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Research Article

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ENCEPHALOCYSTOCELE – DIAGNOSTIC DILEMMA

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ABSTRACT

Background: Encephalocystocele is a congenital brain malformation characterized by brain herniation accompanied with extra-cranial protrusion of the ventricular system. This entity has often been overlooked and insufficiently tagged as encephalocele- wherein the management of both entities differ vastly, hence should be not misdiagnosed by radiologists. **Materals and Methods:** In our study we evaluated a case of polyhydramnios with gross fetal anomalies. **Results:** Detailed evaluation revealed herniation of the dilated ventricles into the cephalocele, on reviewing the literature encephalocystocele was diagnosed and findings were confirmed with the abortus. **Conclusion:** Encephalocystocele is a serious but correctable condition and requires extensive planning and major neonatal brain surgery only after detailed evaluation of the brain tissue by MRI.

KEYWORDS: Neural tube defects, antenatal diagnosis, ventricle herniation.

BACKGROUND

Encephalocele is a rare congenital anomaly where the Cerebral contents along with meninges, and parts of ventricular system herniated from the skull vault. Most encephaloceles originate between foramen magnum and lambda. Although the brain and its ventricular system are enlarged and may protrude out, hydrocephalus is not always seen. Occipital encephalocystoceles are commonly associated with Chiari II and Chiari III malformations. Rare associations include Dandy Walker malformation, cerebellar dysplasias, diastematomyelia and Klippel-Feil syndromes.

Gravid females with history of neural tube defects in the past pregnancies are at a higher risk and hence should be screened in detail for such anomalies.[1] During routine ultrasonography, encephalocystocele may be easily misinterpreted or misrecognized as meningocele or cystic hygroma as all these conditions are often associated with polyhydramnios like other neural tube defects. However careful evaluation of the skull and locating the defect solves this problem.

Yamada K et al first successfully documented encephalocystocele on MRI and confirmed the findings intra-operatively to demonstrate the encephalocele containing horns of lateral ventricle with associated corpus callosum agenesis.
In day to day practice B mode sonography reveals a solid cystic lesion arising through a defect in the occipital region with ventricle protruding into it outside the calvaria. It may contain dysplastic or disorganised brain tissue which is better evaluated on plain MRI images.[2]

MRI plays a role in evaluating intra-cranial brain tissue and helps in ruling out any underlying cortical abnormality like lissencephaly.

MATERIALS AND METHODS

A 32 year old primigravida with 26 weeks of gestation was referred to our department for routine second trimester obstetrics sonography. Clinically the fundal height was more than the gestational age so there was clinical suspicion for polyhydramnios. Patient had no previous scans. Patient was not antenatal care(ANC) registered. Patient was not on any drugs. Patient had spontaneous conception and had no history of IVF or assisted methods. There was no history of consanguineous marriage, or positive family history for genetic disease. After proper requisition, obstetrics sonography was performed using Mindray DC-7 machine with curvilinear transducer (3-5 Mhz).

On delivery the imaging findings were confimed with the abortus after informed consent from the parents.

RESULTS

On B mode sonography using a curvilinear probe -A large well defined solid cystic anechoic lesion measuring

9 x 5.7 cm was noted in the occipital region with no septations and was seen in direct continuity with a 2.5 cm sized defect in the calvarium.[Figure 1]

The ipsilateral ventricle appeared dilated and protruding into this herniated brain tissue indicating encephalocystocele. Rest of the brain parenchyma was normal. No other focal lesion was noted. Posterior fossa structures were not well visualised.[Figure 2].

No malformations of the spine, heart, kidney or other organs were found on sonography.

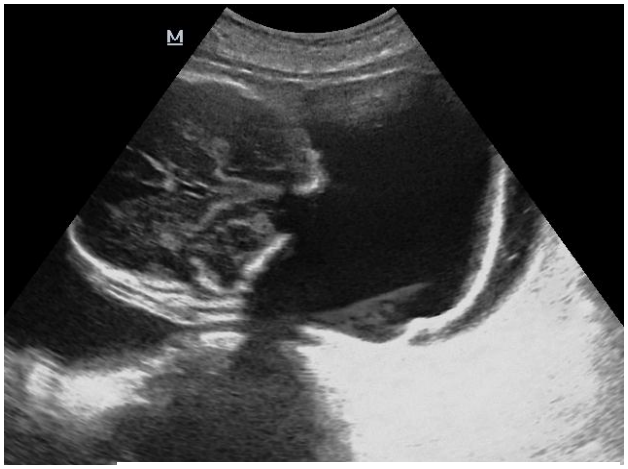

Figure 1: A large solid cystic anechoic outpouching in the occipital region.

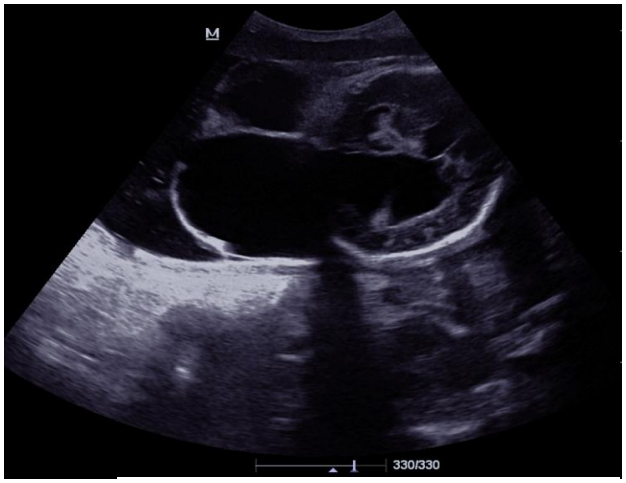

Figure 2: Dilated ipsilateral horn of ventricle which is contained within the out pouching with thinned our brain parenchyma.

CONCLUSION

Encephalocystocele is a serious but correctable condition and requires extensive planning and major neonatal brain surgery only after detailed evaluation of the brain tissue by sonography and MRI. Encephalocystocele does not necessitate termination of pregnancy but a good paediatric surgical setup should be arranged for post natal management.

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