Epilepsy and Agenesis of the Corpus Callosum

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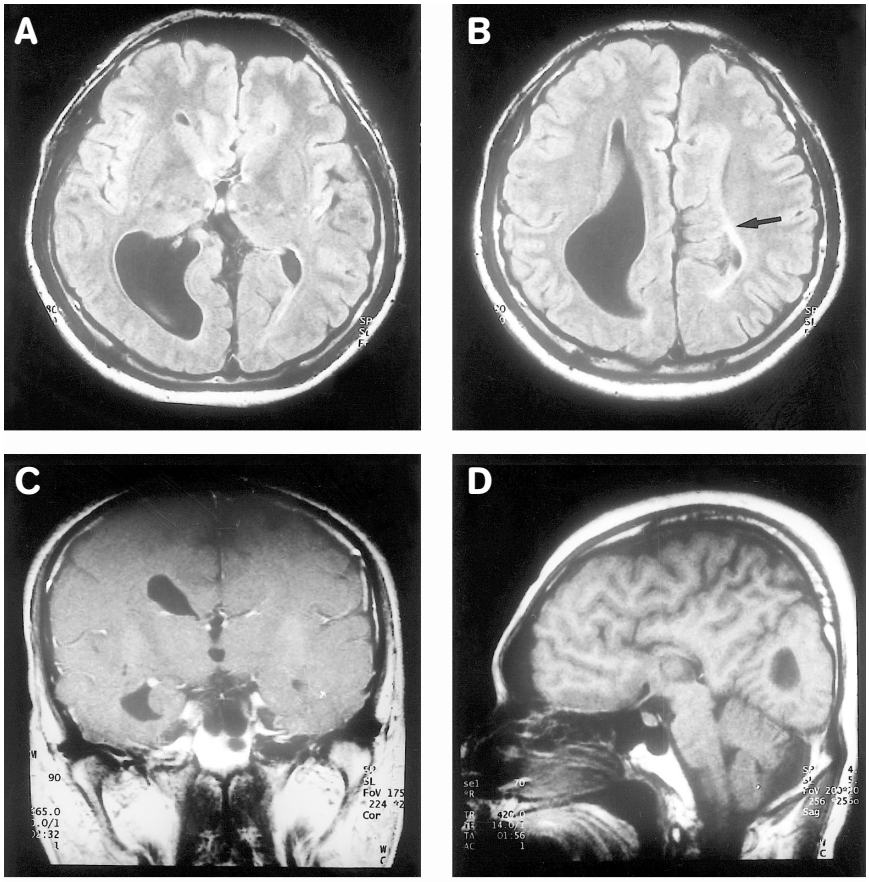

Figure. MRI of the brain in 2002 revealed total absence of the corpus callosum; asymmetric enlargement of right lateral ventricle, especially occipital horn (colpocephaly), and right temporal horn, indicating maldevelopment of right medial temporal lobe; maldevelopment of left lateral ventricle with gray matter abutting left occipital horn; and ectopic gray matter in the regions where frontal horn and lateral ventricle should be located (arrow).

A 26-year-old man had seizure disorder since age of 16 years. The seizures were generalized type with focal onset: They started with head turning to right, right facial twitch and right arm flexion and then there were bilateral arm flexion and leg extension. A mild right Todd's paralysis for about 10 minutes was usually noted in the postictal state.

The seizures were well controlled with carbamazepine 400mg twice a day. He had been free of seizures since 1995.

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The patient was handsome and tall (height 180cm) with no apparent dysmorphic features. He had finished occupational high school with an average grade. He played in the school basketball team. The birth history and family history were unremarkable. His mental and motor development seemed to be normal. His physical and neurological examinations were normal.

Neuropsychological examination revealed a significant depression of verbal-conceptual intellectual function, a mild impairment of core linguistic functioning, and a defect of visual perception of unfamiliar faces in the context of otherwise normal neurobehavioral function. EEGs usually showed mild intermittent diffuse slow waves. CT and MRI showed agenesis of the corpus callosum with associated anomaly and heterotopia mainly in left frontal lobe (Figure).

Agenesis of the corpus callosum is due to defective embryogenesis of the midline telencephalic structures, first described by Reil in 1812[1,2]. This anomaly is most commonly associated with seizures and varying degree of mental retardation[2-4]. Seizures are frequently partial but may be secondarily generalized[4]. Perceptual and cognitive disorders may vary from no apparent defect to various manifestations of the "disconnection" syndrome. Clinical symptoms are usually related to other cranial or spinal abnormalities, i.e. hydrocephalus, heterotopias, brachycephaly, arachnoid cysts, microgyri, spina bifida, meningomyelocele[1]. However, as in this patient, this anomaly may be present without or with only minimal symptoms and signs of neurological abnormality. Although the cause of his epilepsy seemed to be complex, heterotopia in left frontal lobe might be primarily responsible for his seizures with right focal onset.

References:

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