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Cover Page Footnote

This scientific material is a part of a larger paper, a Ph. D. thesis, currently under development by Duncea Roxana-Maria, M.D., Ph. D Student at the Carol Davila University of Medicine and Pharmacy of Bucharest with Prof. Vladimir Belis, M.D., PhD., as thesis coordinator. The thesis has the following title: "The role of forensic investigation in diagnosis, prognosis and estimation of posttraumatic interval in TBI". All authors have read and approved the final version of the text and also declare no conflicts of interest

Review

Forensic and clinical diagnosis in "shaken baby syndrome", between child abuse and iatrogenic abuse

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Abstract

“Shaken baby syndrome” in child abuse cases is a challenge for pediatrician and forensic experts, often a diagnosis of exclusion, with overwhelming moral and legal implications. Diagnosis is based on: subdural bleeding, rupture of retinal vessels, traumatic diffuse axonal injury with diffuse brain encephalopathy in the absence of external traumatic injuries and anamnesis data of an accidentally head injury.

Microscopic findings in diffuse axonal injuries were initially considered as a specific traumatic effect due to unrestricted movement and accelerated rotation of the head. Immunohistochemistry of beta amyloid protein precursor is gold standard method for identifying pathological diffuse axonal lesions, which is however non-specific in brain trauma.

In the diagnosis of this syndrome pediatricians and forensic examiners must take into account the particularities of each case, avoiding scientific speculation, to intuit controversies and always be familiar with the differential diagnosis.

Keywords: forensic, child abuse, iatrogenic abuse, shaken baby syndrome



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Introduction

Today, in the medical world the "Shaken Baby Syndrome" (SBS) is both a challenging diagnostic for pediatrician and forensic experts and a controversial one. The challenges consist of a difficult diagnosis on one hand, and on the other hand the need to consider the complete medical history and police investigation data with careful research of clinical, laboratory and forensic autopsy findings, to reveal the cause of death. In order to establish a definite diagnosis, the pediatrician and forensic expert should exclude other causes of death. The correct diagnosis of child abuse cases is extremely difficult, often is a diagnosis of exclusion, with overwhelming moral and legal implications on the family and society.

Discussion

SBS occurs in children under 3 years, the peak incidence is in boys in the first year of life. The social family environment correlates with the occurrence of SBS, higher in poor families and younger parents with lower educational levels. Death occurs in 10-40% of cases, and survivors can develop serious neurological sequelae: behavioral disorders, cognitive impairment, blindness, and seizures.

- *Pathogenesis*

Classic "Shaken Baby Syndrome" was defined by Guthkelch and Caffey, as a form of physical abuse on a little child, due to a traumatic event in which the baby is shaken, able to generate rapid movement acceleration/ deceleration movement of the skull in relation to the trunk which results in traumatic disruption of the cerebral "bridged" veins associated with subdural bleeding/subdural hematoma, rupture of the retinal vessels with consecutive retinal hemorrhage and traumatic Diffuse Axonal Injury (DAI) with diffuse brain encephalopathy (1, 2). The triad diagnosis enumerated is associated in some cases with old or

recent metaphyseal bone lesions of long bones, rib bone lesions, in the absence of external traumatic injuries and anamnesis data of an accidental head injury.

The "tin-ear" syndrome, first described by Hanigan et al. in 1987, represents a type of SBS with the following major characteristics: contusion at the auricular level, edema on the ipsilateral side with retinal hemorrhages and a subdural hematoma located on the ipsilateral side as well. The typical pattern and onset of the injury might be described by a blow that acts on one side of the neck, in turn generating an accelerated rotation of it around the axis of the neck. This impact must have a certain degree of speed and force, thus being capable of generating serious intracranial injuries but with minimal or absent signs outside (3).

Duhaime et al., based on personal observations and the mechanical experiments that demonstrated that DAI occur not just by shaking and impact but that even minor head injury is needed, concluded that the specific lesions of the SBS are caused from both the impact of head and the brain acceleration. The absence of external injuries is due to a head impact achieved through a soft cloth (blanket, pillow, duvet) (4, 5).

In 2001, Geddes and al. concluded that, on brain tissue in children said to have died as a result of SBS, hypoxia and not necessarily the sheer trauma by itself, combined with an increased intracranial pressure were developing an increased permeability of brain's immature vessels and thus generating the subdural hematoma and retinal bleedings (6). Subdural and retinal hemorrhages were discovered by Geddes mostly lesser in size compared to traumatic bleeding. In a few cases they found DAI in cranio-cervical junction and most of histological brain tissue was similar to the brain tissue in pathological death. As a direct consequence of these findings, there have been many calls for justice of persons convicted of child abuse.

In contrast, other studies have revealed that shaking is not sufficient to produce rupture of the dural "bridged" veins and the rupture of axons, because shaking would likely produce severe neck trauma prior to brain damage (7). Terminology has varied over time and has been frequently modified in the light of new discoveries:

- a) 1970 - "Shaken baby syndrome"/ "Whiplash shaking injury";
- b) 1988 - "Shaken Impact syndrome": brain damage occurring after shaking with impacts through soft surfaces;
- c) 2009 - American Academy of Pediatrics (AAP) - "Abusive Head trauma"; AAP recommends using this more general term encompassing all head trauma as a result of intentional infant abuse, regardless of the production mechanism.

- *Clinical and paraclinical diagnosis*

Nonspecific symptoms: from mild food disorders (vomiting, refusal of food) and various neurological manifestations (drowsiness, apathy, lethargy, convulsions) to severe respiratory disorders. The medical team must perform eye exams and an extended ophthalmoscopy exam to detect retinal hemorrhages and to specify their extension.

Extensive laboratory investigations are recommended to exclude vitamin D deficiency, coagulopathy, genetic disorders, infectious disease, meningo-encephalitis, etc. X-rays of long bones and CT examination are also recommended. When possible, MRI, which is more sensitive than CT in determining diffuse parenchymal lesions and more efficient in assessing posttraumatic interval, should be executed.

- *Forensic autopsy*

In cases followed by death, forensic autopsy is mandatory. Frequently the external examination finds no injuries. Subdural bleeding can be distinguished on a macroscopic level as the subdural bilateral hematoma is often in low amount, present as a thin hematic blade in

children under 1 year and may increase in size at children become older. Microscopic examination of the brain tissue reveals diffuse axonal injury, which occurs most often in children over 1 year, highlighted using immunohistochemical determination of beta amyloid precursor protein (b-APP).

In cases where SBS is suspected, differential diagnosis should identify any situation of repeated physical abuse of the child. The forensic examiner should exclude old hemorrhages in dural spaces, leptomeningeal space, cerebral parenchyma, medullary in the spine or in retina. Microscopic examination of the tissue with Prussian blue staining will highlight chronic repair processes in brain due to macrophages and activated astrocytes.

Autopsy diagnosis is difficult and must link data provided by police with stories of the family and child medical history from birth. Since the classic diagnose triad: hematoma at the subdural space, spots of hemorrhages in the retinal layer and cerebral diffuse encephalopathy is nonspecific and none of the clinical or autopsy findings is pathognomonic, the diagnoses must be made with extreme caution considering both the clinical differential diagnoses and possibility of accidental trauma suffered by the child. For example, Plunket has researched children with accidental drops and discovered that the classical triad used to diagnose SBS may be found after a fall from the same level, followed by a free interval (8).

The interpretation of subdural hematoma found in necropsy requires maximum caution. Subdural hematoma can occur in other kinds of trauma (accidental falls, childbirth, after surgical procedures) and/or may be due to vascular causes, hematologic disease, metabolic disease or infectious. Subarachnoid hemorrhages can be also caused by: coagulopathy, vasculopathy, neonatal alloimmune thrombocytopenia, vitamin K deficiency, cerebral edema, intracranial venous thrombosis.

Intracranial venous thrombosis associate focal edema, focal hemorrhages: sub arachnoidian, sub spinal, subdural (9). Subdural hematoma appears also in infectious diseases: meningitis, hemorrhagic encephalitis.

New research on the etiology of subdural bleeding in young children reveals that subarachnoid and subdural hemorrhages in small amount can occur in young children with extended subarachnoid space, without correlations with head trauma. Subdural benign space enlargement occurs in children with birth macrocrania, and in most cases; it resolves spontaneously by the age of 2-3 years, with a good prognosis and normal neurological development. Subarachnoid hemorrhage can occur at birth (natural or cesarean) in small amounts with posterior location, with spontaneous resorption in about one month after birth. These postpartum hematic accumulations become symptomatic in traumatic births.

Alternative explanations of subdural hematomas in children are: dural venous plexus are more extensive, with thin, drill walls that are playing a role in the absorption of cerebral spinal fluid (CSF) and may lead to chronic subdural hematoma. Intradural leakage from vessels can generate hypoxia and increased intracranial pressure with minimal intradural and subdural bleeding developed posterior frequently. Recent studies have shown that relatively frequently cerebral hypoxia and intracranial hypertension are associated with subdural bleeding (10, 11).

- *Differential diagnosis of retinal hemorrhages.*

Hemorrhages in retina and optic nerve are due to various causes related to increased intracranial pressure: infections, hypoxic encephalopathy, cerebral edema by post traumatic etiology, metabolic or idiopathic. Early retinal hemorrhages are small macular, located in one layer of the retina, but those with severe evolution extend to all layers of the retina and vitreous humor, being distributed more peripherally at ora serrata.

- *The differential diagnosis of encephalopathy associated with diffuse cerebral axonal injury*

Diffuse cerebral axonal injuries are characterized by multiple focal white matter lesions, approximately 1-15 mm, with a characteristic distribution. Microscopic extent of damage usually spreads beyond the lesions detected by imaging.

DAI are suggested in any patient showing clinical symptoms disproportionate to the CT findings. In CT scan on acute phase, only 20% of those with DAI show characteristic petechial hemorrhages situated at the junction of gray matter and white matter, in the corpus callosum and brainstem. For this reason, when CT is negative and clinical symptoms are suggestive, an MRI is recommended. MRI displays many multifocal areas with a pattern of abnormal signal characteristics, such as being bright on T2-weighted images, in the white matter at the corticomedullar junction of the temporal areas, parietal areas or in the splenius of the corpus callosum.

Diffuse axonal injuries were initially considered as a specific traumatic effect due to unrestricted movement and accelerated rotation of the head. Many studies published recently show that DAI may correlate well with other more common diseases that affect the white matter. Amongst them are the demyelinating diseases or conditions such as sepsis that induce brain ischemia and hypoxia. As such, DAI is a generic term that suggests any type of axonal injury both with traumatic and non-traumatic patterns involved.

- *Microscopic diagnosis of diffuse cerebral axonal injury*

DAI diagnosis other than by histopathology methods remains a major challenge. Microscopic changes are various and proportional with the posttraumatic interval. Early, in the first 2 hours post injury, the immunohistochemical methods can highlight b-APP, that can persist for up to 1 month posttraumatic. This technique is extremely sensitive in the detection of DAI

and highlighting the rapid expansion in axonal pathology (12, 13). As such, immunohistochemistry of b-APP is considered the gold standard regarding the best method for identifying clinical and pathological diffuse of axonal lesions. However, the accumulation of b-APP in axons is non-specific and does not occur exclusively in trauma and has also been described as a result of other types of brain damage, including hypoxia/ischemia (14, 15).

In the first 12-hours posttraumatic, axonal retraction "balls" using different silver impregnation techniques, such as Palmgren™, can be revealed. But these changes are also nonspecific for traumatic DAI, as they can appear in non-traumatic brain injury: cerebral infarction in stroke, cerebral parenchymal hematoma etc.

At the two day threshold, posttraumatic mark, it is possible to detect axonal disruptions with reactive microglia, identified with CD68. At the 7th day posttraumatic mark axonal retraction "balls" are heterogeneous and the positivity of reaction for b-APP is reduced (16, 17).

- *The differential diagnosis of fractures commonly associated with SBS*

Both forensic experts and medical clinicians must render with caution metaphyseal fractures of the long bones and to consider possible differential diagnoses with metabolic diseases and chondrodysplastic disorders: vitamin D deficiency, calcium severe deficiency, hypophosphatemia, Jansen Syndrome.

Vitamin D deficiency (Rickets) can cause osteopenia, skull deformation changes, swelling of sternal-rib joints and characteristic metaphyseal lesions in the long bones, pathological fractures. These radiographic changes are transient, asymptomatic and can be interpreted as signs of child abuse in case of a wrong clinical diagnosis.

Conclusions

Medical diagnosis is always at a crossroads of art and science and requires a multidisciplinary approach, objectivity, and consistency with ethics and professional deontology. In the diagnosis of SBS pediatricians and forensic examiners must consider the particularities of each case, interpret literature with caution, avoid scientific speculation, understand controversies, and be familiar with the differential diagnosis of this syndrome.

It is therefore necessary to realize that child abuse is unfortunately a reality, but considering the social, moral and legal aspects of this diagnosis, SBS diagnosis should only be made in certain cases that are unequivocal. Forensic probation of this diagnosis is difficult and requires close collaboration between different specialties in order to avoid iatrogenic abuse, considering that no part of the SBS diagnostic triad is pathognomonic.

Acronyms and abbreviations

SBS: "Shaken Baby Syndrome"; DAI: Diffuse Axonal Injury; b-APP: beta Amyloid Protein Precursor. CT: Computer Tomography; MRI: Magnetic Resonance Imaging; CSF: Cerebral Spinal Fluid.

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This scientific material is a part of a larger paper, a Ph. D. thesis, currently under development by Duncea Roxana-Maria, M.D., Ph. D Student at the Carol Davila University of Medicine and Pharmacy of Bucharest with Prof. Vladimir Belis, M.D., PhD., as thesis coordinator. The thesis has the following title: "The role of forensic investigation in diagnosis, prognosis and estimation of posttraumatic interval in TBI". All authors have read and approved the final version of the text and also declare no conflicts of interest.

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