Acute Chest Syndrome in Sickle Cell Disease in the Setting of Covid-19 Pneumonia

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Introduction

- Sickle cell disease (SCD) is an inherited condition characterized by abnormal B-globin genes in hemoglobin which results in abnormally sickle-shaped red blood cells (RBCs)[1]. A severe complication of SCD is acute chest syndrome, which is a major cause of morbidity and mortality in these patients.
- SARS-CoV2 infection demonstrates respiratory tropism associated with damaged endothelium and hypercoagulability[2].
- There is significant overlap in symptoms of ACS and Covid-19 pneumonia

Case Presentation

63 year old African American female with history of SCD not on maintenance medication and hypothyroidism brought back to ED for worsening SOB after positive Covid test. Patient unvaccinated

Upon presentation, patient was found to meet criteria for severe sepsis with respiratory distress and was admitted for oxygen support and administration of dexamethasone. Patient steadily improved over the first three days of admission.

Patient began to have severe full body pain consistent with acute sickle cell crisis. Patient has never had a crisis before. Patient started IV fluids and morphine for pain, which was escalated to dilaudid by day 5.

Early morning of hospital day 6, on-call resident was called to bedside for acute worsening of hypoxia with change in mental status. Oxygen support was escalated to BiPAP, and she was found to be newly anemic to 5.6, from 10.1. Intensivist was consulted and transfer to higher level of care for exchange transfusion was imitated.

Code blue was called due to respiratory distress and hypoxia on full ventilator support while pending blood transfusion. Second code blue was called for PEA/bradycardia and ROSC was briefly obtained before one more episode of PEA occurred. CPR was done for about 30 minutes without ROSC and patient subsequently expired.

Discussion

There are several documented cases of vaso-occlusive crisis and acute chest syndrome in patients with Covid-19 pneumonia, and these events are generally preceded by hemolytic anemia and painful crises, as seen in this patient.

The hyper-coagulable nature of Covid-19 likely contributed to the patient's presentation, along with the generalized inflammation associated with the condition[3]. Unfortunately, mainstay of Covid-19 treatment includes dexamethasone, which has also been shown to trigger rebound painful crises in these patients.

The patient presented with typical symptoms of worsening pneumonia as well as acute chest syndrome[3], and assessing pain was difficult due to patient's acute change in mental status.

Steroid usage is associated with painful rebound episodes of vaso-occlusive crisis, and risk of administration must be weighed against benefit in this patient population, with vigilant monitoring should usage of dexamethasone be elected.

Given the risk of rapid deterioration in clinical status with Covid-19 pneumonia as well as with acute chest syndrome, appropriate escalation of care and simple or exchange transfusion to reduce the ratio of sickled RBCs to normal RBCs is imperative.

Conclusion

- Patients with SCD and Covid-19 are likely at increased risk for ACS, and both conditions are associated with higher risk of hospitalization, higher risk of development of pneumonia, and higher risk of death.
- This patient was hydroxyurea naïve, and this likely made her more susceptible to acute chest syndrome[2] than patients who had maintenance therapy.
- Patients that have both SCD and Covid-19 should have close monitoring, both in hospital and outpatient. Prevention of Covid-19 and prevention of acute chest crises with maintenance hydroxyurea should also be implemented from the outset, and inpatients should be started on prophylactic anticoagulation with low threshold for escalation of care.

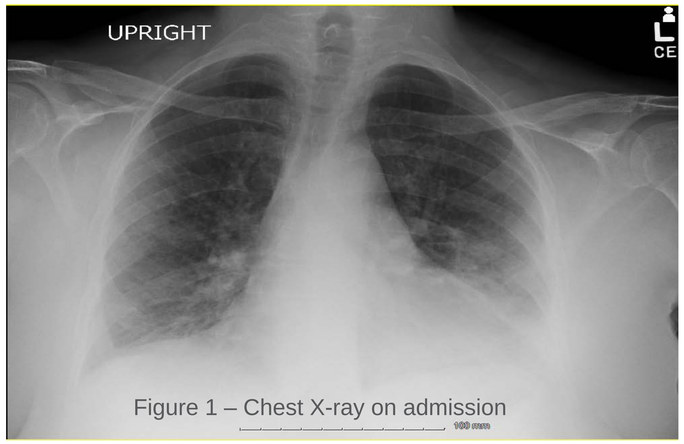

Figure 1 – Chest X-ray on admission

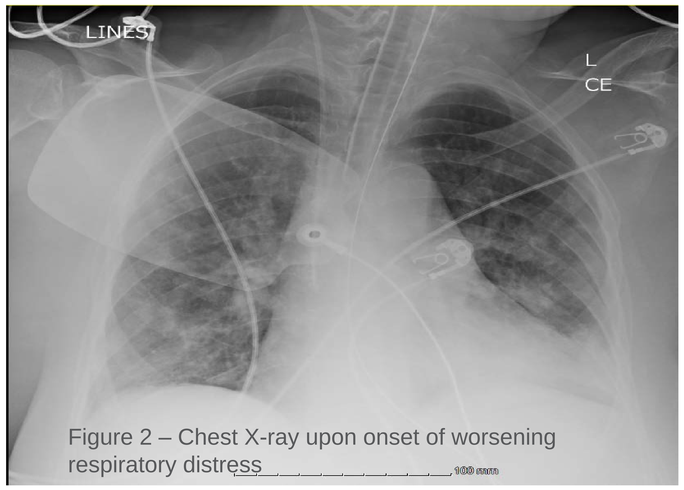

Figure 2 – Chest X-ray upon onset of worsening respiratory distress

References

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