Successful Management of Sickle Cell Disease Patient with Covid-19 Infection in a Low Resource Setting: Case Study

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Authors’ contributions

This work was carried out in collaboration among all authors. Author AK designed the study, performed the statistical analysis, wrote the protocol and wrote the first draft of the manuscript. Authors ECB and AE managed the analyses of the study. Authors EO and BGP managed the literature searches. All authors read and approved the final manuscript.

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ABSTRACT

Background: Sickle cell disease (SCD) patients have greater susceptibility to infections, they are reckoned to be vulnerable patients during the current COVID-19 pandemic. SCD patients are commonly affected by pulmonary complications such as acute chest syndrome (ACS), pulmonary embolism (PE) and pneumonia that contribute significantly to mortality risks.

Aim: The study was aimed at showing the impact of SARS-COV viral pandemic on SCD patients.

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**Presentation of Case:** A 42-year-old male known sickle cell disease patient, who presented with a 5 days' history of chest pain and difficulty in breathing with a pain score of 8/10. Pain was said to be localized and, subside on the ingestion of analgesics (Tab DF118/60mg and PCM 1000mg) with no known aggravating factor, but there was associated history of difficulty in breathing. The patient was being managed as a case of vaso-occlusive crises R/O acute chest syndrome, and was commenced on adequate hydration, oxygen saturation was between 95-85%. On examination, respiratory rate was 20 cycles per minute, pulse rate – 96 beats/minute, BP and chest examination were essentially normal. CBC showed the Packed Cell Volume of 31%, White Blood Cells 15.04 x 10^9/L, Neutrophils 7.51x10^9/µL Lymphocyte 6.50 x10^9/µL, Monocyte 0.76 x10^9/µL Eosinophils 0.20x10^9/µL, Basophils 0.05x10^9/µL, Platelet 358. The electrolytes (Na-135 mmol/L, K 3.5mmol/L, HCO₃⁻20), urea -10 mmol/L and creatinine (88mmol/L) were normal, the chest x-ray showed cardiomegaly but the lung fields were clear. The patient was administered ceftriaxone (prophylactic antibiotics – 1 g daily). The patient tested positive to COVID-19 and was immediately transferred to the isolation centre for proper management. He was commenced on oral medication, azithromycin, dexamethasone, ivermectin, amoxicillin/clavulanic acid, vitamin C, clexane and the analgesic was changed to paracetamol and dihydrocodeine to alternate 3 hourly with accordance to the national guidelines. In addition, he was administered subcutaneous enoxaparin due to the hypercoagulability state of SCD. The patient’s health status improved within 24hours of commencement of the above medications and remained stable all through the period of isolation and a repeat covid-19 test was done 15 days of admission using and reverse transcriptase PCR and was discharged home according to the National protocol.

**Conclusion:** Studies and clinical trials are essential to evaluate effective diagnostic and management options for SCD patients and other high-risk conditions like diabetes hypertension, cancer patients and so on that are associated with fatal complications when infected with COVID-19 and similar diseases.

**Keywords:** Sickle cell; Covid-19 infection; pandemic; high-risk; effectiveness.

1. INTRODUCTION

The recent pandemic of COVID-19 infection (caused by the novel zoonotic SARS-COV-2 coronavirus) [1] that is ravaging the globe is a serious health challenge. Since its emergence at Wuhan, China, the virus has rapidly evolved the entire world due to its momentum, mechanism of transmission, which has not been completely elucidated by the World Health Organization.

Sickle cell disease (SCD) is a heterogeneous group of chronic autosomal recessive disorder [2]. SCD is the commonest genetic disorder in sub-Saharan Africa with Nigeria bearing the highest burden of about 1-2% of her population. SCD is characterized by the substitution of hydrophilic glutamic acid with a less polar hydrophobic valine residue at the position 6 of the β-globin chain. During hypoxic condition there is intra-erythrocytic hydrophobic interaction of the affected haemoglobin causing precipitation and eventually polymer formation that get stuck to small blood vessels and impaired oxygen delivery to tissues. These changes predispose to vaso-occlusive crisis, hyperhaemolysis, hypercoagulability/thrombosis, aplastic anaemia sequestration crisis. Viral infections have been implicated as a precipitator of sickling leading to acute chest syndrome which is fatal [3,4]. COVID-19 is a viral infection with severe respiratory distress with increase thrombosis [5] making a SCD patient at a high risk due to its prothrombotic, pro-inflammatory, immune-compromised state. There is paucity of information on COVID-19 infection and SCD in our environment.

2. CASE PRESENTATION

A 42-year-old male known sickle cell disease patient presented with a 5 days' history of chest pain and difficulty in breathing. Chest pain was dull in nature, so severe that it distorted patient's daily activities, with a pain score of 8/10 (using numerical pain score). Pain is said to be localized and, subside on the ingestion of analgesics (Tab DF118/60 mg and PCM 1000 mg) with no known aggravating factor, there is no history of cough, fever but there was associated history of difficulty in breathing which was said to occur even at rest and was so severe that itdistorts patient sleep, on account of the above symptom. Patient presented to the Haematology Day Care Unit of the University of Calabar Teaching Hospital (UCTH). At UCTH, the patient...
was being managed as a case of vaso-occlusive crises R/O acute chest syndrome, and was commenced on adequate hydration, oxygen saturation was between 95-85%. Respiratory rate was about 20 cycles per minute and pulse rate was 96 beats per minute, blood pressure was normal, chest examination was also essentially normal. He was also commenced on analgesic (rectal diclofenac to alternate with paracetamol 1000 mg 3 hourly). A complete blood count [CBC], peripheral blood film, liver function and electrolytes laboratory investigations were requested. CBC showed the Packed Cell Volume of 31%, White Blood Cells 15.04 x 10^9/L, Neutrophils 7.51x10^3/µL Lymphocyte 6.50 x10^3/µL, Monocyte 0.76 x10^3/µL Eosinophils 0.20 x10^3/µL, Basophils 0.05 x10^3/µL, Platelet 358, the electrolytes (Na^+ 135 mmol/L, K^+ 3.5 mmol/L, HCO^-3 20), urea -10 mmol/L and creatinine (88mmol/L) were normal, the chest x-ray showed cardiomegaly but the lung fields were clear. Patient was also commenced on prophylactic antibiotics ceftriaxone 1 g daily. Despite all these medications, patient’s condition was not improving, chest pain was getting worse and on account of this, patient was asked to do a COVID-19 test which came out positive and was immediately referred to the isolation center after counselling and informed consent.

![Fig. 1. Chest x-ray of the patient](image)

He was commenced on oral medication, azithromycin, dexamethasone, ivermectine, amoxicillin/ clavulanic acid, vitamin C, clexane and the analgesic was changed to paracetamol and dihydrocodeine to alternate 3 hourly. This is the National treatment protocol except for the subcutaneous enoxaparin which was added due to the hyper-coagulability state of SCD. He was also encouraged to take fluid liberally. He was said to had improved within 24 hours of commencement of the above medication and has remain stable all through the period of isolation to date and had tested negative to corona virus using the reverse transcriptase polymerase chain reaction (RT-PCR) conducted on two occasions 48 hours apart after 15 days of admission and was discharged home according to the National protocol.

3. DISCUSSION

This is a case report of COVID-19 infection in a known SCD patient who was initially being managed as a case of vaso-occlusive crisis to rule out acute chest syndrome, which is an acute complication of SCD, it is characterized by chest pain and difficulty in breathing, which is similar to the presentation of our index patient. This is because both condition do have similar symptoms. More so, COVID-19 infection can precipitate pneumonia of variable severity which can mimics acute chest syndrome and other crisis in SCD patient making the diagnosis difficult [6]. Acute chest syndrome is a complex condition often caused by atypical bacterial or less commonly virus. This is because other respiratory virus and influenza viruses are known causes of acute-chest syndrome in SCD patient [7]. There have been several reports on the relationship between influenza viral infection and acute chest syndrome among SCD, it is likely that COVID-19, might cause a similar condition if not possible a more deleterious pulmonary conditions. United Kingdom has also reported an increase mortality in Europe with a much higher level in the Black, Asian and minority ethnic [8], similar finding was also reported in south Asia [8]. Similarly, Beerkken et al. reported a case of acute chest syndrome precipitated by COVID-19 in a SCD patient [9]. Furthermore, Nur et al. reported a case of COVID-19 in SCD patient with vaso-occlussive crisis [10]. SCD is one of those vulnerable medical conditions due to its immunocompromised state, though there are limited data on SCD and COVID-19 in our environment.

COVID 19 is associated with lymphopenia which is contrary to what is seen in other viral infection [11]. Few studies have shown the correlation between lymphopenia and severity of COVID-19 [12]. This is due to the postulation that the coronavirus directly infects the lymphocyte resulting in their death. COVID-19 infection is a prothrombotic condition and characterized by widespread inflammation [13]. A recent
retrospective cohort study has shown that approximately 23% of patient with severe COVID-19 had pulmonary embolism (PE). This could exacerbate the already hypercoagulability and chronic inflammation found in SCD, whose chances of developing, PE is as high as 3.5% [14], thereby predisposing them to a greater risk of developing PE. This was the rationale for the early commencement of low molecular weight heparin. Moreover, low resource countries like ours which lack the technical and treatment availability can render it a challenge in management and distinguishing other pulmonary complication in SCD patient amidst this pandemic. The rate of COVID-19 infection is increasing, with most clime experience a spike during second wave of the pandemic, it is therefore pertinent to evaluate appropriate diagnostic and management strategy in most low resource countries because most account for a higher burden of people living with sickle cell disease. Also further research should be encouraged in understanding the various mechanism and severity of COVID-19 and SCD.

4. CONCLUSION

SCD is a high risk medical condition like diabetes, malignancies, hypertension that are associated with fatal complications when infected with COVID-19 and symptoms are similar. Therefore, a high index of suspicion is needed amidst this pandemic for prompt intervention to avoid morbidity and mortality from COVID-19.

CONSENT

As per international standard or university standard, patients' written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

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COMPETING INTERESTS

Authors have declared that no competing interests exist.

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