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ENCEPHALOPATHIES

AUTOIMMUNE ANTI-HU ANTIBODY LIMBIC ENCEPHALOPATHY

Investigators at the French Reference Center on Paraneoplastic Neurological Syndrome (PNS), Lyon Neuroscience Research Center, and other centers in France performed a retrospective study of children and adolescents with anti-Hu antibodies (Hu-Abs) collected by the Reference Center between Jan 1, 2000 and Dec 31, 2011. Of 251 patients with Hu-Abs, 8 were younger than 18 years. All of the 243 adult patients with Hu-Abs had PNS whereas only 2 of 8 (25%) children had neuroblastoma and opsoclonusmyoclonus. The majority (6 children; 5 female, 1 male) presented with limbic encephalitis manifested by progressive personality changes, memory loss, and seizure, and were free of cancer at a mean follow-up of 50 months. Brain MRI scans were abnormal in 4 of 6 patients and CSF showed oligoclonal bands in 4 patients. Treatment with AEDs and immunotherapy was only partially effective, and 5 children were cognitively impaired. The clinical, radiologic, and biological presentations were similar in this series of patients, suggesting a common neurologic syndrome. (Honnorat J, Didelot A, Karantoni E, et al. Autoimmune limbic encephalopathy and anti-Hu antibodies in children without cancer. Neurology 2013 Jun 11;80(24):2226-2232). (Response: Dr Honnorat. E-mail: Jerome.honnorat@chu-lyon.fr).

COMMENT. Hu-Abs are rarely reported in children, and are found primarily in patients with neuroblastoma and opsoclonus-myoclonus. In the above study, H-Abs are associated with dysimmune nonparaneoplastic limbic encephalitis. In the same time period, these authors identified 35 children with anti-NMDAR antibody encephalitis as compared to only 6 with HU-Abs limbic encephalitis. Hu-Abs should be tested in children with unexplained symptoms of limbic encephalitis.

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