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 involved hemisphere. 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.

REFERENCES

Teaching NeuroImage: Hemiconvulsion-hemiplegia-epilepsy syndrome: Sequential MRI follow-up
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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