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Case Report

Hydranencephaly in a Male Neonate: A Case Report and Literature Review

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ABSTRACT

Hydranencephaly is a rare encephalopathy that occurs in-utero. The aetiology of this anomaly is built around several factors including infarction, leukomalacia, and diffuse hypoxic-ischemic brain necrosis, infection (toxoplasmosis, cytomegalovirus, herpes simplex infections (HSV) and thromboplastic material from a deceased co-twin. It is characterized by destruction of the cerebral hemispheres with transformation into a membranous sac containing cerebrospinal fluid and the remnants of cortex and white matter. The incidence worldwide is 0.2% in infant autopsies. Hydranencephaly is not compatible with a prolonged life after birth. The aim of this report is to alert clinician that antenatal finding of hydrocephalous may be strongly associated with fetal anencephaly, thus routine high resolution anomaly scan should be recommended for detection of such anomaly early in pregnancy.
We report this case of hydranencephaly in a male neonate delivered to a 25 year old divorcee primipara. The baby died within 4 weeks of admission. The father declined autopsy on religious grounds.

Key Words: Hydranencephaly, male, neonate, Sokoto

CASE REPORT

Baby AB, a male neonate was delivered to a 25 year old divorcee primipara. The mother was admitted at 36 weeks due to rapid abdominal growth and, had Abdomino-pelvic ultrasound done which revealed fetal hydrocephalus. She was subsequently delivered of a male neonate by elective caesarean section on account of fetal obstructive hydrocephalus. Apgar scores were 2 and 4 at one and five minutes respectively, the birth weight was 11. 12 kilogram, length was 46 centimeters and the ocipito-frontal circumference was 60centimeters. The baby had transfrontannelle ultrasound scan which confirmed a non-communicating hydrocephalus. There were variable amounts of residual cerebral tissue in the periphery of the tremendously expanded lateral ventricles. Magnetic resonance imaging (MRI) done on the baby also revealed; paucity of the cerebral parenchymal tissue, absence of the midline structures and deficiency of the falx anteriorly. The Baby was managed conservatively but, died within 4 weeks of admission. The father declined autopsy on religious grounds.

Literature review: Hydranencephaly; this relatively rare malformation of the brain is characterized by absence of the cerebral hemispheres and their replacement by sacs

filled with cerebrospinal fluid. It is one of the recognized forms of intracranial malformations associated with intrauterine fetal demise of one of twins in monochorionic twin gestation.[1-3]

Five aetiologies have been described of this condition; infarction.[4] leukomalacia,[5] diffuse hypoxic-ischemic brain necrosis,[6] infections (congenital toxoplasmosis, cytomegalovirus, and herpes simplex infections (HSV)[6] and thromboplastic material from a deceased co-twin [5] These theories have been generally grouped into two; the destructive theory in which the cortex is formed but destroyed in utero and the dysontogenesis in which there is early disruption of organogenesis.[3,7] However, the cause of the condition remains controversial, but since the meninges and cranium are intact, it must be assumed that the anlagen of the neopallium were present during the period of formation of these layers.[4]

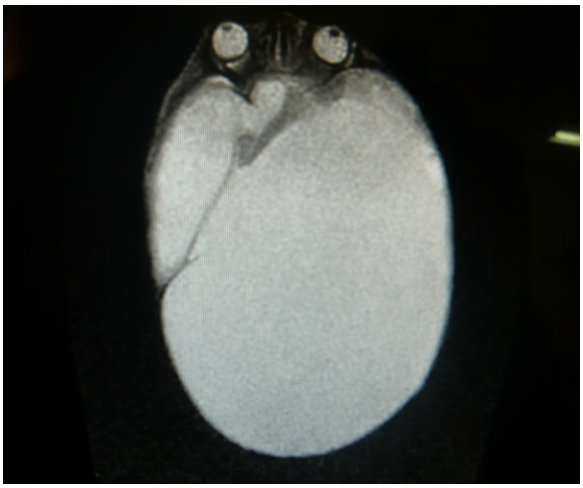

Fig: I. MRI of the infant showing huge hydrocephalus with minimal cortex.

Hydranencephaly can be bilateral or unilateral, in which case only one cerebral hemisphere is involved, leading to hemihydranencephaly.[8] The incidence of bilateral Hydranencephaly is 0.5 per 1000 births[3] and the incidence is lower as the maternal age advances.[9] Diagnosis of hydranencephaly can be made in utero using ultrasonography or magnetic resonance imaging[1] while, CT scan and MRI are important diagnostic tools postnatally.[10] Differential diagnoses of hydranencephaly include bilateral extra cerebral collection of fluid in the skull, severe open lip schizencephaly and extreme Hydrocephalus.[6] These can be differentiated from hydranencephaly by using an EEG as extreme Hydrocephalus will show evidence of cortical activity while hydranencephaly will not and, will give a flat isoelectric recording.[11] Surgery is considered unnecessary as majority of infants with hydranencephaly will die within the first year of life.[2,12] However, survival up to first decade of life has been reported in literature.[13] Children with hemihydranencephaly can lead a normal life however, complete hydranencephaly is not compatible with a prolonged life after birth, with the vast majority of live births dying prior to one year of age.[8] Termination of pregnancy is usually considered justifiable due to this reason.

CONCLUSION

We have reported this rare case of encephalopathy with the aim of alerting clinician that antenatal finding of hydrocephalous may be strongly associated with fetal anencephaly, thus routine high resolution anomaly scan should be recommended for early detection of this anomaly in pregnancy.

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