Epidemiology and Characteristics of Sickle Cell Patients Admitted to Hospitals in Jazan Region, Saudi Arabia

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Abstract:

INTRODUCTION: Sickle cell anemia (SCA) is an autosomal recessive illness caused by the formation of abnormal hemoglobin S. Sickle cell disease has many complications such as vaso-occlusive crisis (VOC), gallstones, stroke, acute chest syndrome (ACS), and others.

OBJECTIVES: This study aimed to describe the epidemiology and characteristics of sickle cell patients admitted to hospitals in Jazan region.

MATERIALS AND METHODS: A cross-sectional study conducted in four hospitals in Jazan region including King Fahad Central Hospital, Prince Mohammed Bin Nasser Hospital, Sabaya, and Abu-Arish General Hospital to estimate the prevalence of admitted sickle cell patients and their characteristics. All patients admitted to medicine and pediatric wards from July 15, 2018, to August 15, 2018, were included in the study. Data were collected by interview.

RESULTS: Four hundred and two of 446 patients have agreed to participate with a response rate of 90%. Ninety-one (22.6%) were SCA patients, among them, 45 (49.5%) were male and 46 (50.1%) were female, with a mean age of 18.81 ± 11.05 years. Most of them were admitted due to VOC (56\%\ (n = 51)) and ACS (12.1\%\ (n = 11)). Almost all sickle patients had VOC (91.2\%\ (n = 83)), and more than half of them had ACS (58.2\%\ (n = 53)) as a complication.

CONCLUSIONS: SCA was the number one cause of admissions in this study. Approximately, one out of each four patients admitted to Jazan hospitals is a sickle cell patient. VOC represents the most frequent complication of sickle disease, followed by ACS. Further studies and interventions to reduce this burden and improve sickle cell patients’ health status are recommended.

Keywords: Complications, Jazan, prevalence, Saudi Arabia, sickle cell disease

Introduction

Sickle cell anemia (SCA) is an autosomal recessive illness caused by the formation of abnormal hemoglobin S.\cite{1} This type of hemoglobin produced by replacement of glutamic acid instead of valine in position number six of the \(\beta\)-chain, which further results in deoxygenation-induced polymerization and finally result in the abnormal crescent shape of red blood cells (RBCs). These abnormal RBCs can occlude small blood capillaries causing vaso-occlusive crisis (VOC), ischemia, and tissue damage; also this sickled hemoglobin makes RBCs vulnerable to broken easily resulting in extravascular and intravascular hemolysis with a result of low hemoglobin level.\cite{2,3} SCA nearly affects all body organs with many complications such as VOC, acute chest syndrome (ACS), gallstones, stroke, ...
and others.[4] Among all hemoglobinopathies, SCA has the highest public health concern.[5] It is the most common hemoglobin abnormality worldwide and one of the most significant single-gene diseases of humanity. Worldwide, SCA carriers are about 7% and 300,000–400,000 babies born yearly with severe forms of SCA. SCA is scattered all over sub-Saharan Africa, the Middle East, and parts of India.[6] In Africa, a number >200,000 of newly born babies are born every year with SCA.[7] SCA is the third principal cause of hospital admissions and mortality among children during the hospital stay in Africa.[8] In Saudi Arabia, SCA is a prevalent disorder. The estimated prevalence of SCA through the Saudi Premarital Screening Program report its prevalence to be 5/1000 for sickle cell trait (SCT) and 0.38/1000 for sickle cell disease (SCD).[9] Newborn screening estimated the prevalence of 21% for SCT and 2.6% for SCD.[10] One of the principal factors that are continuing the occurrence of SCT and increases the risk of homozygous sickle cell patients is the increased frequency of consanguinity.[10] In Saudi Arabia, premarital screening is practically the main preventive measure to prevent the inheritance of hemoglobinopathies including SCA.[10] As there were no existed studies about sickle cell patients’ characteristics and the impact of the disease on the hospitals in Jazan region, we intended this study to explore these points.

Materials and Methods

We conducted a cross-sectional study in four hospitals in Jazan region, including King Fahad Central Hospital (KFCH), Prince Mohammed Bin Nasser Hospital, Sabya General Hospital, and Abu-Areesh General Hospital to estimate the prevalence of sickle cell patients admitted to these hospitals and the prevalence of sickle cell complications among them. The study participants were enrolled in the study using a full coverage method. All patients admitted to medicine and pediatric wards in these hospitals during the 1-month period from July 15, 2018, to August 15, 2018, were included in this study. Data were collected by medical interns working at these hospitals to ensure daily contact with patients’ admissions. Data were collected through an interview using a questionnaire that was constructed by the research team, including questions about sociodemographics of the patients, admission diagnosis, and sickle cell complications. Ethical approval was obtained from the Research and Ethics Committee at KFCH (registry no. 076). Written consents were obtained from the patients.

Statistical analysis

Before data entry, it was managed and checked for any mistakes and missing values. Descriptive statistics was calculated. Categorical variables are presented in frequencies, percentages, and tables. Means and standard deviations were calculated for continuous variables. All statistical tests of the analysis were performed using the Statistical Package for the Social Sciences Program, version 25 (IBM Corp., Armonk, NY, USA). Graphs were built using Microsoft Excel software.

Results

A total of 402 of 446 patients have agreed to participate in this study with a response rate of 90%. Total sample and sickle cell patients’ demographics are shown in Table 1. Among the total sample, 113 (28%) patients have hemoglobin disorders, among them, 91 (22.6%) were sickle cell patients, 15 (3.7%) were sickle cell thalassemia patients, and 7 (1.7%) were beta-thalassemia patients. All causes of admissions were demonstrated in Figure 1. Sickle cell patients who participated in the study represented 11 governorates in Jazan region, most of them were from Abu-Areesh 27.4% (n = 25) followed by Sabya 19.7% (n = 18) then Jazan 18.6% (n = 17), other governorates as shown in Figure 2. Among sickle cell patients, 45 (49.5%) were male and 46 (50.1%) were female.

Table 1: Sociodemographics of the study population

|                          | Total sample (n=402) | Sickle cell patients (n=91) |
|--------------------------|----------------------|-----------------------------|
| **Age*, range (mean±SD)**| 0-100 (27.96±23.57)  | 0-61 (18.81±11.05)          |
| **Gender, n (%)**        |                      |                             |
| Male                     | 206 (51.2)           | 45 (49.5)                   |
| Female                   | 196 (48.8)           | 46 (50.5)                   |
| **Marital status**, n (%)|                      |                             |
| Single                   | 124 (30.8)           | 38 (41.8)                   |
| Married                  | 106 (26.4)           | 15 (16.8)                   |
| Widow                    | 12 (3.0)             | 0                           |
| Divorced                 | 3 (0.7)              | 0                           |
| **Education*, n (%)**    |                      |                             |
| Illiterate               | 134 (33.3)           | 15 (16.5)                   |
| Primary                  | 98 (24.4)            | 22 (24.2)                   |
| Intermediate             | 18 (4.5)             | 6 (6.6)                     |
| Secondary                | 38 (9.5)             | 26 (28.6)                   |
| University and above     | 37 (9.2)             | 9 (9.9)                     |
| **Residency, n (%)**     |                      |                             |
| Rural                    | 230 (57.2)           | 45 (49.5)                   |
| Urban                    | 172 (42.8)           | 46 (50.5)                   |
| **Occupation*, n (%)**   |                      |                             |
| Student                  | 141 (35.1)           | 49 (53.8)                   |
| Employed                 | 49 (12.2)            | 6 (6.6)                     |
| unemployed               | 82 (20.4)            | 16 (17.6)                   |
| **Department, n (%)**    |                      |                             |
| Medicine                 | 188 (46.8)           | 47 (51.6)                   |
| Pediatric                | 214 (53.2)           | 44 (48.4)                   |
| **Consanguinity, n (%)** |                      |                             |
| Yes                      | 202 (50.2)           | 69 (75.8)                   |
| No                       | 200 (49.8)           | 22 (24.2)                   |

*Range (mean±SD); **Age was adjusted and those were <15 years of age excluded from the analysis; †Age was adjusted and those were preschool age excluded from the analysis; *Age was adjusted, and those were <18 years and not currently students excluded from the analysis. SD=Standard deviation
The mean age of sickle patients is 18.81 ± 11.05 years. All sickle patients were admitted through the emergency department with the diagnosis of VOC 56% (n = 51) and ACS 12.1% (n = 11), other causes of admissions as shown in Table 2. They were admitted to medicine and pediatric ward in near equal distribution (medicine 51.6% [n = 47] vs. pediatric 48.4% [n = 44]). Consanguinity was found to be positive among 75.8% (n = 69) of sickle cell patients. Almost all sickle patients had VOC as a complication of sickle disease 91.2% (n = 83), and more than half of them had ACS 58.2% (n = 53), other complications are demonstrated in Figure 3.

Discussion

Prevalence
SCA is the most common hemoglobinopathy globally. In Saudi Arabia. It is most prevalent in Jazan region after the eastern region. In this study, SCA was the most common cause of admissions. More than one-fifth of all patients’ admissions during 1 month were of sickle cell patients. Similar results were also reported in Makkah, Saudi Arabia, in 2013, in which sickle cell patients had the largest portion of patients’ admissions among anemic patients admitted to Al-Noor specialist hospital with a percentage of 38.38%. However, these results do not indicate a higher prevalence in Makkah more than Jazan because they included only anemic patients. In a rural hospital of central India, SCA was the most frequent cause of admission among all hemoglobin disorders, 5.7% of all admissions during 1-year period belonged to SCA.

Patients’ sociodemographics
The vast majority of sickle cell patients were from Abu-Areesh, followed by Sabya. Taking into consideration that Sabya has the greatest number of population, this indicates clustering of the disease in some areas than others, which can be attributed to consanguinity as it plays a major role in sickle cell distribution. Consanguinity has a significant role in the continuity of SCA inheritance in Saudi Arabia. Alhamdan et al., in 2007, reported about 90% of high-risk relatives complete their marriage despite knowing the risk of disease inheritance. The percentage was higher (98%) in the study conducted by Al-Sulaiman et al. in 2010. Reasons for their ignorance of premarital
SCA affects the ability of the patients to work; we found unemployment among 72.7% of sickle cell patients with age >18 years. This finding is consistent with the findings of a previous study that assessed the quality of life among sickle cell patients in Saudi Arabia that found unemployment among 71.4% of sickle cell patients with age >18.18

**Cause of admission**

The most frequent cause of admission was VOC, followed by ACS. This is consistent with most of the previously reported studies.19-22 All patients’ admissions in our study were through the emergency room. It is well-known in the clinical history of SCD that most of their admissions are through the emergency room.23 In this study, we found that the most frequent cause of admission was VOC, followed by ACS. Similar findings were reported in previous studies. The most frequent causes of hospitalization among Yemeni children were VOC (36%), anemic crisis (16%), and ACS (11%).24 In Makkah, VOC was the most frequent reason for admission (47.1%), then SCA alone (23.6%), and ACS (9.3%).25

**Complications**

The most common complications of SCA were VOC and ACS, as reported in many studies.26,27 Our results were consistent with these data as we found the most frequent complication is VOC, followed by ACS then cholecystitis.

**Conclusions**

SCA represented the number one cause of admissions in our study. Approximately, one of each four patients admitted to Jazan hospitals is sickle cell patient. VOC represents the most frequent complication of sickle patients, followed by ACS. Further studies to explore the burden of SCD in Jazan region, consanguinity status after the application of premarital screening and the reasons behind the proceeding of high-risk marriages are needed. The interventions to reduce this burden and improve sickle cell patients’ health status are recommended.

**Limitations**

The short duration of the study together with the limited number of health institutions included may underestimate the burden of SCD in Jazan region. However, we tried to include the largest hospitals in the region where most of the patients admitted to. Furthermore, the time in the emergency room before inpatient admission together with the length of hospital stay was not included in our study.

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**Conflicts of interest**

There are no conflicts of interest.

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