

Fragile brain, handle with care: A case of abusive head trauma and a review of literature

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Abstract

We report an 11-month-old boy who brought with status epilepticus. The presence of bilateral retinal hemorrhages and generalized cerebral atrophy indicating ischemia revealed on imaging tests lead us to keep a differential diagnosis of shaken baby syndrome (SBS). On repeated questioning, mother gave the history of shaking by a relative which further supported our diagnosis. Thus in a child with a nonspecific febrile convulsive state, the treating physician should keep SBS also as a differential diagnosis.

Key words: *Abusive head trauma, Brain injuries, Craniocerebral trauma, Nervous system trauma, Shaken baby syndrome*

Abusive head trauma (AHT), also known as Shaken Baby Syndrome (SBS), is a devastating and potentially lethal form of infant physical abuse. The outcomes are grim. In recognized cases, AHT typically results in death or significant lifelong brain injuries in survivors. Another feature of AHT is the discrepancy between the incidence of clinically recognized cases and the unmeasured burden of the unrecognized cases. Less severe cases may not be brought to medical professionals and may never be diagnosed. Not all the episodes of shaking by a caregiver will result in clinically significant consequences. However, if unrecognized, unknown numbers of infants may develop milder forms of motor dysfunction, sensory compromise, or cognitive losses [1].

The incidence is reported to be 20-30 cases per 100,000 children <1 year of age with a case fatality rate exceeding 20% and significant disability in about two-thirds of the survivors [2]. In India, cases of SBS are very rarely reported and a literature review with the keywords “SBS” and “India” showed only 3 reports [3-5]. We present a case of SBS because we believe there is a gross under-reporting of such cases, and secondly, it is important to elicit the history of SBS for children coming with convulsive state as it can simulate febrile convulsions.

CASE REPORT

The 11-month-old boy was referred with the history of recurrent generalized tonic-clonic convulsions for 5-7 days which was sudden in onset with the movement of upper and lower extremities and up rolling of eyeballs followed by drowsiness. There was a history of previous four such episodes, for which he was treated and was on oral antiepileptics. 2 months back,

the first episode started with fever and non-projectile vomiting followed by tonic-clonic convulsions. Cerebrospinal fluid (CSF) examination done at the time of the first admission was normal. Computed tomography (CT) scan head done at last admission showed diffuse lesions in bilateral cerebral hemispheres with thinning of sulci, cisternae, and ventricles indicative of global ischemia.

There was no history of head trauma or recent vaccination. The birth and developmental milestones history were normal. On admission, the baby was comatose with Glasgow coma scale of E2V1M2, and decerebrate rigidity was present. The funduscopy revealed bilateral retinal hemorrhages. On examination, all the limbs had power <3/5. Deep tendon reflexes were elicited bilaterally, and pupils were responding bilaterally to light. No signs of raised intracranial pressure or meningeal irritation were obtained.

A provisional diagnosis of post-convulsive encephalopathy or cerebral malaria, tuberculous meningitis, or acute disseminated encephalomyelitis was made. His routine laboratory investigations including complete hemogram, platelet counts, coagulation profile, and malarial antigen tests were within normal limits. The chest X-ray was normal. Routine CSF examination and CSF culture were also negative. So, malaria and tuberculous meningitis were ruled out. The portable electroencephalography showed a generalized slowing.

In view of retinal hemorrhages on funduscopy and ischemic changes in previous CT scan, a diagnosis of SBS was considered. On re-interview parents came out with a history that at the time of first seizure episode, a relative took hold

of the baby's neck and chin and shook him vigorously in an attempt to stop the seizure. Following this event, there were multiple episodes of seizures. The magnetic resonance brain showed generalized cerebral atrophy with a little amount of white matter in the fronto-parietal region mostly due to an ischemic insult (Figure 1). An X-ray work-up for fractures was found to be negative. The patient was kept under observation and was discharged following an improvement in the condition after 12 days.

DISCUSSION

SBS was first described by Caffey in 1972 as whiplash baby syndrome where acceleration-deceleration stress is considered to be the cause of the injury [3]. AHT and SBS are forms of premeditated injury inflicted on infants due to violent shaking, the impact to the head, or a combination of both. These mechanisms can result in head trauma such as subdural hematoma, diffuse axonal injury, cerebral edema, retinal hemorrhages, and sometimes fractures of the long bones or ribs, with little or no external findings of trauma [1].

A remarkably constant unexplained finding is that victims are more likely to be males than the females [6]. The possible explanation is the bias among caregivers (particularly fathers) toward male children that "Men do not cry and crying is for women." Therefore, they become more furious and irked by male infant crying rather than female. Confessions from perpetrators have shown that crying is the usual stimulus for AHT and infant physical abuse, and crying was the reported stimulus in 63% of the cases in a study done by Adamsbaum et al. [7].

On the basis of these evidences, crying has been included into the recommendations for prevention of the American Academy of Pediatrics and the Canadian Joint Statement on SBS [8]. Other stimulating factors include socioeconomic standing, societal and family stress, and prematurity, multiple

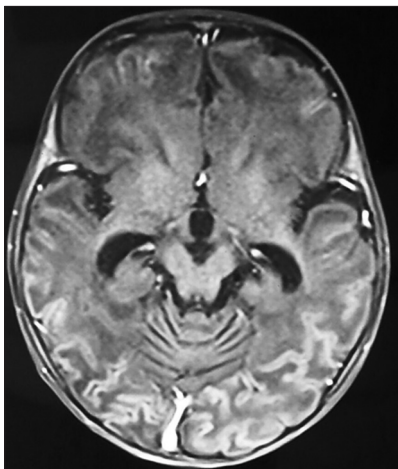


Figure 1: Magnetic resonance imaging brain showing generalized cerebral atrophy with a little amount of white matter in the fronto-parietal region.

births, developmental delays, prior military service, and childhood history of abuse in the perpetrator. Race and ethnicity have not been found to be risk factors [1].

Shaking has its unique effects on the perpetrator and the infant in comparison to other forms of infant abuse. First on slapping, the caregiver might experience a stinging of the hand and might feel guilt or remorse but shaking does not cause that effect. Second, shaking does not leave an external mark on the infant and the perpetrator may not be caught. Third, an infant who is shaken usually stops crying due to concussion-like brain injury, whereas a slap or a hit might cause a further increase in crying. Consequently, shaking reinforces a "positive feedback" cycle in which the caregiver is rewarded by an improvement in the behavior as the infant stops crying and there are no negative consequences for the caregiver [1].

Retinal hemorrhage is one of the characteristic signs of SBS and is known to result from an abrupt rise in intracranial pressure during shaking which was also seen in our patient. Birth trauma can also cause retinal hemorrhage; though, this resolves quickly. Sub-acute bacterial endocarditis, anemia, or thrombocytopenia can also lead to retinal hemorrhage, but these can be ruled out by clinical findings supported by results of laboratory investigations [9,10]. An overt effect of shaking is SBS. However, mild and less violent forms of repeated shaking can lead to the development of mental retardation, learning difficulties, or behavioral problems in later life.

The high prevalence of shaking as a typical caregiving behavior coupled with decreased awareness of its potential dangers indicate that prevention of SBS is an important public health challenge. There is extensive evidence that SBS does occur but how and why the guardians move from caring to abuse remains rudimentary. Thus, early prevention efforts targeting caregiver infant interactions around early crying are encouraging signs that prevention can work and contribute to giving otherwise normal infants a "good start" in life [1].

CONCLUSION

The diagnosis of SBS should be kept in mind while treating young infants with a history of symptoms similar to meningitis or epilepsy. This may help prevent further harm to the patient.

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