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Hydranencephaly: A Case Report

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Abstract | **Case Report**

Hydranencephaly is a congenital neurodevelopmental anomaly in which there is an absence of the cerebral hemispheres and intact infratentorial structures. Diagnosis can be made prenatally or postnatally using ultrasound, CT, or MRI. Imaging findings include the presence of primitive brain structures, variable presence of the falx cerebri, and total or near-total absence of the cerebral hemispheres. We reported a case of newborn hydranencephaly in 1 day old neonate with macrocephaly.

Keywords: Ultrasonnography, CT, hydranencephaly.

INTRODUCTION

The word hydranencephaly is a fusion of hydrocephalus and anencephaly, but the condition actually represents a distinct disorder and is primarily a disease of the fetus; encephaloclastic encephalomalacia can occur in cases of severe perinatal insult [3]. Hydranencephaly occurs in less than 1 in 10,000 births and is characterized by near-total or total absence of the cerebral cortex and basal ganglia. The thalami, pons, cerebral peduncles, and cerebellum are usually present, as may be a small amount of tissue from the occipital, frontal, and temporal lobes [2]. There is no known sex or racial predilection [3]. Imaging including Ultrasonnography, CT and MR plays a key role in diagnosis.

CASE REPORT

The patient was an appropriate-for-gestational-age male infant born at 39 weeks to a 32-year-old mother and unrelated father. Prenatal care was not sought. There were no known teratogenic or infectious exposures during pregnancy. The mother's previous pregnancy had resulted in healthy child. The patient was delivered via induced vaginal delivery with Apgar scores of 7 at 1 and 5 minutes. Birth head circumference was 38 cm. (figure 1).

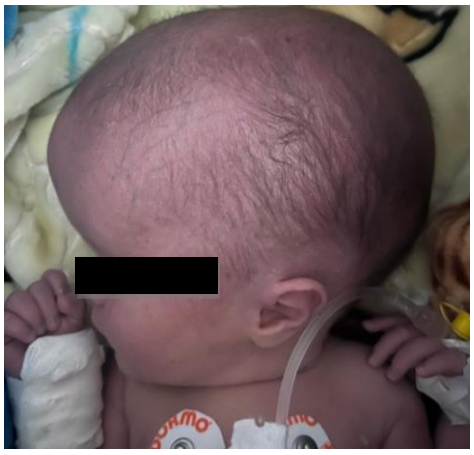

Figure 1: Face of patient with macrocephaly

The anterior fontanelle was soft, and the sutures were split. The eyes were in normal position, with no sundowning. The remainder of the general examination was normal and there were no cutaneous vascular anomalies. The neurologic examination showed a vigorous, alert-appearing infant with mild irritability and a high pitched cry. The baby had a fair suck, normal neonatal reflexes and a mildly decreased tone. The scalp could be transilluminated. A clinical diagnosis of hydranencephaly versus severe congenital hydrocephalus was reached. A ultrasonography performed suggested the absence of cortical mantle, and no brain tissue was seen above the level of the thalamus (figure 2).

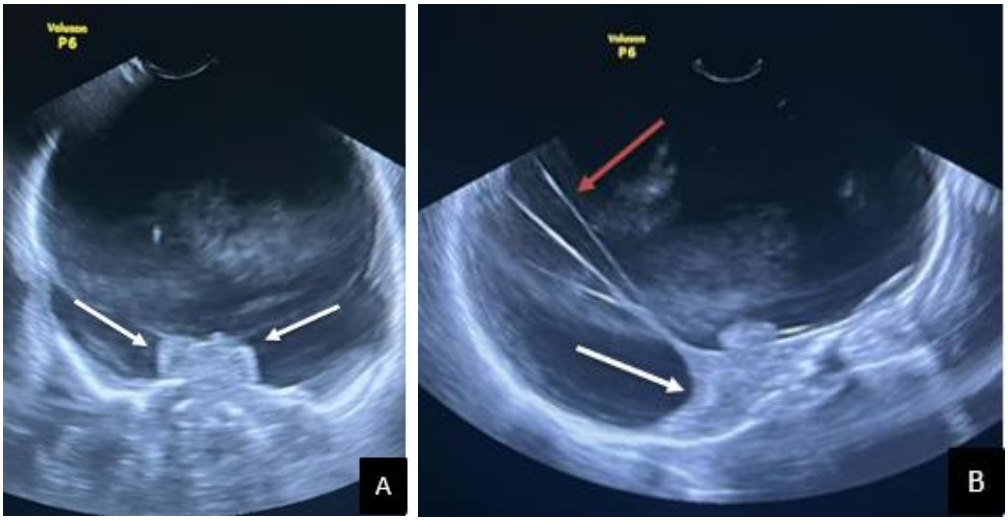

Figure 2: Parasagittal and coronal neonatal head US images shows absence of the cerebral hemispheres, which are replaced by a supratentorial fluid collection. Thalami and cerebellum (white arrows) are intact, Small portion of the falx (red arrow) is also seen

A cerebral CT scan was performed showed the abcense of the cerebral hemispheres which are replaced by a supratentorial fluid collection,With normal appearance of the cerebellum and brain stem (figure 3).

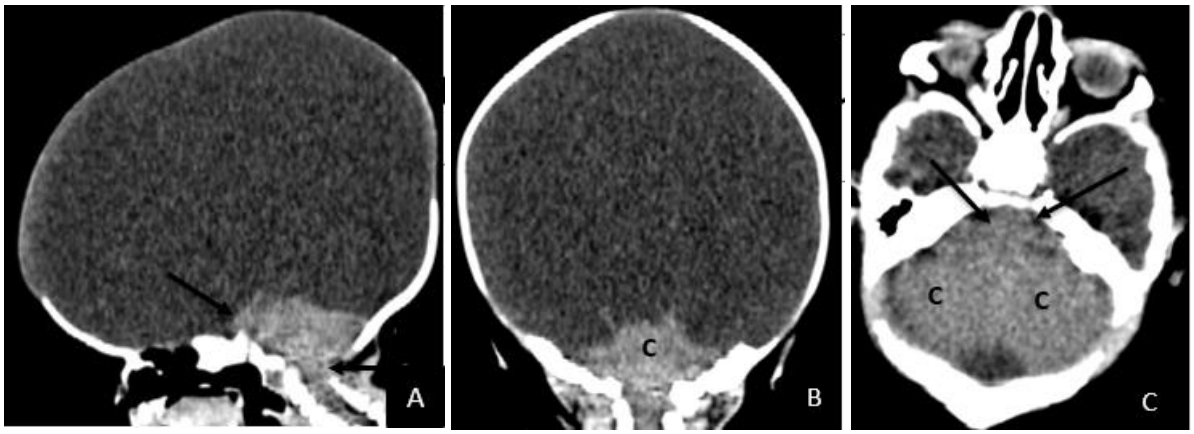

Figure 3: sagittal, coronal and axial non-enhanced cerebral CT images shows replacement of most of the cerebral hemispheres by a supratentorial fluid collection. cerebellar hemispheres (C), brain stem (arrows) are intact

DISCUSSION

Hydranencephaly is a central nervous system disorder in which there is a replacement of the cerebral hemispheres by a thin membranous sac filled with cerebrospinal fluid and debris. It is a rare condition that occurs in approximately 1 per 10,000 births [4,5,7].

Hydranencephaly occurs after the brain and ventricles have fully formed, usually in the second trimester. An infant with hydranencephaly may appear normal at birth or may have some distortion of the skull and upper facial features due to fl uid pressure inside the skull. The infant's head size and spontaneous refl exes such as sucking, swallowing, crying, and moving the arms and legs may all seem normal, depending on the severity of the condition. The exact cause of hydranencephaly is not clear. However, 5 etiologies have been proposed as follows:

- Infarction: Occlusion of the supraclinoid segments of the internal carotid arteries or of the middle cerebral arteries [1,7].
- Leukomalacia: Extreme form of leukomalacia in which there is confluence of multiple cystic cavities [7].
- Diffuse hypoxic-ischemic brain necrosis: Fetal hypoxia due to maternal exposure to carbon monoxide or butane gas resulting in massive tissue necrosis with cavitation and resorption of necrotized tissue [4,5,7].
- Infection: Necrotizing vasculitis or local destruction of brain tissue secondary to intrauterine infection. Fetal infections associated with hydranencephaly include congenital toxoplasmosis, cytomegalovirus, and herpes simplex infections [1-7].

- Thromboplastic material from a deceased co-twin: Release of embolic or thromboplastic material from a deceased twin resulting in liquefaction of the brain tissue in the surviving twin [4,5,7].

Of the proposed etiologies of hydranencephaly listed previously, the first is regarded by many to be the most likely, whereas animal studies have reproduced hydranencephaly in utero by carotid occlusion. All modalities which resolve the brain parenchyma can be used to identify the features of hydranencephaly, including ultrasound (antenatal and postnatal), MRI (antenatal and postnatal), and CT. MRI is the gold standard [11]. In all cases, the anatomical features are the same, although they are demonstrated to a variable degree according to the abilities of each modality:

- Essentially no remaining cortical tissue
  - often islands of residual tissue preserved at occipital poles and orbitofrontal regions
  - medial temporal tissue may be identified, as the medial temporal lobes are supplied by the basilar circulation [11].
- Preserved thalami and posterior fossa
- Falx is usually present
- Hemicranium is filled with fluid, in which choroid can often be identified
- Antenatal ultrasound or vascular imaging demonstrate absence of middle cerebral arteries

There is no standard treatment for hydranencephaly. Treatment is symptomatic and supportive. Hydrocephalus may be treated with a shunt (e.g.a ventriculoperitoneal shunt). The prognosis for children with hydranencephaly is generally quite poor. Death usually occurs in the first year of life [10].

CONCLUSION

Hydranencephaly is a congenital neurodevelopmental anomaly in which there is an absence of the cerebral hemispheres and intact infratentorial structures. It is generally thought to occur as a result of bilateral occlusion of the ICAs. Diagnosis can be made prenatally or postnatally using ultrasound, CT, or MRI. Imaging findings include the presence of primitive brain structures, variable presence of the falx cerebri, and total or near-total absence of the cerebral hemispheres. There is no cure for hydranencephaly, and the prognosis is poor [8, 9].

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