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Case Report

Radiology Section

Role of Diffusion Weighted Imaging in the Detection of the Initial Phase of the Hemiplegia Hemiconvulsion Epilepsy Syndrome: A Case Report

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ABSTRACT

The hemiplegia hemiconvulsion epilepsy syndrome is a unique and rare epileptic syndrome. We are reporting here, a case of a 7 month old female child who presented with right sided hemiconvulsions, followed by ipsilateral hemiplegia. MRI of the brain was done, which showed the classical findings of the hemiplegia hemiconvulsion epilepsy syndrome.

Key Words: HHES, diffusion weighted imaging, MRI, Radiology

INTRODUCTION

The hemiplegia hemiconvulsion epilepsy syndrome is a unique and rare epileptic syndrome which is characterized by prolonged unilateral convulsions, followed by persisting ipsilateral hemiplegia [1]. The term, 'hemiplegia hemiconvulsion epilepsy syndrome' is used to describe a syndrome which is characterized by the appearance of clonic epileptic seizures of long duration, which affected one side of the body, which is usually associated with febrile illness in children who are aged less than 4 years. Subsequently, hemiplegia of varying intensities can develop, which can be permanent [2]. MRI shows the classical features of the hyperintensities which are located throughout the cerebral hemisphere on T2W images and those of restricted diffusion on DWI images in the early stages [3,4]. After a period of several months, these changes disappear and brain atrophy becomes evident, which affects the entire cerebral hemisphere [3,4].

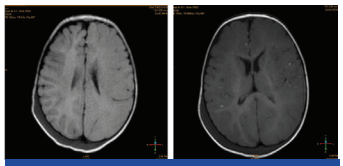
CASE REPORT

A first child of non-consanguineous parents aged 7 months, who was apparently well, presented to us with an acute onset of loose stools, vomiting and fever. The patient was immunized till date and had normal development. She had symptoms of dehydration at admission, which were corrected within two hours. She had the first episode of convulsions, which were right focally located, after which she went into altered sensorium with a Glasgow coma scale of 6/15. The patient had such episodes of seizures, with no regaining of consciousness in between. Her seizures remained refractory despite the treatment with multiple anti-convulsants drugs. Her metabolic work up and CSF analysis were normal. She remained seizure free from day 4 of her admission, but she had right hemiplegia, altered sensorium and oedema which was secondary to hypoalbuminaemia and the inappropriate antidiuretic hormone.

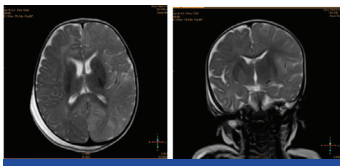
Her sensorium level deteriorated later and she developed hypostatic pneumonia and pulmonary oedema due to fluid retention, for which she required mechanical ventilation on day seven of her admission. She succumbed to it on day eight of her admission.

The child was referred to the Radiology Department on the second day of her admission for MRI of the brain to rule out organic brain disease. MRI was done by using a 1.5 Tesla Philips Achieva machine with multiplanar T1, T2 and FLAIR sequences. Diffusion weighted sequences were also taken, with b value of 1000.

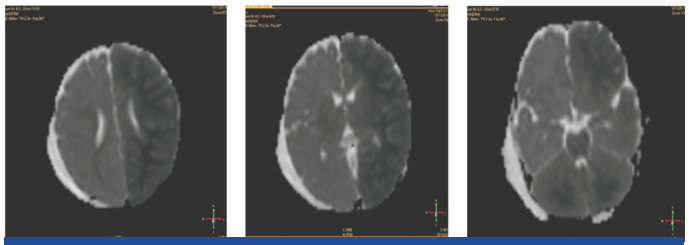
MRI of the brain showed diffuse enlargement of the left cerebral and the cerebellar hemisphere, with effacement of the sulci and the fissures on the same side. The gray and white matter differentiation was lost in the left cerebellar hemisphere.



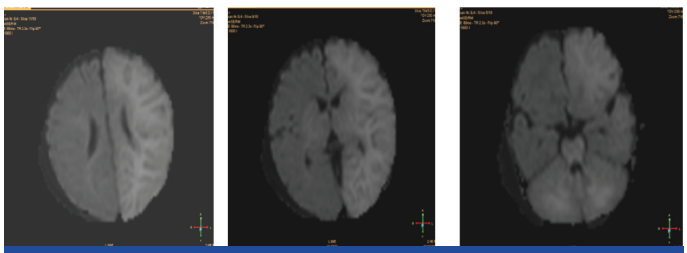
[Table/Fig-1]: Axial unenhanced FLAIR and T1 weighted images showing diffuse enlargement of left cerebral hemisphere with sulcal effacement.



[Table/Fig-2]: Axial & coronal T2 weighted images showing diffuse enlargement of left cerebral hemisphere and diffuse hyperintensity with sulcal effacement.



[Table/Fig-3]: Axial apparent diffusion coefficient images showing diffuse cytotoxic edema in the left cerebral hemisphere and cerebellum



[Table/Fig-4]: Axial diffusion weighted images showing diffuse cytotoxic edema in the left cerebral hemisphere and cerebellum

Restricted diffusion was noted in the white matter of the entire left cerebral and cerebellar hemispheres-which was suggestive of cytotoxic oedema. Sparing of the internal capsule was noted. The ventricles were normal. There was no evidence of any focal mass lesion.

DISCUSSION

The hemiplegia hemiconvulsion epilepsy syndrome is a rare sequence which comprises of sudden and prolonged unilateral seizures which are followed by usually permanent ipsilateral hemiplegia. The hemiplegia hemiconvulsion epilepsy syndrome was first described in 1957 by Gastaut et al [2], which was characterized by the following sequential combination: unilateral or predominantly unilateral clonic seizures of long duration which occurred in the first four years of life and were immediately followed by flaccid hemiplegia which was ipsilateral to the clonic seizures. The hemiplegia hemiconvulsion epilepsy syndrome has a peak incidence during the first two years of life, with 60–85 % of the cases occurring between 5 months to 2 years of age [5,6]. The child in our case report was from the same age group.

Simple partial seizures occur in one third of the patients, partial seizures with secondary generalization occur in 20% of the cases and repeated episodes of status epilepticus occur in approximately 10% of the cases [6]. The child in this case report had repeated episodes of right partial seizures in form of status.

The hemiplegia hemiconvulsion epilepsy syndrome comprises of three phases, with an initial phase of unilateral or predominantly unilateral convulsive seizures of long duration, followed by a second phase of hemiplegia [2,[3]. The third phase is characterized by the appearance of partial epileptic seizures [2,3]. The second phase of the hemiplegia is usually permanent, but however, in 20 % of the cases, it disappears within one to twelve months, with some degree of spasticity, increased tendon reflexes and pyramidal tract signs [2].

According to the aetiology, the hemiplegia hemiconvulsion epilepsy syndrome is divided into two separate categories. The type I category is symptomatic with multiple variable causes like, meningitis, sub-dural effusions, small symptomatic hemispheric lesions of perinatal or prenatal origin, trauma, inherited protein S deficiency and L2 hydroxy glutaric aciduria. The type II category is idiopathic, with no specific causes. In our case, no specific cause was identified and hence, it categorized as type II.

Neuro-radiological studies which were conducted by Toldo et al and Freeman et al demonstrated that in the early stages of the hemiplegia hemiconvulsion epilepsy syndrome, there was enlargement of the cerebral hemisphere with sulcal effacement and diffuse white matter hyperintensities on T2 weighted images, with restricted diffusion on the diffusion weighted images, which were suggestive of cytotoxic oedema. Months later, the cases which were followed up, showed severe gliosis and unilateral cerebral hemispheric atrophy which involved the cortical and the subcortical regions, with dilatation of the ipsilateral ventricular system. The MRI in our case showed similar findings of left cerebral hemispheric enlargement with sulcal effacement and gross cytotoxic oedema on the diffusion weighted images, which involved the left cerebral hemispheric and cerebellar white matter.

The child succumbed due to aspiration pneumonia, three days later to the imaging; hence, a follow up study was not done.

The standard treatment of the hemiplegia hemiconvulsion epilepsy syndrome is medical. However, hemispherectomy and corpus callosotomy have been performed for the reduction of the post gliotic seizures. Surgical and medical treatment are nearly supportive, but unfortunately, there is no curative treatment.

CONCLUSION

MRI of the brain with diffusion weighted imaging is helpful in the early diagnosis of the hemiplegia hemiconvulsion epilepsy syndrome with cytotoxic oedema in the involved cerebral hemisphere. The neuro-radiological findings in our case and in the few hemiplegia hemiconvulsion epilepsy syndrome patients who were reported in the literature seem to be very characteristic and, if these can be confirmed in a larger series of patients, they could permit an early diagnosis and management of the disease.

REFERENCES

[1] Bahi-Buisson N , Kossorotoff M , Barnerias C, Boddaert N , Bourgeois M, et al. A typical case of the hemiconvulsions-hemiplegia-epilepsy syndrome which revealed contralateral focal cortical dysplasia. *Developmental Medicine and Child Neurology* 2005; 47:12:830-34.
[2] Gaustaut H, Poirier F, Payan H, Salamon G, Toga M, Vigouroux M, et al. syndrome; hemiconvulsions, hemiplegia and epilepsy. *Eksp Khirurgiia* 1960; 1: 418-47.
[3] Salih MA, Sallih MA , Kabiraj M , Al-Jarallah AS , El Desouki M, Othman S, et al. The HHE Syndrome. A clinical EEG and neuroradiological study. *Childs Nerv Syst* 1997 May; 13(51):257-63.
[4] Toldo I, Calderone M, Boniver C, Dravet CH, Guerrini R, Laverda AM. The HHE Syndrome: early MRI findings. *Brain Dev* 2007 Mar; 29 (2):109-11.
[5] 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; 247-57.
[6] RogerJ, Iob H, Tassinari CA. Status epilepticus. In:Vinken PJ, Bruyn GW, eds. The epilepsies. *Handbook of Neurology*, Vol. 15 Amsterdam: North Holland. 1974; 145-88.

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