Cerebral microhemorrhages and cerebral fat embolism in hemoglobin S-C disease

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Figure MRI of punctate ischemic lesions

MRI of brain showed numerous punctate ischemic lesions scattered throughout the white matter compatible with a starfield pattern on diffusion-weighted sequence (A, B). Susceptibility-weighted imaging sequence reveals countless microhemorrhages disseminated throughout cerebral cortex and subcortical white matter (C, D) consistent with cerebral fat embolisms.

A 40-year-old man with a history of ventricular septal defect (VSD) and hemoglobin S-C disease presented with progressively worsening headache, followed by sudden thunderclap headache and encephalopathy. The patient became comatose without any focal neurologic deficits and was intubated for airway protection. Chest X-ray was unremarkable.

MRI was performed and showed numerous punctate foci of ischemic lesions and countless microhemorrhages disseminated throughout the brain (figure). The results from lumbar puncture and EEG were unrevealing. The bone marrow biopsy revealed extensive necrosis. The diagnosis of fat embolism was made based on Gurd criteria and the patient was treated with red blood cell exchange transfusions twice suppressing hemoglobin S to <30%. He progressed to a vegetative state requiring tracheostomy and percutaneous endoscopic gastrostomy.

Bone marrow necrosis can occur in sickle cell disease (SCD) or sickle cell variants. Cerebral fat embolism is an uncommon condition and not well-understood because of its rarity. Due to the anatomy of trabecular bone, the release of infarcted necrotic bone marrow tissue into the vasculature is thought to be the source of access to the veins and the systemic circulation.^{1,2} Increased prevalence of right-to-left shunting in SCD and stroke may suggest that paradoxical embolism could be the mechanism.^{3,4} However, an incidental finding of VSD may not explain fat embolism without clear evidence of Eisenmenger syndrome. This case uniquely describes a devastating brain injury in the presence of ventricular septal defect in a patient with hemoglobin S-C disease.

Author contributions

Dr. Cho prepared the first draft. Drs. Ling and Rivera-Lara edited. Dr. Cho finalized the manuscript.

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