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

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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. Brain MRI revealed a bilateral hypothalamic lesion (Figure 1). We found positive AQP4-IgG antibodies in serum and low hypocretin levels (93 pg/mL) in cerebrospinal fluid. 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).

Teaching Slides - http://links.lww.com/WNL/C191
References


Figure. Brain MRI

Brain MRI after two weeks of symptoms, (A) Axial Fluid-Attenuated Inversion Recovery Imaging and (B) coronal T2-weighted image demonstrated a bilateral hypothalamic hyperintense non-enhancing (C) lesion (arrows). (D) and (E) T2-weighted images show resolution of the lesion three months after diagnosis and treatment.
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