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Research Article

Musculoskeletal complications in sickle cell anemia patients: a ten-year retrospective review of hospital-based records (1991-2000) in two Nigerian hospitals

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ABSTRACT

Background: Sickle Cell Anaemia (SCA) presents with various types of clinical manifestations that includes musculoskeletal manifestations that need the intervention of medical personnel including physiotherapy. The study aimed to determine the various musculoskeletal disorders associated with Sickle Cell Anaemia (SCA), types of treatments given including physiotherapy and the mortality rate in SCA patients admitted in two Nigerian hospitals.
Methods: It is a retrospective quantitative review of hospital-based records using patient's files and hospital registers. Ten years case notes of SCA patients were reviewed in two hospitals in Kano, North Western Nigeria. Information on age, gender, types of musculoskeletal disorders, types of treatments received including referrals for physiotherapy and number of deaths recorded within this period.
Results: Records shows a total of 248 patients admitted with SCA over this period. Results revealed 133 (53.6%) males and 115 (46.4%) females. Prevalence of musculoskeletal disorders was found to be 172 (54.1%) out of the 318 SCA cases admitted. The commonest cause of hospital visitation was painful bone crisis (35.5%). Followed by osteomyelitis and dactylitis accounting for 20 (11.6%) of the musculoskeletal disorders respectively. Overwhelming majority 61.7% presenting with musculoskeletal disorders are between the age of 3 months and 10 years. The mortality rate was about 22/248 (8.9%). Only 8.9% of these cases received physiotherapy.
Conclusions: Sickle cell disease affect a wider group of people than is commonly realised, screening for the disease is not universal and this together with a general lack of multi-disciplinary qualitative and clinical evidence-based treatment, leads to many inadequate management of individuals. Physiotherapy is one of the health care services that can be given following musculoskeletal presentations, yet it appropriateness in relation to the disease is rarely being addressed.

Keywords: Sickle cell anaemia, Musculoskeletal complications, Physiotherapy

INTRODUCTION

The term sickle cell disease is the prototype of hereditary hemoglobinopathies characterised by the production of structurally abnormal haemoglobin.[1,2] These hemoglobinopathies include sickle cell anaemia (HbS), sickle cell hemoglobin C disease (HbC) and sickle cell thalassaemia.[3] Only the variant sickle cell (HbS) with genotype SS and (HbC) sickle hemoglobin C are known to cause illness in Nigeria while sickle cell thalassaemia is of two types, the beta plus thalassaemia (Hbs β+ Thal.) and the alpha thalassaemia. The beta thalassaemia gene (usually β+) is very rare, but may occasionally interact

with HbS to cause illness. In contrast α thalassaemia is common but is benign and asymptomatic.

HbSS is clinically the most important of all the genotypes that are encompassed in the broad definition of SCD and its manifestations are more severe. The clinical presentations of the diseases are often similar. Different SCD patients have different disease manifestations.[4-8] Musculoskeletal manifestations constitute up to 80% of indications for presentation to hospital during their life time.[9,10] Pain is the principal complaint either acute following skeletal or soft tissue infarction or chronic secondary to avascular necrosis of bone at various joints.[11]

Generally recurrent vaso-occlussive episodes takes place in most organs of the body including sensitive ones such as the kidneys, lungs and the brain, resulting in chest syndromes, pneumococcal infections, strokes, priapism and renal complications which are clearly life threatening,[12] other disorders associated with SCD mostly musculoskeletal are less life threatening but can drastically reduce quality of life overtime, these may include the hand and foot syndrome, vertebral collapse, painful bone crises, acute and chronic leg ulcers, acute and chronic osteomyelitis and back pain are just a few of the presenting problems[12] in people living with the disease.

The disease can be seen in individuals in the equatorial Africa, Turkey, northern Greeks, eastern province of Saudi Arabia, Italy, parts of India and America.[13] About 300000 infants are born with haemoglobinopathy, 200000 of whom are born in Africa[14] Whereas SCD occurs in 1 in 500 African-American births, it prevalence in countries such as Cameroon, Republic of Congo, Gabon Ghana and Nigeria is between 20-30% while in some parts of Uganda it is as high as 45%.[14] In Nigeria the current accurate figure of individuals with this disorder and associated problems is not known since the majority who were born to rural communities doesn't survive childhood.[15] However, an estimate of 2-3% of the Nigerian population suffers from this disease and about 25% Nigerians are healthy carriers of the abnormal gene.[15] The high incidence of SCD in Africa and other parts of the world is attributed to a hypothesis of "balanced polymorphism" in which the heterozygous HbAS, carriers are protected against the severe effects of malaria in infancy and so have a survival advantage.

SCD causes a heavy burden on the society by the high morbidity and premature death associated with it.[16] Sickle cell disease affect a wider group of people than is commonly realised, screening for the disease is not universal and this together with a general lack of multi-disciplinary qualitative and clinically evidence-based treatment, leads to many inadequate management of individuals. Physiotherapy is one of the health care services that can be given following musculoskeletal presentations, yet it appropriateness in relation to the disease is rarely being addressed. Therefore there is lack of a holistic care towards these individuals even though few studies have elaborated on the role of physiotherapy in SCD individuals.[3] In fact most studies on musculoskeletal presentations of SCA in Nigeria have focused on selected disease conditions.[9,17,18] This study therefore determine the musculoskeletal complications, types of treatments given including physiotherapy and the mortality rate in SCA patients in two Nigerian hospitals.

METHODS

Population and sampling procedure

All admitted SCA cases between 1991 and 2000 were eligible for the study. The study was a retrospective quantitative review of hospital based records. It was carried-out in the medical records departments of Murtala Muhammad Specialist Hospital and Dala Orthopaedic Hospital both in Kano, Nigeria. Two hundred and forty eight cases of SCA were extracted from the hospital registers from 1991 to 2000. Case notes (n=248) of SCA patients who had attended either hospitals were scrutinized and reviewed for participants' medical history. Subjects included in this study were patients aged 3 months to ≥35 years with haemoglobin genotype SS. Case notes containing incomplete data or unclear diagnosis were excluded from this study. Also other types of SCDs that are not SCA were not included in the study.

Ethical consideration

Ethical approval of the institutional review committee of Bayero University Kano was sought and obtained prior to the commencement of this study.

Procedure

SCD patients' case files covering a period of ten years from January 1991 - December 2000 were reviewed in two selected hospitals; National orthopaedic hospital, Dala and Murtala Muhammad Specialist Hospital Kano. The case notes of SCA patients treated and admitted at these hospitals were subjected to detailed scrutiny in the medical records section of each hospital. Ten years records were retrieved and reviewed. Cases of Musculoskeletal complications associated with SCA were extracted and recorded. The following data were collected from the SCA patients' case files; age of the patients, gender, types of disorders which patient was admitted to hospital from which cases of musculoskeletal disorders were extracted and recorded on data collection sheets, types of treatments received including referrals for physiotherapy and number of deaths recorded within this period.

Some of the case files listed on the registers where missing also some of the files has poor documentation or incomplete documentation in such instances there were excluded from the study.

Statistical analyses

Descriptive statistics of percentages and frequency were used to describe the socio demographic characteristics (age, gender, cause of admission and mortality rate), various types of musculoskeletal complications admitted within this period, participants health seeking behaviours including physiotherapy referrals. Tables were used to present the results. The data were analysed using SPSS version 16.

RESULTS

Sociodemographic characteristics: A total of 248 SCA patients were admitted from 1991 to 2000. National orthopaedic Hospital Dala, Kano admitted 35 (14.1%) SCA patients and Murtala Muhammad Hospital, Kano admitted 213(85.9%) SCA patients within this period. Out of the total number of 248 SCA patients, 133(53.6%) were males, while 115 (46.4%) were females. The age of patients ranges from between 3 months - 35 years and above. The highest incidence of admission due to complications of SCA was recorded among the age group of 3 months to 5 years with 92 (37.1%) recorded cases, followed by the age group of 6-10 years with 61 (24.6%) recorded cases. The lowest number of admission was recorded among the age group of 35 years and above with 5 (2.0%) recorded cases. The study observed 318 complications due to SCA within this period. 172 (54.1%) were cases of musculoskeletal disorders while 146 (45.9%) were other complications associated with SCA. A total number of 22 patients (8.9%) were recorded dead and the remaining 226 (91.1%) were discharged alive as of time of research (Table 1).

Table 1: Sociodemographic characteristics of the participants.

Sociodemographic variables	n	Percentage (%)
Age-group (years)		
0.25-5	92	37.1
6-10	61	24.6
11-15	29	11.7
15-20	20	8.1
21-25	24	9.7
26-30	10	4.0
31-35	7	2.8
≥36	5	2.0
Total	248	100
Gender		
Male	133	53.6
Female	115	46.4
Total	248	100
Cause of admission		
Musculoskeletal disorders	172	54.1
Other disorders	146	45.9
Total	318	100
Mortality rate	22	8.9

Table 2 shows the musculoskeletal disorders seen among the SCA patients over ten years period. It shows that painful bone crisis is the most prevalent and cause of admission in these patients (35.5%). Low back pain, osteomyelitis and hand and foot syndrome are the second most common disorders among these patients with 20 (11.6%) each. The third most common cause of admission in this population was found to be (9.9%) avascular necrosis. The study found the least musculoskeletal disorder presenting in this population to be rib infarction and vertebral collapse (1.2% and 1.7%) respectively.

Table 2: Musculoskeletal disorders among SCD patients.

Musculoskeletal disorders	n	Percentage (%)
Avascular necrosis	17	9.9
Painful bone crises	61	35.5
Vertebral collapse	3	1.7
Low back pain	20	11.6
Chronic leg ulcers	13	7.6
Osteomyelitis	20	11.6
Hand and foot syndrome	20	11.6
Rib infarction	2	1.2
Septic arthritis	10	5.8
Priapism	6	3.5
Total	172	100

Table 3 shows the type of treatment received by these patients, out of a total number of 248 patients, all patients (100%) receives various types of medical treatments and only 22 (8.9%) receives physiotherapy treatment. Fourteen (5.6%) also receives traditional treatment in addition to one or both of the stated treatments.

Table 3: Types of treatments sought by the SCD patients.

Type of treatment received	n	Percentages (%)
Overall No. of cases	248	100
Medical management	248	100
Physiotherapy referrals	22	8.9
Traditional treatment	14	5.6

DISCUSSION

The study is a ten-year retrospective study that observes the frequency of musculoskeletal complications among patients with SCA in two hospitals in northern part of Nigeria. The study shows that various musculoskeletal diseases are reported with SCA, which is in line with.[13] The prevalence of musculoskeletal disorders in this study is high 54.1%. This prevalence is higher than that reported by Chinawa et al.,[19] (32.1%) and Balogun et al.,[20] (31.4%) in Nigeria.

The study reported painful bone crisis as the most prevalent musculoskeletal disorder representing 92

(35.5%) of the disorders, this agrees with,[21] who stated that bone pain is the most common and rank as one of the most distressing disorder in SCA individuals and the most common reason for hospital visitation.[22] It also agrees with the study of Chinawa et al.,[19] that reported lower limb and upper limb pain as the common cause of admitted cases of SCA in their study. In the same line[23] reported pain crises including painful bone crisis to occur in all participants in their study as it is the most prevalent cause of health care consultation.

Osteomyelitis 20 (11.6%) is the second most common musculoskeletal complication reported among the SCA patients in the current study. This prevalence is similar with the findings of[19] (12.8%) but at variance with the prevalence of (37.5%) and (18%) reported by[20,24] respectively. Salmonella has been thought to be responsible for osteomyelitis in children with SCA in more than 50% of cases[25] and in over 70% of cases.[26] The disease is thought to have a high tropism for patients with SCA.[23] However, Zanoni et al.,[3] are of the opinion that in addition to the sensitivity of salmonella with SCA patients, the bad hygienic life conditions of the concerned population can explain an endemicity of salmonellosis in some regions especially in the developing countries, therefore buttressing the high prevalence of osteomyelitis among SCA patients in a developing country like Nigeria. The prevalence of septic arthritis in the current study is similar with previous reported prevalence of 6-15%.[16,17,20,27]

The study also reported hand and foot syndrome (dactylitis) and low back pain (11.6%) respectively as one of the leading musculoskeletal disorders seen among these patients. This is in contrast with[20] that reported a lower prevalence of 7.6% among their participants. However the study by[19] did not report any incidence of dactylitis and low back pain in their five year review.

Avascular necrosis is one of the musculoskeletal complications of SCA among this population accounting for 9.9% of the total musculoskeletal complications in this study. Previous studies have reported avascular necrosis to occur in 3-21% of SCA patients.[18-20] Sickle cell disease is the commonest cause off avascular necrosis in Nigeria.[18]

It was also shown that chronic leg ulcers carried about 13(7.6 %) of the cases studied, this prevalence is lower than the (10.7%) reported by[20] and higher than prevalence reported by Chinawa et al.,[19] (1.3%). However the finding is in line with the study by[30] Prasad et al. that reported ulcers to affect about 6% of Nigerian sicklers above the age of 12 years. The present study also reported 6 (3.5%) of the males had priapism, which is in accordance with.[15] however, some previous studies did not document the prevalence of priapism among SCA patients in Nigeria.[19,20] These prevalence is low (6/172) but the impending consequence is very tremendous.

The health seeking behaviour distinctively physiotherapy management is poor among the SCA patients. This might not be unrelated to poor referral system as regards to the sickle cell patients. Even though Physiotherapy has been seen to improve and efficiently treat musculoskeletal dysfunctions in SCA patients.[3,31] According to[32] majority of sickle cell patients managed with chemotherapy alone were seen as very unstable, hence, the recent development and treatment concepts advocate combination of several types of therapies other than chemotherapy alone, especially as regards joints pain and pain crisis. Therefore management of SCA requires holistic interdisciplinary evidence-based approaches.[3] As SCA patients can benefit from physiotherapy there is a need for a functional referral system in these hospitals. There is also a need to develop further programme of physiotherapy care for individuals with SCA, consideration should be tailored toward Facilitation, delivery and exchange of information between those charged with managing the condition, i.e. the individual, physiotherapist and other members of the existing health care team. In addition provision of a functional assessment of the contexts in which the physiotherapist has to function need to be made very clear.

Fourteen cases (5.6%) of the patients in additional to the conventional treatments (medical), also received traditional treatment in their respective community. Traditional medicine or treatments of SCA is a common practice in West Africa. In particular, the root of Fagara zanthoxyloides is widely used in West Africa both to prevent and to treat sickle crises. In vitro, the plant reverses sickling of red blood cells,[33] but there has only been one very small published clinical trial.[34] However, there are anecdotal reports of patients whose lives have been transformed by this treatment. If funding could be found for larger clinical trials to confirm the safety and efficacy of this medicine, this would be an important contribution to the care of sickle-cell patients both in Africa and in the rest of the world.[35]

The mortality rate of the patients as reported in this study is (8.9%), Bacterial infections were found to be the leading cause of death among patients with SCA.[28,29] although recent studies have indicated reduced mortality rate among SCA patients, because of the upward surge in life expectancy of sickle cell patients due to better understanding and correct management.[36-38] In the same line recent studies have reported improvement in the life expectancy of SCA patients over the last century.[37,38] Nevertheless, sickle cell disease causes a heavy burden on the society by the high morbidity and premature death associated with it.[17] It was observed in the course of this study that no carrier state of the disease was admitted within the stated period of the study, which is in accordance with[39] Oni et al., that most carrier state (those with the sickle trait) does not cause clinical pathology.

The study is not without limitations. Not all required information could be obtained from the hospital case files

of the patients this leads to reduced sample size. Another limitation is that the study only covers two hospitals in Nigeria; therefore generalisation of findings should be used with caution. In addition other forms of SCDs were not included in the study therefore limiting findings to SCA patients merely.

The place of genetic counselling is obviously small in Nigeria. Young adults should seek for advice before marriage. Genetic counselling should be done at an age when the significance of counselling is understood but sexual associations and relationships not yet developed. Standardization of haemoglobin genotype determinations in our laboratories should be carried out for accurate and easy diagnosis. Haemoglobin electrophoresis should be made available to as many as required it.

CONCLUSION

The study concluded that various musculoskeletal complications associated with SCA patients exist as documented by previous studies. The most common complications reported is painful bone crises followed by osteomyelitis, low back pain and the hand and foot syndrome. Complication due to rib infarction reported lowest. There is also improvement in their life expectancy. Though there is need for improved and functional referral system as regard these patients.

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