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CASE REPORT

Shaken Baby Syndrome Manifesting as Infantile Spasms Seizure Type

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The diagnosis of child maltreatment leading to head injury is challenging. Here, we present the case of a 3-month-old female infant who presented with focal seizures that lasted for several minutes. After admission, she began to show intermittent clusters of head nods, irritable crying, arching, writhing, stiffening, and jerking of both arms. These results and electroencephalography findings were attributed as the diagnosis of infantile spasms (IS). Brain computed tomography and magnetic resonance imaging (MRI) revealed the presence of chronic subdural hematoma mixed with acute ischemic injuries. Examination of the eye fundus confirmed the presence of retinal hemorrhage. Therefore, all evidence pointed to a diagnosis of shaken baby syndrome (SBS). Based on this case, we suggest that physicians should consider a diagnosis of SBS for children with new-onset IS and that should be evaluated, diagnosed, and treated as promptly as possible.

Key words: Shaken baby syndrome, infantile spasms, electroencephalography, chronic subdural hematoma, acute ischemic injuries

INTRODUCTION

Infantile spasms (IS) is the most common catastrophic epilepsy of childhood.[1] Its incidence is ~3.1-4.2 per 1000 live births and it occurs in ~9% of all children diagnosed with epilepsy.[1,2] It consists of a constellation of presentations, including emotional disturbance and crying during the events, subtle or massive clusters of jerks that occur mainly on awakening, and unusual movements involving head, truncal, and hip flexion or extension or, most commonly, head and body flexion with leg extension. These presentations always start abruptly, worsen gradually, and only briefly interrupt consciousness.[3] In addition; arrest of psychomotor development (e.g., loss of babbling, verbalizations, or head and trunk control) may also be present. IS occurs primarily during the 1st year of life, especially between the 3rd and 8th months. Thus, it is a highly age-specific epileptic response of the infant brain to innumerable insults, such as intrauterine infection or stroke, tuberous sclerosis, perinatal asphyxia, and postnatal insults.[4] Diagnostically, more than 200 structural, metabolic, or genetic abnormalities that involve the central nervous system have been associated with IS, significantly hampering the correct diagnosis and the identification of the etiology of IS.

Head injury is the most common cause of death from physical abuse among children younger than 1 year.[5] Shaken baby syndrome (SBS) is a form of abuse that can cause severe injuries, such as subdural hematoma (SDH), subarachnoid hemorrhage, hemorrhagic contusion, and/or diffuse axonal injury.[6,7]

We describe a case of SBS manifesting as IS that was diagnosed based on clinical presentation, electroencephalography (EEG), and brain computed tomography (CT) and magnetic resonance imaging (MRI). We suggest that SBS should be considered in children with new-onset IS and that patients should be evaluated, diagnosed, and treated without delay.

CASE REPORT

A 3-month-old girl was admitted to a pediatric emergency department in Taiwan with sudden onset of vomiting, irritable crying, tonic-clonic seizures over the left extremities, rightward gaze, and lip cyanosis lasting for 2-3 min. She was born through normal vaginal delivery at 38 weeks and 5 days of gestation (birth weight, 2.50 kg). On examination, this baby had a body temperature of 36.5°C, weighed 9 kg (85th percentile), and measured 66 cm. Her blood pressure, heart rate, and respiratory rate were 96/58 mm Hg, 120 beats/min, and 40 breaths/min, respectively. Her mother had no systemic disease or significant disorder during pregnancy. The patient had no definite history of trauma, had achieved normal developmental milestones, and had no bruises on her skin. She had a soft anterior fontanel

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without prominent bulging, clear lung breathing sound, and regular heart sound. Her abdomen was soft with a hyperactive bowel sound. The cranial nerve examination was difficult. Her muscle tone and deep-tendon reflex were increased. The results of laboratory studies (full septic screen, complete blood counts, levels of serum electrolytes, magnesium, ammonia, and C-reactive protein, liver and renal function tests, arterial blood gas analysis, prothrombin time/international normalized ratio/activated partial thromboplastin time, and urine analysis) were all in the normal range. She was admitted to a general ward under the impression of seizures.

After admission, the infant developed intermittent clusters of head nods, irritable crying, arching, writhing, stiffening, and jerking of both arms. The jerking did not involve her lower extremities. The frequency of head nods and jerking was >50/day on an average (~40-60/day). Sweating, pupillary dilatation, and changes in respiratory or heart rate were observed during these events. Her seizures responded partially to sodium valproate (10 mg/kg/day) and phenobarbitone (5 mg/kg/day), but could be controlled by adding vigabatrin (75 mg/kg/day). Brain CT with contrast [Figure 1] showed symmetrical layers of subdural effusion in bilateral frontotemporal areas. EEG revealed a slow background with burst of sharp waves on the left parietal region and no evidence of hypsarrhythia. Cerebral spinal fluid (CSF) analysis showed a high opening pressure (30 cm H_2O), increased red (3460/μL) and white (40/μL) blood cells, and absence of antigens of herpesvirus, enterovirus, influenza virus, streptococcus, meningococcus, pneumococcus, and *Haemophilus influenzae* type B. Cultures of blood and CSF were devoid of bacteria. An ophthalmologist confirmed severe bilateral retinal hemorrhages and papilledema. Brain MRI coupling showed thick layers of subdural effusion in the supratentorium with a fluid-fluid level in the left parietal region. Falcine SDHs were also noted in the bilateral parietal regions in the subacute stage. Apparent diffusion coefficient map of brain MRI showed diffuse white matter low signals in the bilateral cerebral hemispheres, indicating an acute ischemic change. No midline shift or mass occupying effect was noted. Her consciousness was clear after craniostomy with removal of hematomas and drainage of hemorrhages on day 5. Her head nods and jerking of her upper arms disappeared. As all evidence pointed to a diagnosis of SBS, the father confessed to shaking the patient violently when she was crying and before he gave her a bottle. She was discharged on day 10 with complete recovery. After 3 months, follow-up brain MRI showed resorption of the bilateral SDHs. A mild malacic change and gliosis with some hemosiderin deposition observed at the right parietal lobe was related to prior trauma with hemorrhage. Magnetic response spectroscopy sampling in bilateral hippocampi showed a similar appearance with decreased N-acetyl aspartate peak height. EEG showed better background organization and reduction in epileptiform activity, but persistent slow activity in the right parietal area.

DISCUSSION

The diagnosis of IS is confirmed by EEG and a 30 min recording is often sufficient. Hypsarrhythmia, a typical finding of IS, describes the high-voltage (>200 μV), chaotic nature of epileptiform discharges, lasting several seconds of electrodecrements. Not all patients with IS show hypsarrhythmia on EEG.[8] It is difficult to record the ictal period in patients with IS. Autonomic symptoms, such as skin flushing, sweating, pupillary dilatation, or changes in respiratory or heart rate, can be present (as in the present case). Moreover, the patient's EEG showed a burst-suppression pattern [Figure 2], which is not only supportive of a diagnosis

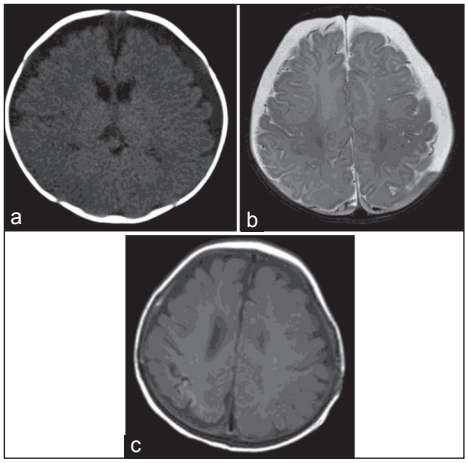

Figure 1. (a) Axial computed tomography study shows fluid collection in subdural spaces over the bilateral frontal convexitis with density slightly higher than intraventricular cerebrospinal fluid. It is suggestive of chronic subdural hematomas. (b) Axial T2-weight image shows subdural hematomas along bilateral cerebral convexities with a blood fluid level on the left. (c) Follow-up axial T1-weighted image shows subsidence of prior subdural hematomas. The brain shows focal atrophy and dystrophic calcification involving the right parietal lobe

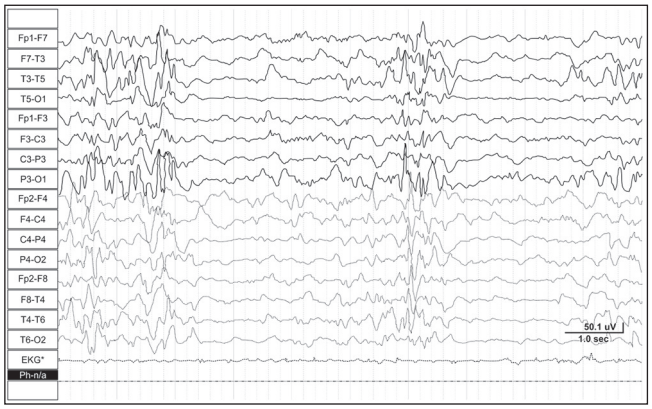

Figure 2. A burst-suppression pattern electroencephalography

of IS, but also indicative of significant underlying brain injury associated with poor outcome.[9]

This girl had no external marks on her body-no bruises, cuts, or fractures, no sign that she was forcefully gripped and no evident neck injury that would seem to result from vigorous shaking- and her mother denied any perinatal asphyxia that might have contributed to the remote IS. Tuberous sclerosis was excluded based on the lack of any signs on neuroimaging, cutaneous slit-lamp examination, and ophthalmologic evaluation. Despite extensive biochemical investigations, no other cause of IS was found. Brain CT revealed symmetrical layers of subdural effusion in the bilateral frontotemporal region; however, MRI showed thick layers of subdural effusion in the supratentorium, with a fluid-fluid level in the left parietal region, which was suggestive of different stages of hemorrhage. Therefore, SBS was suspected.

The injuries that trigger IS are unclear. Although, we did not know the exact time of occurrence of her brain injury, IS typically begins weeks to months after an initiating injury; during these latent period, neural circuits become epileptogenic. Her brain MRI findings were compatible with this hypothesis. A significant brain insult can result in IS and poor neurodevelopmental outcome, despite an early diagnosis of SBS.

CONCLUSION

IS is an epileptic encephalopathy of early infancy with specific clinical and electroencephalographic features and a poor prognosis. However, clinicians must investigate any possible underlying disorders in symptomatic IS, as in this case. Early recognition of the condition, careful diagnostic evaluation, and proper treatment may allow some children to attain seizure control and achieve a normal, or at least much improved, level of development.

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CONFLICT OF INTEREST

The authors have no conflict of interest to report.

REFERENCES

1. Kramer U, Nevo Y, Neufeld MY, Fatal A, Leitner Y, Harel S. Epidemiology of epilepsy in childhood: A cohort of 440 consecutive patients. Pediatr Neurol 1998;18:46-50.
2. Hino-Fukuyo N, Haginoya K, Iinuma K, Uematsu M, Tsuchiya S. Neuroepidemiology of West syndrome and early infantile epileptic encephalopathy in Miyagi Prefecture, Japan. Epilepsy Res 2009;87:299-301.
3. Lux AL, Osborne JP. A proposal for case definitions and outcome measures in studies of infantile spasms and West syndrome: Consensus statement of the West Delphi group. Epilepsia 2004;45:1416-28.
4. Brunson KL, Avishai-Eliner S, Baram TZ. ACTH treatment of infantile spasms: Mechanisms of its effects in modulation of neuronal excitability. Int Rev Neurobiol 2002;49:185-97.
5. Reece RM, Sege R. Childhood head injuries: Accidental or inflicted? Arch Pediatr Adolesc Med 2000;154:11-5.
6. Lo TY, McPhillips M, Minns RA, Gibson RJ. Cerebral atrophy following shaken impact syndrome and other non-accidental head injury (NAHI). Pediatr Rehabil 2003;6:47-55.
7. Geddes JF, Hackshaw AK, Vowles GH, Nickols CD, Whitwell HL. Neuropathology of inflicted head injury in children. I. Patterns of brain damage. Brain 2001;124:1290-8.
8. Karvelas G, Lortie A, Scantlebury MH, Duy PT, Cossette P, Carmant L. A retrospective study on aetiology based outcome of infantile spasms. Seizure 2009;18:197-201.
9. Nunes ML, Giraldes MM, Pinho AP, Costa JC. Prognostic value of non-reactive burst suppression EEG pattern associated to early neonatal seizures. Arq Neuropsiquiatr 2005;63:14-9.