2020

A pediatric patient with sickle cell disease presenting with severe anemia and splenic sequestration in the setting of COVID-19

S. Jacob
Northwell Health

A. Dworkin
Northwell Health

E. Romanos-Sirakis
Zucker School of Medicine at Hofstra/Northwell, eromanos@northwell.edu

Received: 29 May 2020 | Accepted: 1 June 2020

DOI: 10.1002/pbc.28511

Pediatric Blood & Cancer

SOCIÉTÉ INTERNATIONALE D'ONCOLOGIE PÉDIATRIQUE SIOP INTERNATIONAL SOCIETY OF PAEDIATRIC ONCOLOGY

aspho The American Society of Pediatric Hematology/Oncology

WILEY

LETTER TO THE EDITOR

A pediatric patient with sickle cell disease presenting with severe anemia and splenic sequestration in the setting of COVID-19

To the Editor:

Pediatric patients with sickle cell disease have varied clinical experiences. Splenic sequestration is a potentially life-threatening complication of sickle cell disease, estimated to have a prevalence of approximately 12%. Splenic sequestration occurs most commonly in the youngest pediatric patients, with a median age of 1.4 years. The majority of patients with splenic sequestration present with additional clinical symptoms, most commonly fever, and many with a concomitant infection.[1] The effects of COVID-19 on patients with sickle cell disease are not yet well described. We present a case of a pediatric patient with sickle cell disease who presented with severe anemia in the setting of splenic sequestration and COVID-19. We are not aware of any other published cases of COVID-19 presenting in this manner.

The patient is a 2-year 9-month-old male diagnosed with homozygous hemoglobin SS disease. He presented to his hematologist's outpatient clinic with irritability and an isolated fever of 102°F approximately 9 h prior to arrival. He received one dose of ibuprofen at home at the time of fever, with no return of fever prior to presentation to the clinic. Patient's review of symptoms was otherwise negative with no upper respiratory symptoms, no decrease in oral intake, no diarrhea, and no known sick contacts. The patient's medical history is significant for five prior hospitalizations during which he was transfused, but had no history of acute chest syndrome or splenic sequestration, with normal splenic ultrasounds documented on prior admissions. His medications included hydroxyurea, penicillin-VK, and folic acid.

In the clinic, the patient was afebrile, tachycardic to 152 beats per minute, with a blood pressure of 104/61 mmHg and no tachypnea or desaturations noted at room air. His physical exam was significant for irritability and a palpable spleen edge approximately 1.5 cm below the costal margin. The patient had no jaundice or scleral icterus. Initial blood work was notable for an undetectable hemoglobin level. He had total serum bilirubin 2 mg/dL, aspartate aminotransferase 72 U/L, alanine aminotransferase 10 U/L, LDH 815. His serum ferritin was elevated to 4655 ng/mL.

Blood culture was drawn and the patient was empirically treated with ceftriaxone. A rapid respiratory viral panel detected no common viral pathogens. Given the environment of the COVID-19 pandemic, a COVID-19 polymerase chain reaction test was sent that detected viral genome. A urinalysis showed no signs of a urinary tract infection and blood cultures were sterile. An ultrasound showed an enlarged spleen that measured 12 × 3.4 × 9.9 cm. A chest X-ray revealed mild cardiomegaly but no radiographic evidence of an acute pulmonary process.

He was transfused with multiple aliquots of packed red blood cells. The patient's irritability resolved after the initial aliquot of red blood cells and he returned to baseline activity. His fever did not recur and he did not manifest any other symptoms of COVID-19. Prior to discharge, the patient's anemia had improved to hemoglobin 10.3 g/dL and hematocrit 30.4% with reticulocytes 8.2%, and his thrombocytopenia improved to platelet count 109 000/μL from 59 000/μL. His serum ferritin had trended down to 867 ng/mL. On his physical exam, his splenomegaly had improved. He was hospitalized for 4 days, and he remained otherwise asymptomatic during hospitalization and for over 6 weeks posthospitalization.

Since the emergence of COVID-19 in December 2019, patients have been closely followed in an attempt to characterize and treat the clinical manifestations of this pandemic. The number of patients diagnosed with COVID-19 has increased significantly as pathogen detection campaigns have become more widespread. Most children diagnosed with the disease have been reported to present with mild symptoms; fever and cough are the most common clinical manifestations in children with COVID-19. These may be accompanied by symptoms of fatigue, myalgia, upper respiratory tract symptoms, sore throat, headache, dizziness, and gastrointestinal symptoms.[2] While severe complications occur less frequently in the pediatric population with COVID-19, severe morbidity and mortality has been documented in the pediatric population.[3]

Patients with sickle cell disease are prone to infectious complications, however, the effects of COVID-19 on this pediatric population are not yet well documented. Limited descriptions of pediatric patients with sickle cell disease presenting with acute chest syndrome and COVID-19 have been published,[4] however, there have been no documented cases of other complications of sickle cell disease in the setting of a COVID-19 infection in pediatric patients as of yet. In this case of COVID-19, the patient had one isolated fever that did not recur and had no apparent usual symptoms of a viral syndrome. While the patient presented with a severe presentation of splenic sequestration with an undetectable hemoglobin, he had a mild clinical course during hospitalization, with no additional apparent morbidity related to COVID-19. We must keep COVID-19 in the differential diagnosis even without

classic symptoms of the virus, and be prepared for a great variety of clinical presentations.

We document the first case of a pediatric patient with sickle cell anemia who presented in a splenic sequestration crisis in the setting of an infection with COVID-19. This report adds to the developing body of knowledge of the varied presentations of COVID-19 in pediatric patients, especially in those who are immunocompromised.

CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

Smrithy Jacob[1]
Aviva Dworkin[1]
Eleny Romanos-Sirakis[2]

[1] *Department of Pediatrics, Staten Island University Hospital Northwell Health, Staten Island, New York, New York*
[2] *Division of Pediatric Hematology/Oncology, Staten Island University Hospital Northwell Health, Staten Island, New York, New York*

Correspondence
Eleny Romanos-Sirakis, Pediatric Hematology/Oncology, Staten Island University Hospital, 475 Seaview Ave, Staten Island, New York 10305, NY.
Email: eromanos@northwell.edu

ORCID

Eleny Romanos-Sirakis https://orcid.org/0000-0002-5446-7445

REFERENCES

1. Brousse V, Elie C, Benkerrou M, et al. Acute splenic sequestration crisis in sickle cell disease: cohort study of 190 paediatric patients. *Br J Haematol*. 2012;156:643-648.
2. Jiatong S, Lanqin L, Wenjun L. COVID-19 epidemic: disease characteristics in children. *J Med Virol*. 2020. https://doi.org/10.1002/jmv.25807
3. Shekerdemian LS, Mahmood NR, Wolfe KK, et al. Characteristics and outcomes of children with coronavirus disease 2019 (COVID-19) infection admitted to US and Canadian pediatric intensive care units. *JAMA Pediatr*. 2020. https://doi.org/10.1001/jamapediatrics.2020.1948
4. Heilbronner C, Berteloot L, Tremolieres P, et al. Patients with sickle cell disease and suspected COVID-19 in a pediatric ICU. *Br J Haematol*. 2020. https://doi.org/10.1111/bjh.16802