

## Case report



# Radiological aspect of hydranencephaly: a case report

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## Radiological aspect of hydranencephaly: a case report

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## Abstract

*Hydranencephaly is a severe destructive cerebral lesion with absence of the cerebral hemispheres. It can be the consequence of a vascular accident, infection, maternal exposure to toxins, or death of a co-twin in a monochorionic pregnancy. The typical US appearance is a large cystic structure replacing the fetal brain, but the brainstem and the thalami are preserved. It is usually identified at midgestation, but it also can occur later in pregnancy. The prognosis is very poor.*

## Introduction

Hydranencephaly is characterized by the absence of the cerebral hemispheres, with incomplete or absent cerebral falx. The fluid-filled sac that replaces the brain is formed by the leptomeninges and the cerebrospinal fluid [1]. The prognosis is universally poor, and death usually occurs within the first year of life. Termination of pregnancy should be advised if possible. Hydranencephaly is considered to be sporadic, with a recurrence in cases of infectious disorders [2].

## Patient and observation

We present the case of newborn admitted at J7 of life, male, from a followed pregnancy estimated at 40 SA, no notion of consanguinity, delivery by caesarean section of a 35 year old mother; first gestures first parities. The infectious history was positive. Apgar at birth is 10/10. Since birth, the newborn presented with major macrocrania, early vomiting, and a refusal to suckle in a context of apyrexia. Clinical examination finds a subicteric neonate hypotone with a sunset look. Transfontanelle Ultrasonography was objectived an extreme hydrocephalus and cerebellum preserved (Figure 1). Cranial Computed Tomography without intravenous injection identified the hydranencephaly which demonstrates the fluid-filled sac almost completely replacing the cerebral hemispheres. The cerebellum was preserved (Figure 2).

## Discussion

Hydranencephaly is defined as the absence of the cerebral hemispheres; the hemispheres are replaced by a fluid-filled sac [3]. It is rare. The exact cause of hydranencephaly is not clear. However, it has been proposed infarction (Occlusion of the internal carotid arteries or of the middle cerebral arteries), diffuse hypoxic-ischemic brain necrosis and Infection [4, 5]. All imaging modalities, including US, computed tomography, and magnetic resonance, can be used to identify the features of

hydranencephaly. In all cases, the imaging findings are the same. The cerebral tissue is absent. The thalami, brain stem, and cerebellum are preserved. In many cases, islands of residual tissue are seen at the occipital poles and orbitofrontal regions. The falx is usually present, and the cranial cavity is filled with fluid [6-8]. Hydranencephaly is associated with a poor prognosis, and most of the affected infants do not survive beyond the first year of life [9]. Hydranencephaly must be distinguished from extreme hydrocephalus, porencephaly, and alobar holoprosencephaly. The presence of even minimal frontal cerebral cortex is indicative of hydrocephalus. Color Doppler may be useful for differential diagnosis because anterior and middle cerebral arteries are not visualized in hydranencephaly [10, 11].

## Conclusion

Understanding the typical imaging manifestations of hydranencephaly, in the antenatal period, permits a more specific and accurate diagnosis. Wherever, it allows for discussion of medical termination of the pregnancy if possible.

## Competing interests

The author declare no competing interests.

## Authors' contributions

The author read and agreed on the final manuscript.

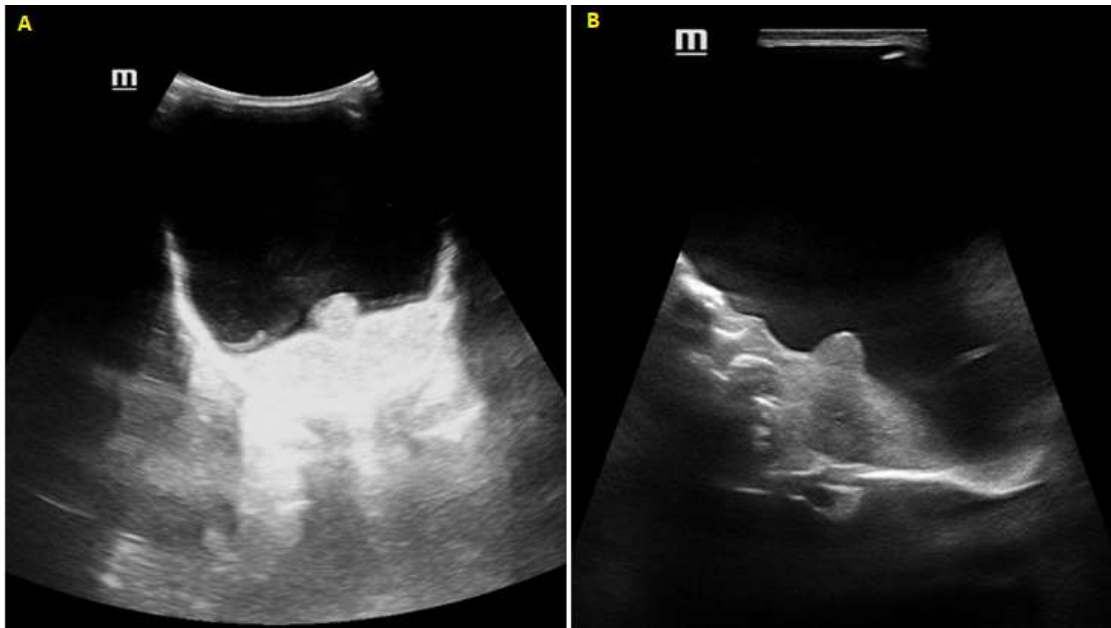
## Figures

**Figure 1:** transfontanelle ultrasonography: coronal and axial section showing the fluid-filled sac almost completely replacing the cerebral hemispheres

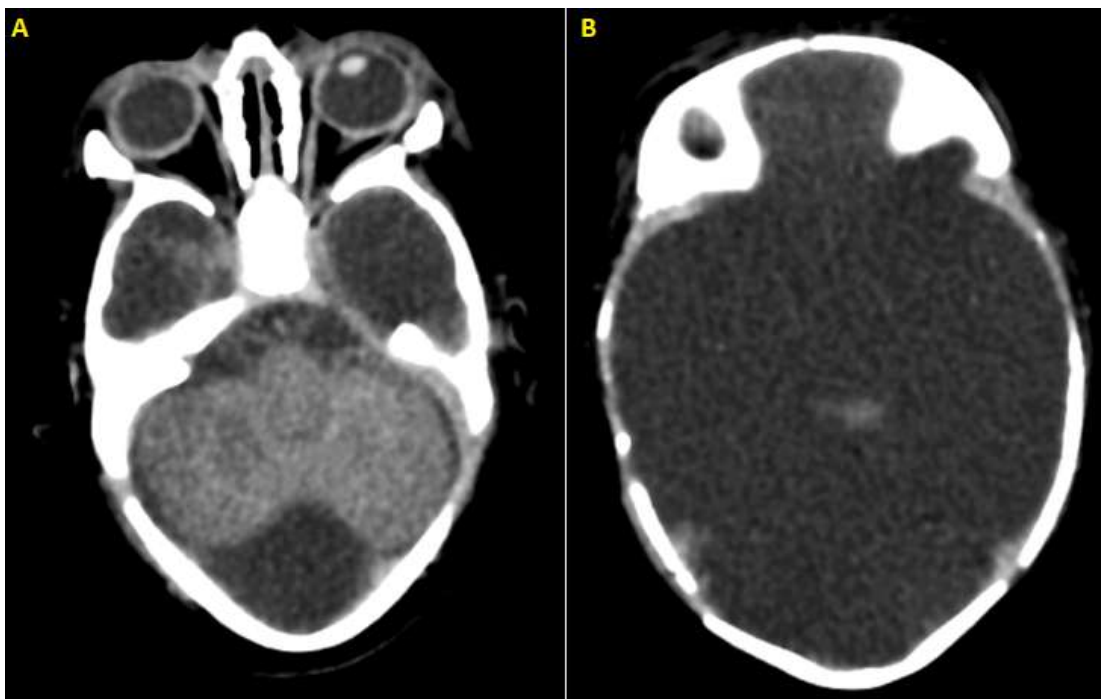
**Figure 2:** cranial computed tomography without intravenous injection (axial section): the fluid-filled sac almost completely replacing the cerebral hemispheres and the cerebellum was preserved

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**Figure 1:** transfontanelle ultrasonography: coronal and axial section showing the fluid-filled sac almost completely replacing the cerebral hemispheres



**Figure 2:** cranial computed tomography without intravenous injection (axial section): the fluid-filled sac almost completely replacing the cerebral hemispheres and the cerebellum was preserved