A case of Hemi-Convulsions- HHE Syndrome

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Abstract:
Hemiconvulsion-Hemiplegia-epilepsy syndrome (HHE) is focal epilepsy, a rare epileptic syndrome characterized by prolonged partial epilepsy producing hemiplegia sometimes followed by epilepsy disorder (1). We report 19 yr male with right hemiplegia, history of prolonged unilateral seizures since 4yrs of age, with evidence of left hemispheric atrophy on magnetic resonance imaging (MRI). The pathogenesis of HHE syndrome is likely to be due to histotoxic epileptic brain damage following prolonged focal convolution due to any cerebral insult (2). Here we present a case of Idiopathic HHE syndrome related to poorly treated febrile seizures in childhood.


was confirmed that patient was having features suggestive of right hemiplegia, during the hospital stay patient suddenly developed jerky movements, involving initially right upper limb and lower limb, later patient developed twitching of face, deviation of angle of the mouth, up rolling of eyes followed by loss of consciousness, patient never developed movements on the left half of the body, clinically diagnosed with partial seizures (hemi convolution) with secondary generalization. Patient is a known case of seizure disorder since the age of 4th year. Patient's first episode of seizures was at the age of 4th year, patient had repeated episodes of focal clonic seizures of the right arm; his consciousness was normal between attacks of partial seizures, as reported by parents. Following the repeated paroxysms of partial seizures, hemi convulsions of the same side appeared involving the right upper limb and lower limb, associated with loss of consciousness; patient is having recurring episodes of similar seizures for past 12years. Patient was born out of a non-consanguineous marriage, his neonatal period was uneventful, his birth weight was around 2.5kg,
with normal developmental milestones and appropriate vaccinations done till age of 5 years. He was further investigated to look for possible cause for seizure disorder, his investigations revealed normal hemogram with HB 13 gm%, TC 5800/cmm, ESR 16 mm at 1 hr, platelet count 3.1 lakh /cmm; normal renal and liver function tests; urine routine examination revealed no abnormalities, His chest X-ray, USG abdomen and pelvis were also normal.

FIGURE 1 FIGURE 2
Patient was also investigated for possible causes for young stroke, patient ECHO/Doppler study revealed trivial regurgitations in tricuspid valve, normal Carotid Doppler with normal carotid intimal thickness, PT/APTT/INR were within normal limits, negative anti nuclear antibodies (ANA), Anti-phospholipid antibodies were negative, anticardiolipin antibodies (IgM and IgG) within normal limits, Homocysteine level is within normal limits [6.8 mol/L normal 4.4-10.8], normal protein C and S, antithrombin III levels [21 mg/dL normal 22-39 mg/dL], his lipid profile was essentially normal. CT imaging revealed atrophy of the left cerebral hemisphere with dilated lateral ventricles (Figure 1, 2).

MRI revealed abnormally small sized left cerebral hemisphere with grossly widened sulci and shrunken gyri suggestive of atrophy and degeneration of cerebral cortex including white matter, thalamus and subthalamic structures suggestive of hemi-atrophy. (Figure 3, 4)

Electroencephalography (EEG) that was done in immediate post ictal state showed few sharp/spike waves with slowed background activity in the left ward placed leads/electrodes, Reported as Electroencephalogram consistent with focal epilepsy.
Discussion:
In 2001, HHE was introduced as a syndrome in the published report of the ILAE Task Force on Classification and Terminology\(^3\). It is the stereotyped sequence of events (hemiconvulsions followed by hemiplegia and leading to a focal epilepsy) that makes HHE “a complex of signs and symptoms that defines a unique epilepsy condition,” that is, a syndrome\(^3\). The term Hemiconvulsion-hemiplegia (HH) is often used to describe the initial stage of the syndrome\(^4,8\). The incidence of HHE has declined considerably, due to effective management of seizures in childhood and due to intravenous or rectal benzodiazepines for the treatment of febrile convulsions has become common practice\(^5\). HHE syndrome is classified based on the aetiology into two types\(^6\): a) Idiopathic hemiconvulsion-hemiplegia, where there is no obvious cause and such cases may represent only prolonged (complicated) febrile convulsions that do not otherwise differ from common febrile convulsions. b) Symptomatic hemiconvulsion-hemiplegia where there is pre-existing asymptomatic lesion, of perinatal or prenatal origin. The ictal discharge consists of rhythmic (2–3/s) bilateral slow waves, with higher amplitude on the hemisphere contralateral to the clinical seizure. High amplitude spikes or sharp waves and recruiting rhythms (10 cycles per second) mainly concerning the posterior regions intermingle with slow waves only on the side contralateral to the clinical jerks\(^7\). Characteristic neuro-radiological findings of uniform hemiatrophy are seen only after the prolonged episodes of hemi-convulsions. In the initial stages CT scan may show swelling and edema of the hemisphere involved in the epileptic discharge\(^1,2,4\). It is important to promptly recognize and apply early vigorous treatment for prolonged infantile seizures of whatever origin, especially of febrile convolution, as this is the most important factor that can influence the long-term outcome and reduce the incidence of post convulsive hemiplegia and late onset partial epilepsy\(^8\). Vigorous and early treatment of the initial episode, usually with rectal administration of a benzodiazepine, is the most appropriate way to reduce neurological sequelae\(^9\). The incidence of HHE syndrome has declined considerably in countries in which emergency care is highly developed\(^10\). We hypothesize that prolonged childhood febrile seizures that are poorly controlled and treated were responsible for the evolution of the HHE syndrome in this patient, as
it is well know that, epilepsy and neurological sequelae might be the direct consequence of seizures *per se*\(^{11,12}\).

REFERENCES:


3) Engel J Jr. A proposed diagnostic scheme for people with epileptic seizures and with epilepsy.


