Case Report ISSN (O):2395-2822; ISSN (P):2395-2814

Covid-19 in a Known 20- Year- Old Sickle Cell Disease Patient

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Received: May 2020
Accepted: May 2020

ABSTRACT

Though experts are predicting that sickle cell disease patients who usually develop complications during episodes of viral infections would have severe COVID-19 infection, we report a case of a 20-yr-old female, who is a known sickle cell disease patient who presented with a mild COVID-19 infection. A combination of high lymphocyte and low neutrophil counts as seen in this patient may explain the mild course of COVID-19 that was observed. Sickle cell disease patients in steady- state with high lymphocyte counts may not be at risk of severe infection with COVID -19 as experts are predicting.

Keywords: COVID-19, Sickle Cell Anemia, high Lymphocyte count.

INTRODUCTION

Coronaviruses are enveloped RNA viruses that cause respiratory illnesses of varying severity from the common cold to fatal pneumonia .The world is battling with COVID-19, a global pandemic caused by the novel coronavirus known as SARS-COV2. The disease was first identified in December 2019 in Wuhan, the capital of China's Hubei province, and has since spread globally. The disease was declared a pandemic by the World Health Organization on March 11 2020.[1] While the majority of cases result in mild symptoms such as fever, sore throat, cough and diarrhea, a severe form of the disease presents as viral pneumonia that can lead to respiratory failure. Severe disease is usually seen in patients with underlying comorbidities such as heart disease, diabetes, and hypertension.

Sickle cell disease (SCD) and its variants are genetic disorders resulting from the presence of a mutated form of hemoglobin, leading to abnormal red blood cells that get stuck to small blood vessels and impede oxygen delivery to tissues.[2] This change leads to "sickle cell crisis" (thrombosis, hyperhemolysis, aplastic anemia, sequestration) that can be precipitated by temperature changes, stress, dehydration, and high altitude. Viral infections in particular precipitate the sickling phenomenon, leading to acute chest syndrome, which is one of the leading causes of death in SCD.[3,4]

There is limited data on COVID-19 and SCD; however, there are speculations that since COVID-19 presents with severe respiratory disease in people with underlying comorbidities, SCD patients with COVID-19 will present with severe disease characterized by frequent and severe sickle cell crisis and acute chest syndrome.

We present the case of COVID-19 in a 20- year- old known SCD patient.

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MATERIALS AND METHODS

A 20 year old female, known sickle cell disease patient who was on her standard of care medications (hydroxyurea, palludrine and folic acid). After returning from a trip to Dubai where she was being worked up for hip replacement, she satisfied the case definition of a suspected case of COVID-19 based on the case definition by the Nigerian Center for Disease Control (NCDC) and tested positive for COVID-19.[5] Though asymptomatic at the time of diagnosis, with her background comorbid condition of sickle cell disease, she was quickly moved into the isolation center of the University of Abuja Teaching Hospital (UATH) Gwagwalada, Abuja, Nigeria for active management.

On arrival at the isolation center, she had no symptoms of COVID-19. The temperature was 36° C, respiratory rate was 18 cycles per minute, pulse rate was 95 beats per minute, blood pressure was normal, and the peripheral oxygen saturation (SpO2) was 99%.

Chest examination and other systemic examinations were essentially normal. Her complete blood count (CBC) showed a packed cell volume of 31%, total white blood count (WBC) was 9.8 x 109/L with a neutrophil count of 36.2% (normal range 40-75%) and lymphocyte count of 52.1% (normal range 25-45%). The electrolytes, urea and creatinine were normal. The Chest X-Ray showed cardiomegaly but the lung fields were clear.

She commenced oral medications; Lopinavir boosted with Ritonavir, Azithromycin, Amoxicillin/ clavulanic acid, Chloroquine, Vitamin C and Paracetamol according to the treatment protocol of

UATH for management of COVID-19. She continued her routine medications for the sickle cell disease and liberal fluid intake.

She remained stable all through the period of admission in the isolation center with peripheral Oxygen saturation (SpO2) ranging from 97% to 99%. She did not manifest any form of sickle cell crisis and did not develop any complication of COVID-19 as anticipated while on admission. She tested negative on two separate occasions to coronavirus consecutively by reverse transcriptase-PCR (RT-PCR) conducted 48 hours apart and after 16 days of admission was discharged home according to the National protocol.

DISCUSSION

Sickle cell disease (SCD) is a collective term for several genetic disorders in which hemoglobin is structurally abnormal, leading to abnormal red blood cells that get stuck to small blood vessels and impede oxygen delivery to tissues. SCD leads to a wide range of clinical manifestations and complications. The disease affects about 25 million people worldwide; the majority of whom are from Africa or of African descent.[4]

Infection has long been recognized as one of the most common precipitants of crisis in SCD. Leucocyte adhesion is the initiating event in a painful crisis, also known as vaso-oclusive crisis.[6] Steady- state neutrophil count correlates with the severity of SCD, and treatment with hydroxyurea, which lowers neutrophil count, reduces the frequency of crisis, painful episodes, and hospital admissions.[7] During an infection, the level of circulating neutrophils increases, predisposing to the painful crisis. Our patient had a low neutrophil count of 36.2% (normal range 40-75%).

Respiratory viruses such as Respiratory Syncytial Virus (RSV) and Influenza viruses are known to cause severe complications such as Acute Chest Syndrome (ACS) in people with sickle cell disease resulting in frequent hospitalization.[8] The American Academy of Pediatrics (AAP) and Advisory Committee on Immunization Practices (ACIP) recommends all children with sickle cell anemia receive influenza vaccine because of the increased risk of hospitalization and severe infection.[9,10] During the 2009 novel influenza A (H1N1) pandemic, children with sickle cell disease had higher hospitalization rates compared to the general population. They experienced more complications such as the acute chest syndrome, acute marrow suppression of red cell production, pain crisis, and hematuria.[11-13]

Though COVID-19 causes mild illness in many, fatal complications have been seen among the old and medically vulnerable such as those with diabetes, hypertension, heart disease, chronic obstructive pulmonary disease.[14] Though it's early and data is limited, experts are predicting that sickle cell disease patients could be at increased risk from COVID-19 with severe complications. We present a case of COVID-19 in a known sickle cell disease patient who had mild disease. The patient was crisis-free throughout hospitalization and did not develop any of the recognized complications associated with COVID-19.

Contrary to what is seen in other viral infections, COVID-19 is associated with lymphopenia.[15] Some studies have shown that it is an indicator of the severity of disease among hospitalized patients with COVID-19. Li Tan et al. in China reported that patients with lower lymphocyte count had a more severe disease.[16] They postulated that the virus might infect lymphocytes directly, resulting in lymphocyte death. Sickle cell disease patients in steady state have been shown to have higher lymphocyte counts than those with haemoglobin phenotypes AA.[17,18] Our patient had a lymphocyte count of 52.1% (normal range 25-45%), which is higher than the normal range for our population.

A combination of high lymphocyte and low neutrophil counts as seen in this patient may explain the mild course of COVID-19 that was observed. Sickle cell disease patients in steady- state with high lymphocyte counts may not be at risk of severe infection with COVID -19 as experts are predicting. However, more data is needed to arrive at this conclusion.

CONCLUSION

COVID-19 caused by the novel coronavirus has been shown to cause severe disease in the elderly and those with underlying health challenges. Experts are predicting that sickle cell disease patients who usually develop complications during episodes of viral infections would have severe COVID-19 infection. However, this may not be the case for sickle cell disease patients on hydroxyurea, who have a high steady- state lymphocyte count before contracting the virus.

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How to cite this article: Kwaghe V, Habib Z, Akor A, Yunusa T, Ekele B. Covid-19 in a Known 20- Year- Old Sickle Cell Disease Patient. Ann. Int. Med. Den. Res. 2020; 6(4):ME10-ME12.

Source of Support: Nil, **Conflict of Interest:** None declared