Shaken Baby Syndrome

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SUMMARY

Shaken baby syndrome represents a specific form of Abused child syndrome. Injury usually concerns the baby’s head and the brain and it is caused by thoughtless treatment accompanied by harsh shaking movements of the head and neck. It can cause a contusion of the cervical spinal cord, a rupture of the bridging veins, intracranial bleeding and a brain tissue impairment either due to the direct axonal damage, or namely by hypoxic-ischaemic insult. The development of the lesion can be fatal, or it can result in permanent impairment of the motor system, in mental or sensory deficits. Occurrence of the syndrome in the Czech Republic is not known, foreign data give 25 cases per 100,000 children below one year of age. Injured babies represent over 1% of those hospitalized at paediatric units of intensive care and more than 10% of the death rate at those departments. Proved abuse has forensic consequences, however, convicive evidence can be difficult to obtain. Article gives a concrete case of a boy with the diagnose Shaken baby syndrome.

Key words: child abuse, subdural haematoma, retina haemorrhages, convulsions in children.

CASE STUDY

Our intensive care unit admitted an eight-month-old boy with vomiting, convulsions, loss of consciousness and hypotonia. The family history was uneventful, the child was from a third, normal pregnancy, born at term by spontaneous delivery, vertex presentation. Birth weight was 2,500 g, and the child adapted well after delivery. The boy was breast-fed for 2 months and then changed to formula milk. Community paediatrician assessed the child’s development as slightly delayed. With the exception of a solitary respiratory infection, the child had not been ill. The social situation of the family was complicated: the parents of the boy lived apart, the mother, however, could not cope with childcare, and the court placed the boy into the father’s custody.

The symptoms of the current disease appeared 4 days before admission to our clinic and were manifested by repeated vomiting. The child was without fever. During examination in the outpatient department increased fluid intake was recommended and a follow-up appointment made for the following day, which the father skipped, because the child’s state had improved. One day later there was a transient rise of temperature. The day after that the temperature was normal again but, from the morning, the boy refused food and drink; in the afternoon the child had convulsions of the whole body, lasting approximately 2 minutes without the child turning blue. After the convulsions the child, generally atonic and unresponsive, was brought to our clinic and admitted. On admission, we stated general hypotonia and impaired consciousness (GCS 5-6) without other characteristic findings; the anterior fontanelle was soft, there were no visible meningeal symptoms. In the first hours after admission, generalized convulsions repeated twice, each attack lasting approx. one to two minutes. A peripheral venous cannula was inserted and midazolam boluses administered repeatedly; phenobarbital was then introduced intravenously. No further convulsions were recorded, and the boy’s state stabilized gradually. In the first six days the patient received ceftriaxon and acyclovir due to suspected encephalitis, which was later excluded. Laboratory investigations showed white blood cells elevated to 23.8 x 10^9/l, elevated lactate levels 3.4 mmol/l, elevated D-dimers 991 µg/l, CRP was normal – 0.7 mg/l. Lumbar puncture produced sanguinolent cerebrospinal fluid with the cytological finding of 106/3 mononuclear cells, 10/3 polymuclear cells, and 48 640/3 erythrocytes. Centrifuged cerebrospinal fluid transformed into clear fluid, spectrophotometric investigation did not point to artificially
introduced blood during the puncture, but to subarachnoid haemorrhage several days old. We performed a brain CT, which revealed a mild cortical atrophy in the frontal area and slight widening of the frontal subarachnoid spaces. MR investigation traced the presence of hygromata along a part of the circumference of the right brain hemisphere and the left frontal lobe, with diffuse changes of the signal in the supratentorial direction, accompanied by atrophy of brain tissue. Signal changes were proof of retarded myelination and possible contribution of hypoxia. EEG investigation repeatedly produced a grossly abnormal recording; after the disappearance of the convulsions, the neurologist evaluated the state as central hypotonic syndrome. The investigation of the fundus oculi revealed numerous peripapillary haemorrhages, which were partly healed by the time the child was discharged from hospital. The follow-up investigation of the cerebrospinal fluid 4 days after the first puncture showed a practically normal cytological finding, with the exception of persisting elevated erythrocyte count at 340/3, but even here the dynamics had a markedly descending trend towards normal levels.

Due to the suspicion of the child having been affected by bad family care, after 15 days of hospitalization and the introduction of oral phenobarbital therapy and after an agreement had been reached with the father, the boy was transferred to an infant nursing home to receive the necessary care while social services continued their investigation.

Later the court issued a judgement for the compulsory placement of the child in an institution and conducted criminal proceedings against the father. The latter was then discontinued due to lack of evidence; the father constantly denied having maltreated the child and demanded that the child be returned into his custody. The judicial proceedings are still under way.

EEG on follow-up has shown no sign of post-traumatic epilepsy and anti-epileptic therapy was withdrawn after several months. The motor and mental development of the boy aged 2 is within the broad normal range.

CT of the brain on follow-up six months later revealed a residual subdural hygroma; at the age of 2 years, the subarachnoid spaces are still wider and the ventricular system more spacious in the presence of slight diffuse atrophy of the brain.

**DISCUSSION**

Although attention has been paid to the issue of the abused child syndrome for a long time now (1-3), SBS incidence in the Czech Republic is unknown. A Scottish study refers to an incidence of about 25 cases per 100,000 children under the age of 1 year (4), another study from the USA states that patients diagnosed with SBS have accounted for 1.4 per cent of all admissions and 17 percent of all deaths in the paediatric intensive care unit (5).

The mechanism of brain injury caused by rough shaking was described first by Guthkelch in 1971 (6), the term “shaken baby syndrome” was first used by Caffey in 1972 (7). Since then it has been defined as a physical injury in the young child (up to 2 – 3 years of age), which is not caused by accident or by beating, accompanied by acute encephalopathy, subdural and retinal haemorrhages. Ambiguities in patient history are common. Although other signs of injury, especially injury to the neck and spinal cord, may be present, and manifestations of the syndrome may be combined with findings of other signs of injury caused by physical attacks, SBS is characterized in particular by a triad of symptoms: subdural (or subarachnoid) and retinal haemorrhage and brain damage (8).

Unlike abuse associated with battering (battered baby syndrome), isolated episodes of SBS need not be accompanied by apparent signs of external injury (haematomas, excoriations, fractures, subconjunctival haematomas, ecchymoses of eye-lids) (9).

Since the mechanism of onset of SBS cannot be restricted to mere shaking, other more general terms have been proposed to name the syndrome, like “inflicted head trauma”, “inflicted childhood neurotrauma”, “childhood head injury”, “inflicted traumatic brain injury”, “nonaccidental neurotrauma”, “nonaccidental head injury”, “shaking-impact syndrome”, etc., pointing generally to brain injury caused by rough handling of the child, not by accidental injury. The original term, however, has been represented most frequently in contemporary literature.

The current concept of SBS onset draws on the idea that head injury is not due to direct blows to the head, but to rapid acceleration - and mainly deceleration - of head and neck movements, and/or by the effect of sharp rotational movements of the head around its centre of gravity. The consequence can be the stretching and rupture of the bridging veins of the brain and occurrence of subdural haemorrhage and multiple retinal haemorrhages. There is simultaneous damage to the cerebral parenchyma, caused according to earlier belief by diffuse axonal damage, but according to more recent reports more probably by ischaemic insults.

There are certain predisposition factors to SBS, including lack of parental education and unfavourable family social and economic circumstances. From the point of view of the child, the occurrence of the syndrome may be supported by circumstances leading to anomalous child behaviour, especially immaturity, frequent crying and colic, difficulty in feeding, retarded development or chronic disease. Apparently the most common situation in which the child is injured during SBS is the inappropriate reaction of the impatient parent or other child-minder to irritating attacks of child restlessness.

The most common symptoms noted in the child’s history by the examining doctor are convulsions, loss of consciousness, apneas, increased irritability, lethargy, vomiting and apnoic pause.

Further development may, in milder cases, lead to a complete resolution followed by normal development of the child. In more serious cases, the resolution of the acute stage may be followed by long-term consequences such as mental retardation, motor disorders and hearing and sight disorders – in the extreme cases even blindness caused usually not by damage to the retina, but rather by central brain lesions. In the most severe cases, SBS ends in death in the acute phase of the disease. In one of the largest studies carried out in Canada, 19 per cent of children with SBS died, 55 per cent had severe neurological consequences after being discharged from hospital, and only 22 per cent of the patients were released without signs of neurological damage (10).

Due to the often unclear patient history, it is difficult, in all the studies, to verify the clinical concept of onset of SBS. That is why it has been questioned recently (11), pointing to the fact that a similar set of symptoms could be caused also by other mechanisms, e. g. by an accidental fall from a small height, mechanisms occurring during cerebral hypoxia, haemorrhagic disease or, rarely, during congenital metabolic disorder (12-14).

The validity of the hypothesis of the origin of SBS has, however, been verified both in animal studies (15) and in studies presenting cases confirmed by independent witnesses of rough handling of children, or by later confession of parents of having maltreated the child (16, 17). Symptoms analogous to SBS have been observed in immature newborns in whom prevention of bronchopulmonary dysplasia was attempted by applying the technique of intensive physiotherapy of the chest, associated with sharp movements of the head (18). It has also been proven that the frequently encountered slight prolongation of coagulation indicators is caused by the rise of
multiple microthrombi during head injury and is therefore usually a secondary finding, not a pre-existing inborn coagulation disorder (19). What is disputable is therefore not the existence of SBS, but rather the ability of doctors to provide reliable proof that the clinical syndrome compatible with SBS was caused in a specific case by this precise mechanism. The only thing available is data showing which findings increase considerably the probability that a case is SBS (20, 21). What is important is also the presence of the basic triad of symptoms; also child abuse is indicated especially by bilateral retinal haemorrhages extending up to the periphery of the retina or even preretinally.

The examination of a child with suspected SBS should include, in the first place, a neurological evaluation, examination of the fundus by indirect ophthalmoscopy (and providing photographic documentation in the case of a positive finding), and CT or MR of the brain.

Long-term care for surviving children involves cooperation between the paediatrician, developmental neurologist, social worker and sometimes also other specialists.

On admission to the ICU, our patient had all the symptoms of the classical SBS triad – impaired consciousness accompanied by convulsions, signs of subarachnoid and subdural haemorrhage and bilateral retinal haemorrhages. Also compatible with the diagnosis of SBS are the social circumstances of the family and data on the mild retardation in the child's development.

The fast healing of the ophthalmic finding pointed to acute, and probably single incidence of retinal haemorrhages.

Definite confirmation of diagnosis of SBS is needed to allow the search for the best possible solution to the social situation of the child and, if necessary, also for forensic purposes; it is, however, very difficult. In the absence of admission by the parents of having maltreated the child, and in the absence of independent witnesses, we maintain, in this particular case, a strong conviction about the validity of the suspected diagnosis.

CONCLUSION

Rough handling of the child may lead to brain damage with symptoms of "shaken baby syndrome". The outcome of babies with the syndrome may be severe. Proving abuse is always difficult and the cases where we suspect SBS are extraordinarily complicated both from the social and forensic points of view. The probability of SBS is increased by a triad of symptoms – impaired consciousness, subdural haemorrhage and retinal haemorrhage. Especially bilateral intraretinal haemorrhages affecting the macula and peripheral retina point strongly to SBS.

Abbreviations

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<tr>
<td>CRP</td>
<td>C-reactive protein</td>
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<tr>
<td>CT</td>
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<td>electroencephalogram</td>
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<td>intensive care unit</td>
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<td>magnetic resonance</td>
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REFERENCES

Comment on Paper by I. Peychl on the "Shaken Baby Syndrome"

The Shaken Baby Syndrome (SBS), as a form of physical child abuse, deserves the special attention of the pediatric community due to its very dangerous possible consequences, since it may imply not only irreversible impairment of the somatic and mental development of the child, but often also the child’s death. Although this type of injury sometimes even may be suspected in older pre-school children, such diagnosis is rare. If it does occur, this is usually in the context of multiple injuries to which a severely physically maltreated child has been exposed for a long period of time. Such a symptomatic picture points out a very advanced process of abuse with all its external and typical signs. Assessment of SBS will, however, usually involve very small children, usually younger than 18 months, although it is most often diagnosed in infants younger than 6 months since toddlers already have a partly developed ability of motor resistance to being handled against their will. Children under one year of age are therefore at greater risk of SBS due to their defencelessness and absolute exposure to the impulses of their parents. The early developmental phase also means that a severe and usually repeated injury of the brain and cervical spine may easily cause a wide range of damage that may lead even to disability (e. g. permanent damage to sight and hearing receptors).

The absence of external injuries allows parents to explain the child’s condition by accidentally adverse circumstances but impact of SBS is so typical that the attention of the diagnosing pediatrician should not be diverted towards possible explanations offered by the parent. On the one hand, the statement made by the parents does not usually meet the criteria of realistic explanation of the child’s injury from the point of view of:

- the place where the injury has occurred,
- the way of the injury,
- the applied force required to cause a certain symptom,
- the scale of the injury,
- the type of symptom,
- the logic of the injury circumstances.

On the other hand, it is typical for a perpetrator to insist on unrealistic explanations to protect Ego, even at the cost of sometimes escalating and naive fabrication. It should be remembered at this point that very small children do not have much opportunity to come by such serious injuries because of the developmental constraints on their motor skills. Nor can serious neglect of basic care of small children cause the configuration of the four symptomatic groups typical of the differential diagnosis in the case of suspected SBS (retinal haemorrhages, subdural or subarachnoid haematomas, symptoms pointing to central damage and manifested, inter alia, by impairment of breathing, activity and consciousness or by epileptic seizures and absence of external signs of injury). The author’s description of the typical SBS course of the child’s in-department treatment and his meticulous summarization of all examination results enable the reader to follow the development and treat of various kinds of symptoms as well as the presentation of convincing arguments for the assessment of the shaken baby syndrome.

Studies conducted in recent decades on child abuse victimizing children younger than 3 years of age have shown that whenever severe and sudden injury or death occur under dramatic circumstances in those children they are usually inflicted by the closest caregivers – often the parents – with the mother being the most common perpetrator. Small children usually have young parents, and it is within this group that one can find persons characterized by extreme and generalized immaturity with signs of disharmonic development of their personality structure, impulsive behaviour and inclination towards uncontrolled expression of apparently hostile, or even explosive behaviour, persons who have been frustrated over a long period of time and who act in circumstances of great stress. Their temperamental characteristics on the one hand, and the previous course of their lives on the other, have made the acceptance and fulfillment of their parental role more difficult. And at the same time they have to rely fully on their own, meagre and inadequate parental skills and knowledge of child development, because a typical feature of child abusers is long-term solitude, sometimes social and inter-generational isolation and inability to acquire and apply the experience of others. This is illustrated very accurately by the case study and its information about the family’s situation.

SBS perpetrators, however, differ from parents developing a system of the child maltreatment. They are impulsive rather than sadistic, helpless rather than violent, and their relationship with the child is marked more by dismay over the overload of duties and at their own helplessness than by the need to express open hostility. The parents who admit their improper treatment of the child usually defend themselves by claiming that the continuous and inconsolable crying disturbed them so much that they applied violent treatment to make the child calm down for at least a while. Such parents perceive psychotherapeutic help as a relief from the feeling they have had of helplessness and exhaustion.

Some groups of small children are more at risk of SBS. Their developmental immaturity makes them more difficult, sometimes very demanding on the parents who are only just coming to terms with their new role. In this respect, too, the presented case study appears to
be very illustrative of the shaken baby syndrome (birth weight at the lower border of the normal range and delayed psychomotor development of the child).

The brief patient history data presented by the author of the article imposes a question why, in such a high-risk family, has there been no timely intervention by the social services. Their assistance and support would have probably reduced the risk of onset of SBS, or may have even prevented it. Primary prevention, carried out by the simple method of distributing to risk parents (especially to mothers in their last months of pregnancy) leaflets on causes of SBS and its danger for the development and life of the child, also containing instructions on simple techniques of coping with irritable reactions of small children, seems to be very effective and is used widely, especially in English-speaking countries.

The article and case study could be useful as material for subsequent presentations of the diagnostic process and course of treatment especially in the Czech context, in the postgraduate training of both pediatricians and pediatric nurses, child clinical psychologists, social workers and other specialists who should be sensitive to issues of child abuse.

REFERENCES

Translation: Naďa Abdallaová