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INFECTIOUS AND AUTOIMMUNE DISORDERS

MENINGITIS AS COMPLICATION OF CURRARINO SYNDROME

Two infants presenting within a period of weeks with gram negative meningitis and diagnosed with Currarino syndrome are reported from Wake Forest University, Winston-Salem, NC, and Eastern Virginia Medical School, Norfolk, VA. Case 1, a 5-month-old girl, had bacteremia 2 months before developing signs of *Escherichia coli* and *Proteus mirabilis* meningitis. MRI spine showed a sickle-shaped sacrum, an anterior sacral meningocele and gastrointestinal fistulous connection. Case 2, a 2-month-old girl, presented with recurrent *Pseudomonas aeruginosa* meningitis. MRI showed a sickle-shaped sacrum, anterior meningocele, and fistulous tract to the rectum. Both patients were treated with antibiotics and surgical repair. (Raczynski RA, Fisher RG, Sass LA. Unusual cases of meningitis as a clue to the diagnosis of Currarino syndrome. **Pediatr Infect Dis** Jan 2010;29:90).

COMMENT. The above letter was written in response to a Mayo Clinic case report of a 20-day-old female infant who presented with meningitis refractory to antibiotics, and was found to have a scimitar sacrum, recto-thecal fistula, anterior sacral meningocele, tethered spinal cord, anal stenosis, and neurogenic bladder. Sequencing of the causative gene for Currarino syndrome, *HLXB9*, on blood leukocytes was positive for a novel nonsense mutation. Family history was positive for the same mutation, mother had a history of Hirschprung's disease, and a cousin had a presacral teratoma. (Kiefer AS et al. **Pediatr Infect Dis** 2009;28(6):547-549).

A Currarino triad, anorectal malformation, sacrococcygeal defect, and a presacral mass, recognized as a caudal regression syndrome and neuroenteric malformation, is due to an anomaly of the notochord in early embryogenesis. The disorder is inherited as an

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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, Université 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.