Teaching NeuroImage: Hypothalamic Involvement in Neuromyelitis Optica Spectrum Disorder in a Child

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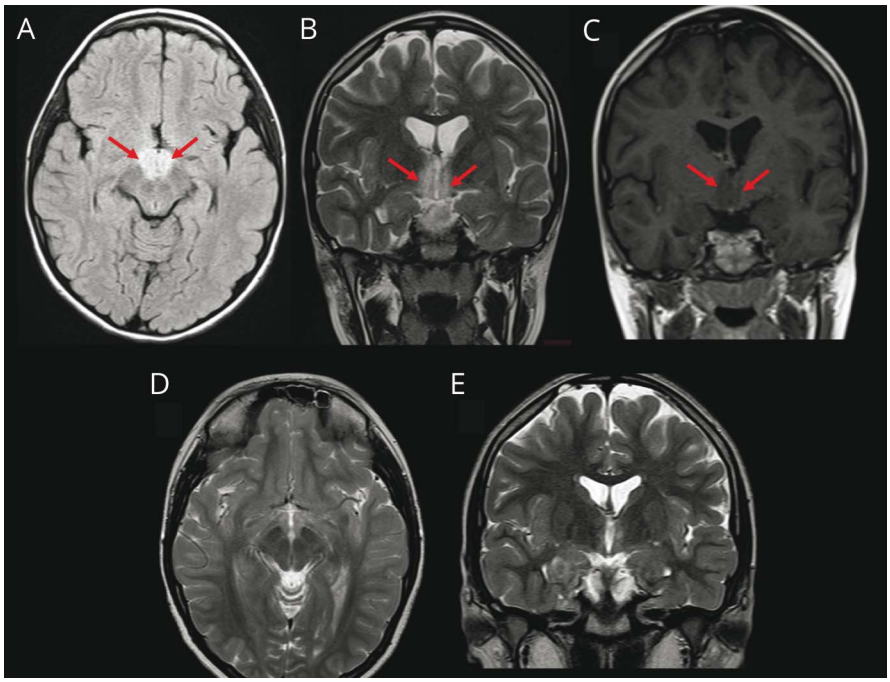
Figure Brain MRI



Brain MRI after 2 weeks of symptoms, (A) Axial fluid-attenuated inversion recovery imaging and (B) coronal T2-weighted image demonstrated a bilateral hypothalamic hyperintense nonenhancing (C) lesion (arrows). (D) and (E) T2-weighted images show resolution of the lesion 3 months after diagnosis and treatment.

A 10-year-old girl presented with a 15-day history of excessive daytime sleepiness and sudden sleep onset, hypnagogic hallucinations, hyporexia, and behavioral changes. An MRI examination of the brain revealed a bilateral hypothalamic lesion (Figure). We found positive AQP4 -IgG antibodies in serum and low hypocretin levels (93 pg/mL) in the CSF. A diagnosis of narcolepsy secondary to neuromyelitis optica spectrum disorder was made. She improved after glucocorticoid administration. After 10 months of immunosuppressive maintenance therapy with azathioprine, she remains asymptomatic without new lesions in the follow-up neuroimages. Any diencephalic clinical syndrome, such as narcolepsy, with hypothalamic involvement, should prompt a serum test for AQP4 -IgG.[1,2]

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