Evolution of epileptic encephalopathy in an infant with non-accidental head injury

Roshan Koul, DM, FRCPCH, Rajesh Poothrikovil, DNT, RPSGT, Faisal Al-Azri, MD, FRCP, Muna Al-Sadoon, MD, PhD.

ABSTRACT
A 5-month-old child, previously healthy, was hospitalized with frequent episodes of tonic seizures. The seizures were controlled with antiepileptic medication. However, the parents did not continue medications after discharge from the hospital. The child was admitted several times with breakthrough seizures. Over time the seizures became refractory to treatment. Neurometabolic work up and imaging studies for uncontrolled seizures revealed non-accidental head injury (shaken baby syndrome) as the underlying cause. His first EEG was normal and changed from normal to an epileptic encephalopathy pattern during his several admissions for uncontrolled seizures. From a normal child at the first admission, the child was severely regressed at the last admission. The present paper highlights the evolution of EEG changes in a child with non-accidental head injuries. This report also highlights considering non-accidental head injury as the underlying cause in younger children presenting with unexplained epileptic encephalopathy.

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**Case Report.** A 5-month-old male child was brought to the emergency services of our hospital with a 10-day history of repeated episodes of stiffness of all 4 limbs, with up rolling of eyes and frothing of saliva. Each attack lasted approximately 2 minutes. After the episodes, the child would develop postictal deep sleep and was noted to be tired and less active after waking up. He had several such attacks daily. He was seen in a peripheral hospital and routine blood tests including blood sugar, calcium, phosphorus, alkaline phosphate, and electrolytes were normal. A CT brain was reported as prominent cerebrospinal spaces. His development was normal for age before the onset of the symptoms. He was born at 41 weeks of gestation with a birth weight of 4.34 kgs, height of 51 cms, and head circumference of 37 cms. He had Apgar scores of 8 at one minute, and 9 at 5 minutes. His development was consistent with age. He was diagnosed at birth as congenital hypothyroidism and was on 37.5 micrograms thyroxine daily. His elder 3 siblings were normal. There was no history of seizures in the family.

In view of the repeated seizures, he was given intravenous phenytoin sodium (18 mg/kg) as an emergency and later observed in the ward. His examination was essentially normal other than mild hypertonia in the lower limbs and exaggerated deep tendon jerks. The blood work up for glucose, calcium, phosphorus, alkaline phosphatase, vitamin D, parathormone, tandem mass spectrometry, lactate, ammonia, and creatine kinase was normal. A routine EEG was normal (Figure 1). He was discharged on levetiracetam and tapering doses of phenytoin. The child presented after 2 months in an obtunded state. The parents had discontinued levetiracetam. The parents also denied any seizures at home. During this time, the child was seen abroad and topiramate was advised. However, the parents were not using any medication. The parents complained that the child had lost milestones and was not smiling. He had poor feeding, a weak cry and had lost weight. On examination, the child was not responding to any painful stimulus. There was no visual following with eyes open. The cranial nerves were normal. There was generalized hypotonia with a power of grade 3/5 in upper and lower limbs. All deep tendon reflexes were exaggerated. Some choreiform like movements were seen in the hands and sometimes in the legs. An urgent metabolic work up in the form of tandem mass spectrometry, lactate, ammonia, and creatine kinase was carried out to rule out a metabolic encephalopathy. All the results were normal. A repeat EEG showed bilateral high amplitude slow waves. A brain MRI revealed a right side subdural hematoma (Figure 2). In view of the subdural hematoma, other metabolic work up (serum copper, urine organic acids) and coagulation work up were carried out, which were all normal. Microscopic examination of the hair (kinked hair) was normal. In view of no underlying cause found, a suspicion of non-accidental head injury (shaken baby syndrome) was considered. Ophthalmic examination was normal. He was restarted on levetiracetam and observed in the ward. The parents refused further work up for shaken baby syndrome, particularly skeletal survey, and left against medical advice. Two months later, the child presented with tonic clonic status epilepticus and swelling of

**Figure 1** - Ten seconds EEG epoch, sleep record at 5 months age, shows a normal sleep EEG with mildly asynchronous sleep spindles.

**Figure 2** - Axial MRI brain (FLAIR), showing right subdural hematoma (arrow).
the right knee. The parents had discontinued the oral antiepileptic drug (levetiracetam). On this admission, he was given intravenous phenytoin sodium 18 mg/kg followed by daily maintenance. Since the seizures were not controlled, intravenous midazolam infusion was also started. Levetiracetam was administered via nasogastric tube. The epileptic status was refractory, and topiramate was also added to the treatment. The status epilepticus became controlled over 5 days time. On further questioning regarding the type of seizures, the parents reported that in addition to generalized stiffness, the child was getting brief myoclonus as well. This time, the parents agreed to further work up for shaken baby syndrome. The skeletal survey revealed an old healed fracture in the right radius (Figure 3), and a recent fracture of the right lower end of the femur (Figure 4). An EEG revealed features of epileptic encephalopathy in the form of hypsarrhythmia, and burst suppression (Figures 5 & 6). This EEG was carried out at the age 10 months and 20 days, 5 months after the first EEG. The brain MRI was repeated and it showed brain atrophy and resolution of the previous subdural hematoma (Figure 7). On monitoring the anthropometry, the head size increase was slow. At birth it was 37 cms, at first

Figure 3 - Plain x-ray of forearm shows old healing fracture at the lower end of the radius (arrow).

Figure 4 - Plain x-ray of right thigh shows recent fracture of the femur (arrow).

Figure 5 - Ten second sleep EEG epoch (10 months and 20 days age) of a hypsarrhythmia pattern, consists of high amplitude chaotic slow wave background with intermixed sharp waves.

Figure 6 - Sleep record at the age of 10 months and 20 days showing burst-attenuation pattern in a 10 seconds epoch. Bursts consist of high amplitude spike, polyspike, and slow wave discharges.
Discussion. Child abuse or maltreatment is seen throughout the world and can present as physical, sexual, or emotional abuse, and neglect. Over the years cases of child abuse have been reported from this region, considered to be uncommon in the past. Non-accidental head injury (shaken baby syndrome) is a form of child abuse and a relatively new diagnosis, predominantly affecting neurological functions. Shaken baby syndrome is mainly seen in infancy and these children present with neurologic features depending on the severity and continuation of the abuse. Irritability, bulging fontanel, hemiparesis, seizures, developmental regression, and even death are reported. Symptoms usually occur immediately after the insult, an important indicator of diagnosis. Shaken baby syndrome is under recognized and under reported and remains a diagnostic challenge. The diagnosis must be considered in any infant or young child who collapses with no obvious cause.

A careful medical and social history supplemented by appropriate investigations will help in diagnosis. The diagnosis is usually based on patient history that does not correlate with the clinical features. In addition, supportive laboratory and imaging studies suggest child abuse. Seizures at presentation are uncommon, though epilepsy is reported in 20% children on long-term follow-up. If shaken baby syndrome is diagnosed early and protection is given, chances of further injury and insult to the brain could be prevented. Any type of seizures could be seen at the onset in shaken baby syndrome. Infantile spasms and epileptic encephalopathy are rare. Only two reports describe an association between infantile spasms and shaken baby syndrome. We observed progressive EEG changes with each admission. From a normal EEG at first admission, hypsarrhythmia and burst suppression (epileptic encephalopathy) were noted on the last admission. Was this a natural evolution of the epileptic encephalopathy after the first abuse, or occurred after repeated abuses? We believe it was later, based on observations and different age long bone fractures. The parents did not continue antiepileptic medications after each discharge from hospital. This could be another form of child abuse and a reason for the seizures to become refractory. The child returned back to normal condition after first admission and had normal baseline EEG. After the second admission following seizures, the child started lagging behind in milestones. Long bone fractures of different ages favors repeated abuse of the child. The diagnosis of shaken baby syndrome was based on parents not giving a clear history, and poor correlation between history and examination. Non-treatment of a child with antiepileptic drugs at home and evidence of subdural hematoma on MRI confirmed the diagnosis of child abuse and shaken baby syndrome. All the causes for seizures including metabolic disorders were ruled out on detailed metabolic investigations when the child presented at the initial 2 admissions. Retinal hemorrhages were not seen in our child, important evidence to confirm the diagnosis of shaken baby syndrome. However, 20% of children with shaken baby syndrome do not have eye abnormalities. Other rare conditions like glutaric aciduria type I, and bleeding diathesis were excluded before shaken baby syndrome was diagnosed.

Figure 7 - Repeat MRI brain showing bilateral cortical atrophy and disappearance of subdural hematoma.
Evolution of the EEG in shaken baby syndrome in our case is an interesting electro-clinical association. It was by coincidence and with repeated EEGs on each admission that we could see the changes developing in EEG after each admission following seizures and insult. Evolution of EEG from normal to epileptic encephalopathy is not uncommon in this age group irrespective of underlying cause.\cite{10} Our case highlights the use of sequential EEGs in the management of seizures in infancy.\cite{11}

A wide range of neurologic sequelae have been seen in survivors of shaken baby syndrome in the form of cognitive and behavioral disturbances, cerebral palsy, blindness, and epilepsy. One third of shaken baby syndrome patients die, and 60% of survivors have moderate to grave morbidity.\cite{1,5,12} Severe cerebral atrophy is seen in these children on follow-up, as was found in our case.\cite{12}

In conclusion, we did not find any other cause in this child for epileptic encephalopathy other than shaken baby syndrome. Child abuse should be considered in the underlying causes when an infant presents with refractory seizures, and the known causes are excluded. It is worthwhile to perform skeletal survey to rule out non-accidental head injury.

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References


CASE REPORTS

Case reports will only be considered for unusual topics that add something new to the literature. All Case Reports should include at least one figure. Written informed consent for publication must accompany any photograph in which the subject can be identified. Figures should be submitted with a 300 dpi resolution when submitting electronically or printed on high-contrast glossy paper when submitting print copies. The abstract should be unstructured, and the introductory section should always include the objective and reason why the author is presenting this particular case. References should be up to date, preferably not exceeding 15.