Limbic Encephalitis in a Boy with N-Methyl-D-Aspartate Receptor Antibodies

A 6-year-old previously healthy boy had a 48-hour history of severe disorientation and bizarre behavior (agitated, fearful), preceded by a mild prodromal 10-day headache. Vital signs and neurologic examination were unremarkable. Results of preliminary laboratory investigations, including full blood count and smear, inflammatory markers, urea, electrolytes, virology, thyroid function, thyroid peroxidase antibodies, baseline metabolic screen, throat swab, urine culture, and urine toxicology, were all reported as normal. With an electroencephalogram, a slow background and a focal electrographic seizure was shown. The boy was treated with a loading dose of fosphenytoin, empiric ceftriaxone, and acyclovir.

With magnetic resonance imaging (day 2), bilateral symmetrical signal hyperintensity in the hippocampi consistent with limbic encephalitis was revealed (**Figure**). Autoimmune encephalitis was presumed, and the boy immediately was treated empirically with intravenous immunoglobulin (0.4 g/kg) for 5 days. During hospital admission, he exhibited ongoing memory loss, aggression, and mutism, and on day 13, he was further treated with pulsed methylprednisolone (30 mg/kg) for 3 days, followed by oral prednisolone for 4 weeks. The results of a cerebrospinal fluid infectious screen and oligoclonal bands were negative. N-methyl-D-asparate receptor antibodies were positive in serum and cerebrospinal fluid. Tumor search results were negative. The boy gradually improved, and at 9 months he is almost completely recovered with occasional mild deficits in attention. Repeat neuroimaging results (day 20) were normal.

The autoimmune encephalitides are an important group of treatable conditions. Neuropsychiatric presentations are common, but the spectrum of disease presentation and outcome in the pediatric population is still emerging. Limbic encephalits is often defined by the clinical features of limbic dysfunction, evidence of cerebrospinal fluid inflammation, and abnormalities in the limbic regions on electroencephalogram or magnetic resonance imaging.[1] Although rare, classic limbic encephalitis is increasingly recognized in children, associated with anti-N-methyl-D-aspartate receptor, anti-voltage-gated potassium channel, anti-glutamic acid decarboxylase, and paraneoplastic antibodies.[1,2] Early recognition and treatment of autoimmune encephalitis is associated with better outcomes.[1,2] ■

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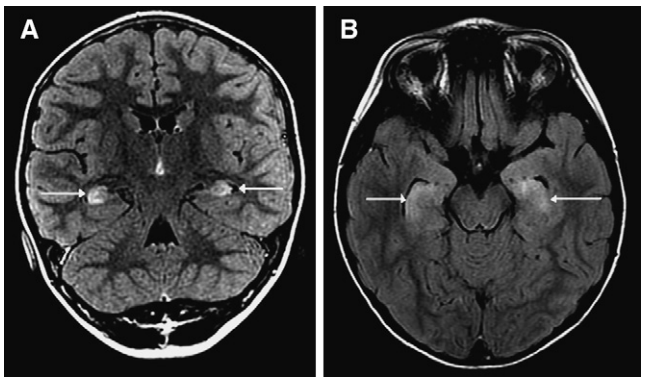

Figure. T2 fluid attenuation inversion recovery magnetic resonance imaging of the brain. **A,** Coronal image demonstrating bilateral symmetrical increased signal intensity in the region of the hippocampus. **B,** Axial image demonstrating increased signal in the mesiotemporal lobes bilaterally.

References

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2. McCoy B, Akiyama T, Widjaja E, Go C. Autoimmune limbic encephalitis as an emerging pediatric condition: case report and review of the literature. J Child Neurol 2011;26:218-22.

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