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# HEMICONVULSION HEMIPLEGIA EPILEPSY (HHE) SYNDROME : MAGNETIC RESONANCE IMAGING IN ACUTE PRESENTATION.

Radiodiagnosis

**Dr. Sucheta Thakur*** MD; Department of Radio Diagnosis, Indira Gandhi Medical College, Shimla, HP, India.*Corresponding Author

**Dr. Praveen** MD; Department of Radio Diagnosis, Indira Gandhi Medical College, Shimla, HP, India.

**Dr. Suresh Kumar** DM, Department of Radio Diagnosis, Indira Gandhi Medical College, Shimla, HP, India.

## ABSTRACT

Hemiconvulsion hemiplegia epilepsy (HHE ) syndrome is characterized by initial prolonged unilateral seizures in infancy and early childhood which is followed by ipsilateral hemiplegia. After a variable seizure free period, most patients develop intractable epilepsy. Neuroimaging in acute phase shows evidence of cytotoxic edema followed by hemi atrophy of the affected hemisphere. HHE can occur in patients with previous structural brain abnormalities or can occur without any underlying pathology which is called as idiopathic HHE. We report three cases of HHE, with prolonged right hemiconvulsion followed by right hemiplegia in two patients and left hemiconvulsion with left hemiplegia in one patient. Magnetic resonance imaging (MRI) of both patients showed characteristic neuroimaging findings of acute phase of HHE including diffusion restriction on diffusion weighted images involving left cerebral hemisphere

## KEYWORDS

Hemiconvulsion hemiplegia epilepsy syndrome (HHE), Magnetic resonance imaging.

## INTRODUCTION

Hemiconvulsion- hemiplegia- epilepsy (HHE) syndrome is characterized by a prolonged unilateral seizure followed by the development of hemiplegia on the same side of the convulsion which is initially flaccid and is gradually replaced by spastic condition which become less marked with time. Children younger than 4 years of age are commonly affected and common onset is between 5 months to 2 years of age. Seizures can be simple partial seizures (33%), partial seizures with secondary generalisation (20%) and repeated episodes of status epilepticus (10%).[1]

We report three cases of Hemiconvulsion -hemiplegia -epilepsy syndrome . All three presented in acute phase of HHE with hemiconvulsion and were shown to have characteristics neuroimaging findings suggestive of diffuse cytotoxic oedema involving the one of the hemisphere .

### Case 1

A 19- months old patient presented to pediatric emergency room with altered sensorium and continuous right sided seizure for 10 hours. Patient was euglycemic at the time of admission and was managed as a case of status epilepticus. His birth history was normal. There was no family history of epilepsy and febrile seizure. Cardiovascular, respiratory and abdominal examinations were normal. Central nervous system examination revealed paucity of movement on right side of body with grade 0 power in right upper and lower limb with reflexes being brisk on the right side. Laboratory tests revealed normal blood urea , creatinine , serum calcium and serum electrolytes level. CSF examination was also normal . Child was started on continuous midazolam infusion after mechanical ventilator support. The seizures aborted and the child was gradually tapered from midazolam infusion and weaned from ventilator support in the next 2 days. On Magnetic resonance imaging ( MRI )evaluation, there was presence of diffuse gyral swelling and effacement of sulci in the left cerebral hemisphere along with diffusion restriction [Figure 1 (a, b, c & d)]. MR angiography was normal. The child was started on the oral carbamazepine and physiotherapy of the right side of body was advised. At the time of discharge, the child had right sided hemiparesis with grade 1 power, hyperreflexia.

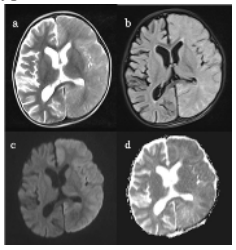

**Figure 1.** Magnetic Resonance Imaging (MRI) Brain showing gyral swelling and effacement of sulci in left cerebral hemisphere on **(a)** T2 Weighted axial Images (T2WI) , **(b)** FLAIR axial images along with diffusion restriction on diffusion weighted images **(c & d).**

### Case 2

A 2- years old male child with history of Right sided focal seizure involving the right upper , lower limb and right side of face presented to the paediatric emergency room in status epilepticus. Birth history was normal. Cardiovascular, respiratory and abdominal examinations were normal. On central nervous system examination there was decreased tone and power in the right upper and lower limb with brisk reflex. CSF examination was normal. Laboratory investigation including complete hemogram revealed microcytic hypochromic anaemia with thrombocytopenia. Other routine blood test were normal. Child was started on iv phenobarbitone and phenytoin . On further evaluation with computed tomography (CT) head, there was effacement of sulci and loss of grey and white matter differentiation in left cerebral hemisphere (Figure 1a). On evaluation with MRI, brain there was T2/FLAIR hyperintensity involving the cortex of left cerebral hemisphere along with gyral swelling, effacement of sulci and diffusion restriction [Figure 1(b,c,d,e &f)]. MR angiography was normal.

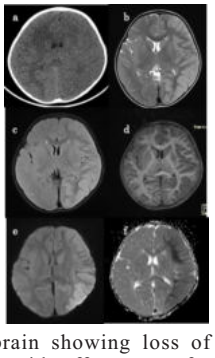

**Figure 2.** NCCT brain showing loss of grey white matter differentiation along with effacement of sulci in left cerebral hemisphere **(a)**. MRI Brain showing hyperintensity along the cortex in left cerebral hemisphere along with gyral swelling and effacement of sulci in left cerebral hemisphere on T2WI (**b**) , FLAIR axial images **(c),** T1 weighted images (**d**) with diffusion restriction on diffusion weighted images **(e & f )**

### Case 3

4 years old male child came in paediatric emergency room with left sided multiple seizures . Cardiovascular , respiratory and abdominal examinations were normal . On central nervous examination patient had decreased power in left upper and lower limb with brisk deep tendons reflex. On evaluation with MRI right cerebral cortex showed T2/FLAIR hyperintensities and effacement of sulci , gyral swelling

along with diffusion restriction in the involved right cortex [ figure 3(a, b, c & d )]. MR angiography of the patient was normal.

The clinical presentation and MRI findings of all three patients were proposed to be most consistent with acute phase of hemiconvulsion - hemiplegia -epilepsy syndrome.

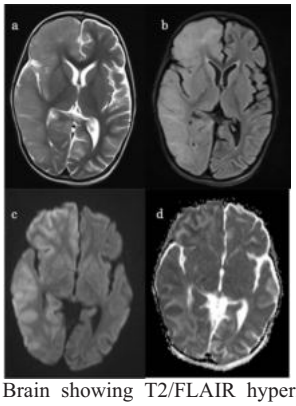


**Figure 3.** MRI Brain showing T2/FLAIR hyperintensity , gyral swelling and effacement of sulci in right cerebral hemisphere on T2WI **(a) ,** FLAIR axial image **(b)** along with diffusion restriction on diffusion weighted images **(c & d)** .

## DISCUSSION

HHE is characterized by the clonic epileptic seizure of long duration affecting one half of the body followed by ipsilateral hemiplegia of varying intensity. After a variable duration most patients develop epilepsy [2]. In many patients no obvious cause was documented (idiopathic) but there can be multiple causes of the initial seizure in HHE have been documented previously ; meningitis , subdural effusions, small asymptomatic hemispheric lesions of perinatal or prenatal origin , trauma , inherited protein S deficiency [3] and L2 Hydroxyglutaric aciduria [4].

The characteristic neuroimaging findings associated with HHE syndrome include effacement of sulci with gyral swelling along with diffusion restriction suggestive of cytotoxic oedema involving the affected hemisphere which is often followed by the cerebral hemi atrophy after variable duration of time.

Toldo et al [5], demonstrated that the imaging abnormality on T2WI and diffusion-weighted image (DWI) was limited to the white matter of the affected hemisphere after seven days following hemiconvulsion , and after a month severe gliosis with unilateral brain atrophy were present. The standard treatment of HHE syndrome involves medical treatment which aims at satisfactory control of seizures. Carbamazepine and/or phenobarbitone have been used with good control of seizures. Surgical treatment with hemispherectomy and corpus callosotomy has been found successful in refractory cases.[6]

## CONCLUSION

The neuroimaging findings presented in our cases and in the other HHE patients studied in literature seem to be very characteristics and if confirmed in larger series, could be helpful in an early diagnosis.

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