Monochorionic Twin Pregnancy with Hydranencephaly and Co-Twin Death: A Case Report

ABSTRACT:

Background:Hydranencephaly is a rare congenital brain malformation with an incidenceestimated to be between 1 in 10000 and 1 in 5000 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, with antenatal nuclear MRI being 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. The prognosis for children with hydranencephaly is generally poor, with death usually occurring within the first year of life.

Methods:We conducted a thorough clinical examination and neurological assessment. Imaging studies (CT, MRI) confirmed the diagnosis of hydranencephaly. Data were collected from medical records and interviews with the patient's parents.

Results: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 intrauterine growth restriction (IUGR) detected on prenatal ultrasound. Clinically, she had microcephaly, and postnatalimaging revealed hydranencephaly.

Conclusion and Recommendations:hydranencephaly is a rare and serious malformation of the nervous system with a guarded prognosis.The etiopathogenesis is still poorly understood. During the antenatal period, it is important to discuss the possibility of medical termination of pregnancy if feasible. Otherwise,cephalocentesismight be considered to avoid a cesarean section. It is crucial to distinguish this condition, which has a poor prognosis, from extensive hydrocephalus, which has the potential for an improved prognosis with early shunting procedures.

Key words: Hydranencephaly, Monochorionic Twin Pregnancy, Female newborn, Cephalocentesis

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 appear remarkably normal at 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, delivered by caesarean section due to

in utero fetal death ofthe co-twin.The mother had an O+ blood type, her infectious history was negative, and there was no significant 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 examinationrevealed 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.

All laboratory data were within normal limits.The TORCH serology for our patient was negative.A brain computed tomography (CT) scan demonstratedsignificant biventricular hydrocephalus, compressing the brain parenchyma, with evidence of encephalomalacia.Brain magnetic resonance imaging (MRI) revealed hydranencephaly with bilateral frontal cystic encephalomalacia lesions.

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

Upon admission,thepatient received rehydration; the evolutionwas favorable in terms of hydration with weight gain and an increase in cranial circumference 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 theparents was provided.

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

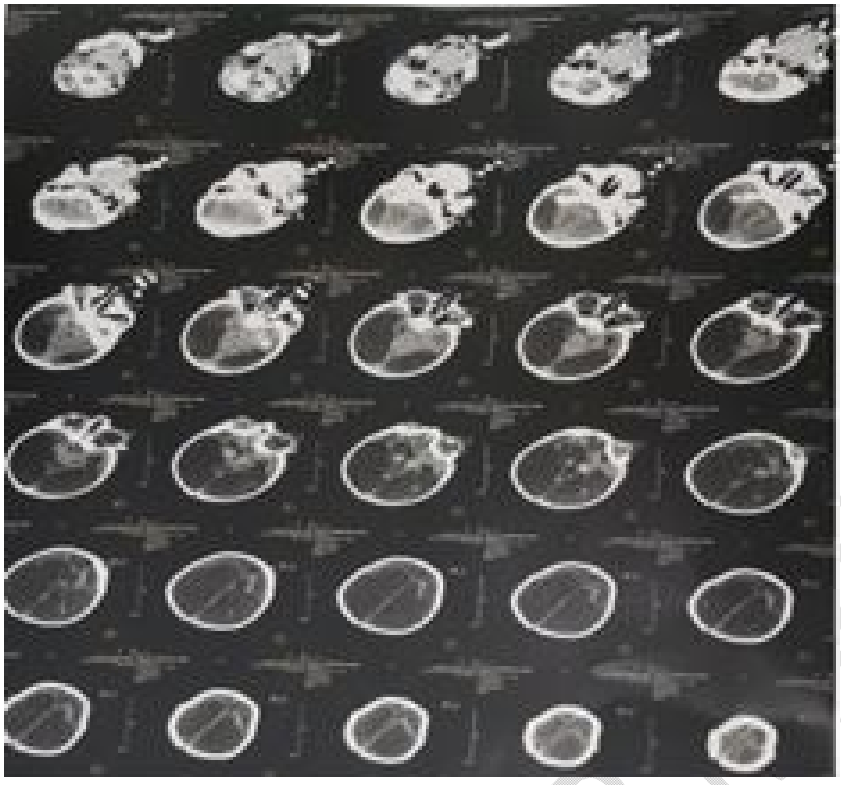

Figure 2:Brain CT of our patient: significant hydrocephalus lamining the brain parenchyma

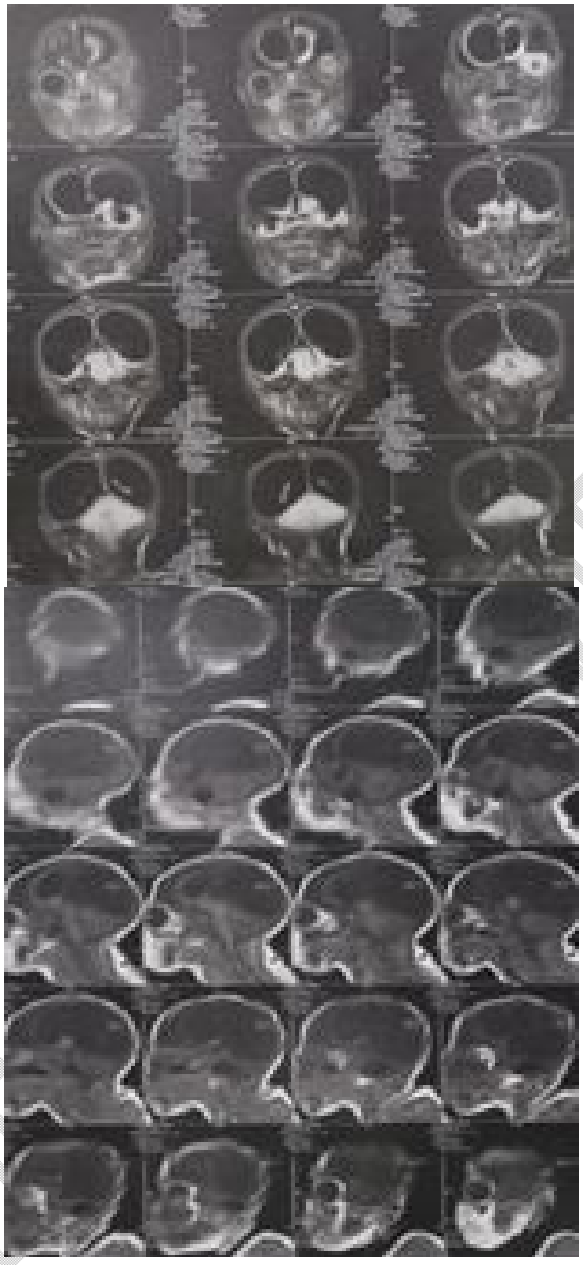

Figure 3: brain MRI indicative of hydranencephaly with bilateral frontal cystic encephalomalacia lesions

DISCUSSION:

Hydranencephaly was fully described as a different entity from hydrocephalus in 1972 by Crome **[1].**It is a rare brain malformation,with an incidence estimated to be between 1in 10000 and 1 in 5000 pregnancies **[2-3].** It accounts for 1% of diagnosed hydrocephalus cases**[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 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 the internal carotid arteries are not always occluded at autopsy" **[5].**"Intrauterine infections, particularly toxoplasmosis and viral infections (enterovirus, adenovirus, parvovirus, cytomegalovirus, 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 the 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 a 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, with the subcortical elements such as the thalami generally preserved"**[11],** evoked potentials showed the absence of any cortical activity **[12].**

"The differential diagnosis includes 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 shunting after birth, cognitive function may, in some cases, be normal. In contrast,hydranencephaly has an irretrievably poor prognosis, with only brain stem function remaining.

"The 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 the diversion of cerebrospinal fluid to attempt to preserve subcortical tissues and prevent massive macrocephaly. This approachmay prolong

survival but does not influence neurodevelopment”**[14].“**In surviving cases, the 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].No treatment seems to be effective at the moment.Antenatal diagnosis allows for the discussion of medical termination of the pregnancy or a cephalocentesisavoiding a futile caesarean section due to the poor prognosis” [20-22]
“If hydranencephaly has been definitively diagnosed in utero, cephalocentesis may be offered to decompress the fetal head, allowing for vaginal delivery. Although this procedure may further damage the fetal head, it will not change the outcome and,most importantly, will spare the mother from an unnecessary operation”**[16-19-20].**

“Cephalocentesis,though not routinely used, is a destructive procedure that still has an important role in modern obstetrics. It can be utilized to drain excessive cerebrospinal fluid (CSF) from a fetus with hydrocephalus”**[28, 29],**particularly in cases with severe associated abnormalities incompatible with survival (e.g. thanatophoric dysplasia), those compatible with survival but resulting in the potential absence of cognitive function, or those associated with severe neurological abnormalities (e.g. Edwards’ syndrome (Trisomy 18), alobar holoprosencephaly, or Dandy-Walker malformation),or in cases of intrauterine fetal demise. It is also usedin nonviable fetuses with hydrocephalus to facilitate normal vaginal delivery and avoid maternal morbidity associated with cesarean delivery **[23-26-27]**There are no fetal benefits to the procedure, as cephalocentesis almost always results in perinatal death due to intracranial bleeding. Chervenak et al**. [24]** performed cephalocentesis in eleven patients with hydrocephalus to promote vaginal birth, and ten out of these eleven had perinatal deaths due to intracranial bleeding. Perinatal death following cephalocentesis has been reported in over 90% of cases. **[30]** If decompression is performed in a controlled manner, mortality may be reduced.

“The cephalocentesis technique varies depending on the fetal presentation. In cephalic presentation, it can be performed transvaginally,but in breech position, it is wiser to perform it transabdominally. To make the procedure safer and easier, it can be done under ultrasound guidance” [20]. Ultrasound guided transabdominal cephalocentesis was first performed by Osathanondh**[21]**in 1980. It is a simple procedure without serious complications and,in cases of obstructed labor due to a hydrocephalic fetus, can easily be performed.This procedure can prevent needless cesarean sections and obstetric complications such as trauma to the birth canal or uterine rupture.

CONCLUSION:

Hydranencephalyis a rareand serious malformation of the nervous system with a guarded prognosis;its etiopathogenesis is still poorly understood. During the antenatal period, it allows for discussion of medical termination of pregnancy if possible. Otherwise,cephalocentesiscan be considered to avoid a cesarean section. The treatment focuses on palliative care and psychological support for parents.

Disclaimer (Artificial intelligence)

Option 1:

Author(s) hereby declare that NO generative AI technologies such as Large Language Models (ChatGPT, COPILOT, etc) and text-to-image generators have been used during writing or editing of manuscripts.

Consent

As per international standards or university standards, patient(s) written consent has been collected and preserved by the author(s).

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