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Case Report
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Hemiconvulsion Hemiplegia Syndrome with Iron Deficiency Anemia in a Child with Review of Literature

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Abstract

Hemiconvulsion Hemiplegia Epilepsy (HHE) syndrome is a rare complication of prolonged focal seizures in children and usually idiopathic but may be associated with structural, infective, traumatic and degenerative diseases. We present a three years male child with iron deficiency anemia, who had left sided focal status epilepticus clonic type for nearly 12 h, and later developed left sided hemiparesis. This is the first association of iron deficiency with hemiconvulsion-hemiplegia-syndrome; and illustrates the importance of neuroimaging in each case of status epilepticus. More importantly, the case also raises the possibility that deficiency in iron could be a contributing factor in cases of hemiconvulsion-hemiplegia-syndrome.

Keywords: Hemiconvulsion Hemiplegia Epilepsy (HHE); Febrile seizure; Status epilepticus; Iron deficiency anemia

Introduction

Hemiconvulsion-Hemiplegia-Epilepsy (HHE) is a rare outcome of prolonged focal status epilepticus that usually occurs in children below 4 years of age [1]. This entity starts with a focal status epilepticus and concurrent febrile illness, which subsequently evolves to ipsilateral hemiparesis of the convulsing side; at follow-up over months to years, two-third of the patients develops epilepsy. Magnetic Resonance Imaging (MRI) of the brain in the acute stages characteristically shows gross cerebral edema in the contralateral hemisphere, which later turns into atrophy [2]. At present, the underlying etiology of HHE remains poorly understood. Several etiologies for the initial seizures in HHE syndrome have been proposed and include viral infections, meningitis, subdural effusion, protein S deficiency, cyanocobalamin deficiency, L2 hydroxy glutaric aciduria and mutations SCN1A and CACNA1A [3-6]. However, it is an established but under diagnosed and potentially preventable sequel of prolonged status epilepticus in children younger than 4 years. Here we present a case of 3 years male child of hemiconvulsion hemiplegia syndrome with iron deficiency anemia.

Case Presentation

A-3-years male child born to a nonconsanguineous marriage *via* normal vaginal delivery and cried immediately after birth came to the emergency with complaint of sudden onset abnormal body movement followed by unconsciousness 7 days back and weakness of left side of body for 3 days. Child was apparently alright 7 days back and he developed sudden onset left sided clonic convulsions involving both upper and lower limbs while playing with friends at his home. After few minutes the child had drooping of head and frothing from his mouth followed by unconsciousness. The above episode was associated with one day of fever only. There was no history of trauma, headache, vomiting, cough, cold, blurring of vision, rash, animal bite, any unknown substance ingestion, bluish discoloration of body. There was no past history of any similar episodes, any seizures or previous hospitalization. His developmental milestones were achieved according to the age.

Before arrival in emergency, the child received treatment for meningitis for 4 days and status epilepticus (Intravenous lorazepam, phenytoin, sodium valproate, levetiracetam and continuous midazolam; 6 μg/kg/min infusion for 12 h). After that the seizures got controlled and the child was conscious but in altered sensorium. The finding of CSF was normal. They took the child to home by own. In home the caretakers noticed that the child could not able to move his left half of body for which they came to us after 3 days.

On examination, the child was conscious, irritable, lying supine in bed. His vitals were stable. Glasgow Coma Score was 11/15. There was left sided upper motor neuron type of 7th cranial nerve

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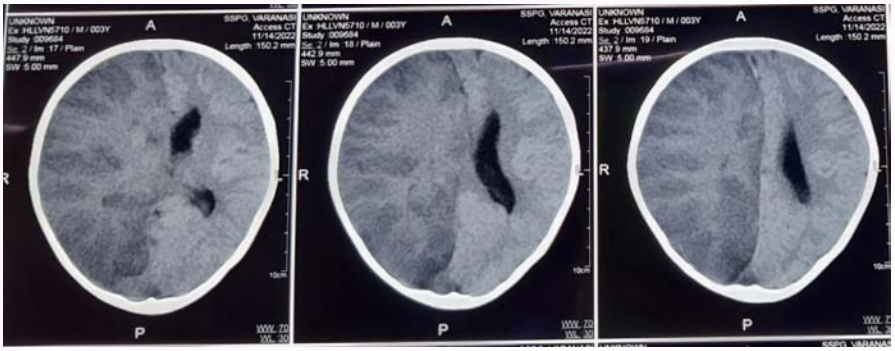

Figure 1: NCCT Head, showed wedge shaped hypodensity involving complete right cerebral hemisphere with mass effect and effaced right ventricle.

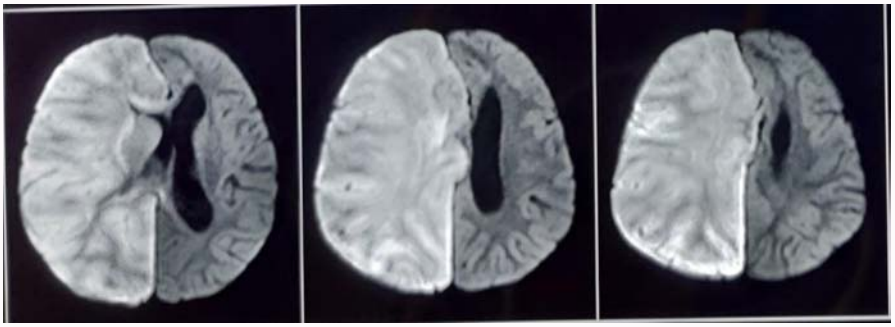

Figure 2: MRI BRAIN, showed edematous right cerebral cortex, subcortical and deep white matter along with mass effect and dilatation of contralateral lateral ventricle.

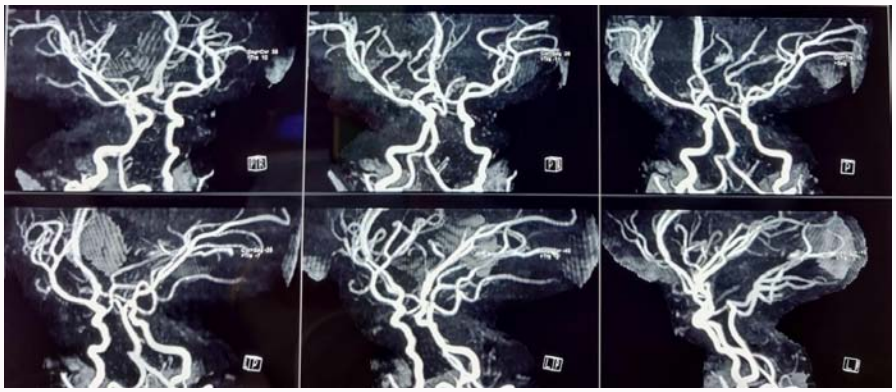

Figure 3: MR Angiography, revealed no significant abnormality in cerebral vessels.

palsy with left sided hemiplegia. He had no loss of sensation and signs of meningeal of irritation.

His CBC revealed TLC: 6.76 × 10^3/µl, DLC (N:51, L: 35, E7, M6 AND B1%), RBC: 4.05 × 10^6/µl, Hb: 6.8 gm/dL, MCV-72 fl, MCH-16.8 pg, MCHC- 23.3 gm/dl, platelets- 156 × 10^3/µl. Serum Iron, ferritin, TIBC were 11 µg/dl, 9.9 ng/ml and 499.93 µg/dL respectively. Mentzer index and Transferrin saturation was 17.7 and 2.20% respectively. Serum cyanocobalamin (314 pg/mL) and homocysteine (10 µmol/L) was normal. His tuberculin test and CBNAAT for CSF and gastric aspirate came out to be negative. Liver function tests, renal function test, PT, aPTT, INR reports were normal. Urine for organic acid screening came to be normal.

His NCCT Head (Figure 1) showed wedge shaped hypodensity involving complete right cerebral hemisphere with mass effect and effaced right ventricle.

MRI BRAIN (Figure 2) - showed edematous right cerebral cortex, subcortical and deep white matter along with mass effect and dilatation of contralateral lateral ventricle.

MR Angiography (Figure 3) revealed no significant abnormality in cerebral vessels.

We treated the child with acetazolamide (50 mg/kg/day), oxcarbazepine (10 mg/kg/day) and low dose aspirin (3 mg/kg/day to 5 mg/kg/day). The child's general condition improved gradually. He started recognizing his uncle and aunt. Facial palsy improved gradually by the time of discharge but weakness of left side of body persisted. The child did not develop any episode of fever or seizure during the period of hospitalization. The child was discharged after 7 days with a diagnosis of hemiplegia hemiconvulsion syndrome with iron deficiency anemia.

Discussion

Hemiconvulsion-Hemiplegia/Hemiconvulsion-Hemiplegia-Epilepsy (HH/HHE) syndrome was first reported by Gastaut et al. in 1957. Pathophysiology of this syndrome still remains poorly understood and the long-term cognitive outcomes are still unclear [1].

Many literatures have confirmed it that iron deficiency has a strong correlation with febrile seizures and pediatric stroke [7,8]. Iron Deficiency Anemia (IDA) hold a 10-fold increased risk of acute stoke in well toddlers [9]. Studies reported frequency of iron deficiency in patients with febrile seizures is around 63% in India [10]. Iron is required for hemoglobin synthesis as well as for enzymes participating in neurochemical reactions. Various hypotheses about iron deficiency and iron deficiency anemia in inducing seizures are decrease of GABA inhibitory neurotransmitter, change in neuron metabolism, reduction of enzymes such as monoamine and aldehyde oxidases and impairment in oxygenation and energy metabolism of the brain [11]. Studies also revealed that low iron status as well as ferritin concentration may be a significant risk factor for the development of febrile seizures [12].

Conclusion

As Iron Deficiency Anemia (IDA) is already an established risk factor of febrile seizure and stroke in children, we would like to highlight that IDA may be a potential trigger to develop seizure and subsequently Status Epilepticus which may lead to a catastrophic entity like Hemiconvulsion-Hemiplegia Syndrome. But whether IDA is a chance association or causation to HH/HHE syndrome will require further confirmation by more case reports or studies.

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