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Cerebellar lesions in pediatric abusive head trauma

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ABSTRACT

Pediatric abusive head trauma (AHT) or non accidental head trauma (NAHT) is a major cause of death from trauma in children under 2 years of age. Main etiological factor for non accidental head trauma is shaking a baby, causing brain injury by rotational head acceleration and deceleration. The consequent brain damage as shown by magnetic resonance imaging (MRI) is subdural haemorrhage and to a lesser extent parenchymal injuries of variable severity. Involvement of the cerebellum has very rarely been described.

We report the clinical history and the development of cerebral magnetic resonance imaging findings in two children with serious brain injury following probable shaking who presented the typical "triad" with subdural haematoma, retinal haemorrhage and encephalopathy. We want to draw attention to cerebellar involvement characterized by cortico-subcortical signal alterations most prominent on T2w images following diffusion changes during the acute period. We discuss cerebellar involvement as a sign of higher severity of AHT which is probably underrecognized.

1. Introduction

The current definition of pediatric abusive head trauma (AHT) includes an injury of the skull/intracranial contents of a child up to the age of five years due to intentional abrupt impact and/or violent shaking.[1] This term is accepted for the intracranial and spinal lesions in abused infants and children.[2] NAHT (non accidental head trauma) is often used synonymously. Shaking is supposed to be the main mechanism of death from AHT in babies between three and six months.[3,4] In Western countries the incidence of AHT in children under one year ranges from 14 to 28 per 100.000 live-births and AHT represents the major cause of head injury in children under one year.[5,6]

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AHT is characterized by a triad of subdural haematoma, retinal haemorrhages and encephalopathy.[3,4,7] Additionally there is a high incidence of cervical spine injury.[8] AHT can be caused by shaking alone. External signs of injury are often missing and symptoms can be nonspecific and similar to viral infection or minor illness. Only severe cases show life-threatening symptoms.[2,9–11]

The consequent brain damage as revealed by magnetic resonance imaging (MRI) is well described mainly consisting in subdural haemorrhage and subsequent development of hygroma. To a lesser extent reports refer to cerebral parenchymal injuries. And to our knowledge to date MRI findings showing involvement of the cerebellum in AHT have very rarely been reported and not specially described with respect to follow-up.

In many cases, the crucial factor for the outcome of affected children is not the subdural and/or subarachnoidal bleeding but the subsequent effects and severity of the intraparenchymal injuries.[2]

We report two cases of children with AHT and present their clinical history and the evolution of their severe brain lesions as seen by MRI and draw attention to the associated cerebellar lesions. We collect evidence that additional cerebellar lesions in AHT indicate a higher severity of the brain damage.

2. Case 1

This boy was a healthy term born first child of young parents. Apgar score was 8/9/10 and birth weight 2950 g. First milestones were normal. He was admitted to our hospital at the age of 4 months with apathy, bradypnoea, facial haematoma in stripes on the left ear and cheek. No specific trauma was reported by the caregiver, who stated that he had tried to feed the baby who was not reacting and had a seizure. He then called the emergency and started resuscitation. The emergency physician found a baby breathing insufficiently with bradycardia and no reaction to stimuli.

Computed tomography (CT) brain scan on admission showed bilateral subdural haematoma and some evidence for bilateral brain oedema. Intracranial pressure (ICP) was raised to 30 mmHg and external drainage of the cerebrospinal fluid was performed. Ophthalmological investigations showed multiple bilateral retinal and subretinal haemorrhages.

MRI on the next day (Fig. 1a, b, e, f) showed widespread multifocal cortico-subcortical changes of the supratentorial parenchyma, on diffusion weighted imaging (DWI), not corresponding to specific vascular territories and sparing the diencephalon. In addition, there were bilateral widespread diffusion changes of the cerebellar hemispheres (Fig. 2b). Bilateral subdural haemorrhage hyperintense on T1weighted (T1w) images indicated a minimum age of 2 days (according to Ref. [12]). There was no evidence for spinal lesions.

No additional abnormalities were seen on whole-body MRI. Metabolic testing for glutaric aciduria type 1 and Menkes disease was normal.

MRI three weeks after admission (Fig. 1c, g) showed expansion of the subdural space and evidence for formation of neo-membranes in the subdural space indicating a transformation from subdural hygroma into chronic encapsulated subdural haematoma. The telencephalon showed severe cortical and subcortical atrophy with in part multicystic transformation. T2w hyperintensity was now evident in the former areas of DWI changes, while the diencephalon did not show clear abnormalities. Signal changes were also seen in the cerebellum affecting the cerebellar foliae cortico-subcortically with predominance in the depth of the foliae (Fig. 2c).

Last MRI at the age of 11 months still showed chronic encapsulated subdural haematoma. The main finding was severe global supratentorial brain atrophy (Fig. 1d, h) as well as cortico-subcortical signal changes in the cerebellar foliae (hyperintense on T2w) (Fig. 2d, e).

This boy was last seen in our hospital at the age of three years and nine months and showed microcephaly, severe bilateral spastic movement disorder not allowing any gross nor fine motor function and profound intellectual deficiency with only simple communication (reacting to contact with smiles, neither language production nor comprehension), severe cerebral visual impairment and epilepsy with generalized tonic-clonic seizures, dysphagia and drooling due to neurological dysfunction. He was living in an institution for severely disabled persons without contact to his parents.

3. Case 2

This term born girl, first child, was born by uncomplicated vacuum extraction, Apgar 7/8/9, birth weight 3.870 g, Her first milestones were normal and head circumference (HC) was between P50 to P75. At the age of 5 months 2 weeks the caregiver reported that she found her vomiting, then the eyes deviated upwards followed by hypotonia, perioral cyanosis and poor breathing. Cardiac massage was started. The emergency was called and found her breathing sufficiently with normal oxygen saturation and normal heart rate. After hospital admission she was intubated and presented a focal left-sided seizure with bilateral propagation which necessitated antiepileptic treatment. Bilateral retinal as well as preretinal haemorrages were found. EEG showed low voltage. HC was 1 cm above P97.

Metabolic testing for glutaric aciduria type 1 and Menkes disease was normal.

A CT scan on the day of admission showed bilateral subdural hygroma.

MRI on next day showed widespread and multifocal areas of signal changes in the cerebrum and cerebellum on DWI. T2w sequences also showed first mild signal changes in these areas (Figs. 1a, b, e, f, 2a, b), the diencephalon was nor affected. In addition, there was evidence for fresh blood in the subdural and subarachnoidal spaces along the frontal and parieto-occipital lobes as well as behind the cerebellum (seen on T2*, and to a minor extent on T1w). This finding can be summarized as haematohygroma. There was a haematoma in the spinal cervical CSF space not affecting the spinal cord.

MRI on follow-up (1 week, 3 weeks, 3 months and 19 months after admission, examples in Fig. 1c, d, g, h) showed slow resorption of the haemorrhages, but transformation of the hygroma into a chronic encapsulated subdural haematoma with neo-membranes and microhaemorrhages. In the

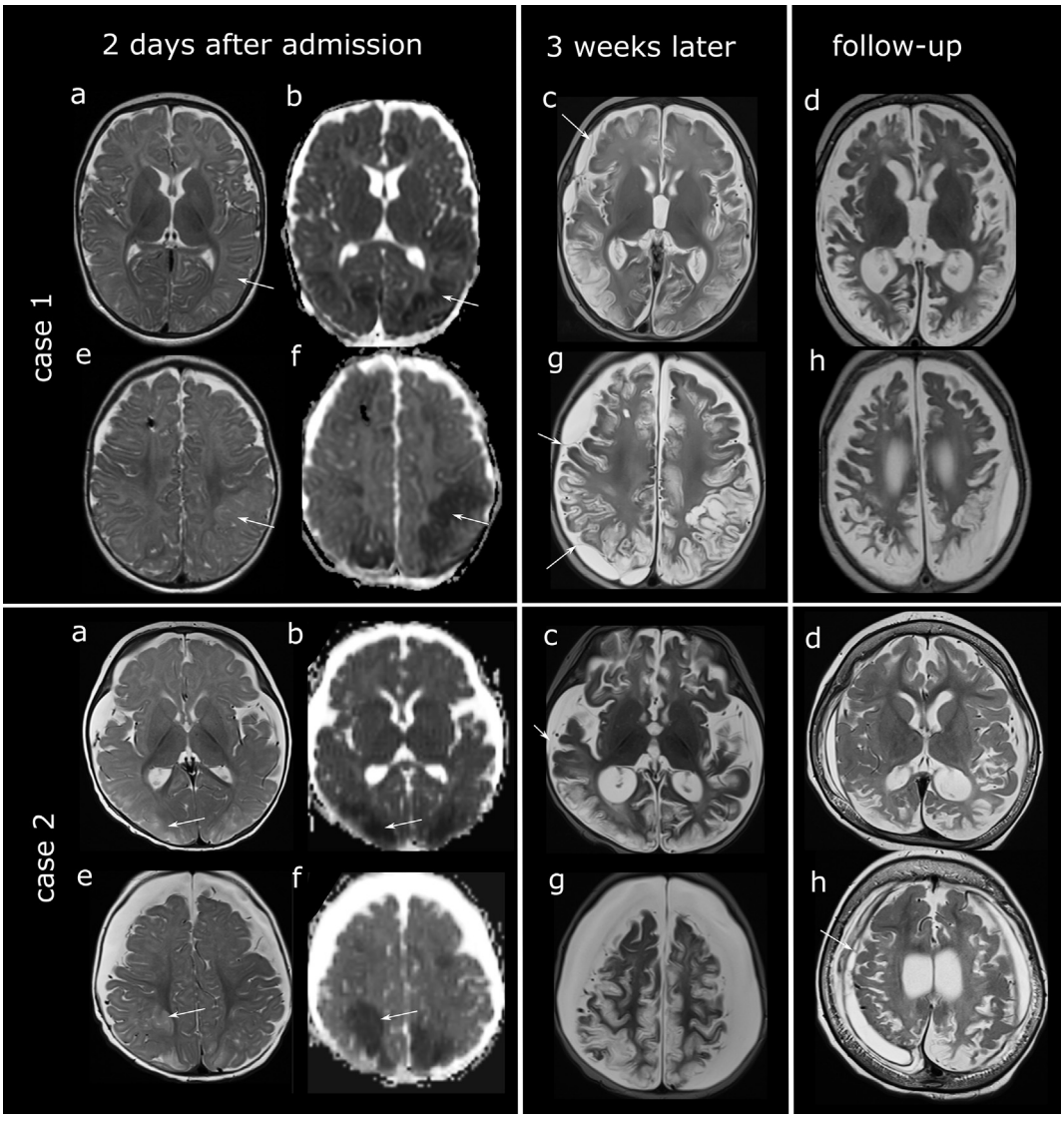

Fig. 1 – MRI of supratentorial lesions in both cases. In the acute phase, 2 days after admission, axial DWI/ADC images show wide spread multifocal changes (hypointense in ADC) (arrows b, f) with some mild blurring already on T2w images (arrows, a, e). Axial T2w images three weeks later and on follow-up (case 1 at 11 months, case 2 at 22 months) show severe cortical and subcortical atrophy with in part polycystic transformation; the diencephalon is spared; there is evidence for the formation of neo-membranes in the subdural space (arrows).

cerebrum and cerebellum the changes on DWI slowly disappeared (still seen 1 week after the insult), but severe multifocal cortico-subcortical atrophy developed. In the cerebellum, signal changes were located cortico-subcortically in the depth of the foliae (Fig. 2c–e). The diencephalon did not show clear pathological changes.

Follow up at the age of 3 years and 6 months showed a severely handicapped child with a HC at P3. She could follow objects with the eyes, reacted to tactile stimuli, vocalized and laughed in response. She had axial hypotonia, but dystonic posturing, hypertonia and mild spasticity of the extremities. She was able to grasp nearby objects and to sit by herself. Epilepsy was controlled with antiepileptic drugs.

Taken together, in addition to severe supratentorial damage (cerebral pathology in AHT), our patients both showed cerebellar involvement characterized by cortico-subcortical signal alterations most prominent in the depth of the foliae and best seen on T2w images following diffusion changes during the acute period.

4. Discussion

These two patients showed the characteristic triad of severe AHT with subdural haematoma, retinal haemorrhages and encephalopathy.[11] MRI on follow-up in both showed the development of chronic subdural haematoma with neo-membranes and micro-haemorrhages. More importantly, it showed widespread multifocal cerebral and cerebellar injury, first seen on DWI then mainly on T2w images, leading to

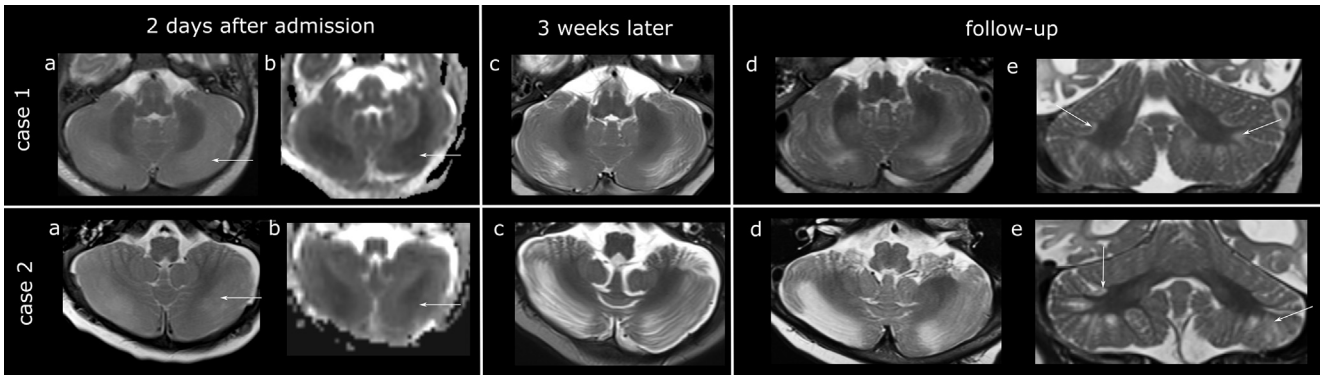

Fig. 2 – MRI of cerebellar lesions in both cases. In the acute phase, 2 days after admission, axial DWI/ADC images show pathological signal change (hypointense in ADC) (arrows b) in the hemispheres, with some mild blurring already on T2w images (arrows a). T2w images three weeks later and on follow-up (case 1 at 11 months, case 2 at 22 months) show cortico-subcortical signal changes (hyperintense) in the depth of the foliae, best seen on coronal images (arrows e).

severe atrophy, especially of the telencephalon. Signal changes of the cerebellum were bilateral and symmetric and located cortico-subcortically in the depth of the foliae. The diencephalon was spared in both cases.

In children with AHT, subdural haemorrhages and their resorption and in part transformation into hygroma is well known. A transformation into chronic subdural haematoma with development of neo-membranes and neo-vascularisation with the risk of microhaemorrhages as seen in our cases has also been described in rare and severe cases.[13]

Here, however, we want to put emphasis on the intra-parenchymal sequelae of AHT as shown by serial MRIs with special reference to the cerebellar involvement and its evolution.

The cerebral changes corresponded to what has been typically reported in MRI studies of AHT.[9,14] In the acute phase, both of our cases showed a pathology best seen on DWI sequences, e.g. hyperintense signal changes with corresponding low signals in ADC-maps. These changes were multifocal and bilateral, and did not correspond to specific vascular territories. On T2w images, first changes with mild hyperintensity in the affected regions of the cortex and white matter were seen. On follow-up changes on DWI slowly disappeared, and changes on T2w images became more marked with hyperintensities of cortex and subcortical regions. Basal ganglia and thalamus were spared, a feature which is also typically reported in AHT. Lesions then changed into severe multifocal cortico-subcortical cystic lesions and atrophy.

In addition, both our children showed cerebellar lesions, which were first evident as bilateral widespread diffusion changes of the cerebellar hemispheres, on follow-up signal changes involved the cortico-subcortical domain predominantly in the depth of the cerebellar foliae.

Cerebellar changes following shaking in AHT have only rarely been described and even less reported. In two reports on patients with AHT, three figures showed additional cerebellar lesions in 3 patients on DWI similar to the ones we found in our patients: Two cases with cerebellar involvement were reported by Zimmerman et al. 2007.[14] They described five different imaging patterns on DWI in 33 infants with NAHT. One pattern showed supratentorial watershed lesions, and 2 of these cases (6% of the total group - a girl, four month old, Fig. 2c, and a boy, seven month old with multiple trauma, figure 3a and 3b) also had what the authors called cerebellar watershed lesions, as they involved the cerebellar hemispheres between the three vascular territories supplying the cerebellum. In a recent review by Wright on characteristic intracerebral patterns of injury in AHT, one figure (6d) shows bilateral cerebellar DWI changes in addition to cerebral pathology on DWI, but these were neither specifically mentioned nor described with respect to the clinical situation.[9]

To our knowledge the evolution of these cerebellar DWI changes on follow-up were never reported. In our two patients, after bihemispheric DWI changes in the acute phase, a particular pattern of signal changes appeared affecting the cerebellar foliae cortico-subcortically particularly in the depth of the foliae, best seen on T2w, but also visible on T1w.

Similar patterns of cortico-subcortical signal changes in the depth of the cerebellar foliae have been described also in other pathologies. A first is thiamine-transporter defect. Supratentorial changes, however, are completely different from those in AHT. They typically involve basal ganglia and thalami as well as cortico-subcortical areas.[15] A second is cerebellar bottom-of-fissure dysplasia, without any additional cerebral pathology.[16] Its clinical correlate is unclear, as the finding is often incidental. In none of the cases described with these two pathologies, changes on DWI have been reported.

There is an ongoing discussion about the pathogenesis of the brain lesions in AHT. Different mechanisms are involved following shaking, e.g. loss of cerebral autoregulation resulting in increased blood flow and disruption of the blood–brain barrier followed by cellular toxicity due to defective ion transport and increased excitatory amino acids.[8,19] Brain regions with defective autoregulation are at greater risk for ischaemic and hyperaemic injury. Especially infants are more sensitive for this mechanisms because of their immature

autoregulation and a greater risk for apnoea and hypotension.[17]

Regarding the cerebellar lesions we can only speculate on the pathomechanisms. Our two cases and the cases from the literature describe these cerebellar lesions associated with severe supratentorial parenchymal lesions. Thus, cerebellar involvement in AHT is probably a sign of high severity and widespread involvement of the brain. Global hypoxic-ischaemic injury due to systemic perfusion failure usually does not involve the cerebellum, but typically affects the diencephalon: Severe hypoxic-ischaemic injuries, for instance following cardiac arrest, after the neonatal age but before one year of age mainly affects supratentorial structures especially the basal ganglia (predominantly posteriorly), medio-lateral thalami and dorsal midbrain and are symmetric.[18] Thus the pathomechanism in AHT may indicate a more sustained or repetitive and multifocal instead of a more acute and global tissue perfusion failure.

Neuropathological findings in AHT support our hypothesis that the cerebellum is affected in only the most severe cases: Multicystic encephalomalacia (MCE) is known as an end-stage finding in fatal AHT. In 4 of 5 cases, autopsy showed that the cerebellum was affected with changes which may correlate with the MRI pattern found in our patients, e.g. irregular loss of neurons, predominantly in the depths of the cerebellar foliae with reactive glial changes, patchy degeneration of the cortical neurons with remaining neurons on the outers sections of some cerebellar foliae, loss of the Purkinje cells and small neurons of the granular layer in the depths of the cerebellar foliae.[20]

Parenchymal brain injuries in AHT are the decisive markers for functional outcome in the affected children.[10] Our two children both showed a severely compromised development on follow-up with severe spastic and dystonic movement disorder, severe intellectual disability, epilepsy and decreased head growth. This corresponds to the extent and topography of the lesions which were widespread and multifocal.

5. Conclusion

We draw attention to a pattern of a severe brain damage in children with AHT following shaking characterized by additional involvement of the cerebellum. We assume that cerebral and cerebellar parenchymal lesions in children with AHT following shaking have a complex mechanism. Cerebellar parenchymal injury can be part of the brain trauma in cases of pediatric AHT and is to date probably underestimated and underrecognized.

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Conflicts of interest

No conflicts of interest.

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