Teaching NeuroImage: Hemiconvulsion-hemiplegia-epilepsy syndrome

Sequential MRI follow-up

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A 42-month-old boy presented with frequent left partial motor seizures of 3 months duration. The symptoms began after an acute episode of fever, encephalopathy, vomiting, and left partial seizures lasting 1 week. CSF was unremarkable and negative for HSV PCR. Evaluation for procoagulant states was negative. He recovered with left hemiparesis. MRI during the acute illness (figure 1) and at 3 months (figure 2) suggested a diagnosis of hemiconvulsion-hemiplegia-epilepsy (HHE) syndrome.

HHE syndrome is characterized by prolonged unilateral convulsions with fever in children under 4 years of age, who subsequently develop hemiplegia, partial epilepsy, and extensive atrophy of the in-

volved hemisphere.^{1,2} The pathogenesis is believed to be an interplay among genetic predisposition; viral infection (e.g., influenza, HHV 6) or toxin (theophylline) exposure; excitotoxicity due to prolonged ictal activity; and contributory systemic factors such as cytokine excess, hypoxia, ischemia, and fever.²

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Figure 1 T1 axial (A), T2 axial (B), and coronal (C) (FLAIR) sections of brain show thickening of cortical gray matter with increased signal intensity on T2W and FLAIR images with effacement of sulcal spaces and midline shift suggestive of unilateral encephalitis

Figure 2 T1 axial (A), T2 axial (B), and coronal (C) (T2) sections of brain showing right hemispherical atrophy with gliotic changes and ventricular dilation, as well as right subdural hematoma

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Disclosure: The authors report no disclosures.

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Neurology 2008;71:e28

DOI 10.1212/01.wnl.0000325475.04616.e3

This information is current as of September 8, 2008

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