Videos and Images in Neuropediatrics

Starfield Pattern on Brain MRI in a Patient with Duchenne Muscular Dystrophy

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An 18-year-old man with Duchenne muscular dystrophy (DMD) presented with encephalopathy 20 minutes after falling from a sitting position. X-ray of his left femur showed a minimally displaced acute fracture. On initial examination, he was obtunded with minimal withdrawal to painful stimuli with a Glasgow coma scale (GCS) score of 9. Computed tomography (CT) of the brain was without acute intracranial pathology and electroencephalogram showed diffuse slowing. Magnetic resonance imaging (MRI) of the brain demonstrated innumerable punctate foci of T2 hyperintensity (¬Fig. 1A) with associated diffusion restriction (¬Fig. 1B, 1C) and increased susceptibility (¬Fig. 1D) in a "starfield pattern" characteristic of a diagnosis of cerebral fat embolism syndrome (FES).

FES classically presents as a triad of encephalopathy, respiratory failure, and rash, ¹ although not all three are required for diagnosis. This commonly follows a fracture where lipid microparticles are released and occlude microvasculature throughout the body. ² Fat microemboli to the brain lead to foci of cytotoxic edema, which appear as scattered areas of diffusion restriction ³ likened to a starfield. Patients with DMD are more susceptible to fractures even with minor trauma due to osteoporosis from reduced ambulation and chronic corticosteroid use. A high index of suspicion for FES should be maintained for patients with DMD presenting with altered mental status following trauma.

Conflict of Interest None declared.

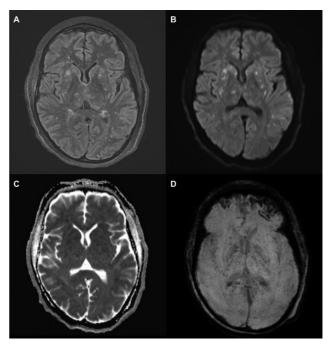


Fig. 1 (A) Magnetic resonance imaging (MRI) of the brain axial T2 fluid attenuated inversion recovery (FLAIR) sequence showing innumerable hyperintense punctate foci throughout the deep gray structures and cerebral white matter with corresponding diffusion restriction on (B) diffusion weighted imaging (DWI) and (C) apparent diffusion coefficient (ADC) sequences and increased susceptibility on the (D) susceptibility weighted imaging (SWI) sequence.

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