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Case Report

Infantile Spasm with Computed Tomographic Features of Bilateral Intracerebral Hemispheric Infarcts: A Case Report

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Abstract: Infantile spasm also known as West syndrome is a devastating form of an early infantile encephalopathy, and belongs to the group of drug resistant epilepsy, with an incidence of about 0.25-0.42 in every 1000 children. This is an eight-month-old male infant who was referred for a contrast enhanced computed tomography scan (CECT) of the head on account of frequent stiffness of the both upper and lower limbs and the trunk since birth. He also has poor developmental milestones; poor suckling, neck control, social smile and inability to sit or crawl. The CECT showed an elongated and summit shaped skull on the scout image, with associated multiple non-enhancing oval areas of hypodensities in both cerebral hemispheres, poor grey-white matter interphase, prominent sulci, gyri, ventricular system and basal cisterns, with thickened gyri-pachygyria. The report of the electroencephalogram (EEG) showed spikes of activity in the brain that is referred to as hypsarrhythmia. Infantile spasm was diagnosed in a child with triad infantile spasms, neurodevelopmental retardation and EEG findings of hypsarrhythmia most likely from multiple cerebral infarcts was established. We report the case of infantile spasm with CECT features of cerebral hemispheric infarcts due to its rare nature and peculiar presentation.

Keywords: Cerebral infarcts, infantile spasms, Paraparesis, Milestones.

INTRODUCTION

Infantile spasm (IS) is an epileptic syndrome that occurs in children less than 1 year and rarely more than 2-years of age and often characterized by clinical spasms, hypsarrhythmia in an electroencephalogram and most times the spasms are associated with developmental delay or regression [1, 2].

The international league against epilepsy (ILAE), the 2017 operational classification of seizures described IS as epileptic spasms occurring at infantile age [3, 4]. These spasms are further defined as sudden flexion, extension or mixed extension-flexion of predominantly proximal and truncal muscles [3, 4].

Infantile spasms have been classified as idiopathic in the absence of an accompanying neurologic disorder, cryptogenic with a presence of a suspected etiology which is yet to be identified and symptomatic when a definite etiology can be demonstrated [5].

Infantile spasm has been reported to have numerous etiologies, but the etiology is detected in about 60-75% of cases with relevant radiological, infectious, and metabolic investigations [3, 6].

Some etiologies which are Neurocutaneous syndromes, brain malformations, neonatal strokes and sequelae of hypoxic ischemic encephalopathy are most often obvious on imaging such as magnetic resonance imaging [3, 6].

The world health organization defines stroke as a clinical syndrome of rapidly developing focal or global disturbance of brain function lasting more than 24-hours or leading to death with no

obvious nonvascular cause [7, 8]. The entity stroke in childhood is rare and has an estimated incidence of about 2.5-13 per 100,000 per year, with an estimated mortality rate of 0.6 per 100,000 death per year [9, 10].

Pediatric stroke is a neurological injury caused either by occlusion or rupture of a cerebral vessel, and can either be ischemic, hemorrhagic or both [1, 11].

The ischemic form of stroke Frequently occurs from cerebral arterial vascular occlusion than venous occlusion of cerebral veins or sinuses. The hemorrhagic stroke occurs from bleeding from a ruptured cerebral artery or from bleeding into site of an acute ischemic stroke [11].

The most frequent form of stroke in the pediatric age group is the acute ischemic stroke (AIS) and responsible for about half of the cases, this is in contrast to the adult population in whom 80-85% of all cases are ischemic [11-13].

Multiple etiological factors or Risk factors have been associated with pediatric stroke, these may be cardiac disease (cardiomyopathies), drugs, hematologic conditions (Sickle cell disease), trauma (head injury), infection (human immunodeficiency virus infection), vascular causes (arteriorvenous malformation), syndromic and metabolic disorders (Marfan syndrome) and vasculitis(idiopthic and cerebral vasculitis) [11-18].

The diagnosis of IS according to the National Infantile Spasms Consortium has observed that about 55% of cases reach an etiological diagnosis following clinical examination and MRI [3]. Neuroimaging among which is PET(Positron emission tomography) scan also becomes vital in delineating forms of cortical dysplasia and structural lesions in Individuals with IS [4, 19].

The prognosis and developmental outcome of IS is Rather poor and nonencouraging, it has been observed That varying forms of associated defects like auditory and visual defects, psychiatric disorders, epilepsy and demise of patients at early age has also been reported [20-22].

CASE REPORT

This is an 8-month-old male infant that was referred for a contrast enhanced computed tomogram of the head and brain on account of seizures/jerky and spastic movements that became intense at the age of about 4 months, poor and delayed developmental milestones, mental and growth retardation and an oblonged shaped skull.

The patient is the 5th child of his parents, the Mother aged 37-years of age and had no formal education and also Not gainfully employed. She denied history of similar occurrence or family history of sickle cell disease and bleeding disorders. She admitted to delayed onset of antenatal care and also intake of lots of local herbs during the index pregnancy.

The mother described the spasms as clusters of sudden and repeated bowing/bending of the head and trunk with stiffening of the upper limbs;arms and lower limbs;legs lasting for about 1-2 seconds per episode and appear repititive and alternative almost about 10-15 times in an episode not lasting about 3-5 minutes, and the patient may have about 3 or more attacks every day most especially after the child wakes up.

The mother admitted to poor developmental milestones; failure to achieve neck control, sitting and crawling, there is accompanying loss of social smiles and interaction, and increased fussiness or silence.

The patient had a summit skull with increased vertex-basion ratio/distance, the orbits are normal but with a suggestion of bilateral lateral squints, drooling of saliva but patient appeared conscious, not pale, anicteric, not dehydrated or in any form of respiratory distress and had no obvious neurocutaneous affectation; no skin pigmentation or cutaneous nodules. The muscle tone were low; grade II, the tendon reflexes were markedly reduced and some were absent. No obvious spinal abnormality or mass demonsrated.

The patient had a normal packed cell volume of about 37%, had increased white blood cell count of 19000/mm^3 in favor of the lymphocytes, normal ESR of 7mm/hr, the urinalysis showed pus cells, traces of protein and red blood cells. The urine micoscopy yielded the growth of Escherichia coli.

The blood pressure was normal; 90/55mmHg and the pulse rate was also normal wiith a value of 100beats per minute.

The patient had an EEG done; presented with the results that was reported as having spikes called hypsarrhythmia; this belongs to the triad of IS.

The CECT done demonstrated a summit skull with prolonged vertex-basion distance(figure 1), there are areas of non-enhancing Cerebrospinal fluid density (Hounsfield Unit:4) spaces in both Cerebral hemispheres; the chronic infarcts, there is prominence of the demonstrated lateral ventricles, the Sulci and gyri with the basal cisterns, arachnoid

and Subarachnoid spaces also showed prominence due to loss cortical volume (figures 2&3). Prominence and abnormal pattern of the sulci lead to a suspicion of gyrial malformation; pachygyria (figure 2&3). The cerebellum showed normal appearances and morphology (figure 4). No masses to rule out Tuberous sclerosis and other forms of congenital or acquired brain infections or malignant masses.

The outcome of the computed tomographic scan; multiple cerebral infarcts most likely from congenital cerebrovascular insults, these are most likely the cause of the infantile spasm in the index case.

The index case had a combination of the spasms;seizures, hypsarrhythmia on EEG and neurodevelopmental retardation, hence the diagnosis of infantile spasm.

The parents of the patient admitted to been adequately counselled by the referring physician, also has been placed on drugs; ACTH (adrenocorticotrophic hormone), Vigabatrin and Pyridoxine. The parents admitted to the occasional use of injectables at the hospital to abort seizures which are most times difficult to abort once commenced.

The patient although passed on at the age of 2-years; this information was from the parents, and at the time of the demise most neurodevelopmental milestones were not achieved.

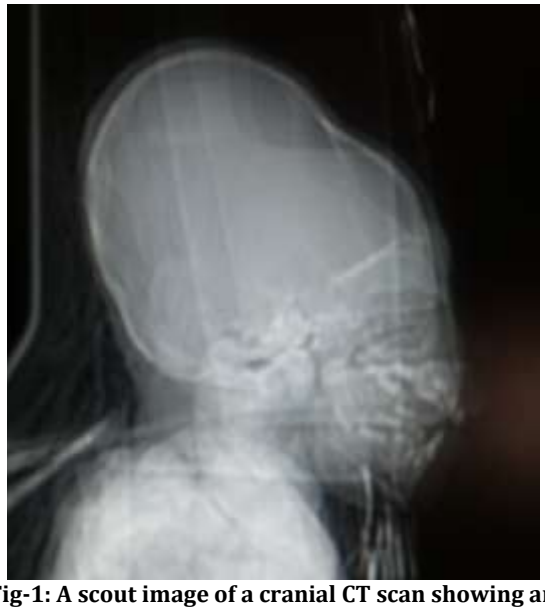

Fig-1: A scout image of a cranial CT scan showing an elongated skull; the summit skull with an elongation of the vertex-basion diameter. The Sella and sutures with the facial skeleton show normal appearances

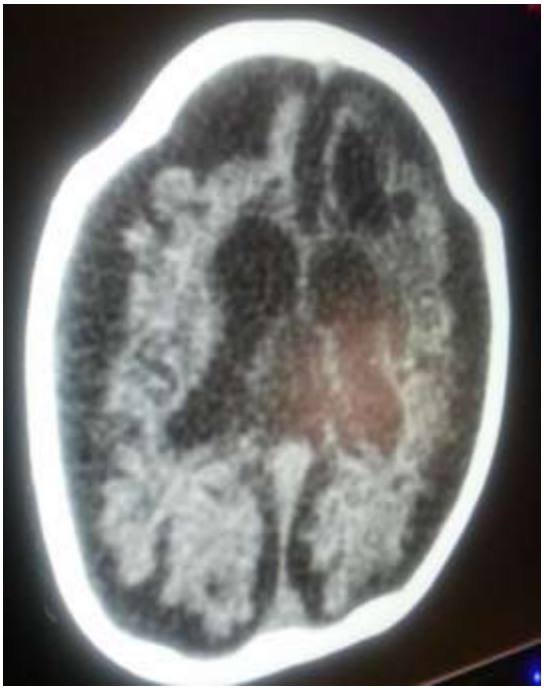

Fig-2: Axial image of CECT at the level of the lateral ventricles showing areas of hypodensity, prominent lateral ventricles, prominent subarachnoid spaces with reduced cortical volume and thickened gyri

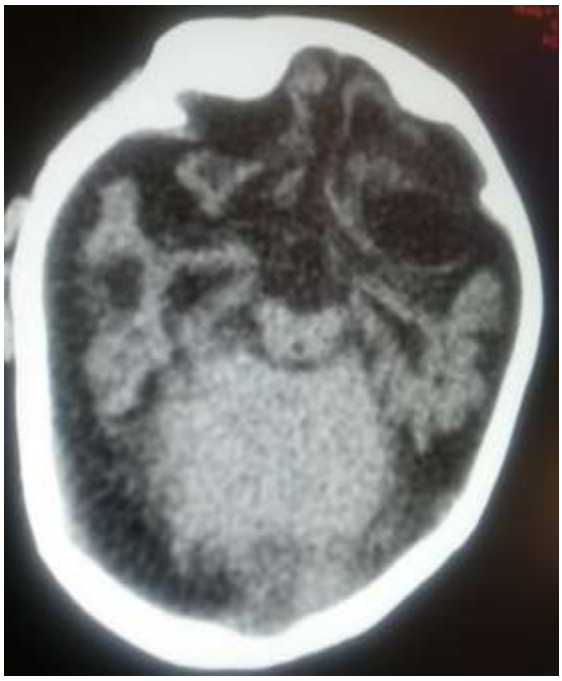

Fig-3: Axial CT scan image of the brain lower slice at the level of the cerebellum showing the thickened gyri; pachygyria, prominent sulci, prominent basal cisterns, prominent arachnoid and subarachnoid spaces and the oval hypodense area of chronic infarct

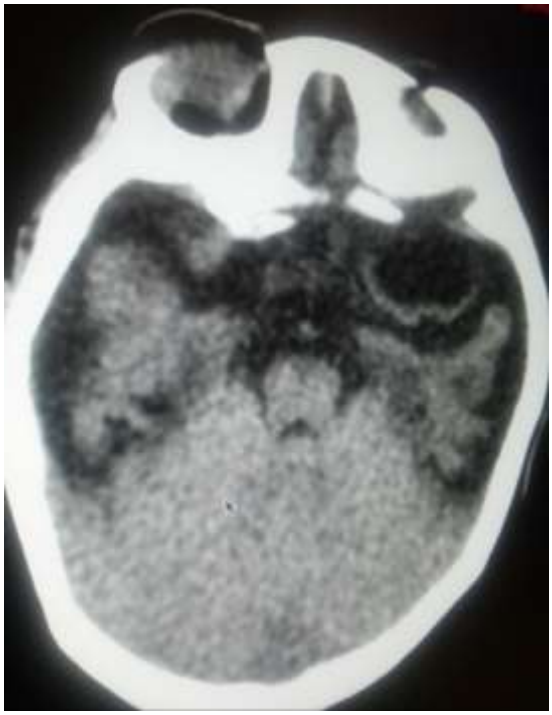

Fig-4: Axial image of the CECT at the level of the ventricle showing a normal cerebellum devoid of any hypodense areas; the entertained chronic infarcts

DISCUSSION

Infantile spasms belong to one of the most devastating childhood epilepsies due to the difficulty of controlling seizures and the association with mental retardation, and was first described in 1841 by Dr West WJ [4, 23, 24]. The patient under review had episodes of difficult to control seizures and mental retardation in conformity to these literatures.

Infantile spasm or West syndrome is characterized by the triad of infantile spasms, hypsarrhythmia (EEG pattern described by Gibbs), and neurodevelopmental retardation [4, 20, 25]. The index case also had infantile spasms, similar EEG finding and growth with mental retardation in conformity to these literatures.

West syndrome has no significant gender preference, but a slight male preponderance has been reported with about 90% of cases seen in infants aged less than 12-months with a peak of onset between 4-6months [26]. The case under review happens to be a male infant with the age of onset of about 5months, thereby conforming to this literature.

This group of epileptic syndrome; IS has been etiologically classified as either; symptomatic with an identifiable cause prenatal, perinatal and postnatal cause, and; cryptogenic with an unknown cause with a normal neuroimaging, neurological and metabolic evaluation, and finally; idiopathic form with pure functional cerebral dysfunction having complete recovery, no residual dysfunction, normal neuroimaging, etiologic evaluation and neurodevelopment [4, 20]. The case under review is most probably the symptomatic group having features on the neuroimaging; cerebral hemispheric infarcts with gyrial and skull abnormality diagnosed postnatally, thereby conforming to these literatures.

Infantile spasms have also been classified as either focal and diffuse groups with respect to varying lateralizing signs, these provides a relative effect on outcome and treatment strategy [20, 27]. The index case is most probably amongst the diffuse group, with respect to the multiple groups of muscles involved and forms of presentation, thereby conforming to these literatures.

Seizures in West syndrome have been further classified based on the type of muscles involved, the flexor spasm; regarded as most characteristic seizure type, and accounting for 42% of cases, we also have the mixed flexor-extensor spasms noted in about 50% of cases, the flexor spasm is predominantly featured in naming the syndrome [20, 21]. A third form may also consist of mainly the extension of muscles and termed extensor spasms [28]. The index case shows both flexion and extension of the muscles and belongs to the mixed flexor-extensor spasms thereby conforming to these literatures.

West syndrome by large has varying etiologies which mechanism are not fully understood, the causative disorders are classified as prenatal, perinatal and postnatal [20, 22]. The index case is most likely from prenatal cause and presented with some form of structural abnormalities from birth; some of these were summit skull, pachygyria, poor grey-white matter interphase with multiple chronic cerebral infarcts on CT scan, thereby conforming to these literatures.

West syndrome has a poor prognosis, and has a peak onset of the seizures at the age of 3 to 7 months, and usually occurs before the age of 2-years in about 93% of affected individuals [4, 20]. The index case was not an exception; spasms had a peak around the fifth month of life, thereby conforming to these literatures.

Neuroimaging plays an important role in the diagnosis of IS, this often leads to detection of an etiology in about 70% of cases. The neuroimaging modalities are mainly MRI and CT scan for structural abnormality, while PET Scan for detecting focal CNS lesions that may warrant surgical resection [20, 21]. The index case had only CT done as a neuroimaging, this demonstrated structural abnormalities like

summit skull shape, pachygyria, prominent sulci, gyri with subarachnoid spaces and bilateral multiple cerebrospinal fluid density areas; chronic infarcts, thereby conforming to these literatures.

Infantile spasms are known to be resistant to most conventional antiepileptic drugs and prove difficult to manage, most researchers show combination of drugs which include ACTH, corticosteroids and vigabatrin, these have shown some effectiveness with ACTH been the most prescribed [20, 27]. The index case also had antiepileptic combination of ACTH, corticosteroid and vigabatrin conforming to that reported by most literatures.

Patients with IS do have a poor prognosis, it has been observed that a third of the sufferers die before the age of three-years and about 50% of cases die before the age of ten-years, some of the victims do present later in life with visual and auditory defects in about a third-half of them and about 90% of these cases had mental retardation [20, 21].

CONCLUSION

Pediatric patients with recurrent seizures of early onset especially in the infancy period should be examined clinically and by neuroimaging to rule out the possibility of infantile spasm so as to commence adequate management in order to reduce the progression of neurodevelopmental retardation in this group of patients.

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