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Hydranencephaly in the neonatal period: a case report

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***Abstract**: Hydraencephaly is a rare congenital disorder of the central nervous system that affects about 1/5000 newborns, characterized by the complete absence of the cerebral hemispheres replaced by cephalospinal fluid and debris of the cerebral cortex [1]. Usually the cerebellum and brainstem are intact. Hydraencephaly can affect only one hemisphere (hemihydraencephaly) which is even rare and generally has a better prognosis. Its pathophysiology is still poorly understood and subject to multiple studies, In the literature, the most proposed etiology is bilateral thrombosis of the carotid arteries [1,2].*

Keywords: Hydranencephaly-UTF-CT-scann

INTRODUCTION:

Hydraencephaly is a rare congenital disorder of the central nervous system that affects about 1/5000 newborns, characterized by the complete absence of the cerebral hemispheres replaced by cephalospinal fluid and debris of the cerebral cortex [1]. Usually the cerebellum and brainstem are intact. Hydraencephaly can affect only one hemisphere (hemihydraencephaly) which is even rare and generally has a better prognosis. Its pathophysiology is still poorly understood and subject to multiple studies, In the literature, the most proposed etiology is bilateral thrombosis of the carotid arteries [1,2].

CASE REPORT:

Newborn baby, 3 days old, male, from a first degree consanguineous marriage of a 29 year old mother and a 46 year old father. The pregnancy was poorly monitored and the patient was referred to our hospital after the gynecologist noted an increase in head circumference during an antenatal ultrasound, the delivery was performed by cesarean section, the Apgar at birth was 10/10, the newborn presented a macrocrania at birth with a PC at 44 cm without signs of intra-cranial hypertension syndrome or the sensitivomotor deficit. The transfontaneous ultrasound showed a significant hydrocephalus (Fig.1) and the CT confirmed the diagnosis of hydranecerephaly by showing the subtotal absence of the supratentorial cerebral parenchyma with the respect of the posterior cerebral fossa (Fig.2), an external ventricular bypass was performed with cephalo spinal fluid output in hyperpressure, the patient died on the 7th day of life.

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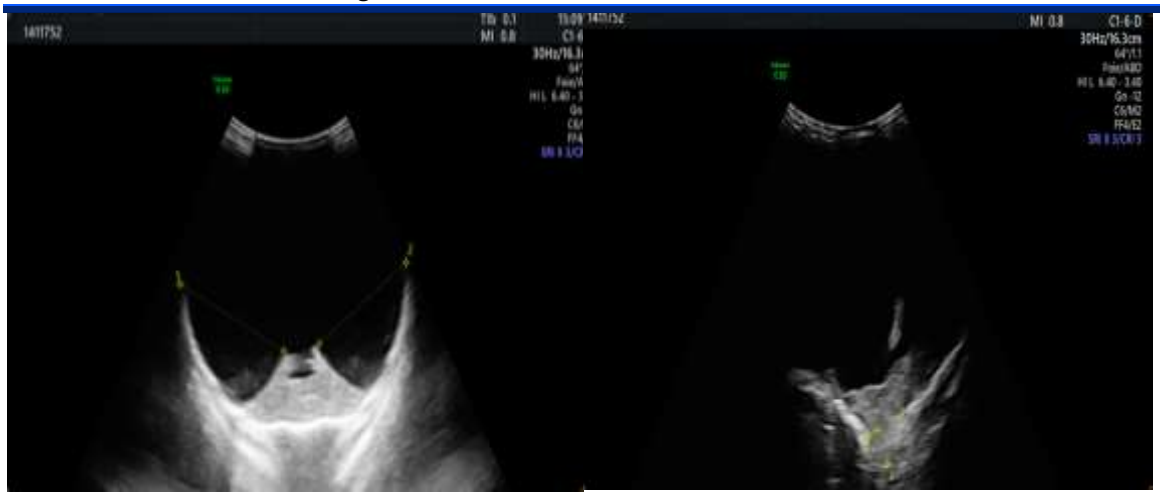

Fig. 1. transfontanellar ultrasound: Triventricular hydrocephalus, laminating the cerebral parenchyma with mass effect on the posterior cerebral fossa and ptosis of the cerebellar amygdala.

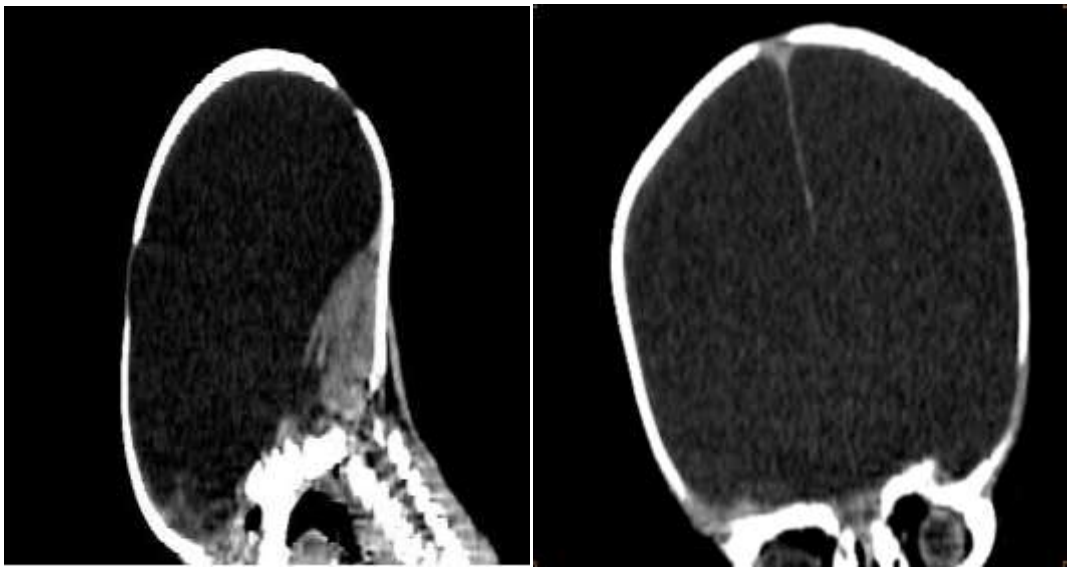

Fig. 2. Cerebral CT coronal and saggital recontruction. Scannopgraphic aspect compatible with hydranencephaly: importent triventricular hydrocephalus, laminating the cerebral parenchyma

Discussion:

Hydranencephaly is a rare congenital neurological disorder, characterized by the absence of the supratentorial brain parenchyma replaced by CSF with preservation of the posterior cerebral fossa [1,3].

The causes of central nervous system (CNS) malformations are both genetic and environmental, often interrelated, multifactorial, with different types of interactions between genes and environmental factors. It is commonly described as a consequence of bilateral thrombosis of the internal carotid arteries of infectious (toxoplasmosis, CMV), malformative, or hemodynamic origin, which explains the presence of a subtentorial parenchymal tissue [2,4].

The postnatal diagnosis is made by imaging, in particular CT or MRI [1,3].

The antenatal diagnosis can be made by ultrasound as early as the 13th week of pregnancy. In our case, the diagnosis was made in the neonatal period on the 3rd day of delivery with transfontanellar ultrasound and a cerebral CT.

At birth, the clinical examination of the newborns did not reveal any clinical sign that could distinguish hydranencephaly from hydrocephalus.

Macrocrania requires a paraclinical exploration, such as electroencephalogram, which allows to differentiate between hydrocephalus and hydranencephalus by the presence of electrical activity in hydrocephalus and the absence of electrical activity in hydranencephalus [1,5].
The differential diagnoses are severe hydrocephalus, alobar holoprosencephaly and schizencephaly [1].
Holoprosencephaly results from a defect in the induction of neurectoderm by the prechordal plate.

The prognosis of hydranencephaly remains poor and the majority of patients do not survive beyond the neonatal period, however cases are described in the literature that have survived to the adolescent age and and a woman described in the literature survived until the age of 32 years with a some autonomy. These cases of survival observed are due to the preservation of the subcortical and brainstem regions that contain the neural circuitry necessary to maintain body temperature, blood pressure, cardiorespiratory and other vital functions [6]. Cephalocentesis can be proposed to the mother if the diagnosis of hydranencephaly is made antenatally, thus allowing decompression of the fetal head and avoiding an unnecessary cesarean section [5].

CONCLUSION:
Hydranencephaly is a rare neurological disorder, its physiopathology is still a subject of scientific research, its diagnosis relies mainly on imaging, no treatment seems to be effective at the moment and the majority of the cases do not go beyond the neonatal period, the ante-natal diagnosis allows to discuss a medical interruption of the pregnancy or a cephalocentesis avoiding a useless caesarean.

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