Open Access Library Journal
2024, Volume 11, e10941
ISSN Online: 2333-9721
ISSN Print: 2333-9705

Hydranencephaly in an Infant: About a Case at Medical Imaging for All (IMT) in Kinshasa/The Democratic Republic of Congo

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How to cite this paper: Diallo, B.I., Ndudi, P., Mazoba, T.K., Mbongo, A.T., Mvila, G.L., Tshibola, J.M., Kuzoma, L., Minouche, C., Anelia, P.L., Kola, R.S., Tshikwela, M.L. and Molua, A.A. (2024) Hydranencephaly in an Infant: About a Case at Medical Imaging for All (IMT) in Kinshasa/The Democratic Republic of Congo. *Open Access Library Journal*, **11**: e10941.
https://doi.org/10.4236/oalib.1110941

Received: October 30, 2023
Accepted: February 25, 2024
Published: February 28, 2024

Abstract

Hydranencephaly corresponds (in its major and bilateral form) to the total ischemic destruction (most often occurring between 3 and 6 months of pregnancy) of the normally developed cerebral hemispheres which are replaced by a vast liquid cavity surrounded by the meninges. However, there are the basal ganglia, the brainstem and the cerebellum. We report and describe a case of an infant, aged 9 months received in Medical Imaging for All for a brain scan on macrocephaly. In the CT-scan, we note the absence of the cerebral hemispheres replaced by LCS, without peripheral cortical plate except at the medial part of the temporal lobes, constituting a cerebral water pocket. The thalamus, cerebellum, brain stem and falx cerebri are intact. The residual temporal and occipital lobes. Exaggerated opening of the fontanelles. We concluded with hydranencephaly and we completed by transcranial ultrasound. We find an anechoic aspect of the cranial cavity, the presence of the falx of the brain; the persistence of cerebral remnants in the occipital region, medial of the temporal lobes vascularized by the anterior and posterior cerebral arteries and their anastomoses, the posterior fossa, the tent of the cerebellum, the basal nuclei and the brainstem which are preserved; the cranial vault is deformed with macrocephaly by continuous production of LCS.

Subject Areas

Clinical Medicine

Keywords

Hydranencephaly, IMT, Kinshasa

1. Introduction

Hydranencephaly corresponds (in its major and bilateral form) to the total ischemic destruction (most often occurring between 3 and 6 months of pregnancy) of the normally developed cerebral hemispheres which are replaced by a vast liquid cavity surrounded by the meninges. However, there are the basal ganglia, the brainstem and the cerebellum [1] [2]. It can fit into a polymalformative syndrome or not.

Hydranencephaly is a rare malformation of the central nervous system [3]. It represents 1% of diagnosed hydrocephalus [4]. Its incidence would be between 1/10,000 and 1/5000 of pregnancies [4] [5] [6]. In the USA the incidence would be 1.4 to 2.8 per 100,000 births [7]. In Japan, it would be 2.1 per 100,000 births [3].

Diagnosis is facilitated by imaging and electroencephalogram (EEG) [8]. Its management remains poorly codified, and poses an ethical problem with regard to the quality of life with or without treatment of its children [7]. Its pathophysiology remains unclear. It would result from an anomaly in embryogenesis, after the formation of the neural plate. The main characteristic of hydranencephaly is an absence of cortical mantle, replaced by cerebrospinal fluid (CSF); when it exists, it is non-functional. It is secondary to internal carotid thrombosis itself secondary to an infection (toxoplasmosis, CMV), a vascular malformation or hemodynamic disorders. It can become part of Fowler's syndrome (lethal affection *in utero*) which is characterized by glomerular vascular proliferation, hydranencephaly/hydrocephalus and fetal akinesia. Hydranencephaly is exceptionally unilateral [9]. We report and describe a case of hydranencephaly diagnosed in a resource-limited country.

2. Methods

This is a case-report carried out at the "Medical Imaging for All" Hospital Center in June 2023. It is a center based in Kinshasa, renowned in the DRC for the diagnostic management of diseases and in ultrasound monitoring pregnant women. A Toshiba 16-bar scanner and a high-end Philips IU22 ultrasound scanner were used for the examination of our patient.

Ethics approval

The data were treated anonymously and strictly confidential:

- This protocol was then presented to the Local Committee of the Medical Imaging Department of the Faculty of Medicine of the University of Kinshasa, and received a favorable opinion.

- The rules of benevolence and ethical respect as prescribed in the guidelines described in the Declaration of Helsinki were scrupulously followed.
- The Helsinki principles were respected [anonymous and strictly confidential treatment of data].
- The case was presented to the staff and received a favorable opinion (cirimed.007/2023).

3. Presentation of the Case

3.1. Patient Information

It was an infant, aged 9 months, living in the Mont-Ngafula commune in Kinshasa, received in Medical Imaging for all for a brain scan on macrocephaly. She is the second in a family of two children, the other of whom is in apparent good health.

There was no history of malformation or consanguinity in the family.

He had never performed the imaging examination for this pathology since birth.

His birth weight was 3900 g. The mother had not performed the fetal ultrasound during the pregnancy.

3.2. Scan Result

On the cerebral scanner taken in helical acquisition, seen in parenchymal and bone windows, followed by multiplanar reconstruction (MPR) without injection of iodinated contrast product: we note the absence of the cerebral hemispheres replaced by LCS, without peripheral cortical plate except at the medial part of the temporal lobes, constituting a cerebral water pocket. The thalamus, cerebellum, brain stem and falx cerebri are intact. Figure 1 provides on this information. We concluded to hydranencephaly and we completed by transcranial ultrasound.

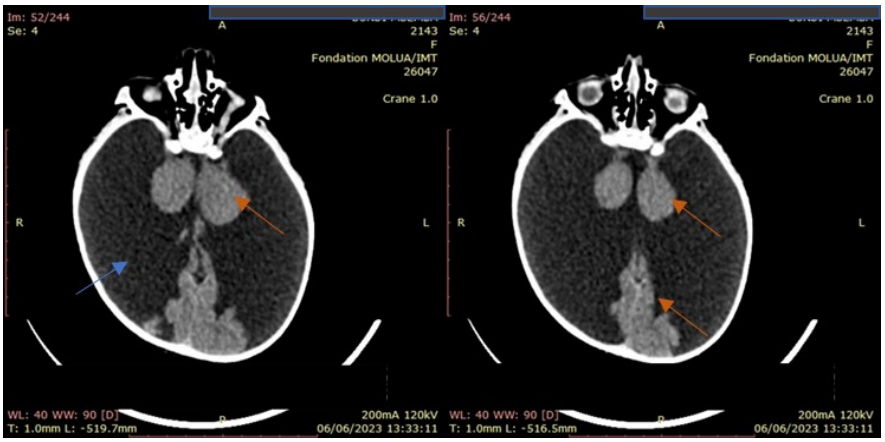

Figure 1. Cranio-encephalic scanographic images in axial section the absence of the cerebral hemispheres replaced by LCS (blue arrow). The thalamus, cerebellum, brain stem and falx cerebri are intact. The residual temporal and occipital lobes (red arrow).

3.3. In Standard Transcranial Ultrasound and in Color Doppler Mode

We find an anechoic aspect of the cranial cavity, the presence of the falx of the brain; the persistence of cerebral remnants in the occipital region, medial of the temporal lobes vascularized by the anterior and posterior cerebral arteries and their anastomoses, the posterior fossa, the tent of the cerebellum, the basal nuclei and the brainstem which are preserved; the cranial vault is deformed with macrocephaly by continuous production of LCS. Figure 2 provides on this information.

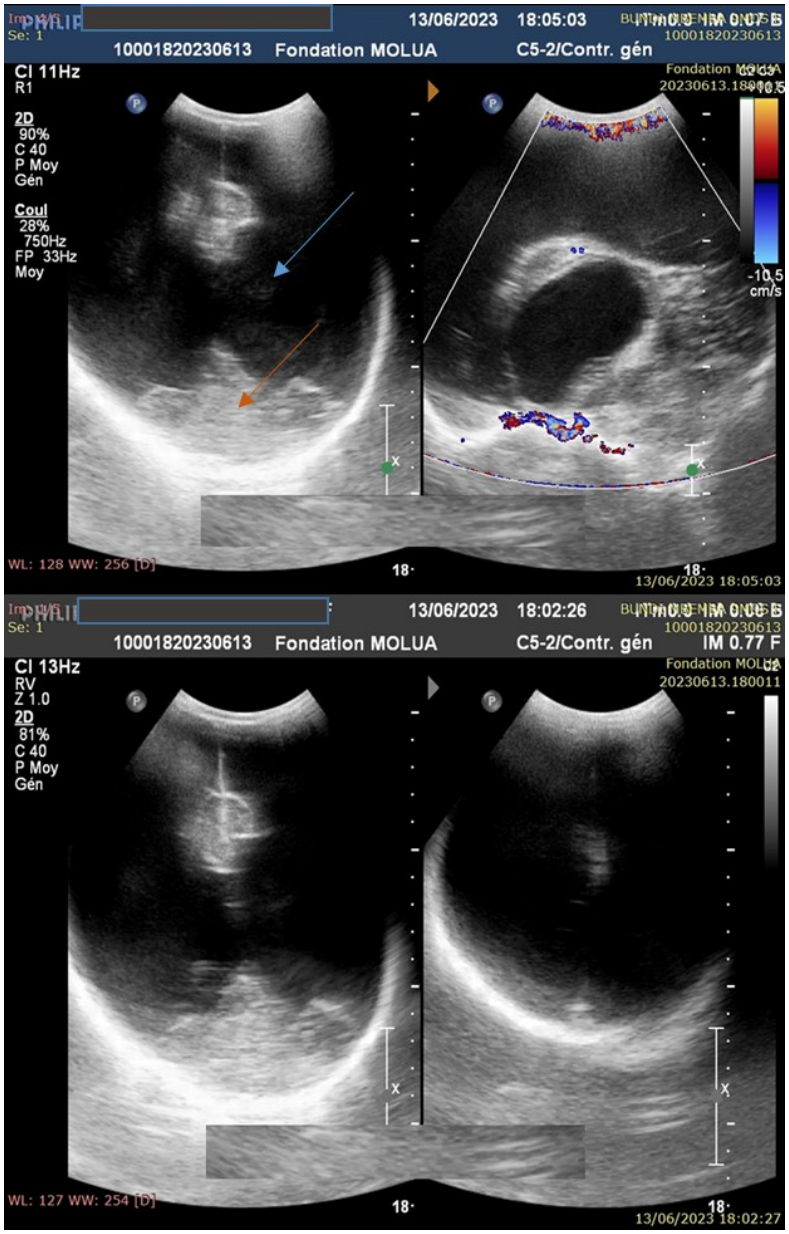

Figure 2. Ultrasound images showing an anechoic aspect of the cranial cavity (blue arrow), the presence of the false brain; the persistence of cerebral remnants in the occipital region (red arrow).

4. Discussion

In our case, the diagnosis was based on the use of cerebral scanner coupled with transcranial ultrasound. The use of cMRI is recommended although access is more difficult, especially in countries with limited resources such as the Democratic Republic of Congo (DRC). The antenatal diagnosis was not found in our study. The notion of parental consanguinity was also not found in our case, unlike Mbaye M *et al.* [9] in Senegal, who report 41% of inbreeding cases, while Malheiros *et al.* [3] report 17.6% in Brazil.

We noted a delay in consulting parents, which we explain by the fact that the population is poorly informed about this pathology and its clinical manifestations, but also by the low socio-economic level of this family. This observation coincides with that of Mbaye M *et al.* [9]. Indeed the average cranial circumference of the study shows the lack of responsiveness of the parents faced with the evolution of the pathology in their child.

The mean age in most literature data does not exceed 1 year for those who did not die in utero [9] [10]. It has been described in the literature of cases having progressed up to the age of 32 years [5]. This poses, on the one hand, the problem of ethics in the face of the use of medical termination of pregnancy as a therapeutic means, and on the other hand the high cost of taking care of these children who have an almost zero intellectual development [11] [12]. The absence of realization of ultrasound during pregnancy in the DRC seems to be an explanation vis-à-vis our case.

Psychological support and counseling for families can constitute means of prevention for subsequent pregnancies: fetal ultrasound which is thus presented as a "must pass".

5. Conclusion

Hydranencephaly is a serious malformative pathology, diagnosed both prenatally and after birth using medical imaging methods, including ultrasound, CT scan and MRI. This pathology has a poor prognosis, and poses a major socio-economic and ethical problem.

Conflicts of Interest

The authors declare that they have no conflict of interest.

Authors' Contributions

Conception: Diallo Bomane and Antoine Molua. Writing and submission: Tacite Kpanya. Proofreading of plates and manuscript: All authors have read and approved.

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