Fat Embolism Syndrome as the Presenting Syndrome of Undiagnosed Sickle Cell Disease in a Middle-aged Adult

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Introduction

Fat embolism syndrome (FES) represents a myriad of clinical signs and symptoms that occur following the release of fat emboli into systemic circulation including hypoxemia, neurologic changes, petechial rash, and multi-organ dysfunction. FES often occurs post-orthopedic trauma but can be a complication of sickle cell disease (SS).[1]

Case Report

An otherwise healthy, 44-year-old female presented to our emergency department (ED) with worsening, atraumatic back and knee pain after an ED visit three days prior where x-rays and blood-work were unremarkable. She was short of breath, hypoxic, tachycardic and hypotensive. Her mental and respiratory status deteriorated requiring intubation. Laboratory work was significant for hemoglobin 8.5 g/dL, hematocrit 24.0%, WBC 14.8x10^9/L, and platelets 59x10^9/L including 11% bands and 15% nRBCs (baseline CBC had no abnormalities). A chest x-ray revealed bilateral, fluffy infiltrates consistent with pulmonary edema. Further blood-work revealed acute renal and liver failure, lactic acidosis, and troponinemia. CT imaging revealed patchy, widespread pulmonary disease and splenomegaly. She underwent resuscitation with fluid, blood products, vasopressor support, and broad spectrum antibiotics. All infectious disease workups were negative. The patient was ultimately transferred elsewhere for whole blood exchange but expired. Autopsy revealed vertebral and long-bone marrow infarction and evidence of FES. She was discovered to be hemoglobin SS post-mortem and no trigger for this episode, including infectious disease, was identified.

Discussion

Sickle cell disease screening is routine in the United States but less standardized in other nations.[2] Most sickle cell disease patients exhibit disease signs by age 4-5 months making her middle-aged presentation uncommon.[3] Our patient, a Haitian immigrant, as well as her husband, adamantly denied prior SS symptoms or crisis. Moreover, according to a recent systematic review, FES is a rare complication of sickle cell disease with a predilection for heterozygous patients.[4] Of all FES cases, only 15% of patients were homozygous SS (n = 13).[3] It is rare for FES to be the presenting syndrome of a previously undiagnosed, middle-aged, homozygous SS disease patient.

Conclusion

We present a rare case of FES as the presenting syndrome of a previously undiagnosed, middle-aged, SS disease patient. Prompt consideration and treatment of FES should be initiated to avoid complications.

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