%) were male and 40 (48.2%) were female give male: female ratio of 1.1. The age ranged between 9 months to 14 year with mean (SD) 6.4 (3.2). The mean (SD) age at diagnosis was 3 (1.1) years old. A family history with SCD was reported by 53 (63.8%), and 9 (10.8%) reported the death of a family member from SCD. The mean (SD) of times of hospitalization was 1.1 (0.9) and within the preceding 12 months, 36 (43.4%) had been hospitalized once and 7 (8.4%) had been hospitalized 2 or more times. The most prevalent prior complication of SCD was a vaso-occlusive episode which had been diagnosed in 61 participants (73.5%). Using logistic regression analyses there was significant association between rural residence (CI= 1.5 – 22, OR= 17.1, P= 0.015), older age ≥ 6 year (CI= 5.1 – 11.6, OR= 5.5, P= 0.001) and SCD complications.

Conclusion: Extensive effort is needed to improve the quality of life among sickle cell disease children.

Keywords: Sickle cell, epidemiology, children, anemia, Gadarif state, Sudan.

1. Introduction

Sickle cell disease is a common and severe genetic disease affecting the children (Hamideh and Alvarez 2013). It is an autosomal recessive condition, caused by a mutation in the β globin gene, which changes the sixth amino acid from glutamic acid to valine (De Montalembert et al. 2011). Clinical features of sickle cell disease include anemia, infections, painful crises and central nervous system complications such as stroke. It is a major health problem in developing countries (Brousse et al. 2017). Hemoglobin SS, in Africa, accounts for 75% of 300,000 annual affected births (Piel et al. 2013). In Sudan sickle cell disease is a major health problem and it is more common in western region (Ali et al. 2014). The first report of the presence of the HbS gene among Sudanese people was reported early in 1950 (Abbott et al. 1950). The magnitude and the problem of the sickle cell disease becomes worse because of little knowledge among Sudanese people, consanguineous marriage, and few data in the literature (Mohammed et al. 2006). Therefore, this study aims at filling a gap in knowledge of the epidemiology of sickle cell disease, in eastern Sudan, a high burden setting of infectious diseases (Adam et al. 2016). Information presented in this study is expected to offer potentially useful insights to those aiming to provide comprehensive care for the children and the health care planners to implement strategic plan to improve the quality of care for sickle cell disease patients in particular and for children as general.

2. Materials and Methods

2.1 Study design and data collection

This was a descriptive cross sectional, facility-based study conducted from 1st January 2016 to 31st December 2016 to investigate the epidemiological factors of sickle cell disease among children in Gadarif hospital, eastern Sudan. A total of 83 children (<18 year old) attended Gadarif hospital with confirmed diagnosis of sickle cell disease (HbSS genotype) was selected through non probability- convenient sampling method for this study. A structured questionnaire was used to gather data from parents or guardian. The information sought by the questionnaire include: the socio-demographic characteristics (age, gender, residence, ethnicity, mean age at diagnosis family history of SCD, history of death due to SCD among the family members), duration of the illness, reason of clinical presentation (fever, fatigue, weakness, loss of appetite and weight loss), complications, and prior therapies received by participants in the clinic or during prior hospitalization, clinical sign (pallor, jaundice, enlarged spleen and liver), patient outcome (improvement, death). Proper systemic examination was performed to each patient by a paediatrician including cardiovascular system, respiratory system, abdomen, musculoskeletal system and central nervous system. Basic tests were performed for every patient on admission and repeated when clinically indicated. These included complete blood count, urine analysis, and blood film for malaria, stool analysis and abdominal ultrasound. All patients were under multidisciplinary care and were closely followed during their hospital stay and then every month in the referred clinic. Complications of SCA used in this study were those proposed by the Comprehensive Sickle Cell Centers in 2010 which include: I) Acute chest syndrome: acute respiratory illness with fever and/or respiratory symptoms such as cough, dyspnea, tachypnea, or hypoxia requiring hospitalization, II) Dactylitis: a new episode of acute pain and swelling in the fingers or toes with no clear source other than vaso-occlusion, III) Hyperhaemolysis: an episode of marked anaemia with evidence of increased red blood cell destruction, IV) Leg ulcers: ulceration of the skin of the legs with prolonged wound healing requiring medical attention for wound treatment, debridement, or dressings, V) Priapism: painful persistent, prolonged erection of the penis, VI) Sequestration crisis: an episode of marked anemia and enlargement of the spleen requiring hospitalization, VII) Stroke: acute focal neurological deficit in a pattern consistent with a stroke syndrome, VIII) Vaso-occlusive episode: a new episode of acute pain with no clear source other than vaso-occlusion that requires hospitalization and IX) Vision problem: participant complaining of vision problem that

was confirmed by any defect in visual field testing (Ballas et al. 2010). All patients received folic acid and instructions to drink extra fluids. Pneumococcal vaccination was not given to any subject, and neither prophylactic penicillin nor Hydroxyurea were given to any participants.

2.2 Ethical clearance

The study was approved and received ethical clearance from the Research Board at the faculty of medicine and health science, Gadarif University.

2.3 Statistical analysis

Data was entered into a computer database and SPSS software (SPSS Inc., Chicago, IL, USA, version 21.0) and double checked before analysis. Analysis of t was used to compare means and x2 was used for categorical variables. Univariable and multivariable logistic regression were used to identify factors associated with complications. For associated factors, odds ratios (OR) were determined with 95% confidence intervals [95% CI] and P value < 5 was considered significant.

3. Results

3.1 Baseline Characteristics

Among the total 83 investigated children, 43 (51.8%) were male and 40 (48.2%) were female give male: female ratio of 1.1. The age ranged between 9 months to 14 year with mean (SD) 6.4 (3.2). The age distribution of the studied cases was as follows: infants (0 to 2 years) constituted 9 (20.9%) cases; toddlers (3 to 5 years) 11 (25.6%) cases; children (6 to 9 years) 14 (32.6%) cases and ten- to fourteen-year-old constituted 9 (20.9%) of the cases. Approximately one half (50.6%) were of rural residence and more than one ethnic groups were represented. Hawsa tribe was the most prevalent (46.9%) tribe followed by Dabina (26.5%), Miseria (20.7%) and Kenana (6%) tribes. The mean (SD) age at diagnosis was 3 (1.1) years old. A family history with SCD was reported by 53 (63.8%), and 9 (10.8%) reported the death of a family member from SCD.

3.2 Clinical Evaluation and Hospitalizations

Thirty five children (42.2%) presented for follow up however, pain (33, 39.8%) followed by fatigability (8, 9.6%), fever (5, 6%) and jaundice (2, 2.4%) were the reported clinical presentation among the participants. Pallor was detected in 71 (85.5%), while hepatomegaly and skin ulcer were identified in 12 (14.5%) and 3 (3.6%) child respectively.

All of the participants (100%) reported a history of being hospitalized at least once for the management of their disease. The mean (SD) of times of hospitalization was 1.1 (0.9) and within the preceding 12 months, 36 (43.4%) had been hospitalized once and 7 (8.4%) had been hospitalized 2 or more times. Among children hospitalized in the past 12 months (43 children) the most common cause of hospitalization was a vaso-occlusive crisis in 20 (46.5%) children. Fever was the reason for hospitalization in 8 (18.7%) children, jaundice in 5 (11.6%), congestive heart failure in 5 (11.6%) epistaxis in 3 (6.9%) and acute chest syndrome in 2 (4.7%) participants. The number of hospitalizations in the preceding 12 months did not differ according to the socio-demographic data, table I.

3.3 The Prevalence of SCD-Complications among the Participants

The most prevalent prior complication of SCD was a vaso-occlusive episode which had been diagnosed in 61 participants (73.5%). Acute chest syndrome had been diagnosed in 8 participants (9.6%), and stroke had been diagnosed in 1 participant (1.2%).

3.4 Factors Associated with Complication

Using logistic regression analyses there was significant association between rural residence (CI= 1.5 – 22, OR= 17.1, P= 0.015), older age ≥ 6 year (CI= 5.1 – 11.6, OR= 5.5, P= 0.001) and SCD complications, (Table II).

Table I: Comparison between hospitalized and non-hospitalized children with SCD in the preceding 12 months in Gadarif hospital, Eastern Sudan, 2016.

Variables	Hospitalized	Not hospitalized	*P*
	(*N*=43)	(*N*=40)	
Age at diagnosis, years	3.1(1.0)	3.0 (1.2)	0.273
Ethnicity, Hawsa	20 (46.5)	19 (47.5)	0.622
Gender, male	23 (53.4)	20 (50)	0.121
Rural residence	22(51.2)	20(50)	0.855
Family history, yes	22(51.2)	21(52.5)	0.346

Data of age, is shown as mean (SD) while data of other variables are shown as number (%) as applicable.

Table II: Factors associated with SCD- related complications among children in Gadarif hospital, eastern Sudan, using univariate and multivariate analyses.

Variable	Univariate analyses			Multivariate analyses		
	OR	95% CI	*P*-value	OR	95% CI	*P*-value
Age ≥ 6 year	5.2	2.5-10.3	0.000	5.5	5.1-11.6	0.001
Residence, rural	6.1	2.8-10.5	0.000	17.1	1.5-22	0. 015
Gender, male	2.4	1.4-4.0	<0.001	1.3	0.7-2.4	0.213
Hawsa tribe	0.6	0.3-1.6	0.524	0.8	0.8-1.9	0.577
Family history, yes	1.9	0.9-2.3	0.06	1.2	0.8-2.0	0.221

Abbreviations: OR, Odds Ratio; CI, confidence interval

4. Discussion

This study described the epidemiological factors of SCD in eastern Sudan. Also it highlighted the frequency of hospitalizations and factors affecting the SCD-related complication. Sickle cell anaemia is one of the major health concern in the world (Brousse et al. 2017). In Sudan sickle cell anaemia is highly prevalent (Ali et al. 2014). Similarly SCD is a major health problem in countries sharing Sudan the same socio-demographic characteristics. For example Tanzania has the 3rd highest number of sickle cell anaemia in Africa after Nigeria and the Democratic Republic of Congo (Makani J et al. 2011). In the current study Hawsa tribe are predominantly affected by the disease. Many authors and observers believed that the sickle cell gene has been brought to Sudan through immigrants from West African tribes, especially from Hawsa, Folani and Bargo races (Bereir et al. 2007). And this of no doubt explained the Hawsa tribe being the dominant racial group among our participants. The male to female ratio in this study is 1:1 and the mean age of the children included in the study is 6.4 (3.2) year. A similar age distribution has been described in children with sickle cell disease in neighbouring countries. In Nigeria, the median age was 5.9 years while in Tanzania it was 6 years. In contrast to our study Saidi et al. found the male gender is predominantly affected among the sickle cell disease Tanzanian children (Saidi et al. 2016, Adegoke et al. 2015). The variation in the prevalence of the clinical presentation, the complications and lifetime hospitalization among the sickle cell disease population may be partially attributed to differences in the study age group as well as the socio-economic determinants. Socio-economic factors are important factors affecting the situation of health in all people. For instance there are many studies proved that increased poverty is associated with worse health (Marmot 2005). This is particularly relevant to our current study in which significant association between rural residence and SCD-related complications was found. In our case the rural people are of low educational level and they are of low daily income when compared with the urban residents.

Our results failed to find any statistical association between the frequency of hospitalization and socio-demographic data but on the other hand a significant statistical correlations were observed between SCD-related complications, rural residence and increased age. This contradiction may be attributed to the culture of the people in our setting who tried to treat the minor conditions without medical consultation. This will make the problem more worse by triggering sickle cell crisis and more complications. The poverty is one of the determinants that may make little if no improvement in the care that should be provided to children. Amendah et al. estimated that the overall annual economic cost per child for the provision of outpatient clinic care for children with SCD at Kilifi District Hospital (KDH), in coastal Kenya ranged from USD 94 to USD 229 in 2010 (Amendah et al. 2013).

One of the major limitations of this study: the sample size was small and the sample size was determined by convenience sampling method thus the variability and bias cannot be measured or controlled. Also it is a facility based study and not reflect the burden of the problem in the whole community.

5. Conclusion

While there is growing awareness about sickle cell disease our results indicate that SCD is a major public health problem with its severe complications among older children and those from rural area. Extensive effort is needed to improve the quality of life among sickle cell disease children through health education and economic support in particular in rural areas.

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Competing interests

The authors declare that they have no competing interests.

Authors' contribution AMA AEG, OSM and have designed the study. AMA and MAM carried out the work and gathered the data. AAA made data analysis and interpretation and wrote the manuscript. All authors revised and approved the manuscript.

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