



Vaso-occlusive crisis and acute chest syndrome in sickle cell disease due to 2019 novel coronavirus disease (COVID-19)

Erfan Nur  | Aafke E. Gaartman  | Charlotte F. J. van Tuijn | Man Wai Tang | Bart J. Biemond

Department of Clinical Hematology, Amsterdam University Medical Centers, Amsterdam, The Netherlands

Correspondence

Erfan Nur, Department of Clinical Hematology, Amsterdam University Medical Centers, Amsterdam, The Netherlands.

Email: e.nur@amc.uva.nl

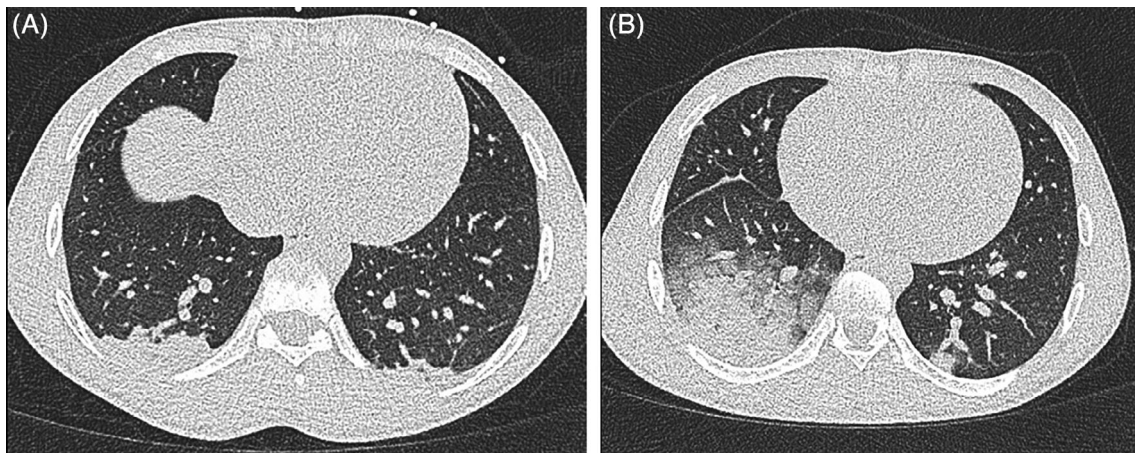


IMAGE 1 A, Chest CT imaging of patient 1 at first presentation to the emergency room (ER) showing infiltrates at basal fields. B, CT imaging at second ER presentation of patient 1, showing an increase in double-sided infiltrates

In March 2020, during the 2019 novel coronavirus disease (COVID-19) pandemic, caused by the newly emerged virus SARS-CoV-2, two patients with homozygous sickle cell disease (SCD) were admitted to our hospital with a painful vaso-occlusive crisis (VOC) triggered by COVID-19. Both patients had no flu-like complaints characteristic of COVID-19 during or preceding the VOC episode.

Patient 1, a 24-year-old man with a previous medical history of minor pain episodes without indication for hospitalization, presented with severe right thoracic pain for 3 days. At presentation he had a temperature of 37.6°C; pulse of 76/minute, blood pressure of 106/65 mmHg; respiration rate of 18/min and a peripheral oxygen saturation (SpO₂) of 97%. A non-contrast chest CT showed double-sided infiltrates without ground-glass opacities or crazy paving and

was not characteristic of COVID-19 (Image 1A). Throat and nose swabs were negative for SARS-CoV-2.

A diagnosis of VOC complicated by acute chest syndrome (ACS) was made. Treatment with oxygen, intravenous morphine with patient-controlled analgesia (PCA), fluid replacement therapy and amoxicillin/clavulanic acid was initiated. After 1 day, the level of pain had decreased significantly (numeric rating scale decreasing from nine to two) and the patient remained respiratory stable throughout his hospital stay. He was discharged with amoxicillin/clavulanic acid continued at home. However, the next day he returned to the emergency room with increased pain, dyspnea, respiration rate of 20/minutes, SpO₂ of 93% and a temperature of 38.9°C. Chest CT imaging showed progression of the double-sided infiltrates in the lower lobes of the lungs

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(Image 1B). A second PCR, this time of a sputum sample, was positive for SARS-CoV-2. He was treated with oxygen (up to 5 L/min) and morphine (PCA) while amoxicillin/clavulanic was continued. The patient had a smooth recovery, without a need for exchange transfusion. He was discharged again after 3 days and was instructed to stay at home (in isolation) until 24 hours after becoming completely symptom-free.

Patient 2, a 20-year-old woman with a history of frequent VOCs presented with severe pain of the back and extremities for 1 day. She had no respiratory or gastro-intestinal complaints. Because of a dip in SpO₂ to 88% after a 100 µg parenteral fentanyl bolus in the ambulance, the suspicion of COVID-19 rose which led to performing a chest CT and a throat and nose swab for a SARS-CoV-2 PCR. While the CT imaging did not show any pulmonary abnormalities, the PCR was positive for SARS-CoV-2. She remained hospitalized to treat her VOC without developing any respiratory symptoms.

The World Health Organization (WHO) recently declared SARS-CoV-2 infection a pandemic. Severe respiratory illness occurs in approximately 15%-20% of infected patients.¹ As of March 31, 2020, 800,049 laboratory-confirmed cases and 38,714 deaths have been documented globally.² In SCD, COVID-19 can potentially cause severe (pulmonary) complications, either by directly causing severe pneumonia or by triggering a VOC and/or ACS. While further experience regarding the clinical presentation of COVID-19 in SCD needs to be awaited, the following important points need to be taken into consideration based on the above described patients.

Similar to what we know from other viral infections,^{3,4} SARS-CoV-2 can also cause ACS in SCD. Furthermore, as can be seen in patient 1, an ACS can develop without the typical pulmonary complications that can be seen with COVID-19. Both patient 1 and especially patient 2 illustrate that COVID-19 might trigger a VOC without the presence of flu-like symptoms of COVID-19. With respect to diagnosis, the history of patient 1 illustrates the low sensitivity of the

PCR of the throat and nose swabs in the primary diagnosis of COVID-19, which is estimated to be around 70%. We therefore suggest to perform a second PCR, preferably on a sputum sample, and non-contrast chest CT imaging when there is no alternative explanation for VOC or when the clinical suspicion for COVID-19 remains high.

Based on these two patients, at our center we decided to include SARS-CoV-2 PCR in the evaluation of SCD patients presenting with VOC.

ORCID

Erfan Nur  <https://orcid.org/0000-0002-7069-930X>

Aafke E. Gaartman  <https://orcid.org/0000-0002-7534-5084>

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