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Case Report

Hydranencephaly in an Infant: The Radiologic Findings and a Case Report

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Abstract: Hydranencephaly (HE) is a rare congenital brain disorder in which the cerebral hemispheres are absent and replaced by sacs filled with cerebrospinal fluid (CSF). This entity is rarely seen in postnatal life and has an incidence of less than 1 per 10,000 live births. This is a 5-day-old neonate brought in for a transfontanelle ultrasound scan (TFUS), referred from a peripheral health facility on account of relatively large head, sudden onset of fever, poor cry after birth and a low Apgar score. The TFUS was done through patent anterior and posterior fontanels in sagittal and axial planes and through the temporal bone interrogation. The TFUS demonstrated absence of the two cerebral hemispheres; these are replaced by hypoechoic fluid (CSF), the presence of an echogenic midline falx cerebri, the presence of the cerebella hemispheres and midbrain with normal morphological appearance. A diagnosis of hydranencephaly in a neonate (5-day-old) was established following the aforementioned findings from TFUS; the parents were advised to consult a neurosurgeon in a tertiary health facility for further expertise management. We report a case of 5-day-old neonate with TFUS findings conforming to Hydranencephaly due to its rare nature and peculiar presentation.

Keywords: Hydranencephaly, CSF, TFUS, Cerebral Hemispheres.

INTRODUCTION

Hydranencephaly (HE) is a rare congenital disorder characterized by absence and subsequent replacement of the cerebral hemispheres by marked volume of CSF and usually associated with intrauterine fetal death and rarely seen in postnatal life [1-4]. This is a rare entity with an incidence of less than 1 per 10,000 live births, and suspected to be caused by bilateral carotid arteries occlusion in the prenatal life most often in the second trimester following varying causes [1-5].

Hydranencephaly is regarded as one of the most common and severe form of bilateral cerebral hemispheric anomaly, in which a membranous sac filled with CSF, replaces the cerebral hemispheres which are either partially or completely absent [6-8].

In HE, the cranial cavity may retain remnants of glial tissue and ependymal, most often along the falx and close to the diencephalon, consequently having an intact cranial vault and meninges, with presence of midbrain structures like the basal ganglia, brainstem and posterior fossa structures [9, 10].

Regarding HE, most patients often die prenatally, though some that survive may be asymptomatic at birth, while others may be symptomatic with increasing intracranial pressure at later weeks of life and may present with hyperirritability, affectation of muscle tone(hyper/hypotonia), increase in head circumference, widened or persistent fontanelles, late on set of seizures, hydrocephalus, hearing and visual loss [6-11].

The condition hydranencephaly was first described in 1904 by Turnbull, but the term HE, was first introduced by Spielmeyer in 1905 when he reported the malformation in twins [12, 13]. Some of the etiologies of HE, may include infections, toxins, genetics, vascular factors, maternal hypoxia and twin-twin transfusion syndrome [12-15].

In diagnosis of HE, following clinical suspicion, imaging plays a vital role, the use of fetal ultrasonography which most often times demonstrates sacs of fluid replacing the cerebral hemispheres, later confirmed by cranial computed tomography scan or magnetic resonance imaging [12-15].

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CASE REPORT

This is a 5-day-old neonate brought in for a transfontanelle ultrasound scan (TFUS), referred from a peripheral health facility on account of relatively large head, sudden onset of fever, poor cry after birth and a low Apgar score.

The child appears irritable, lethargic with a relatively large head, reduced muscle tone, mildly dehydrated with a widened anterior fontanelle. The child had a complete skull vault and no any-other associated physical anomaly demonstrated.

The child is the 8th child of the mother with healthy surviving seven siblings; the mother never attended any antenatal care, confessed to history of multiple episodes of illness during the index pregnancy, multiple drug and herbal concussion use.

The TFUS was done through patent anterior and posterior fontanels in sagittal and axial planes and through the temporal bone interrogation. The TFUS demonstrated absence of the two cerebral hemispheres; these are replaced by hypoechoic fluid (CSF), the presence of an echogenic midline falx cerebri (figures 1&2), the presence of the cerebella hemispheres and midbrain with normal morphological appearance. The deeper structures such as the basal ganglia, cerebella hemispheres and midbrain structures were demonstrated. A diagnosis of hydranencephaly in a neonate (5-day-old) was established following the aforementioned findings from TFUS; the parents were advised to consult a neurosurgeon in a tertiary health facility for further expertise management.

We report a case of 5-day-old neonate with TFUS findings conforming to Hydranencephaly due to its rare nature and peculiar presentation.

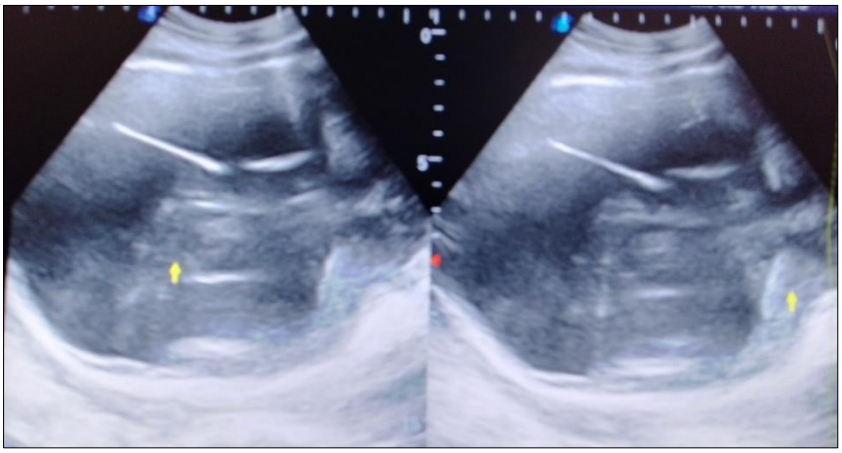

Figure 1: Both images are sonograms from TFUS through the anterior fontanelle, demonstrating absence of the cerebral hemispheres which are replaced by extensive amount of CSF, presence of an echogenic midline falx cerebri and the peripheral echogenic outline; the skull vault. The CSF replaced cerebral hemispheres is termed Hydranencephaly

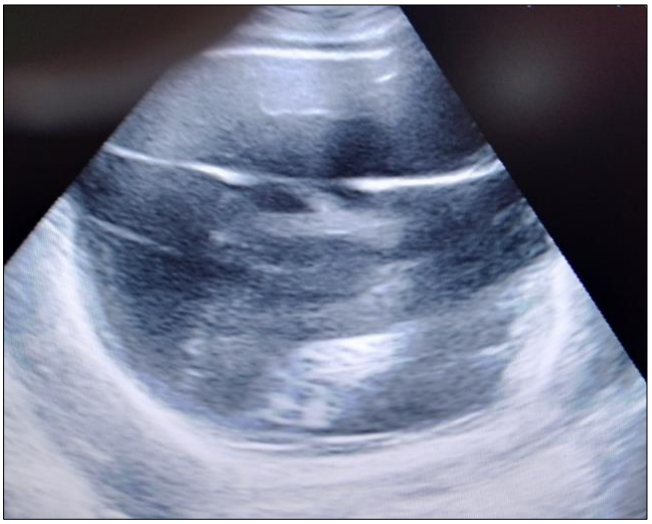

Figure 2: Sonogram from a parasagittal plane TFUS demonstrating fluid filled cranial cavity, absence of the cerebral hemispheres, centrally placed falx cerebri, hyperechoic basal ganglia tissue faintly demonstrated (inferior and lateral to the falx cerebri) and peripherally located echogenic skull vault

DISCUSSION

Hydranencephaly (HE) is a rare congenital disorder characterized by absence and subsequent replacement of the cerebral hemispheres by marked volume of CSF and usually associated with intrauterine fetal death and rarely seen in postnatal life [1-4]. The case under review is a 5-day-old neonate and had absence of cerebral hemispheres which are replaced by marked amount of CSF.

In HE, the cranial cavity may retain remnants of glial tissue and ependymal, most often along the falx and close to the diencephalon, consequently having an intact cranial vault and meninges, with presence of midbrain structures like the basal ganglia, brainstem and posterior fossa structures [9, 10]. The case under review had an intact cranial cavity, falx cerebri, basal ganglia, brainstem and posterior fossa structures, thereby conforming to these literatures.

Some of the etiologies of HE, may include infections, toxins, genetics, vascular factors, maternal hypoxia and twin-twin transfusion syndrome [12-15]. The patient under review had no known cause, but suspected to be from carotid arterial occlusion, maternal infections and toxins, thereby conforming to these literatures.

Regarding HE, most patients often die prenatally, though some that survive may be asymptomatic at birth, while others may be symptomatic with increasing intracranial pressure at later weeks of life and may present with hyperirritability, affectation of muscle tone(hyper/hypotonia), increase in head circumference, widened or persistent fontanelles, late on set of seizures, hydrocephalus, hearing and visual loss [6-11]. The patient had a postnatal life, with large head, irritability, hypotonia and widened anterior fontanelles, thereby conforming to these literatures.

In diagnosis of HE, following clinical suspicion, imaging plays a vital role, the use of fetal ultrasonography which most often times demonstrates sacs of fluid replacing the cerebral hemispheres, later confirmed by cranial computed tomography scan or magnetic resonance imaging [12-15]. The case under review was diagnosed following fetal ultrasonography; TFUS, this confirmed the fluid filled sacs replacing the cerebral hemispheres, conforming to these literatures.

CONCLUSION

Suspected cases of HE, should have ultrasonography either during pre or post-natal period; this will demonstrate the so-called fluid-filled sacs replacing the cerebral hemispheres characteristic of the condition, and also will exclude associated differential diagnosis of HE especially in resource limited environments like ours.

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