Hemiconvulsion-hemiplegia syndrome

A 6-year-old female child presented with right focal convulsions for the past 8 days (lasting for more than 8 hours), followed by right-sided weakness. She had a past history of perinatal asphyxia, developmental delay, and seizure disorder. Her growth parameters were below the 3rd percentile with obvious microcephaly. Her neurological examination revealed normal sensorium, absence of neck rigidity, right hemiplegia, and right upper motor neuron facial palsy. Magnetic resonance imaging of the brain revealed atrophy of the entire left cerebral hemisphere not limited by any vascular territory, predominantly involving the left temporal lobe, with dilatation of the left lateral ventricle [Figure 1]. Based on the clinicoradiological picture, the child was diagnosed as hemiconvulsion-hemiplegia syndrome (HHS).

The differential diagnosis of acute postictal focal weakness includes vascular insults, central nervous system infections, and developmental brain malformations.[1] HHS is a rare sequel to protracted febrile focal convulsive seizures of

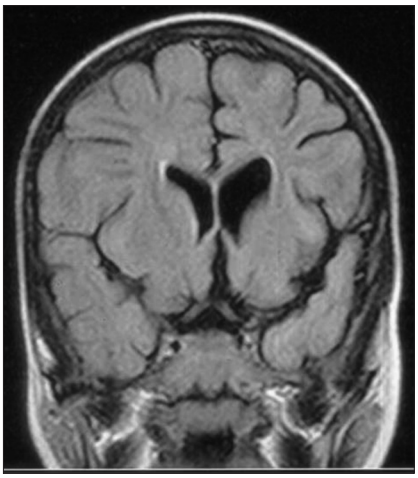

Figure 1: Magnetic resonance imaging fluid-attenuated inversion recovery coronal view; note the atrophy of the entire left cerebral hemisphere, not confined to any single vascular territory, with ex-vacuo dilatation of the left lateral ventricle

early childhood.[1,2] Neuroimaging hallmarks include acute edema followed by chronic atrophy of the entire epileptic hemisphere, independent of any vascular territory.[1,2] This is usually followed years later by the development of intractable epilepsy; the complete triad is better known as hemiplegia-hemiconvulsion-epilepsy syndrome (HHE).[1,2] While the exact pathophysiology remains elusive, the role of central nervous system developmental abnormalities predisposing to the injurious effects of prolonged seizures seems important.[2] Early recognition of the syndrome can provide better counseling to the family regarding the subsequent risk of developing epilepsy.[2]

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Conflicts of interest
There are no conflicts of interest.

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