

OCULAR MANIFESTATIONS IN SHAKEN BABY SYNDROME

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ABSTRACT

In the absence of external physical signs, child abuse is not easy to diagnose. Shaken baby syndrome is a unique form of child abuse where the only consistent external physical signs are its ocular manifestations. We report two cases which illustrate the typical presentation, with hallmarks of this syndrome, namely intraocular and intracranial haemorrhages. The visual prognosis of these infants are usually poor.

Keywords: child abuse, intracranial haemorrhage, intraocular haemorrhage, visual prognosis

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INTRODUCTION

Shaken baby syndrome is a form of child abuse where there is intracranial injury associated with intraocular haemorrhage in absence of external physical signs of head injury. In Singapore, an average of 53 cases of child abuse were reported to the authorities annually, most of these being cases with physical evidence of abuse⁽¹⁾. Although the entity of shaken baby syndrome has long been established, there has not been any reported cases of shaken baby syndrome in Singapore. We present 2 cases of shaken baby syndrome.

CASE REPORTS

Case 1

A 6-month-old Malay male infant was transferred to NUH Paediatric ICU from a private hospital on July 27, 1993 with severe raised intracranial pressure (ICP) and impending tentorial herniation. The child was noted to be lethargic 3 days prior to admission. There was no history of head injury or physical abuse. The baby's birth history was unremarkable and had a normal head circumference at birth.

On examination, the baby had a temperature of 38.4°C, heart rate was 187/min and BP was 133/82 mmHg. He was in a state of obtundation, with dystonic posturing of the limbs bilaterally. He exhibited brainstem releasing phenomena with lips smacking, mouthing movements and eye deviation. The anterior fontanelle was tense and sutures widely separated. Head circumference was increased, measuring 45cm. There were no external physical signs of injury.

Eye examination revealed a fixed and dilated left pupil. The right pupil was 4mm and reactive to light. There were bilateral widespread intra- and subretinal haemorrhages, more marked in the left. A massive vitreous haemorrhage was noted in the left eye, obscuring details of the disc, macula and retinal vasculature.

There was macula oedema in the right eye. The right optic disc was not swollen, however. The anterior segments appeared normal.

A head CT scan done revealed bilateral gross subdural effusion with evidence of fresh bleeding into the subdural space and adjacent to falx cerebri. In view of his condition on admission, an emergency subdural tap was done and 25ml of blood stained fluid was obtained. ICP was more than 15 cm of water. Because of evidence of rapid reaccumulation of subdural fluid despite repeated subdural draining, a subdural-peritoneal shunt was placed on 30 July. The shunt worked well and the patient's condition stabilised.

Subsequent ophthalmic review 6 days later revealed left rubeosis iridis, subretinal, peripapillary and vitreous haemorrhages and a total retinal detachment. The haemorrhage in the right eye had resolved. In view of the extent of ocular damage and poor visual prognosis, he was managed conservatively. He was later discharged and last seen on 17 August, exhibiting conjugate eye deviation to the left and Marcus Gunn pupil in the left eye. He was later brought away to another country and defaulted follow-up.

Case 2

A 3-month-old Chinese male infant was admitted to the paediatric ward on 22 March 1994 for one episode of non febrile tonic seizure lasting about 5 minutes while being rocked in a swing. His birth history was unremarkable and there was no previous history of seizures. Developmental milestones were normal. There was no family history of seizures. During admission, he had 2 other episodes of fits lasting less than a minute each, and was treated with rectal diazepam.

On examination the child was sedated, afebrile, heart rate was 128/min and blood pressure was 110/56 mmHg. There were no external signs of head or soft tissue injuries. The anterior fontanelle was bulging. His muscle tone was normal but the deep tendon reflexes were brisk. Fundoscopy revealed bilateral diffuse intraretinal, subretinal and vitreous haemorrhages. The retina was oedematous but the macula was spared. Disc details were obscured by the haemorrhages. A CT scan of the head was done showing subdural blood along falx cerebri and along the convexity of the right temporal lobe.

He was seen by a neurosurgeon who recommended that the child be managed conservatively. His condition stabilised and he was later discharged with phenobarbitone. When seen again on 12 April he seemed to be interested in large and colourful toys. Ophthalmoscopy showed a band of preretinal haemorrhage overlying the left macula with partial posterior vitreous detachment temporal to the macula. There was a band of

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subretinal fibrosis running vertically temporal to right macula. The intraretinal haemorrhages had resolved.

The child was last seen on 5 May. His visual acuity was 6 / 60 in the left and 2 / 60 in the right on Catford drum. There was a large collection of vitreous haemorrhage overlying the left macula. The right eye was stable. However, the VA prognosis is uncertain.

DISCUSSION

Although child abuse has been recognised for a long time, formal modern discussion of battered baby syndrome was not done until 1946 when Caffey reported six infants who suffered from the combination of subdural haematomas and characteristic bone lesions^(2,3). Since then it was known that the intracranial haemorrhages and characteristic bodily changes in these children were caused by trauma, and Kempe et al in 1962 first used the term "battered baby syndrome" to describe them. Gilkes and Mann in 1967 reported the occurrence of retinal haemorrhages in these cases, and Mushin reported 12 out of 19 battered babies with ocular damage have permanent vision impairment^(3,4).

In order to account for the absence of evidence of external head injuries in this group of infants, Caffey in 1974 introduced the concept of whiplash shaken infant syndrome, suggesting that violent shaking of the infants while grabbing them by their extremities or thorax resulted in whiplashing of the head onto the thorax. This, together with factors like relatively heavier infant heads, laxity of neck muscles, pliable sutures and fontanelles, supple infantile brains made them susceptible to tearing of the bridging vessels of the brain⁽⁵⁾. Such injuries often give rise to unexplained acute neurological signs, infantile fatalities and delayed neurological deficits and mental retardation^(2,5,6).

The cases presented illustrate the typical presentation of infants with shaken baby syndrome. There is usually very little direct evidence of trauma through admission of parents or guardians and no witness is available. Physical signs of head injuries or injuries to the soft tissues of the face and neck are lacking. Similar to other reported cases, the cases under discussion were below the age of one; and this diagnosis should be considered in all infants presenting with lethargy, irritability, poor feeding, seizure, failure to thrive, vomiting, hypothermia, bradycardia, hyper- or hypotension, respiratory irregularities and coma^(7,8).

The hallmarks of shaken baby syndrome, as shown in the above cases are intracranial haemorrhages, commonly subdural and / or subarachnoid and retinal haemorrhages. Because a history of shaking is often lacking, the diagnosis is usually based on these clinical and radiological findings. Ocular manifestations of the shaken baby syndrome consist of bilateral diffuse subretinal, intraretinal, preretinal, and vitreous haemorrhages and cotton wool spots. Other less common manifestations include intraretinal blood-filled schisis cavity, and bilateral symmetrical ring-shaped retinal folds^(7,10). Anterior segment is normal. Differential diagnosis of the ocular findings in this syndrome are Terson's syndrome, Purtscher retinopathy and retinal central vein occlusion⁽¹⁰⁾.

The mechanism of injury to the brain is thought to be due to rapid, repeated, to and fro acceleration-deceleration forces occurring in shaking⁽⁵⁾. While this alone is considered sufficient to cause serious intracranial injury and deaths^(5,11,12), recently there have been concepts suggesting that the shaken baby syndrome, at least in their most severe form, may have been ascribed to direct blunt impact⁽⁹⁾. However, Alexander et al concluded that shaking in and of itself is sufficient to cause serious intracranial injury or death, although a substantial number of cases in his study represent a combination of both shaking and direct impact⁽¹¹⁾.

The presence of retinal haemorrhage implies a shaking component of injury, as they are often seen in the shaken baby syndrome but not often seen in cases due to road traffic accident with skull fractures due to direct trauma⁽¹¹⁾. One must be aware of the prevalence of idiopathic retinal haemorrhages of the newborn infant in the first 5 days of life but they are rarely associated with intracranial bleeds. Other possible causes of intraocular haemorrhage include cardiopulmonary resuscitation, haematologic disorder, and ruptured cerebral vascular malformation or aneurysm⁽¹²⁾. Shaking manoeuvre is also postulated to cause displacement of vitreous, and traction produced may displace the neurosensory retina, thus creating folds⁽⁹⁾.

Caffey believed that habitual, manual, casual whiplash shaking of infants may produce an insidious, progressive clinical picture of small but cumulative intracranial and intraretinal bleeds that may result in mental retardation and impairment of vision⁽⁵⁾. Duhaime et al conclude that fatal cases of shaken baby syndrome are not likely to occur from the shaking that is associated with play, feeding or in a swing unless a child has predisposing factors such as subdural hygroma, brain atrophy, or collagen-vascular disease⁽⁸⁾.

Wilkinson et al⁽⁶⁾ noted that diffuse fundus involvement, vitreous haemorrhage and large subhyaloid haemorrhage were associated with more severe acute neurological injury. Long term neurological injury, however, was not predictable.

In conclusion, unless one is familiar with this and the other conditions which may produce the similar constellation of signs discussed, the infant with shaken baby syndrome may remain undiagnosed or be misdiagnosed in view of the paucity of external clinical signs, lack of and often unreliable history, and the varied clinical presentations. Although the visual outcome may range from normal to completely blind, the prognosis of shaken baby syndrome is generally poor^(5,10). A substantial number of infants will eventually have residual neurological deficits and permanent impairment of vision^(3,10,12).

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