

Was the Hydranencephaly Defense Valid for the Death of a Child?: A Case Report

هل كان الدفاع في قضية موه انعدام المخ صحيحاً لوفاة الطفل؟ دراسة حالة

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Abstract

Hydranencephaly is a central nervous system disorder at birth in which brain's cerebral hemispheres are absent and replaced by sacks filled with CSF. The prevalence of hydranencephaly is less than 1 in 10,000 births, with 0.2 percent of children autopsies showing the condition.

A case of hydranencephaly is described in this report. This was discovered during the medicolegal autopsy of a newborn male infant who was accidentally found dead in a wastebasket. Presented here are the results of external examination, autopsy, histological, and toxicological findings. The fact that hydranencephaly was the accused mother's sole and most potent defense in proving her innocence was underlined here, since the presence of such an argument was enough to arouse suspicion that it was a case of infanticide committed by the mother. المستخلص

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موه انعدام الدماغ هو غياب نصفي الدماغ عند الولادة مع امتلاء القحف الدماغي بالسائل الدماغي الشوكي؛ إذ إن معدل شيوع موه انعدام الدماغ هو أقل من 1 لكل 10000 مولود و 0.2 % من حالات تشريح الأطفال ذوي الفئة العمرية الأقل من سنة.

تم تقديم حالة من موه انعدام الدماغ في هذا التقرير ، وقد تم اكتشافها بشكل غير متوقع أثناء تشريح طبي شرعي لطفل رضيع وُجد ميتًا في حاوية قمامة.

وتم توضيح الفحص الخارجي، التشريح، دراسة عيناتها النسيجية ونتيجة عينات السموم. والحقيقة أن اضطراب موه انعدام الدماغ كان المدافع الوحيد والأقوى.

المتاح للأم المتهمة لإثبات براءتها تم التركيز عليها هنا؛ لأن وجود مثل هذه الحجة كان كافيًا لإثارة الشك في أنها كانت حالة قتل أم لطفلها.

الكلمات المفتاحية: علوم الأدلة الجنائية، العلوم الجنائية، علم الأمراض الشرعي، موه انعدام المخ، قتل الرضع، التشريح.

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1. Introduction

Hydranencephaly is a congenital cerebral condition that is relatively uncommon [1]. The hemispheres of the brain are entirely or almost completely absent. In their position, there is a membranous sac filled with cerebrospinal fluid (CSF), glial tissue, and ependymal cells [2]. The hallmarks of hydranencephaly are the replacement of brain matter with CSF and the retention of posterior cranial fossa structures [3].

A case of hydranencephaly is described in this report. This was discovered during the medicolegal autopsy of a newborn male infant who was accidentally found dead in a wastebasket. The prosecutor requested a medico-legal autopsy after suspecting infanticide.

This is the third case study in forensic literature that we are aware of [4,5].

2. Case report

A corpse of an initially putrefied male baby was discovered dead in a garbage bin, wrapped in a black plastic bag. The prosecutor requested a medico-legal autopsy after suspecting infanticide.

3. Forensic Examination Revealed

Normal-looking baby boy except for a slightly elongated head. On the body, there are neat colorful clothes and a diaper with a small amount of dark brown stool. The baby's growth parameters were within the normal range. He was 48 cm tall, weighed 2.85 kg, and had a head circumference of 34.5 cm, a chest circumference of 31 cm, and an abdominal circumference of 28.0 cm. The fontanelles were open, suture lines were separated, and there were no visible or palpable masses; the head was covered by black hair about 1 cm long. Both corneas were opaque, and there were no petechial hemorrhages on the sclera, conjunctiva, on either eye or face. There was no pallor around the mouth or on the nasal orifices, and the mouth's frenums were intact. There was no evidence of blunt force applied to the body.

Owing to initial putrefaction, the abdominal wall at the right lower part was green-stained. The umbilical cord was dry, stiff, 2.5 cm long, still attached to the abdominal wall, with a sewing thread tightening its margin. In the scrotum, the testes could be palpated. The fingernails and toenails were noticeable. There were no congenital abnormalities or deformities found.

The external surface of the skull was intact, but there was diffuse scalp edema with contusions on the right side of the anterolateral and posterior surfaces.

The lack of the cerebral hemispheres was the most startling finding (Figure-1). The supratentorial cranial cavity was filled with an abundance of murky dark-red fluid containing residue-like necrotic debris. Both the falx cerebri and the tentorium cerebelli were intact. There was a small tissue mass near the inferior part of the falx cerebri. The existence of fragile liquified infratentorial structures was discovered after the fluid was extracted and the tentorium cerebelli was removed.

The lungs were edematous, filling the thoracic cavity, dark-red colored, and solid inconsistency. Both kidneys had a nodular surface with normal gross anatomy, stomach contained curdled milk. No other abnormalities were found; organs were all present and macroscopically intact without any putrefactive changes.

4. Histological Findings

The small tissue mass near the inferior part of the falx cerebri showed normal cerebral tissue with no pathological finding (Figure-2). The cerebellum was normal. Lung samples showed aerated alveoli, no inflammation, no signs of acute respiratory distress syndrome, no pathological changes except for congestion. Kidneys showed normal architecture



Figure 1-Hydraencephaly showing the lack of the cerebral hemispheres.



Figure 2- The small tissue mass near the inferior part of falx cerebri shows normal cerebral tissue (H&E stain, at magnification A- 4×, B- 10×, C- 20×).

with no cystic or inflammatory changes. The liver showed normal architecture with no inflammatory changes or any pathological findings.

Other organs showed no pathological findings and no significant autolytic changes were seen.

5. Toxicology Screen

Ethyl alcohol was found in the blood sample (19 mg/dl). Other toxicological screening of blood & liver samples were clear. Also, DNA matching helped to confirm the mother.

6. The Cause of Death

The cause of death was identified as a congenital malformation of the central nervous system, which is hydranencephaly, and infanticide was excluded.

7. Maternal History

The accused woman who was confirmed by DNA matching to be the biological mother of this baby boy by DNA matching was married previously and her marriage resulted in three healthy children, all of them delivered by normal vaginal delivery with no history of abortion. Now she works as a prostitute. This pregnancy was her fourth pregnancy, which was an illegitimate, smooth pregnancy with no antenatal care follow-up. Due to social embarrassment and legal issues, the delivery was normal vaginal at home without any assistance from medical professionals. She had no psychiatric illness.

She didn't notice any abnormality in her baby in the first few days, but suddenly he became completely unconscious and pulseless. She waited 2 days to confirm his death, and then left him in the nearest wastebasket.

8. Maternal Clinical Forensic Examination

Maternal clinical forensic examination could not done as it was not requested by the prosecutor. Actually it was not needed in this case, either medically or legally.

9. Discussion and Conclusion

Hydranencephaly is the absence of the cerebral hemispheres at birth, with CSF filling the gap in the cranium [2,6]. The prevalence of hydranencephaly is less than 1 in 10,000 births, with 0.2 percent of children's autopsies showing the condition [7]. The cause is unclear, but it has been linked to infectious, toxic, aplastic, genetic, or vascular causes, as well as maternal hypoxia and fetal transfusion syndrome. Secondarily, occlusion of cerebral arteries above the supraclinoid level, mainly affecting the major vessels of the anterior circulation, is the most frequently suggested mechanism for growth [8,9].

Infants with hydranencephaly are thought to have a shorter lifespan, with survival rates ranging from weeks to months [3]. Those who survive, display no obvious neurological or psychiatric symptoms at first, and the infant's head size and spontaneous reflexes including sucking, swallowing, crying, and moving the arms and legs may appear normal at birth. Respiratory infections, drug-resistant seizures, developmental delay, spastic diplegia, and extreme growth failure are all common causes of death in these patients [2].

In this case, many shreds of evidence were present, enough to prove that this baby was alive for several days after birth. These included the complete absence of vernix caseosa, complete absence of meconium, the presence of an aerated respired lung, the presence of curdled milk in the stomach, the presence of healing signs at the remnant of the umbilical cord. He seemed completely normal at birth, so his mother did not seek any medical help, which clarifies why he was not diagnosed during his life.

However, its discovery in unusual circumstances (dead in a plastic bag in a garbage can) sparked suspicions of infanticide.

The tightening of the umbilical cord margin by sewing thread, together with the presence of scalp edema (caput succedaneum), indicates that this baby was delivered vaginally in the absence of any medical help. Scalp contusion with no external scalp injuries or any evidence of head trauma makes normal vaginal delivery the most probable explanation. Otherwise, no signs of trauma were present. Toxicology screening was clear as ethylene alcohol at this low level is explained by postmortem microbial fermentation [10]. Also, histopathological examination showed that all organs were normal with no pathological findings. According to autopsy findings, the exact cause of congenital hydranencephaly in this baby was not known. Therefore, the act of infanticide in this case was ruled out. Hydranencephaly was declared as the only cause of baby's death after exclusion of any other pathological, traumatic or toxicological possibilities. Therefore, hydranencephaly was the only and most powerful defense available for the accused mother in proving her innocence.

The cause of death was identified as a congenital malformation of the central nervous system, which is hydranencephaly, and infanticide was excluded.

Ethical Approval

The informed consent and ethical approval in this case was obtained from the prosecutor.

Conflict of interest

The authors declare that they have no conflict of interest.

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References

- Rossi A. Hydranencephaly. InBrain Imaging with MRI and CT: An Image Pattern Approach 2010 Jan 1 (pp. 195-196). Cambridge University Press. https://doi.org/10.1017/ CBO9781139030854.096
- Pavone P, Praticò AD, Vitaliti G, Ruggieri M, Rizzo R, Parano E, Pavone L, Pero G, Falsaperla R. Hydranencephaly: cerebral spinal fluid instead of cerebral mantles. Italian journal of pediatrics.

2014 Dec;40(1):1-8. https://doi.org/10.1186/ s13052-014-0079-1

- Cecchetto G, Milanese L, Giordano R, Viero A, Suma V, Manara R. Looking at the missing brain: hydranencephaly case series and literature review. Pediatric neurology. 2013 Feb 1;48(2):152-8. https://doi.org/10.1016/j.pediatrneurol.2012.10.009
- Verzeletti A, Leide A, De Ferrari F. Hydranencephaly in a case of suspected infanticide. Med Sci Law 2015. https://doi. org/10.1177/0025802414536153.
- Ohshima T, Kondo T, Lin Z, Nagano T. Suspected maternal infanticide in a case of hydranencephaly. Int J Legal Med 1993. https://doi. org/10.1007/BF01222120.
- Crome L, Sylvester PE. Hydranencephaly (hydrencephaly). Arch Dis Child 1958. https://doi. org/10.1136/adc.33.169.235.
- Vaneckova M, Seidl Z, Goldova B, Vitkova I, Baxova A, Petrovicky P, et al. Post-mortem magnetic resonance imaging and its irreplaceable role in determining CNS malformation (hydranencephaly) - Case report. Brain Dev 2010. https://doi. org/10.1016/j.braindev.2009.03.011.
- Quek YW, Su PH, Tsao TF, Chen JY, Ng YY, Hu JM, Chen SJ. Hydranencephaly associated with interruption of bilateral internal carotid arteries. Pediatrics & Neonatology. 2008 Apr 1;49(2):43-7. https://doi.org/10.1016/S1875-9572(08)60011-X
- Lindenberg R, Swanson PD. "infantile hydranencephaly"-a report of five cases of infarction of both cerebral hemispheres in infancy. Brain 1967. https://doi.org/10.1093/brain/90.4.839.
- Ziavrou K, Boumba VA, Vougiouklakis TG. Insights into the origin of postmortem ethanol. Int J Toxicol 2005. https://doi. org/10.1080/10915810590936391.