The “Starfield” Pattern of Cerebral Fat Embolism From Bone Marrow Necrosis in Sickle Cell Crisis

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Abstract

Sickle cell disease may manifest with cerebrovascular and systemic complications. Sickle crisis that results in avascular necrosis of long bones with resultant cerebral fat embolism syndrome is rare and has a characteristic “starfield” pattern on MRI. This “starfield” MRI pattern should raise suspicion for sickle cell crisis in patients without a known history of the disease, which can lead to earlier sickle cell red blood cell exchange transfusion and treatment. We present a case of a male who presented emergently with acute seizure, coma with a characteristic MRI pattern, which lead to the diagnosis of avascular bone marrow necrosis and cerebral fat embolism syndrome from sickle cell crisis

Keywords

cerebral fat embolism syndrome, bone marrow necrosis, sickle cell disease, seizure, magnetic resonance imaging (MRI)

Case Summary

A 23-year-old man with sickle cell disease presented with a 2-day history of dehydration, progressive behavioral agitation, and generalized tonic-clonic seizures. Cerebrospinal fluid and infectious disease workups were unremarkable. His hemoglobin was 8.1 g/dL, and multiple sickle cells were seen on his peripheral blood smear. Hemoglobin S was elevated to 51% (normal: 0%). An emergent packed red blood cell exchange transfusion was initiated. Initial brain magnetic resonance imaging (MRI; Figure 1) showed multiple strokes. Cardiac, hereditary hypercoagulation profile, autoimmune, and vasculitis workups were unremarkable. Ten days later, he remained comatose without ongoing activity of seizure. A repeat brain MRI (Figure 2A and B) showed evolution of the diffuse microvascular pattern.

Cerebral fat embolism syndrome (FES) was suspected, and an MRI of his pelvis showed patchy bone marrow abnormalities (Figure 3). Formal bone marrow biopsy and aspirate showed extensive intramedullary necrosis (Figure 4). He remained in a persistent coma.

Discussion

Fat embolism syndrome typically presents with a pulmonary, central nervous system and cutaneous manifestation triad.¹ However, acute/subacute mental status changes and seizures without the classic FES triad are possible and could pose a diagnostic dilemma. Seizure can cause acidosis and hypoxia, which can paradoxically worsen the microvasculopathic

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Figure 1. Initial brain magnetic resonance imaging (MRI) reveal multiple, diffuse punctate, and patchy foci of restricted diffusion and gradient echo peppered throughout the white matter in the frontal, parietal, and occipital lobes, the posterior limb of the internal capsules, and the cerebellar white matter bilaterally. Postgadolinium images reveal multiple punctate foci of enhancement corresponding to the areas with restricted diffusion suggestive of subtle blood brain barrier breakdown, consistent with diffuse microvasculopathic infarctions. The MRI of the brain shows (A) numerous diffuse subcortical “starfield” pattern of diffusion-weighted images (DWIs), which correlate with apparent diffusion coefficient (ADC; B). C and D show the gradient echo (GRE) sequence and microhemorrhages throughout the cerebral and cerebellar regions.

Figure 2. Repeat magnetic resonance imaging (MRI) with restricted diffusion and T2/fluid-attenuated inversion recovery (FLAIR) hyperintensity throughout the bilateral caudate, putamen, and medial thalamic nuclei was consistent with a delayed anoxic–hypoxic injury that likely occurred during his initial generalized seizures. A, Diffusion-weighted image (DWI) showing restricted diffusion of bilateral caudate and (B) apparent diffusion coefficient (ADC) showing corresponding decrease in signal consistent with delayed anoxic brain injury.

Figure 3. Magnetic resonance imaging (MRI) pelvis—axial T1 postgadolinium images demonstrate heterogeneous hyperintensities of the femur with subtle enhancement (A), consistent with avascular necrosis (AVN).

Figure 4. Bone marrow aspirate ($\times 4$ magnification) shows (A) normal cellular bone marrow intermixed with extensive intramedullary necrosis (B).

crisis, causing delayed hypoxic–ischemic injury. With cerebral FES, brain MRI typically demonstrates multiple non-confluent areas with a “starfield” pattern.² Appearance of long bones on MRI can vary with different stages of bone infarction.³ This case demonstrates the need for a high degree of clinical suspicion, as early aggressive hydration and RBC exchange transfusions are fundamental to optimize outcomes.¹

Authors' Note

LPD contributed to writing of the article and collection of the patient data and radiographic records. KB contributed to bone marrow sampling and input regarding the pathology report. KMB contributed to critical revision of the article. WDF contributed to critical revision of the article. All authors had final approval of the article.

Declaration of Conflicting Interests

The authors declared the following potential conflicts of interest with respect to the research, authorship, and/or publication of this article. Dr Dhakal has nothing to disclose. Dr Bourgeois has nothing to disclose. Dr Barrett and Dr Freeman are associate editors for *The Neurohospitalist* and recuse themselves from this article's review process.

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References

1. Eriksson EA, Schultz SE, Cohle SD, Post KW. Cerebral fat embolism without intracardiac shunt: a novel presentation. *J Emerg Trauma Shock*. 2011;4(2):309-312.
2. Cox G, Tzioupis C, Calori GM, Green J, Seligson D, Giannoudis PV. Cerebral fat emboli: a trigger of post-operative delirium. *Injury*. 2011;42(suppl 4):S6-S10.
3. Jain R, Sawhney S, Rizvi SG. Acute bone crises in sickle cell disease: the T1 fat-saturated sequence in differentiation of acute bone infarcts from acute osteomyelitis. *Clin Radiol*. 2008;63(1):59-70.