Childhood Anti-NMDA Receptor Encephalitis

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Abstract

Anti N-methyl-D-aspartate receptor (NMDR) encephalitis has been recognized as the most frequent autoimmune encephalitis in children after acute demyelinating encephalomyelitis (ADEM). However, due to the variable clinical presentation, the paucity of specific findings on standard laboratory and radiological investigations remains under recognized. First described in 2005, most commonly affected are children and young adults; 80% patients are females. It has characteristic evolution in several stages, in prodromal phase appears fever, malaise, nausea, vomiting, diarrhea, or upper respiratory tract symptoms in about 70%, then gradually manifest psychotic symptoms (delusions, hallucination and mania) that in children can be present as temper tantrum, hyperactivity, irritability, neurologic presentation as seizure, status epilepticus and dystonia. NMDA receptors are ligand cation channels involved in synaptic glutaminergic transmission that plays a key role in functions such as memory, learning, behavior and cognition. Anti-NMDA encephalitis is associated antibodies against the NRI1 subunit of NMDA receptors. MRI changes are non-specific; EEG changes are slowing and background activity; CSF abnormalities are common, mild lymphocytic pleocytosis, elevated protein and positive oligoclonal bands in 60% of patients. Definitive diagnosis is based on demonstration of anti-NMDA antibodies in CSF or serum. Management includes symptomatic therapy, definitive immunotherapy, and tumor surveillance. Early diagnosis and aggressive immunotherapy are important. In this presentation, I introduce a patient with anti-NMDA encephalitis.

Keywords: Childhood, Patients, Management

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