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Suspected maternal infanticide in a case of hydranencephaly

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Summary. A medico-legal autopsy case of hydranencephaly in a male infant which was first suspected of maternal infanticide is reported. The infant was 48 cm in height, weighed 2.86 kg and the circumference of the head, the chest and the abdomen was 32.2 cm, 31.0 cm and 30.4 cm, respectively, with no deformities of the head or body. Autopsy examination, however, revealed a severe defect in the central nervous system. The cranial cavity was filled with a cloudy dark red fluid (ca. 310 ml) instead of the cerebral hemispheres. The residual central nervous tissues were mostly subtentorial structures from the midbrain to the spinal cord namely, corpus mamillare, corpora quadrigemina, corpus pineale, crus cerebri, pons, cerebellum, medulla oblongata and spinal cord. The basal ganglia, thalamus, hypothalamus and chiasma opticum could not be found, although atrophic hypophysis, eyeballs and optic nerves were present. The usual distribution of cerebral blood vessels, especially the branches of the anterior and middle cerebral arteries and Willis' ring, was absent despite the presence of the internal and external carotid arteries. Other organs were, in general, congestive. The marked cortical atrophy of the adrenal glands (left 0.5 g, right 0.6 g), especially the zona fasciculata, was characteristic. The hydrostatic lung test gave partially positive results, but this was considered to be due to artificial respiration by an ambulance man because amniotic fluid components were microscopically noted and fully expanded alveoli were not found. In conclusion, the cause of the infant's death was diagnosed as stillbirth due to aspiration of amniotic fluid caused by the severe defect of vegetative hypothalamic function through hydranencephaly.

Key words: Infant death – Hydranencephaly – Suspected maternal infanticide – Stillbirth – Malfunction of vegetative hypothalamus

Zusammenfassung. Ein Fall einer rechtsmedizinischen Obduktion an der Leiche eines männlichen Kindes mit Hydranencephalie wird beschrieben, in welchem zunächst der Verdacht einer Kindstötung bestand. Das Kind war 48 cm lang und 2,86 kg schwer, die Umfänge des Kopfes, des Brustkorbs und des Abdomens waren 32,2 cm, 31,0 cm, und 30,4 cm. Kopf mit Körper waren normal geformt. Die Obduktion deckte jedoch einen schweren Defekt des Zentralnervensystems auf. Die Schädelhöhle war mit einer trüben, dunkelroten Flüssigkeit (ca. 310 ml) ausgefüllt. Die Gewebsreste des Zentralnervensystems waren überwiegend subtentoriale Strukturen des Hirnstamms, speziell Corpus mamillare, Corpora quadrigemina, Corpus pineale, Crura cerebri, Pons, Cerebellum, Medulla oblongata und Rückenmark. Die Basalganglien, Thalamus, Hypothalamus und das Chiasma opticum konnten nicht gefunden werden, obwohl eine atrophische Hypophyse sowie Augenbulbi und Sehnerven in den Augenhöhlen vorhanden waren. Die normale Verzweigung der cerebralen Blutgefäße, speziell der Äste der vorderen und mittleren Cerebralarterien und des Circulus arteriosus Willisi, fehlten trotz Vorhandensein der Arteriae carotides internae et externae. Andere Organe waren im allgemeinen gestaut. Die ausgeprägte Rindenatrophie der Nebennieren (links 0,5 g, rechts 0,6 g), spezielle der Zona fasciculata, war charakteristisch. Die Lungenschwimmprobe gab teilweise positive Resultate, dies war jedoch die Folge einer künstlichen Beatmung durch einen Mann des Rettungsdienstes zu erklären, weil Fruchtwasserbestandteile mikroskopisch festgestellt und vollständig entfaltete Alveolen nicht gefunden wurden. Im Resultat wurde eine Totgeburt des Kindes diagnostiziert. Todesursache war die Aspiration von Amnionflüssigkeit, verursacht durch einen schweren Defekt der vegetativen hypothalamischen Funktion infolge Hydranencephalie.

Schlüsselwörter: Kindstod – Hydranencephalie – Verdacht der Kindstötung – Totgeburt – Fehlfunktion des vegetativen Hypothalamus

Introduction

Hydranencephaly is a rare and severe condition of the central nervous system in infants where nearly the entire cerebral hemispheres have been destroyed by the encephaloclastic process in the uterus. The brain is largely

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replaced by a fluid-filled membranous sac resembling a large cyst [1–3].

In this report, a case of this hydranencephaly is presented. This was unexpectedly found during the medicolegal autopsy of a newborn male infant who was left illegally by his mother after the delivery at home and was first suspected of maternal infanticide. This kind of discovery of the lesion seems very rare and, to the best of our knowledge, is only the second published case report found in Japan [4].

Case report

1. Case profile. A 44-year-old female delivered a male baby in the lavatory at home, suddenly and unexpectedly while she was sitting on the toilet. She lost consciousness and was found by her son when he returned home. She was brought to hospital in a state of shock. The infant was found wrapped in a bath towel in the hallway beside the lavatory and artificial respiration was unsuccessful.

A past history on pregnancy revealed 2 normal deliveries and 2 abortions (including one induced abortion). The last menstruation was reported to have been in November of the previous year and she was unaware of the present pregnancy. On this basis, the delivery must have occurred at least at the end of the ninth month or early in the tenth month of gestation.

Approximately 5 hours after the discovery of the dead infant, a judicial autopsy was performed on the grounds of suspected infanticide by the mother.

2. Physical findings. Initially, no congenital anomaly or deformity was observed and there was no evidence of pressure on the mouth or nasal orifices or of blunt force to the body (Fig. 1).

The infant was 48 cm in height and weighed 2.86 kg. The circumference of the head, the chest and the abdomen was 32.2 cm, 31.0 cm and 30.4 cm, respectively. The longitudinal and transverse lengths of the head, and the widths of the shoulder and pelvis were 10.5 cm, 9.0 cm, 11.0 cm and 8.0 cm, respectively. The head hair was approximately 2 cm long and no abnormal bulging was palpable at the anterior and posterior fontanelles. The diameter of the ossification center at the lower end of the femur and calcaneus were 0.5 cm and 0.7 cm, respectively. The testes were palpable in the scrotum.

The umbilical cord was torn at the 52 cm distal portion from the umbilicus of the infant, probably due to extreme stretching. The remnant of the cord attached to the placenta (775 g in weight, 23 cm × 16 cm × 2–2.5 cm) was 19 cm long. The total length of the cord was therefore 71 cm, indicative of precipitate labor and was consistent with the mother's description of the circumstances at parturition.

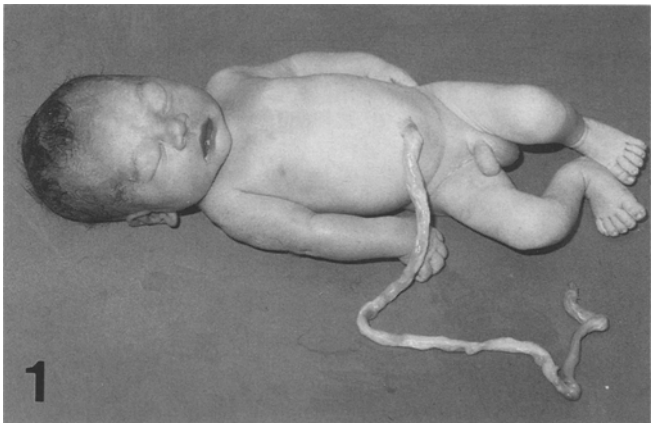

Fig. 1. Macroscopic view of the male infant with hydranencephaly in the present case showing no anomaly or deformity in appearance and being suspected of maternal infanticide

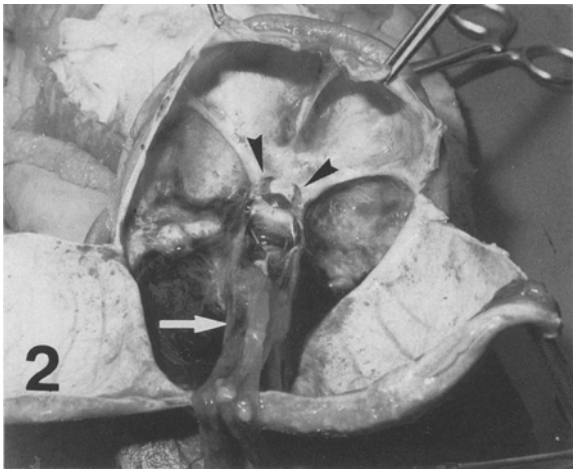

Fig. 2. In the subtentorial fossa, the remnant of the central nervous system was found *(arrow)*. The hypophysis and the optic nerves in the orbits were noted despite their atrophic changes *(arrow heads)*

All of these physical observations were within the normal ranges for mature infants indicating a full term pregnancy and were consistent with the presumption that this delivery occurred at least after the end of the ninth month of gestation.

3. Autopsy findings. The most characteristic and unexpected finding was found in the central nervous system. When the cranial cavity was opened, a membranous sac or cyst filled with a cloudy dark red fluid was noted in place of the cerebral hemispheres. The fluid (ca. 310 ml) contained the residue-like necrotic debris of the cerebrum. Dura mater encephali, including the falx cerebri, was intact.

After the fluid was drawn off and the tentorium cerebelli removed, the residual central nervous tissues could be observed (Fig. 2). The hypophysis and the structures from the midbrain to the spinal cord, namely, corpus mamillare, corpora quadrigemina, corpus pineale, crus cerebri, pons, cerebellum, medulla oblongata and spinal cord, were confirmed. However, basal ganglia, thalamus, hypothalamus and chiasma opticum could not be found. The eyeballs and the optic nerves were noted in the orbits, and cranial nerves except the bilateral olfactory nerves were correctly located (Fig. 3).

The distribution pattern of the cerebral blood vessels was abnormal, and the anterior and middle cerebral arteries and Willis' ring could not be found, although the branches of the vertebral arteries in the cranial cavity were barely observed and the internal and external carotid arteries were found in the neck.

The heart (18.0 g) showed no congenital abnormality and contained dark red blood (ca. 29 ml) with small amounts of fragile clots. The lungs were dark red, congestive (left 25 g, right 36 g) and solid in consistency with numerous petechial hemorrhages in the visceral pleurae. The hydrostatic lung test gave partially positive results, but this was interpreted as a resuscitation artifact, since typical crepitance was not heard on rubbing gently between the fingers.

The liver (130 g), the pancreas (2.1 g) and the kidneys (left 11.7 g, right 11.8 g) demonstrated mild to moderate congestion with no pathological findings. The spleen, in contrast, was severely congestive (20.5 g; normal value, 8 ± 3.1 g in Japanese infants) [5].

The marked cortical atrophy of the bilateral adrenal glands (left 0.5 g, right 0.6 g) was most conspicuous, weighing about 12–17% of the mean value for Japanese infants (left 4.1 ± 1.79 g, right 3.6 ± 1.37 g) [5].

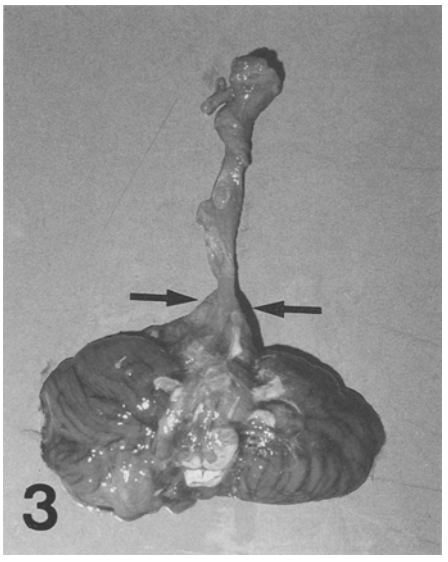

Fig. 3. Ventral view of the remnant central nervous system. The *arrows* show the most rostral end of the midbrain. The cerebellum and the brain stem appear normal at first sight, but they were really atrophic. The cranial nerves except the olfactory nerves were correctly located.

The thymus was slightly increased in weight (13.7 g; normal value 12.1 ± 4.8 g) [5] and was solid in consistency with marked lymphocyte proliferation (see below). The thyroid gland was slightly congestive.

4. Histopathological findings. In the remnants of the central nervous system, the most rostral portion, such as the cut surface of corpus mamillare and corpora quadrigemina, had no ependymal cell layer, and the glial cell layer was directly exposed. The shape of nerve cells in the midbrain, pons and medulla oblongata was relatively well preserved, but there were also nerve cells in which Nissl granules were not conspicuous. The nerve fibers in the ventral portion of the pons and medulla oblongata were atrophic. In summary, there was no neuropathological finding which was incompatible with the encephaloclastic process. The cerebellum was unremarkable.

The lungs were microscopically observed as parenchymatous and amniotic fluid components were found in the respiratory bronchioles, alveolar ducts and alveoli. The expansion of air passages, especially in their peripheries, such as alveolar ducts and alveoli, was barely noticeable. Other pathological changes, for example bronchitis or pneumonia, were not noted except for congestion.

The cortex of the bilateral adrenal glands, especially the zona fasciculata, was markedly atrophic. The thymus was, in contrast, parenchymatous and the lymphocyte proliferation in the cortex was remarkable. Hassall's bodies seemed somewhat proliferative.

Other organs showed no remarkable pathological changes.

Discussion

The first pathological report on hydranencephaly was presented by Cruveilhier in which he described the gross anatomic features of this lesion under the term "anencéphalie hydrocéphalique" [2]. Spielmeyer [6] used the term "Hydranencephalie" and characterized the disease as the condition where large portions of the cerebral hemispheres were replaced by a fluid-filled membranous sac within a relatively normal-sized cranium.

Hydranencephaly was previously considered to be a malformation, although it is now widely accepted as a type of encephaloclastic process of the infant's brain in the later intrauterine period, resulting from various causes, i.e. fetal encephalitis, circulatory disturbances, intracranial hemorrhage, trauma, etc. [3]. According to the description by Escourolle and Poirier [7], the inner surface of the membranous sac is lined not by ependymal cells but by a glial border, in contrast to the porencephalic cavities and hydrocephalus. However, this pathological description seems insufficient because the existence or non-existence of the ependymal cell layer in porencephaly or hydrocephaly is not one of the criteria for a differential diagnosis between these lesions and hydranencephaly. Furthermore, hydranencephaly and porencephaly belong to the same encephaloclastic spectrum [3; Hori A, personal communication].

The diagnosis of hydranencephaly in the present case is conclusive from the autopsy and histopathological findings described [1–3]. According to Lange-Cosack [8], the present case seems to belong to the second group because the basal ganglia are lost, and the pons, medulla oblongata and cerebellum are present. The disorder of vegetative neural function becomes manifest because the thalamus, hypothalamus and a most of the midbrain are destroyed, leading to death at birth or at least within 3 months [9].

The etiology of hydranencephaly has not yet been determined. Possible factors such as circulatory disturbances (embolism, vasculopathy, etc.), intracranial hemorrhage, trauma and infection are now considered [1–3]. The possibility of vascular lesion is based on the finding that the major encephaloclastic process occurs in the distributional area of the internal carotid arteries [1–3]. In fact, Myers [10] reported that this kind of lesion was experimentally produced in fetal monkeys by bilateral ligation of the carotid arteries and jugular veins. For the possibility of fetal infection, cases associated with fetal toxoplasma or cytomegalovirus infection have been reported [3, 11].

In the present case, the mother was admitted to hospital in a state of shock and was diagnosed as a possible HELLP syndrome [12]. This term derives from such findings as hemolysis, elevated liver enzymes and low platelet count, and the syndrome often occurs as a severe consequence of hypertension in pregnancy. According to the mother's description, she was already edematous in May of this year, probably after the seventh month of gestation, suggesting that she suffered from the toxemia of pregnancy. Therefore, as a possible etiology of the present case, a circulatory failure might have occurred due to toxemia, and embolism might have happened at the peripheries of the internal carotid arteries. The possibility of toxoplasmosis could be excluded by the serological and histopathological examinations.

It was very interesting that the adrenal cortex was markedly atrophic, especially the zona fasciculata. This finding corresponds to the theory that the zona fasciculata is responsible for mineral corticoid production which chiefly depends on adrenocorticotrophic hormone

(ACTH) [13] and the secretion of ACTH was probably defective in this case due to the destruction of the hypothalamic region.

From the medico-legal point of view, the cause of death of the male infant was interpreted as asphyxia due to amniotic fluid aspiration into the airways caused by marked malfunction of vegetative hypothalamus (respiration, heat production, etc.). A disorder of the adrenocortical response to "stress" seems to have also been involved in the infant's death. This means that the death was essentially based on "hydranencephaly", a severe defect of the central nervous system. On the other hand, there are many cases of hydranencephaly without damage of the hypothalamus which show similar clinical and pathophysiological manifestations, and the encephaloclastic process with preserved hypothalamus can also be a cause of death [3; Hori A, personal communication].

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References

1. Swaiman KF, Jacobson RI (1984) Developmental abnormalities of the central nervous system. In: Baker AB, Baker LH (eds) Clinical neurology, vol. 4, revised edn. Harper and Row, Philadelphia, Chap. 55
2. Halsey JH Jr, Allen N, Chamberlin HR (1971) The morphogenesis of hydranencephaly. J Neurol Sci 12:187–217
3. Friede RL (1989) Developmental neuropathology, 2nd edn. Springer, Berlin Heidelberg New York, pp 28–43
4. Ueda M, Ishida N, Hamana S, Egi H (1958) A case of hydranencephaly – an autopsy case suspected of an infanticide. Jpn J Leg Med 12:839–849
5. Investigation Committee of the Medico-Legal Society of Japan (1992) Reports on medico-legal data from the massive investigation performed by the medico-legal society of Japan – Weight and size of internal organs of normal Japanese today. Jpn J Leg Med 46:225–235
6. Spielmeyer W (1905) Ein hydranencephales Zwillingspaar. Arch Psychiatr Nervenkr 39:807–819
7. Escourolle R, Poirier J (1978) Manual of basic neuropathology, 2nd edn. Saunders, Philadelphia, pp 178–181
8. Lange-Cosack H (1944) Die Hydranencephalie (Blasenhirn) als Sonderform der Großhirnlosigkeit. I. Anatomischer Teil. Arch Psychiatr Nervenkr 117:1–51
9. Lange-Cosack H (1944) Die Hydranencephalie (Blasenhirn) als Sonderform der Großhirnlosigkeit. II. Klinischer Teil. Arch Psychiatr Nervenkr 117:595–640
10. Myers RE (1969) Brain pathology following fetal vascular occlusion: an experimental study. Invest Ophthalmol 8:41–50
11. Altshuler G (1973) Toxoplasmosis as a cause of hydranencephaly. Am J Dis Child 125:251–252
12. Weinstein L (1982) Syndrome of hemolysis, elevated liver enzymes, and low platelet count: a severe consequence of hypertension in pregnancy. Am J Obstet Gynecol 142:159–167
13. Deane HW, Greep RO (1946) A morphological and histochemical study of the rat's adrenal cortex after hypophysectomy, with comments on the liver. Am J Anat 79:117–146