

autosomal dominant, and mutations of the gene HLXB9 are found in 50% of patients. The diagnosis should be suspected in infants with recurrent or polymicrobial, especially gram negative, meningitis, persistent CSF pleocytosis, and a history of bowel abnormality.

PET SCAN AND AUTOIMMUNE FOCAL ENCEPHALITIS

The value of the PET scan in the diagnosis of autoimmune focal encephalitis is reported in a 22-month-old girl who presented with involuntary movements, hemiparesis, and behavioral changes at Juntendo University School of Medicine, Tokyo Metropolitan Institute for Neuroscience, Japan. On admission, she had low-grade fever, choreic movements in the right arm, and mild ataxia. MRI and EEG were unremarkable. CSF cell count was 19 per mm³ and protein 15 mg/dL; viral isolation was negative. Symptoms deteriorated and methylprednisolone therapy was ineffective. She developed hemiparesis, aphasia, agitation and temper outbursts. PET scan showed hypermetabolism in the left temporal lobe, caudate and lentiform nuclei, and prefrontal area. Autoimmune focal encephalitis was suspected and treatment with IV immunoglobulin begun. Behavior improved, phonation improved, and at 18 months after onset, hemiparesis and involuntary movements lessened. Repeat PET scan at 3 months after onset showed hypometabolism in the left temporal lobe. Aphasia and temper outbursts persisted, and her developmental quotient at 28 months was 54. Immunohistochemical analyses on sera showed immunoglobulin G autoantibodies that reacted specifically with cytoplasm of neurons in prefrontal and temporal lobes, globus pallidus, and putamen, corresponding with the neurologic symptoms. At 3 months after IVIG therapy, serum autoantibodies had disappeared and symptoms improved. Autoantibodies were important in the etiology of the focal encephalitis. (Sekigawa M, Okumura A, Nijima S-i, Hayashi M, Tanaka K, Shimizu T. Autoimmune focal encephalitis shows marked hypermetabolism on positron emission tomography. *J Pediatr* Jan 2010;156:158-160). (Reprints: Mariko Sekigawa MD. E-mail: m-seki@med-juntendo.ac.jp).

COMMENT. PET scan is considered a useful adjunct in the diagnosis of autoimmune focal encephalitis, when MRI is unremarkable. FDG-PET detects inflammatory changes with more sensitivity than SPECT, which reflects regional cerebral blood flow. Serum immunohistochemical analyses are required to detect specific autoantibodies.

Autoimmune limbic encephalitis is discussed by Jerome Honnorat, Universite Claude Bernard, Bron, France. (Honnorat J. *Lancet Neurol* Jan 2010;9:24-25). Limbic encephalitis is an inflammatory disorder affecting the hippocampi, amygdalae, and fronto-basal and insular regions. Originally considered rare, paraneoplastic, and unresponsive to treatment, a new subtype is described with autoantibodies against a Gaba subunit receptor, that is not always associated with cancer and may be treatable. (Lancaster E et al. *Lancet Neurol* Dec 3, 2009). The antibodies are directed against neuronal cell-surface antigens, and patients may improve with immunotherapy.

Paraneoplastic limbic encephalitis with antibodies against intracytoplasmic antigens is not susceptible to immunotherapy.

HUMAN HERPESVIRUS 6 INFECTION AND FEBRILE SEIZURES

Frequency and clinical characteristics of primary human herpesvirus 6 (HHV-6) infection in hospitalized children with febrile seizures (FS) were investigated at Aghia Sophia Children's Hospital and Laiko General Hospital, Athens, Greece. Of 130 children with FS admitted during the study period, 65 returned for follow-up and were included; 55 had a first FS and 10 the second. The mean age was 21.35 months; 36 were male and 29 female. Using PCR in acute phase plasma and indirect immunofluorescent assay for antibody titers in acute and convalescent sera, primary HHV-6 infection was verified in 10 (18%) with a first FS. Of 10 with primary HHV-6 infection, 6 had typical roseola, and 6 presented with respiratory or gastrointestinal symptoms. No child with a second episode of FS had primary HHV-6 infection. HHV-6 was type B in 8 children and type A in 2. HHV-6 DNA was not detected in 3 CSF samples examined. None of 85 control subjects had primary HHV-6 infection, but 49% had immunoglobulin G antibodies against the virus. Testing for HHV-6 infection is recommended as a routine laboratory test in young children admitted to the ED in Greece for first occurrence of a FS. (Laina I, Syriopoulou VP, Daikos GL et al. Febrile seizures and primary human herpesvirus 6 infection. *Pediatr Neurol* Jan 2010;42:28-31). (Respond: Dr Syriopoulou. E-mail: vsyriop@med.uoa.gr).

COMMENT. Primary HHV-6 infection is frequently associated with first febrile seizures (FS). HHV-6 infection is nearly universal in infancy or early childhood, and in the US it accounts for one third of all FS in children up to the age of 2 years. (Hall CB et al. *New Engl J Med* 1994;331:432-438). No increase or decreased incidence of recurrent FS occurs in children whose first FS is caused by HHV-6 vs other infections (Jee SH et al. *Pediatr Inf Dis J* 1998;17:43-48). The definitive role of HHV-6 infection in the etiology of FS is inconclusive, and multiple factors may be involved (Millichap JG, Millichap JJ. *Pediatr Neurol* 2006;35:165-172): 1) the height of the temperature (usually >39C); 2) encephalitis (mean frequency of virus detection in CSF is 14.5% of 138 samples in 9 studies of HHV-6 associated FS; range 0-100%); and 3) viral neurotropism (HHV-6 encephalopathic factor). Further studies of CSF HHV-6 DNA are required. Evidence suggests that the generally accepted benign nature of FS may need modification, and the definition of FS changed to include a transitory encephalitic process in some more complex cases.

Magnetoencephalography showed equivalent current dipoles as evidence of focal interictal epileptic discharges in 8 of 15 children following a febrile seizure (Anninos P et al. *J Child Neurol* Jan 2010;25:61-66). The authors propose the MEG as a prognostic indicator of development of epilepsy in FS patients.