MOVEMENT DISORDERS

NEURAL BASIS OF TICS: A FUNCTIONAL MRI STUDY

Event-related functional MRI (fMRI) was used to study the neural basis of spontaneous motor and vocal tics in 10 patients with Tourette syndrome, at the National Institute of Neurological Disorders and Stroke, Bethesda, MD. fMRI activities were analyzed 2 seconds before and at tic onset, by synchronized video/audio recordings. Before the onset of tics, activated paralimbic areas included anterior cingulate and insular cortex, supplementary motor area and parietal operculum. At tic onset, fMRI activity occurred in sensorimotor areas including suprior parietal lobules and cerebellum. The paralimbic and sensory association areas involved in tic generation are similar to those implicated in movements triggered internally by unpleasant or emotional sensations, such as pain and itching. (Bohlhalter S, Goldfine A, Matteson S et al. Neural correlates of tic generation in Tourette syndrome: an event-related functional MRI study. **Brain** August 2006;129:2029-2037). (Respond: Mark Hallett, Human Motor Control Section, National Institute of Neurological Disorders and Stroke (NINDS), NIH, Building 10, Room 5N226, 10 Center Drive, MSC-1430, Bethesda, MD 20892).

COMMENT. The authors postulate a limbic overdrive of the motor system and activation of sensorimotor areas underlying the pathophysiology of tic generation. The unpleasant urge and psychic tension that precede and trigger tics are similar to those of pain or itching, and involve the same paralimbic and sensory areas demonstrated by fMRI for tics. The distinction between the neural basis of involuntary motor tics and tics voluntarily acted out to bring momentary relief of the unpleasant sensation needs further study.

GENETICS OF INFANTILE BILATERAL STRIATAL NECROSIS

The gene mutation causing autosomal recessive infantile bilateral striatal necrosis (IBSN) was identified in eight consanguineous Israeli Bedouin families, in a study at Schneider Children's Medical Center, Petah Tikva, Israel, and other centers. The age of onset of the disease in the 12 affected individuals ranged from 7 to 15 months. Choreoathetoid movements of the face, trunk and extremities, dystonia, pendular nystagmus, optic atrophy, and spastic quadriparesis were associated with gradual disappearance of the basal ganglia on serial MRI scans. At 10 to 11 years old, the MRI showed a small, residual caudate nucleus and putamen with abnormal signals. Metabolic workup was normal. Sequencing of the nup62 gene showed a missense mutation in all patients mapped to the chromosomal region 19q13.33. Five prenatal diagnoses were made in 3 at-risk families. The p62 protein is involved in the basal ganglia degeneration. (Basel-Vanagaite L, Muncher L, Straussberg R et al. Mutated nup62 causes autosomal recessive infantile bilateral striatal necrosis. **Ann Neurol** August 2006;60:214-222). (Respond: Dr Basel-Vanagaite, Department of Medical Genetics, Schneider Children's Medical Center of Israel and Rabin Medical Center, Beilinson Campus, Petah Tikva, 49100 Israel).