



A Comprehensive Study on the Hematological Progression of Sickle Cell Disease Patients with COVID-19 at the Center for Research and Control of Sickle Cell Disease, Bamako

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Received date: 29 December, 2023, Manuscript No. JBRHD-24-124036;

Editor assigned date: 01 January, 2024, Pre QC No. JBRHD-24-124036 (PQ);

Reviewed date: 15 January, 2024, QC No. JBRHD-24-124036;

Revised date: 22 January, 2024, Manuscript No. JBRHD-24-124036 (R);

Published date: 30 January, 2024 DOI: 10.4172/jbrhd.1000194

Abstract

The Coronavirus disease 2019 pandemic is a real crisis that has exposed the unpreparedness of many healthcare systems worldwide. Several underlying health conditions have been identified as risk factors, including sickle cell disease, a chronic illness with various complications that can increase the risk of severe COVID-19 infection. Our study aimed to investigate the profile of sickle cell patients diagnosed with COVID-19 and explore any potential relationship between these two conditions. We analyzed data from 11 sickle cell patients who contracted COVID-19 between June and December 2020 and were treated at the CRLD (Center for Sickle Cell Disease and Research). The patients' COVID-19 diagnosis was confirmed using the (Real-Time Reverse Transcriptase-Polymerase Chain Reaction) RT-PCR technique on nasopharyngeal swab samples and/or based on clinical and radiological findings, including CT scans. The patients consisted of 7 males and 4 females, with a mean age of 40 ± 12 years. The sickle cell phenotypes observed were SC (45.4%), SS (36.37%), and S β \pm thalassemia (18.2%). During the COVID-19 infection, we observed a slight increase in white blood cell and platelet counts, but a decrease in mean hemoglobin levels and red blood cells. Only 3 out of 11 patients (28%) had a fever at the time of diagnosis. Three patients required red blood cell transfusions due to severe anemia, and 7 out of 11 patients (63.6%) were hospitalized, with one patient admitted to the intensive care unit due to pulmonary embolism. All patients recovered from COVID-19.

Keywords: Sickle cell disease; COVID-19; CRLD (Center for Sickle Cell Disease)

Introduction

The Coronavirus pandemic has caused numerous deaths and disrupted healthcare systems, particularly for chronic diseases like sickle cell disease. Sickle cell patients constitute a vulnerable group who require lifelong appropriate medical care. The Coronavirus disease (COVID-19), caused by Severe acute respiratory syndrome corona virus two, has become a pandemic affecting over 170 countries worldwide [1]. The heightened susceptibility to bacterial and viral infections poses a comorbid risk in this coronavirus pandemic. The increased susceptibility of sickle cell patients is attributed to compromised immunity resulting from Functional Hypersplenism and Systemic Vasculopathy, which predisposes them to organ damage and dysfunction, with a risk of thrombosis [2,3]. It is currently known that pulmonary thrombosis is a major comorbid factor of COVID-19. Several anecdotal reports have suggested an increased risk of COVID-19 complications in sickle cell patients [4,5]. General population data have shown that advanced age and the presence of medical conditions such as cardiovascular diseases, hypertension, diabetes, and pre-existing respiratory insufficiency pose a high risk for severe COVID-19 complications [6]. This preliminary work aims to report some clinical, hematological, and evolving characteristics of sickle cell patients with COVID-19 in a regular follow-up context.

Methodology

Our study is a matched case-control study where the cases were their own controls, focusing on sickle cell patients followed at the CRLD in Bamako from June to December 2020. During this period, the Center recorded 11 sickle cell patients who were diagnosed with COVID-19 as part of their regular follow-up program. The diagnosis of COVID-19 was confirmed using the RT-PCR technique on nasopharyngeal swab samples in 9 patients, while in cases where RT-PCR results were inconclusive, diagnosis was based on clinical and radiological (CT scan) characteristics. This inconclusiveness can be attributed to the currently accepted sensitivity of the RT-PCR technique, estimated at 70% [2]. Baseline hematological parameters were compared to those during the COVID-19 infection period. The outcomes of the affected sickle cell patients were also documented in collaboration with the treatment center.

Results

A total of 11 sickle cell patients with COVID-19 were identified at the Center for Research and Control of Sickle Cell Disease. Among these 11 sickle cell patients who tested positive for COVID-19, 7 (63.6%) were male and 4 (36.4%) were female. The average age at diagnosis was 40 ± 12 years, ranging from 30 to 66 years. The distribution of sickle cell phenotypes was as follows: Five SC (45.4%), four SS (36.4%), and two S β \pm thalassemia (18.2%). None of the S β 0-thalassemia cases were affected by COVID-19. One of the eleven patients had pre-existing comorbidities such as hypertension, diabetes, and early-stage nephropathy. None of the patients were enrolled in a blood transfusion program. However, four patients had a history of Acute Chest Syndrome (ACS) and were receiving treatment with

hydroxyurea (Hydrea®) at a dose of 20 mg/kg/day. Three patients were diagnosed during a vaso-occlusive crisis of sickle cell disease. These vaso-occlusive crises of varying severity were likely exacerbated by the coronavirus infection. Other clinical manifestations included cough, fever, ageusia, dyspnea, rhinorrhea, hypoxia, and anosmia. These clinical signs varied from patient to patient, with combinations of symptoms observed in the same patient. Seven out of the eleven patients (63.6%) required hospitalization, including one in the intensive care unit due to a pulmonary embolism. The other four patients, who did not exhibit major symptoms, received home monitoring by the COVID-19 focal point team at the CRLD, following the established management protocol in Mali. Patients No. 8 and No. 9 presented with a severe clinical form, experiencing all the associated symptoms of COVID-19. Details of the clinical and radiological characteristics upon admission of sickle cell patients with COVID-19 are summarized in Table 1.

The most frequently reported symptoms among these patients were hypoxia (9/11), anosmia (9/11), ageusia, and rhinorrhea (8/11). Fever was the least common symptom, observed in only 3 out of 11 subjects. One patient was asymptomatic, while another exhibited only hypoxia.

Four out of the eleven subjects had at least 7 symptoms, indicating a significant disparity among individuals. Comparative analysis of the mean levels of baseline hematological parameters and those determined upon admission for COVID-19 infection showed variable changes. There was a slight increase in white blood cell and platelet counts during coronavirus infection. However, there was a decrease in the mean levels of hemoglobin and red blood cells. The average values will be summarized in Table 2.

Therapeutically, all patients with hypoxia received oxygen therapy to maintain saturation levels $\geq 95\%$. Three patients were transfused with packed red blood cells due to deoxygenation. Six (6/11) patients experienced vaso-occlusive crises associated with COVID-19 and received analgesics based on the intensity of pain, in addition to 200 mg of chloroquine phosphate per day for 10 days and 500 mg of azithromycin on the first day, followed by 250 mg from the 2nd to 4th day, following national guidelines for the management of coronavirus disease. Three patients received thrombosis prophylaxis with enoxaparin 40 mg per day for three days during their hospitalization. The average length of hospital stay was 6.2 days. All patients had a favorable outcome (Table 3).

Patients	Cough	Fever*	Hypoxia*	Dysgeusia*	Anosmia	Dyspnea	Rhinorrhea	Thoracic imaging
1	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes
2	No	No	Yes	No	No	No	No	No
3	No	No	Yes	Yes	Yes	No	Yes	No
4	No	No	Yes	No	Yes	No	Yes	No
5	Yes	No	Yes	Yes	Yes	Yes	Yes	Yes
6	No	No	No	Yes	Yes	No	Yes	No
7	No	No	No	No	No	No	No	No
8	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes
9	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes
10	Yes	No	Yes	Yes	Yes	Yes	Yes	Yes
11	Yes	No	Yes	Yes	Yes	Yes	No	Yes

Note:*Hypoxia: $\text{saO}_2 < 95\%$; *Ageusia: loss of taste; *Fever: Temperature $\geq 38^\circ\text{C}$

Table 1: Clinical and radiological features in patients with COVID 19 infection on admission.

Constants	Values before COVID-19	Values during COVID-19 infection	P-value
Erythrocytes (millions/mm ³)	3.38 \pm 0.97	3.27 \pm 0.88	0.77
Hemoglobin (g/dl)	11.09 \pm 1.87	10.89 \pm 2.20	0.58
Leucocytes (/mm ³)	8.86 \pm 2.39	12.04 \pm 4.11	0.04
Platelets (G/L)	324.40 \pm 128.06	457.40 \pm 185.10	0.07

Table 2: Mean haematological parameters of patients before and during coronavirus infection.

Patients	Age	Sex	Phenotype	Complication	RT-PCR	TDM	Hb	GB	Day on treatment
1	34	M	S β	No	Yes	Yes	11	6.14	5
2	30	M	SS	No	Yes	No	9.4	12.3	5

3	32	F	SS	No	Yes	No	7.5	8.63	6
4	34	M	SS	No	Yes	No	10.5	11.9	6
5	59	F	Sβ	No	Yes	Yes	8.8	19.6	5
6	36	F	SC	No	Yes	No	12.1	13.6	6
7	31	F	SS	No	Yes	No	9.5	15.6	6
8	50	M	SC	No	No	Yes	13.2	11.2	6
9	34	M	SC	No	No	Yes	12.1	14.5	5
10	41	M	SC	No	Yes	Yes	14.8	6.9	6
11	66	M	SC	Yes	Yes	Yes	10.8	5.3	10

Table 3: Sociodemographic and biologic characteristics of hospitalized patients with COVID-19.

Discussion

The number of people affected by COVID-19 is increasing due to screening campaigns. Eleven (11) sickle cell patients monitored at the Center for Research and Control of Sickle Cell Disease tested positive for COVID-19, with 7 males and 4 females. Studies on non-sickle cell subjects have reported an association between age and the severity of COVID-19. The majority of studies conducted have shown that an age above 60 increases the risk of complications from COVID-19 [7]. The average age at diagnosis in our study population was 40 ± 12 years, ranging from 30 to 66 years. Another study reported an average age of 35.5 [8]. The sickle cell phenotypes observed were as follows: five SC (45.4%), four SS (36.37%), and two Sβ ± thalassemia (18.2%). Comparative analysis of the biological characteristics revealed a significant increase in leukocytes and a tendency for increased platelet levels during COVID-19 infection. The elevated mean platelet count observed in these patients with COVID-19 may pose a potential risk of thrombosis. Therefore, thromboprophylaxis would be desirable for infected sickle cell patients hospitalized for coronavirus. Among the sickle cell patients with COVID-19, none were enrolled in a transfusion program, which rules out the hypothesis of any transfusion-related transmission of the virus. Eight out of the eleven sickle cell patients (73%) did not have a fever. In our study, the absence of a specific and dominant symptomatology of COVID-19 infection in these sickle cell patients suggests a need for differential diagnosis [2,6]. The outcome was favorable for all these cases, as no deaths were recorded. Despite this finding, we recommend the implementation of hygiene measures in this population and better organization of care. Although sickle cell disease is associated with a high risk of bacterial and viral infections, we were unable to demonstrate any severity of COVID-19 infection in sickle cell patients monitored at the CRLD in Bamako. A recent study conducted in Bahrain also found no increased severity of COVID-19 infection in sickle cell patients compared to non-sickle cell patients [8].

Conclusion

A lot remains mystery regarding the actual impact of COVID-19 infection on the symptomatology of SCD. The limited sample size of our observation does not allow for definitive conclusions to be drawn, however, we can note that fever is not a systematic symptom that

prompts further investigation of potential clinical signs (such as loss of taste), and considering the greater vulnerability of these patients, protective measures should be proposed as soon as the condition is suspected.

Notably, fever does not consistently emerge as a symptom that triggers further investigation into potential clinical signs, such as loss of taste. Given the heightened vulnerability of SCD patients, it is crucial to propose protective measures at the early suspicion of the condition. Despite the uncertainties, acknowledging these observations underscores the need for cautious consideration in managing COVID-19 in individuals with SCD.

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