Hydranencephalyin monochorionic twin pregnancy with in utero fetal death of the co-twin: A case report

ABSTRACT:

Hydranencephaly is a rare congenital brain malformation with an incidenceestimated to be between 1/10000 and 1/5000 of pregnancies. It is characterized by the absence of development of the cerebral hemispheres, which are replaced by cerebrospinal fluid.
The diagnosis of hydranencephaly can be made from 15 weeks of amenorrhea via obstetric ultrasound;however, due to the operator-dependent nature of this method, antenatal nuclear MRI remains the definitive test to establish the diagnosis. The condition results from an anomaly in embryogenesisafter the formation of the neural plate,though the exactpathophysiology of hydranencephaly is not yet clear.
There is no definitive treatment for hydranencephaly,treatment is symptomatic and supportive. Hydrocephalus may be treated with a shunt,such as ventriculoperitoneal shunt. This condition poses anethical problem regarding the quality of life and the treatment of affected children.The prognosis for children with hydranencephaly is generally poor; with death usually occurring within the first year of life.
It is importantto distinguish this condition, which has an associated poor prognosis, from extensive hydrocephalus, which has the potential for improved prognosis with early shunting procedures.
We reporta case of a female newborn, from a monochorionic twin pregnancy with an in utero fetal death of her twin, admitted for the exploration ofcongenital hydrocephalus and IUGR detected on prenatal ultrasound. Clinically, she had microcephaly, and postnatalimaging revealed hydranencephaly.

INTRODUCTION:

Hydranencephaly is the most severe form of bilateral cerebral cortical destruction, characterized by the almost complete absence of the cerebral hemispheres.In this condition, the cerebral hemispheres are replaced by sacs filled with cerebrospinal fluid.
Hydranencephalic infants may look remarkably normal after birth, but structural defects can be identified using modern neurological techniques.
There is no definitive treatment for hydranencephaly. The prognosis for children with hydranencephaly is generally poor, and many children with this disorder die before their first birthday.

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CASE PRESENTATION:

We report the case of a female newborn,born to a 26-year-old motherat 36 weeks' gestation, gravida 1 para 2,from a monochorionic twin pregnancy, by caesarean section due to in utero fetal death ofco-twin, the mother had an O+ blood type, and her infectious history was negative, with no special medical history, no history of taking toxic or medical drugs, and no consanguinity,Apgar scores were 10 at both 1 and 5 minutes, the newborn had a birth head circumference of 28 cm, which is below the 10^{th} percentile.

Congenital hydrocephalus and intrauterine growth restriction (IUGR) were detected on prenatal ultrasound, and the infant was therefore transferred to the neonatal intensive care unit for evaluation after birth.She was admittedto our Neonatology service on the 10^{th}day of life.

Physical examination revealed a dehydrated newborn with a 16% weight loss over 10 days (admission weight was 1900g versus 2300g at birth), her height was 43 cm,below the 10th percentile(-3 SD). Microcephaly was noted, with a head circumference of 28 cm, also below the10^{th} percentile(-3 SD), No other craniofacial dysmorphism , extremity deformities, or other congenital malformations were observed. Primitive reflexes, including sucking and grasping reflexes, were intact.

Laboratory data were all within normal limits.The TORSCH serology for our patient was negative.A brain computed tomography scan demonstratedsignificant biventricular hydrocephalus, compressing the brain parenchyma, with evidence of encephalomalacia.Brain magnetic resonance imaging revealed hydranencephaly with bilateral frontal cystic encephalomalacia lesions.

As part of the malformation assessment, the abdominal-renal and cardiac ultrasound was normal.

Upon admission,the patient received rehydration; the evolutionwas favorable in terms of hydration with weight gain and an increase in cranial perimeter by 0,5 cm in 15 days.No surgical treatment is planned, particularly no external diversion.After discussion with the parents, palliative care was offered on day 25 of life at home. Psychological monitoring for the parents was provided.

Figure 1: Images of our patient showing microcephaly: a)before rehydration b) Afterrehydration

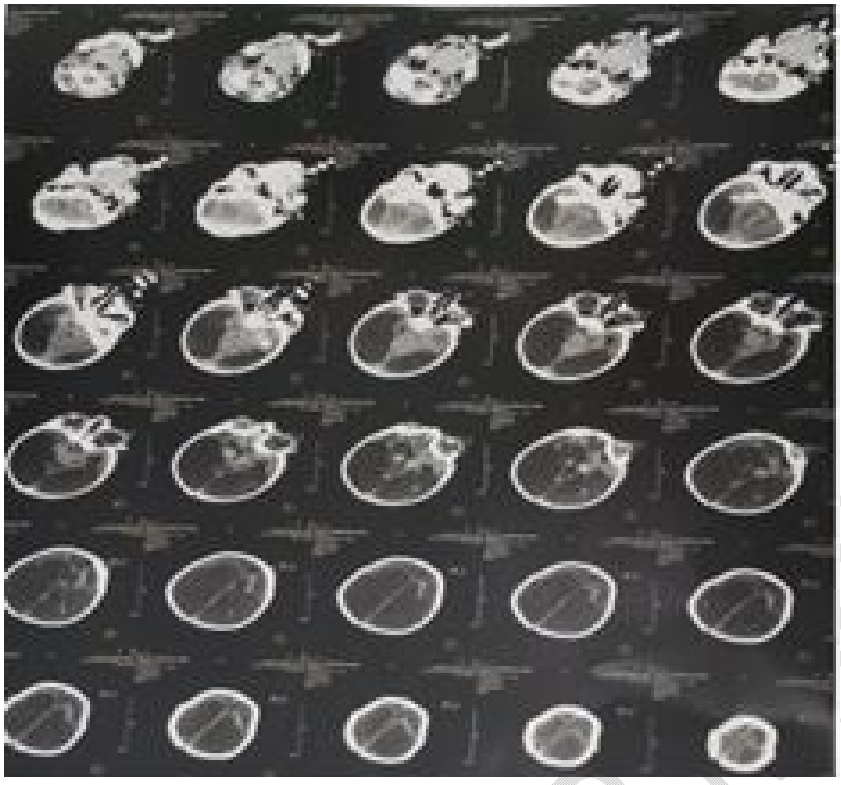

Figure 2:Brain CT of our patient: significant hydrocephalus lamining the brain parenchyma

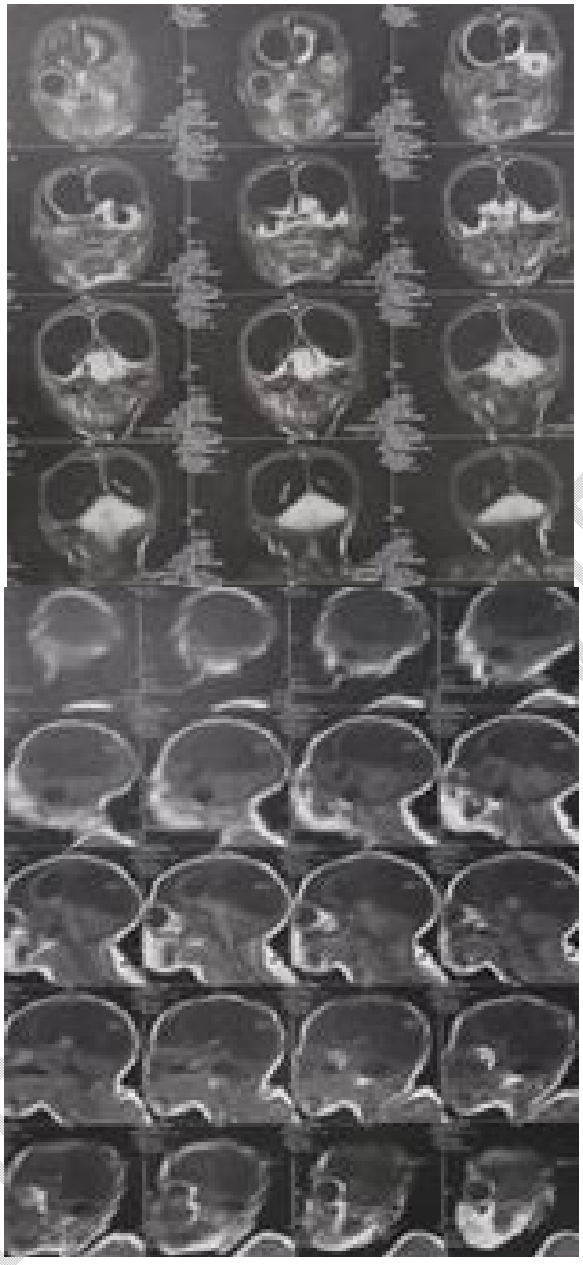

Figure 3: brain MRI indicative of hydranencephaly with bilateral frontal cystic encephalomalacia lesions

DISCUSSION:

Hydranencephaly was fully described as a different entity from hydrocephalus in 1972 by Crome **[1].**It is a rare brain malformation,its incidence would be between 1/10000 and 1/5000 of pregnancies **[2-3].** It represents 1% of diagnosed hydrocephalus **[2].**

Hydranencephaly occurs after the brain and ventricles have fully formed, usually in the second trimester. The brain destruction is complete or almost complete in a bilateral internal carotid artery distribution, with the cerebral hemispheres replaced by fluid covered with leptomeninges and dura. During the destructive phase, unusual ‘‘masses’’ of hemorrhage and soft tissue may be seen **[4].** Because the ventricles have already been formed, the falx cerebri is present. The cerebellum, midbrain, thalami, basal ganglia, choroid plexus, and portions of the occipital lobes, all fed by the posterior circulation, are typically preserved.

While the pathogenesis of hydranencephaly is thought to be a vascular accident, this cannot always be confirmed because internal carotid arteries are not always occluded at autopsy **[5].**Intrauterine infections, particularly toxoplasmosis and viral infections (enterovirus, adenovirus, parvovirus, cytomegalic, herpes simplex, Epstein-Barr, and respiratory syncytial viruses), have been implicated in a number of cases. Toxic exposures and cocaine abuse have been reported, and hydranencephaly has been described in rare syndromes **[6].** An extreme form of leukomalacia formed by confluence of multiple cystic cavities **[7].**Diffuse hypoxic-ischemic brain necrosis **[8].**In monochorionic twin pregnancies, death of one twin in the second trimester may cause a vascular exchange to the living twin through the placental circulation, leading to hydranencephaly in the surviving fetus **[9].**
In our case, we observed the presence of encephalomalacia and monochorionic twin pregnancy with intrauterine death of one twin.

The diagnosis of hydranencephaly can be made from 15 weeks of amenorrhea by obstetric ultrasound, but given its operator-dependent nature, antenatal nuclear MRI remains the definitive test to establish the diagnosis **[10].**

The postnatal diagnosis of hydranencephaly is made by brain CT scan and magnetic resonance imaging shows the absence of brain parenchyma which is replaced by cerebrospinal fluid, the subcortical elements of which the thalami are generally preserved**[11],** evoked potentials showed the absence of any cortical activity **[12].**

The differential diagnosis is major hydrocephalus and holoprosencephaly**[12].**

There are important reasons to differentiate hydranencephaly from hydrocephalus; these reasons relate to prognosis and management **[17, 18].** Children with hydrocephalus, without chromosomal or other structural abnormalities, have an unpredictable prognosis. With proper ventricular shunt after birth, mentation may in some cases be normal. In contradistinction, hydranencephaly has an irretrievably poor prognosis, with only brain stem function remaining.

Treatment of hydranencephaly remains poorly codified, and poses an ethical problem regarding the quality of life with or without treatment **[13].** The diagnosis of hydranencephaly is a crucial time for parents to fully understand, before deciding on pursuing medical treatment that is palliativeand relies on diversion of cerebrospinal fluid to attempt to preserve subcortical tissues and prevent massive macrocephaly and may prolong survival but not influence neurodevelopment **[14].**In surviving cases, preservation of subcortical brainstem regions that contain neural circuitry is necessary to maintain body temperature, blood pressure, cardiorespiratory, and other vital functions. While most patients do not survive beyond the neonatal stage, some have been able to live for years and even into adulthood **[15].** If hydranencephaly has been definitively diagnosed in utero, cephalocentesis may be offered to decompress the fetal head, allowing vaginal delivery. Even if this may further damage the fetal head, it will not change the result and above all will spare the mother from an unnecessary operation **[16].**

CONCLUSION:

hydranencephalyis a rareand serious malformation of the nervous system with a guarded prognosis; the etiopathogenesis is still poorly elucidated. The antenatal period, it allows us to discuss the medical termination of pregnancy if it is possible. Otherwise decide on a cephalocentesis to avoid a cesarean section. The treatment depends on palliative care and psychological support for parents.

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