

more effective diagnosis of patients with suspected CDA and congenital hemolytic anemia.

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SICKLE CELL DISEASE

PP 74

How to treat and manage covid19 in SCD patients

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Objective: Coronavirus disease 2019 (COVID-19) is an infectious disease caused by severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2). It was first identified in December 2019 in Wuhan, China, and has resulted in an ongoing pandemic.

Case report: A 24-year-old man with a history of SCD (HbS/β0-thalassemia) on maintenance hydroxyurea therapy presented to our hospital, with a complaint of pain in the extremities and chest over two days. The patient with mild cough and high fever was hospitalized. Blood tests and lung CT were performed. Result of blood test show evidence of systemic hemolysis with a decrease in hemoglobin from 8.9 g/dL to 6.7 g/dL. His white blood cell count was $25.2 \times 10^3/\mu\text{L}$, CRP 243.21 mg/L. CT scans of the lungs showed a consolidated area where air bronchograms were observed in and around the medial segment of the middle part of the right lung and the posterobasal segment of the lower part of both lungs, and an icy glass landscape was observed. Lung damage is 1–5% (grade I). His oxygen saturation SpO₂ was normal (98%). The SARS-CoV-2 PCR nasopharyngeal swab testing was sent and returned negative on hospital day one after which the patient was started on antiviral and antibiotic for severe COVID-19 pneumonia. An improvement in blood counts was observed 4 days after starting treatment (WBC $16.93 \times 10^3/\mu\text{L}$, CRP 100.31 mg/L). On day ten, after normalization of all symptoms and blood values the patient was discharged home.

Methodology: In this study we selected 1 patient with SCD followed in Thalassemia Center of Azerbaijan.

Results: Given the higher likelihood of ACS it is possible that SCD patients are also at higher risk of such complications from COVID-19, particularly those with a history of pulmonary comorbidities. However, it is unclear if the SARS-CoV-2 pandemic will lead to increased rates of ACS for sickle cell patients. Still, hospitalized sickle cell patients should be monitored closely for development of ACS and if this occurs, exchange transfusion should be promptly initiated.

Conclusion: COVID-19 pneumonia as a cause of acute chest syndrome in an adult sickle cell patient. Patients with sickle cell disease (SCD) who are infected with COVID-19 may have a significant risk of developing acute chest syndrome (ACS), a potentially life-threatening complication. In this case we will present how manage COVID 19 in patient with SCD.

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STEM CELL TRANSPLANTATION

PP 75

High-dose methyl prednisolone in veno-occlusive disease

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Objective: Veno-occlusive disease (VOD) is a serious complication of hematopoietic stem cell transplantation (HSCT). If it is not identified and treated earlier, mortality is high. Combination usage of high-dose methyl prednisolone (MPZ) and defibrotide in VOD treatment have been described in some studies. Here, we present a patient with VOD who responded well to high-dose MPZ.

Case report: 14-month-old girl, diagnosed with thalassemia major, received HSCT from her sibling donor with busulfan and cyclo-phosphamide conditioning. On day +11, the patient experienced painful hepatomegaly and elevated total bilirubin (2.25 mg/dL) with 7% weight gain from baseline and respiratory distress while under defibrotide prophylaxis. VOD was diagnosed according to the modified Seattle criteria. Fluid and salt restriction were performed, spironolactone was started, and defibrotide was continued. Due to lack of significant improvement in the patient condition after 4 days of defibrotide, HDM was started at dose of 250 mg/m² per dose every 12 h on day +15.

Methodology: A day after MPZ, the patient's condition started to improve. After six doses of methylprednisolone, the dose was reduced to 2 mg/kg. Then, the dose was reduced by decreasing to half-dose in three-day periods. The defibrotide was discontinued on day +36, and the patient was discharged on day +45. The patient is currently being followed problem-free after 2 years of transplantation with 100% donor chimerism.

Results: VOD treatment response with high-dose MPZ and defibrotide combination can be better than treatment response with defibrotide alone. The easier and cheaper supply of steroids also prevents the treatment delay. In a study, it was shown that receiving high-dose MPZ without defibrotide was also found to be effective in the VOD treatment. The mortality rate in patients with multiple organ failure symptoms in VOD is between 50% and 100%. However, mortality rate can be decreased by early detection of VOD symptoms such as of painful hepatomegaly, weight gain and ascites. This findings may develop before hyperbilirubinemia especially in pediatric patients. Knowing this is important for early diagnosis and treatment of VOD.

Conclusion: As a conclusion; high-dose MPZ was found to be an effective treatment in VOD even at a dose of 250 mg/m² per dose every 12 h in our patient. High-dose MPZ might be an alternative treatment to defibrotide in early phase VOD. Further studies are needed on the efficacy and dosage of MPZ in VOD.

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