



Sincipital Encephalocele, a Rare Anomaly Diagnosed Antenatally in the Late Pregnancy

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Abstract A case of sincipital encephalocele with intracranial cyst is presented which was diagnosed at 35 weeks gestational age in a patient who had undergone no ultrasound examination earlier. The purpose of this article is to make the radiologists and the obstetricians aware of this condition, prognosis, and complications.

Keywords Sincipital encephalocele · Ultrasonography · Fronto-nasal encephalocele

Introduction

Encephalocele, a type of neural tube defect is defined as herniation of the meninges and the brain parenchyma through a defect in the cranial vault. The various types of encephaloceles include occipital which is the most common (70–80%), followed by sincipital (15%) and basal (10%) [1]. Sincipital encephaloceles are anterior encephaloceles that involve the frontal, nasal, orbital and mid-facial regions.

Case Report

A 23 year old primigravida of approximately 35 weeks gestation was referred to the department for ultrasonography of the fetus. She had not undergone any sonographic examination prior to this. No biochemical screening had been done to exclude neural tube defects (NTD's).

The USG examination showed a viable 35 weeks intrauterine fetus as per the femur length and biparietal diameter. On examining the head, a cystic structure was noted in the right frontal region measuring about 2.62 cm, with normally visualized posterior fossa structures and thalami (Fig. 1). On further evaluation, a large defect was noted in the fronto-nasal region with herniation of a sac with no vascular supply, and frontal lobe parenchyma was seen herniating into it (Fig. 2). No other intracranial or extra-cranial abnormality was noted. The diagnosis of frontonasal sincipital encephalocele with intracranial cyst was documented. The female delivered the baby 2 weeks later by LSCS and the findings of the sonography were confirmed thereof (Fig. 3). The baby was referred to the neurosurgeon for further management.

Discussion

The various types of sincipital encephaloceles include: frontonasal, nasoethmoidal and naso-orbital (lateral). The present case belongs to the frontonasal sincipital encephalocele type. This classification is based on the bones forming the roof and floor of the entity. The herniation of the brain parenchyma through the foramen cecum and the fonticulus frontalis between the nasofrontal sutures into the glabella along the margins of nasal bridge with roof being formed by the frontal and floor by the nasal

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Fig. 1 Axial ultrasonographic image of the fetus at the level of thalami showing the intracranial cystic lesion in the right frontal region (calipers) and normal cerebellum and thalami (TH)

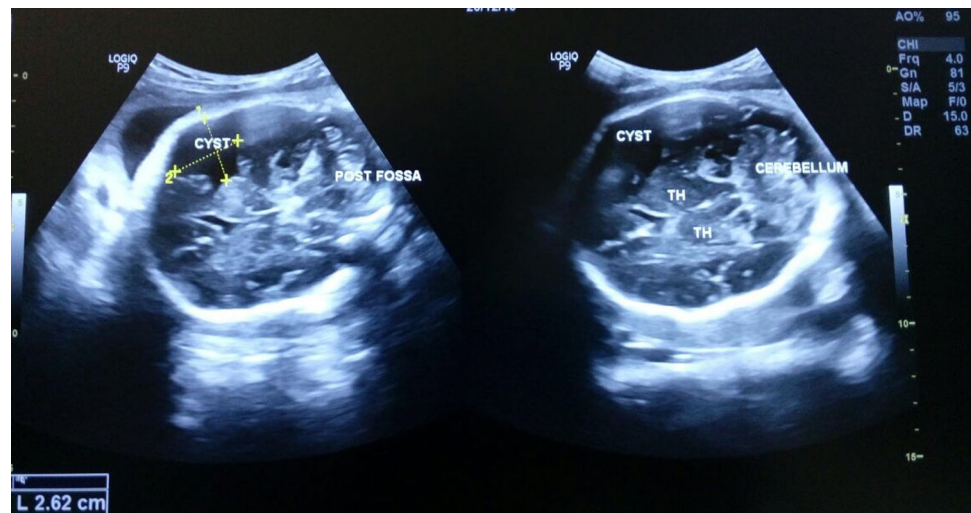
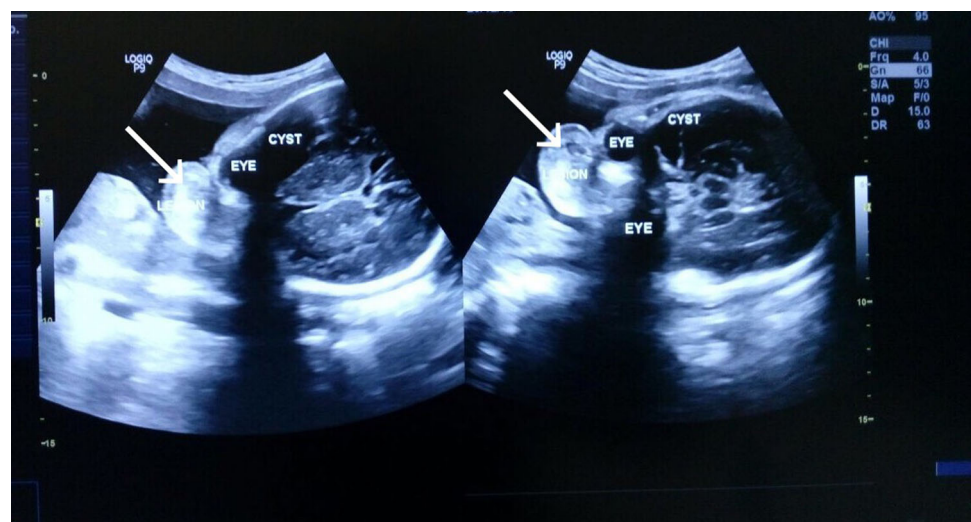


Fig. 2 Frontal region of the fetus with a large sac herniating through the fronto-nasal defect containing the right frontal brain parenchyma (arrows)



bones, defines the frontonasal type of sincipital encephalocele. The antenatal diagnosis of this entity is important by measuring maternal serum alpha fetoprotein and ultrasonography at 18–22 weeks of gestation. However, the maternal and amniotic fluid serum AFP may be normal if the lesions are completely epithelialized. It is usually associated with other anomalies of the central nervous system like intracranial cysts, partial callosal agenesis, facial clefts, interhemispheric lipomas and migration abnormalities. If this condition is encountered in early gestation, proper counseling of the patient is required regarding the prognosis and the mental deficit that may remain even if proper surgery is carried out. The future offspring have up to 6% risk of abnormality of the central nervous system [2]. Studies suggest that there is abnormal cognitive development in about 52% of patients having encephaloceles [3, 4].

The encephalocele form due to the non-separation of the neuroectoderm from the surface ectoderm resulting in a bony defect which facilitates the herniation of the meninges and brain parenchyma. However, the anterior encephaloceles are due to the failure of the mesodermal migration into the midline leading to the herniation of the brain matter and resulting failure of closure of anterior neuropore. The uniqueness of these encephaloceles is that they are post-neurulation defects unlike other types of encephaloceles. In many countries, obstetric protocols have been established to identify the major congenital anomalies in fetuses early in the pregnancy using biochemical as well as ultrasonographic studies. The most common methods used to screen for NTD's are serum and amniotic fluid AFP, amniotic fluid N-actylcholinestrerase, and USG examination in early gestation. The anomaly scan which is done at 18–22 weeks of the gestation forms the backbone of the diagnosis of various congenital anomalies. The



Fig. 3 Post-natal photograph of the fetus displaying the large right fronto-nasal encephalocele

present patient missed this important step and the anomaly went undetected till late pregnancy. The encephaloceles can be diagnosed in the second trimester, and also in the first trimester. Sonography helps in the proper delineation of the calvarial defects and the nature of the herniating contents. The fetal MRI is supreme to the USD in view of higher resolution and lack of shadowing by the calvarium and low amniotic fluid volume.

Occipital encephaloceles are associated with syndromes like Meckel–Gruber syndrome (Occipital encephalocele, enlarged kidneys and polydactyly), so evaluation of all the fetal organs is necessary to rule out any syndromic pathology.

If the size of the sac is large and encephalocele is bulky or there is association with lethal defects, termination of the pregnancy is the choice, due to significant morbidity and mortality. If the lesion is detected late in pregnancy as in the present case, the mode of delivery is also to be decided as in large lesions, vaginal delivery will be difficult so LSCS is preferred.

Post-delivery, surgery is the only option for the patient. Post natal evaluation includes MRI for proper delineation of the sac and contents and CT for the bony defect [5, 6]. The radiologist's role is to properly define the type of encephalocele; size, contents, extension and to rule out other abnormalities.

Conclusion

Sincipital encephaloceles are rare anomalies that belong to the important group of neural tube defects which may have association with many anomalies and have many deleterious long term effects on the fetal development and cognition. They should be identified early in gestation and the couple should be counseled regarding the prognosis. The only way to diagnose these and other anomalies early in gestation is by following the set protocols for fetal sonography i.e, biochemical screening in the first and second trimester and most importantly to emphasize the importance of second trimester anomaly scan at 18–22 weeks among the pregnant females visiting the obstetricians and ultrasonographers. Every pregnant women deserves a routine anomaly scan.

Compliance with Ethical Standards

Conflict of interest All the authors declare that they have no conflict of interest.

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