CASE REPORT

HEMICONVULSION-HEMIPLEGIA-EPILEPSY SYNDROME

M. R. Ashrafi MD [1],
J. Tafarroji MD [2],

Abstract

Objective

Hemiconvulsion-Hemiplegia-Epilepsy syndrome (HHE) is an initial phase of unilateral or predominantly unilateral convulsive seizures usually of long duration, with a second phase of hemiplegia (usually permanent), immediately following the hemiconvulsions;and then a third stage, characterized by the appearance of partial epileptic seizures. The causes of the initial convulsions in HHE syndrome are multiple but in many patients no cause in obvious.

Neuroimaging studies demonstrate an acquired atrophy, that often is preceded by swelling and edema of the hemisphere. Here we report a case of 18 month-old boy, with an initial phase of repeated episodes of partial seizures, followed by hemiparesis and epileptic seizures, with one episode of generalized convulsive status epilepticus . Minor head trauma was the trigger factor of the initial attack. Brain neuroimaging showed generalized atrophy , that was more prominent in the left hemisphere.

Key words: HHE syndrome, Hemiplegia, Hemiconvulsion, Status Epilepticus

1.Associate professor, Department of pediatric Neurology, Children's Medical Center,Tehran University of Medical Sciences
2.Resident of pediatrics, Children's Medical Center,Tehran University of Medical Sciences

Corresponding Author :
J. Tafarroji MD
Children's Medical Center
Tel:+982166935848
Fox:+982166935848
E-mail: javad883@yahoo.com

Introduction

HHE Syndrome comprises an initial phase of unilateral or predominantly unilateral convulsive seizures that are usually of long duration (hemiconvulsions), followed immediatedely by a second phase of hemiplegia (usually permanent), and then a third stage that is characterized by the appearance of partial epileptic seizures (1,2). The first two stages constitute a particular form of status epilepticus rather than a distinct syndrome.The third phase of partial epilepsy, in conjunction with the history of the temporal sequence of hemiconvulsions and hemiplegia , represents a characteristic epilepsy syndrome(3).The hemiplegia that immediately follows the convulsions, is initially flaccid and fairly massive, but tends to become spastic and less marked as time passes. The minimum duration of the hemiplegia is arbitrarily set at more than 7 days to separate it from the more common postictal or Todd paralysis. In 20% of the cases the hemiplegia was not permanent, and disappears within 1 to 12 months(1), but some degree of spasiticity, increased deep tendon reflexes and pyramidal tract signs persist.

Two mechanisms playing a role in the development of a later epilepsy are induced by cytotoxic edema and thalamic dysfunction causing a disruption of the thalamo-cortical circuit (4).The incidence of the syndrome has considerably declined over the past 20 years in industrialized countries but cases are still common in third world countries (5). Presented here is the case report of an 18 month-old boy with the

diagnosis of HHE syndrome, triggered by a mild head trauma.

Case report

An 18 month-old boy, was referred to Children's Medical Center for the evaluation of right side hemiparesis and seizure disorder. The second child of consanguineous parents, he was delivered at term, by repeated cesarean section, delivery with an unremarkable birth history. At 45 days of age, the patient 's first seizures began as repeated episodes of focal clonic seizures of the right arm; the infant's consciousness was normal between attacks of partial seizures, as reported by their parents. Immediately following the repeated paroxysms of partial seizures, hemi convulsions of the same side appeared. After 2 weeks of hospitalization, seizures were controlled with Phenobarbital, but gradual upper limb paresis was observed.

Two days before the initial seizures the patient had a mild head trauma, without loss of consciousness. His gross motor development was normal with ability of sitting at 6-7 months of age and standing at 11-12 months, but weakness of the right arm persisted. He was able to say 2-3 words until 1 year of age, and his social and language development seems normal. Routine laboratory exams including, cerebrospinal fluid tests were normal. Brain CT scan and EEG were unremarkable. The patient was discharged with phenobarbital and physical therapy was recommended. At one year of age, the parents discontinued phenobarbital and a one month later, the seizures started again as partial seizures with one episode of generalized convulsive status epilepticus . After his admission, the patient's right arm palsy changed to right hemiparesis. The patient was discharged 2 weeks later after seizure control with a prescription of Phenobarbital. At 18 months of age, the patient was able to walk, only with support, and right side paresis was prominent. Head circumference,weight and length were 49.5 cm, 11kg and 79 cm respectively.

Neurological exams showed spasticity of right upper and lower extremities with brisk DTR and upward plantar reflex. Fundoscopic examination was normal. Hematological, lipid profile and biochemisteric laboratory findings were normal. Blood gas parameters , blood ammonia and blood lactatet levels were normal . Urine nitroprusside test was negative. ProteinC, proteinS,anti thrombin C and antiphospholipid Ab were within normal limits. Brain CTscan and MRI showed generalized brain atrophy with dilatation of lateral and third ventricles and prominent cortical sulci .brain atrophy was prominent on the left frontotemporal sregions. Hyper intense abnormal signals in the white matter of the left hemisphere were seen, in favour of gliosis (Figures 1,2,3).

EEGs were done several times with digital and analog (vega 10) techniques, and except for a few sharp waves on the right side and mild asymmetry, no other paroxysmal activities were seen (Fig 4).

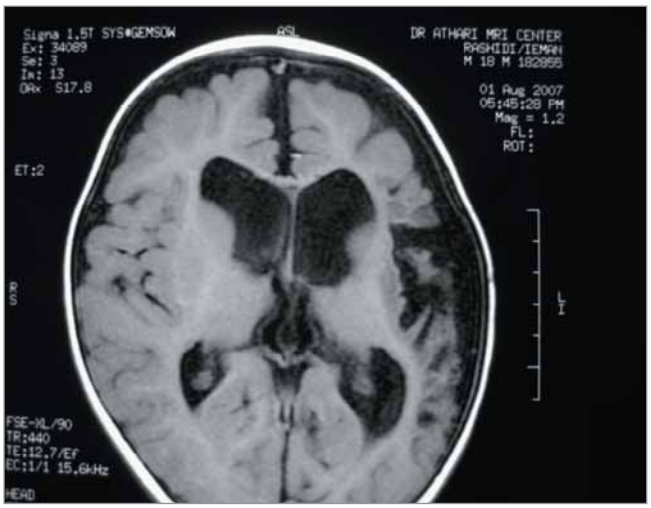

Fig 1: T1-weighted axial magnetic resonance imaging (MRI) shows dilatation of both ventricles and cortical atrophy. Hemiatrophy of left hemisphere is prominent.

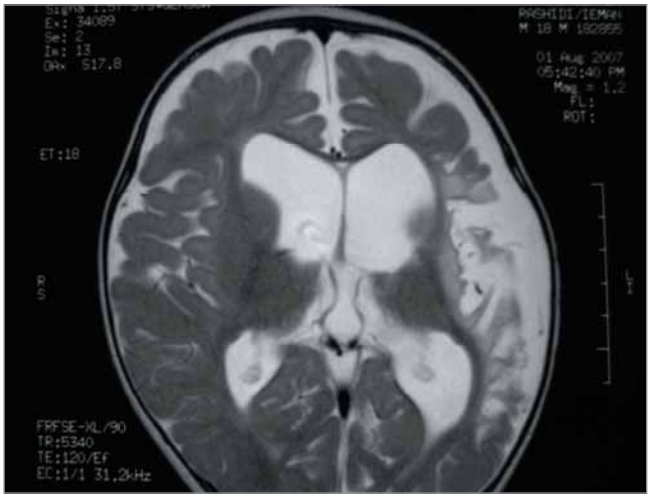

Fig2: T2-weighted axial MRI shows generalized atrophy a ventriculomegaly. Left side hemiatrophy with cortical atrophy and hyperintense signals of subcortical white matter are seen.

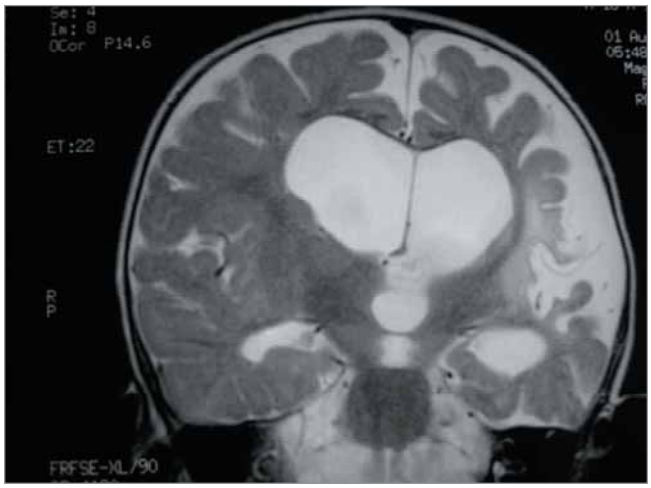

Fig 3: T2-weighted coronal MRI shows prominent left hemisphere cortical atrophy and hyperintense abnormal signals of subcortical white matter indicate gliosis .

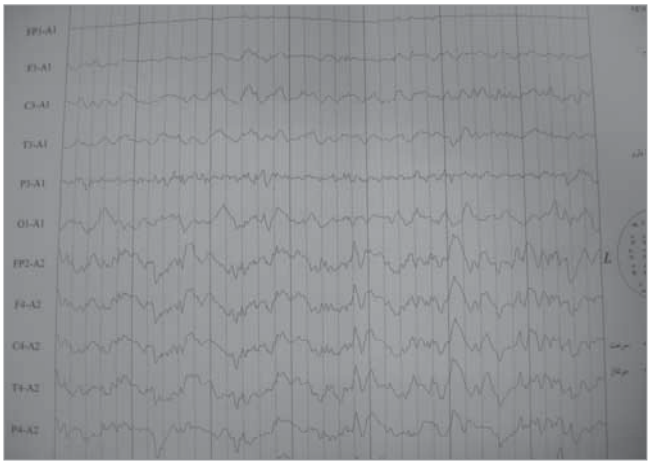

Fig 4: EEG shows few sharp waves on the right side.

Discussion

HHE Syndrome, first described in 1960 by Gastaut et al (1), has a peak incidence during the first 2 years of life, with 60-85% of cases occurring between 5 months and 2 years of age. Only few cases have been reported at 4 years or older (6,7). Our patient became symptomatic from 1.5 months of age, which to our knowledge is the youngest patient to have been reported yet. Simple partial seizures occur in approximately one third of the patients, partial seizures with secondary generalization in 20% and repeated episodes of status epilepticus in approximately 10% (7). In our patient, repeated episodes of partial seizures, occurred as focal clonic and one episode of status epilepticus.

The causes of the initial seizures in HHE syndrome are multiple; meningitis,subdural effusions, small asymptomatic hemispheric lesions of perinatal or prenatal origin, trauma (8), inherited protein S deficiency (9) and L2 Hydroxyglutaric aciduria (10) have been documented, but in many patients no cause is obvious(8). Post convulsive hemiplegia is rarely associated with vascular obstruction (11). In our case a mild head trauma was reported 2 days before the onset of the seizures but no other predisposing factors including metabolic or protein C and S deficiency were found. The pathogenesis of HHE syndrome is likely to be caused by prolonged focal convulsions responsible for a hypoxic cerebral edema(12) or the result of the epileptic activity itself, as has been observed in febrile status epilepticus (7). In this case, initial phase of repeated focal seizures and one episode of status epilepticus resulted in hemiparesis although phenobarbital withdrawal may have played a role. Swelling and edema of the involved hemisphere that was rapidly followed by atrophy is the main neuroimaging finding at the time of initial seizures(4,13).

Eventually, when the hemiparesis persists, generalized atrophy of the hemisphere is seen(14).

Many neuroradiologic studies, such as that of Toldo et al, demonstrated that seven days following hemiconvulsion, the imaging abnormality on T2 and diffusion-weighted image (DWI) was limited to the white matter of the left hemisphere and, a month later, severe gliosis and unilateral brain atrophy were already evident(2,13,15). Neuroimaging studies in our case showed generalized brain atrophy with ventriculomegaly that was more prominent in the left hemisphere. Hemiatrophy of the left hemisphere with hyper intense abnormal signals in subcortical white matter were other findings. The occurrence of seizure is usually preceded or accompaind by the appearance of paroxysmal EEG abnormalities, especially in the temporal area. EEG showed ipsilateral slowing and low voltage of background activity(2). Although EEGs were taken several times, except for a few sharp waves on the right side, no other paroxysmal activities were seen. While the standard treatment of HHE syndrome is medical, but hemispherectomy (16) and corpus callosotomy (17,18) have been performed and reduction in seizures reported(16). Unfortunately there is no curative treatment for HHE syndrome and

surgical and medical treatments are merely supportive. It has been demonstrated that in acute presentations, the use of anti-edema therapy should be considered to prevent cell injury (4).At the present time, our patient's seizures are controlled by phenobarbital, but must be followed. To our knowledge this is the first Iranian case HHE Syndrome to be reported.

References

1. H . Gaustaut ,F.Poirier,H.Payan,G.Salamon, M. Toga and M. Vigouroux. H.H.E. syndrome;hemiconvulsions,hemip legia,epilepsy.EkspKhirurgiia 1960; 1: 418-447.
2. Salih MA,Sallih MA , Kabiraj M ,Al- Jarallah AS ,EI Desouki M ,Othman S ,et al. HHE Syndrome .Aclinical EEG and neuroradiological study . childs nerv syst 1997 may ;13(51):257-63.
3. Chauvel P, Trottier S, Vignal JP. Somatomotor seizures of frontal lobe origin. In:Chauvel P, DELGADO-Escueta AV, Halgren E, et al.,eds. Frontal lobe seizures and epilepsy. New York :Raven Press. 1992. p.185-232.
4. Auvin S,Devism L, Maurage CA ,Soto- Ares G, Cuisset JM ,Leclerc F ,et al. Neuropathological and MRI findings in an presentation of HHE :areport with pathophysiological implications seizure 2007 Jun ;16(4): 371-6.
5. Arzimanoglou A ,Dravet CH. Hemiconvulsion – hemiplegia - epilepsy syndrome ,Available at: www. medlink.com / Accessed October , 2003.
6. Aicardi J ,Chevrie JJ. Febrile convulsions: neurological sequelae and mental retardation. In:Brazier MA ,Coceani F, eds. Brain dysfunction in infantile febrile convulsions .New York:Raven Press.1976.p.247-257.
7. RogerJ,lob H,Tassinari CA. Status epilepticus. In: Vinken PJ, Bruyn GW,eds. The epilepsies. Handbook of neurology, Vol. 15 Amsterdam:North Holland. 1974;145-188.
8. Aicardi J, Chevrie JJ . Consequences of status epilepticus in infants and children . In :Delgado-Escueta AV,Waster-lain CG ,Treiman DM ,Porter RJ ,eds.Status epilepticus : mechanisms of brain damage and treatment. Advances in neurology, Vol. 34. New York: Raven Press.p. 115-125.
9. Mondal RK, Chakravorty D, Das S. HHE Syndrome and protein S deficiency . Indian J Pediatr 2006 Feb; 73(2):157-9.
10. Lee C, Born M, Salomons GS, Jakobs C, Woelfle J. HHE Syndrome as a presenting feature L-2-hydroxyglutaric aciduria. J Child Neurol 2006 Jun; 21(6):538-40.
11. Norman RM. Neuropathological findings in acute hemiplegia in childhood with special reference to epilepsy as a pathogenic factor . In :Bax M ,Mitchell RG, eds. Acute hemiplegia in children. London :Heinemann. 1962. p.37-48.
12. Herbst F, Heckmann M, Reiss I, Hugens-Penzel M, Gortner L, Neubauer B. HHE Syndrome Klin padiatr 2002 may-jun; 214(3) :126-7.
13. Freeman JL. Coleman LT, Smith LJ, Shield LK. HHE Syndrome;characteristic early magnetic resonance imaging findings:J Child Neurol 2002 Jan ;17 (1):10-60.
14. Tan N, Urich H. Postictal cerebral hemiatrophy with a contribution to the problem of crossed cerebellar atrophy .Acta neuropathol (berlin) 1984; 62:332-339 .
15. Toldo I ,Calderone M, Boniver C, Dravet CH , Guerrini R , Laverda AM. HHE Syndrome :early MRI findings ,Brain Dev 2007 Mar ;29 (2):109-111.
16. Devlin AM, Cross JH, Harkness W, Chong WK, Harding B, Vargha-khadem F,et al. Clinicall outcomes of hemispherectomy for epilepsy in childhood and adolescence. Brain 2003 Mar ;126(pt3):556-66.
17. Kwan SY, Wong TT, Chang KP ,Lee YC, Guo WY, Su MS. postcallosotomy seizure outcome in HHE Syndrome. Zhonghua Yi Xue Za Zi (taipei) 2000 Jun; 63(6) :503-11.
18. Kwan SY ,Wong TT , Chang KP ,Chi CS ,Yang TF , Lee YC ,et al. seizureoutcom after corpus callosotomy . Childs Nerv syst 2000 Feb ;16 (2):87-92.