Acute phase of Hemiconvulsion-Hemiplegia Epilepsy Syndrome: Magnetic Resonance Imaging Findings and Review of literature

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ABSTRACT
HHE syndrome is characterized by the occurrence of prolonged clonic seizures with unilateral predominance occurring during fever in infancy, who subsequently develop a transient or definitive hemiplegia. Later partial epilepsy is often observed. We report a case of a four-year and nine-month-old boy with hemiconvulsion–hemiplegia–epilepsy (HHE) syndrome documented by magnetic resonance imaging (MRI) in its acute phase. In particular, T2 and DWI abnormalities appear to be well correlated with parenchymal damage that results from sustained ictal activity.

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Introduction
HHE syndrome is characterized by prolonged unilateral convulsions during fever in infancy and early childhood, followed by the development of transient or definitive hemiplegia. Neuroradiological studies showed unilateral edematous swelling of the epileptic hemisphere at the time of initial SE [1]. Later partial epilepsy is often observed and an extensive atrophy of the involved hemisphere is also documented by neuroimaging [2]. The mechanisms underlying the HHE are still unclear. Longitudinal MRI studies are rare and the pathogenesis of the abnormalities observed in the early and in the later stages is still poorly understood [3–4].

We describe the case of a five years old child, who was hospitalized in our structure for convulsion and the imaging confirmed a HHE syndrome .

Case report
We report the case of a four-year and nine-month-old boy with no significant medical history, who was admitted to the emergency for a prolonged status epilepticus with fever.

He was given diazepam and phenobarbital and then he was intubated and transferred to the pediatric intensive care unit. Routine laboratory investigations were normal and cranial CT on the day of admission revealed no abnormal tissue densities.

Extensive investigation showed no evidence of central nervous system infection. At Day 2, a left hemiplegia was noted which led to a second cranial CT (third day) that showed signs of unilateral cerebral edema. A MRI five days after admission showed sulcal effacement of the right hemisphere on T1WI (fig.1), a slightly increased signal on T2WI/FLAIR (fig. 2) and hyperintensity of white matter and thalamus on DWI of the right cerebral hemisphere predominantly involving the subcortical U-fibers (fig. 3) with signal loss on ADC, indicating diffusion restriction consistent with edema. The affected areas are independent of vascular territories. No vessel or flow abnormalities on TOF angiography (fig. 4), nor focal abnormalities or atrophy.

Fig. 1. Sulcal effacement of the right hemisphere with slight grey-white dedifferentiation on T1WI.

Fig. 2. Axial and coronal T2WI (a,b), coronal FLAIR (c) : diffuse swelling of the right cerebral hemisphere and slightly increased signal of the whole right cerebral hemisphere including the subcortical U-fibers with significant mass effect causing a slight midline shift (5mm).
In our case, among the known risk factors for the development of HHE, there were young age and prolonged febrile status epilepticus. The finding of early DWI abnormalities including axonal damages in the thalamus of the epileptic hemisphere illustrating the involvement of the whole hemisphere in this condition supporting the idea that prolonged focal febrile seizures produce or contribute to the development of brain damage.

We excluded in our patient an underlying condition such as brain malformation, cortical dysplasia or thrombosis (Fig 3). A thalamic dysfunction induced by cell damages can be responsible for disruption of the thalamo-cortical circuit and can play a role in the later epilepsy [1]. Auvin S et all’s findings suggest that the use of anti-edema therapy or NMDA-type glutamate receptors antagonists should be considered to prevent the cell injury in HH syndrome.

An EEG recording is important at the acute phase of HH syndrome to exclude an ongoing subtitle status epilepticus. However the diagnosis of HHE does not require EEG to be confirmed [1].

**Conclusion**

HHE syndrome could be diagnosed by MRI in its acute phase. In particular, T2 and DWI abnormalities appear to be well correlated with parenchymal damage that results from sustained ictal activity, independent of any vascular territory with subsequent appearance of epilepsy. Although experimental data do not suggest a particular vulnerability to cell injury during brain development, prolonged seizure activity, hyperthermia, inflammation and blood brain barrier damage seems to contribute to the pathogenesis of HHE syndrome.

**References**