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Orthopedic Complications of Sickle Cell Disease: A Retrospective Evaluation of 469 Patients in Port Harcourt, Nigeria

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Abstract

Background: Sickle cell anemia (SCA) is a hereditary blood disorder characterized by abnormal hemoglobin, leading to the deformation of red blood cells. There is growing impact of this condition on the musculoskeletal system. This study aimed to evaluate the Orthopedic complications in patients with SCA in Port Harcourt between

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July 2013 and June 2023.

Materials and Methods: A retrospective analytical study of Orthopedic complications in patients with SCA who presented to the Hematology clinics of two teaching Hospitals was conducted. Collected data was analyzed with the Statistical Package for Social Sciences.

Results: Among the 469 patients with SCA, 198 were males. The mean age of the study population was 25.21 years. The Orthopedic complications assessed included Avascular Necrosis of the Femoral head, Chronic leg ulcers (CLU), Osteomyelitis, and Osteoarthritis. A vascular Necrosis (AVN) of the Femoral head was observed in 50 (44.6%) patients. Chronic leg ulcers were present in 57 (50.9%) participants, while Osteomyelitis (OM) was identified in 14 (12.5%) patients. Osteoarthritis (OA) was the least common complication, occurring in only 3 individuals. The age group above 25years was statistically associated with development of Orthopedic complications especially for CLU and OM.

Conclusion: Orthopedic complications are common in SCA patients, with a vascular Necrosis of the Femoral head, Chronic leg ulcers, Osteomyelitis, and Osteoarthritis being the most frequently observed complications. These findings underscore the need for comprehensive management strategies and multidisciplinary care to minimize the impact of these complications on patients' quality of life.

Keywords
Orthopedic Complications; Sickle Cell Disease; Port Harcourt; Nigeria

Introduction
Sickle cell anemia (SCA) is a hereditary blood disorder characterized by abnormal hemoglobin, leading to the deformation of red blood cells. This disease is most found in sub-Saharan Africa, East Mediterranean areas, Middle East, and India [1-2]. There has been a 13·7% global increase in the births of babies with sickle cell disease over a 21-year period from year 2000 and 2021 [3]. While the primary manifestations of SCA are related to the hematological system, there are divers clinical presentations with likelihood of the sickle cell disease patient being seen by the hematologist for outpatient care, the pediatrician or physician at the Emergency Department for sickle cell crisis, [4-5] by a General Surgeon for acute abdomen, [6-7] by a Nephrologists / Urologist for hematuria, [8] by the Orthopedic Surgeon for Orthopedic complications,[9-12]etc. Additionally, in managing any surgical condition in a patient with sickle cell anemia, special considerations or precautions are often taken for good outcome.[13-17]

While the standard care is known to be the use of analgesia, oxygenation, hydration, blood transfusions, and prevention of triggers of sickle cell crisis, [18] the use of a drug – Voxelotor, an inhibitor of polymerization of Hemoglobin S by increasing the oxygen affinity, has recently been shown to be very useful [19-22]. Orthopedic presentations and complications of sickle cell anemia has been reported in Eastern Nigeria, [23-24] Western Nigeria, [25-26] southern Nigeria, [27] and Northern Nigeria [28]. There

is therefore growing recognition of the impact of this condition on the musculoskeletal system. Orthopedic complications among others are common among sickle cell disease patients in our practice, as reported in some studies in Nigeria [24-25, 27, 29]. Nigeria is known globally to have the largest cohort of sickle cell anemia (SCA), [27] and Port Harcourt is a cosmopolitan city with a high traffic of Nigerians due to petroleum oil and gas exploration and processing activities. This makes Port Harcourt to be home to many Nigerians, with the potential of physician encountering sickle cell disease patients and their complications. This study therefore explored orthopedic complications of sickle cell anemia patients as seen at the Hematology Clinics of Two Teaching Hospitals, Port Harcourt from July 2013 to June 2023.

Materials and Methods

Study Area: The study was carried out at The Rivers State University Teaching Hospital and the University of Port Harcourt Teaching Hospital, both being tertiary healthcare facilities in Port Harcourt, the capital of Rivers State, South-South of the Federal Republic of Nigeria.

Study Sites: The study site/setting was the Hematology Clinics of both Teaching Hospitals in Port Harcourt Nigeria.

Research Design: A retrospective analytical study.

Study Population: All patients (469) who were seen and treated at the study sites with orthopedic complications of Sickle Cell Anemia were included.

Sampling Method: Total population of sickle cell anemia patients with orthopedic complications as seen in the registers was used.

Study Instrument: The registers of the Hematology Clinics were used to obtain data that was imputed into a preformed designed for the study.

Study Variables: Patients' age, sex, disease history (age of SCA diagnosis, known steady-state packed cell volume (PCV), history of blood transfusion, presence of siblings with SCA), and the orthopedic complications were the variables of interest.

Validity/Reliability of Instrument: The study data was scrutinized by all the authors for authenticity or otherwise before use.

Data Analysis: Data obtained was formed into tables and analyzed using the Statistical Package for the Social Sciences (SPSS) version 20.0. The Fishers' Exact (p-value) test was used for the association between age / sex and orthopedic complications.

Results

Variable	Frequency (n = 469)	Percent (%)
Sex of Patients		
Male	198	42.2
Female	271	57.8
Age group of Patients (Years)		
≤ 15	17	3.6
16 – 20	150	32
21 – 25	133	28.4
26 – 30	65	13.9
>30	104	22.2
Mean age: 25.21 ± 8.784 years		
Age Group at Diagnosis (Years) (n = 302)		
<1	22	7.3
1 – 3	105	34.8
4 – 6	85	28.1
7 – 9	34	11.3
10 – 12	16	5.3
>12	40	13.2
Steady State PCV (n = 153)		
15 – 19	19	12.4
20 – 24	82	53.6
25 – 29	42	27.5
>29	10	6.5
Mean PCV: 23.43 ± 3.622		
History of Transfusion (n = 347)		
Yes	246	70.9
No	101	29.1
Only Child with HBSS in Household (n = 293)		
Yes	214	73
No	79	27

Table 1: Socio-demographic Characteristics and Disease History.

There were more females (271, 57.8%) than males (198, 42.2%) in the study; most of whom were within the age categories of 16 – 20 and 21 – 25. The mean age was 25.21 ± 8.784 years. The mean steady state PCV was 23.43 ± 3.622. Those who have had blood transfusion consisted 246 (70.9%) while households that have or have had only one HBSS child accounted for 214 (73.0%).

Variable	**Frequency**	**Percent**
	Multiple Response (n = 124)	
Complications		
Avascular Necrosis	50	44.6
Chronic Leg Ulcer	57	50.9
Osteomyelitis	14	12.5
Osteo-Arthritis	3	2.7

Table 2: Orthopedic complications.

Chronic leg ulcer and a vascular necrosis were the more common orthopedic complications, accounting for 50.9% and 44.6% of orthopedic complications among patients.

Variable	**Sex of Patients**		**Fishers' Exact**
			(p-value)
Complications	Male	Female	
	n (%)	n (%)	
Avascular Necrosis	19 (9.6)	31 (11.4)	0.549

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Chronic Leg Ulcer	23 (11.6)	34 (12.5)	0.777
Osteomyelitis	3 (1.5)	11 (4.1)	0.168
Osteo-Arthritis	2 (1.0)	1 (0.4)	0.576

Table 3: Association between Sex and Orthopedic Complications.

The association between sex and orthopedic complications were not statistically significant, as shown by Fishers' Exact test.

Variable	**Age Group of Patients (Years)**		**Fishers' Exact**
	Merged		**(p-value)**
Complications	≤ 25	>25	
	n (%)	n (%)	
Avascular Necrosis	26 (8.7)	24 (14.2)	0.085
Chronic Leg Ulcer	28 (9.3)	29 (17.2)	0.018
Osteomyelitis	5 (1.7)	9 (5.3)	0.044
Osteo-Arthritis	1 (0.3)	2 (1.2)	0.296

Table 4: Association between Age Group Orthopedic Complications.

Fishers' Exact test shows a statistically significant association between orthopedic complications and age group of patients, this includes chronic leg ulcer and osteomyelitis, having p-values of 0.018 and 0.044.

Discussion

A wide range of the Nigerian population have been reported to have poor knowledge of sickle cell disease including secondary school students, [30-31] undergraduates, [32-33] local government workers, [34] and women of childbearing-age [35-37]. The fact that Sub-Sahara Africa has the highest prevalence of this abnormal hemoglobin disease (with Nigeria having the largest population of the patients), [38] did not change the experience of poor knowledge. The abnormality associated with the beta chain alteration (valine replaces glutamic acid in position six) is responsible for the path physiologic changes seen when

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the red cell sickles following exposure to triggers such as low altitudes, infections, extremes of temperature amongst others [27]. This change has considerably negative impact on the patients' organs / systems including the bones and joints, irrespective of the patient's sex. In this study, there were more females with orthopedic complications. This finding differs from the reports from Enugu and Lagos Nigeria where majority of the patients were males, [23-25] but share similarity with a South-South Nigeria study where female patients predominated [27]. Majority of our patients were in their young age (mean age was 25.21 ± 8.784 years), as seen in most other studies.

No statistically significant relationship was found between patients' gender and orthopedic complications. Our finding is in agreement with another report from a Nigerian study where no relationship was found with gender [23]. However, a statistically significant relationship was found between the age group of these patients and orthopedic complications (Chronic leg ulcer /Osteomyelitis - p-values of 0.018 and 0.044 respectively), making our study finding different from that of Enugu Nigeria study [23]. The mean steady state packed cell volume of the patients was 23.43 ± 3.622. This value is close to the value of 24.46±4.76 reported in a Lagos Nigeria Study, [39] but lower than the value of 27.18 ± 5.35% from North Maharashtra in India [40]. However, the steady state packed cell volume in our study is higher than the value of 20.7 reported in a Zaria (Nigeria) study for SCA patients with Chronic leg ulcer [41]. Majority (70.9%) of the households of these patients had only one child with HB SS, while the rest had more than one. This finding may vary depending on the society in question and the genotype of the parents.

Chronic leg ulcer was prevalent in about half (50.9%) of our patients, and a vascular necrosis in 44.6% of the patients, followed by Osteomyelitis. These findings highlight the significant burden of orthopedic complications in patients with SCA. A vascular necrosis of the femoral head, a condition characterized by the death of bone tissue due to poor blood supply, was observed in a considerable number of individuals. Chronic leg ulcers, which can significantly impact mobility and quality of life, were also prevalent. Osteomyelitis, a bone infection, was identified in a subset of the study population. The presence of osteoarthritis further emphasizes the long-term impact of SCA on joint health. Osteomyelitis was the commonest orthopedic complication of the disease as seen in Lagos some 12 years ago [25]. There were higher number of patients with chronic leg ulcer and a vascular necrosis than was reported in Lagos, [25] and relatively similar to the report outside Nigeria for osteonecrosis [42] However, a review article published in year 2015 reported vascular necrosis, osteomyelitis, septic arthritis, leg ulcer, etc. as the most common orthopedic complications of sickle cell anemia [23].

Study Limitations

The findings of this study were derived from a retrospectively collected data from medical records. It is therefore limited by the general demerits of this type of study.

Conclusion

Orthopedic complications are common in adults with Sickle Cell Anemia (SCA), and a vascular Necrosis of the Femoral head, Chronic leg ulcers, Osteomyelitis, and Osteoarthritis were the most frequently observed complications seen in Port Harcourt. These findings underscore the need for comprehensive

management strategies and multidisciplinary care to minimize the impact of these complications on the quality of life of individuals with SCA.

Other Information

Ethical considerations: The approval of the Research Ethics Committee of the Rivers State University Teaching Hospital was obtained. This study involved only contact with medical records with no contact with the individual patients.

Source of Funding: The study was self-funded by the researchers.

Conflict of Interest: None declared.

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