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CASE REPORT

Prenatal Diagnosis of Atretic Parietal Cephalocele

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ABSTRACT

Introduction: Cephalocele is the herniation of intracranial structures such as arachnoid, glial and central nervous system rests through a fetal skull defect. Although the estimated incidence of cephaloceles is 0.8-4 per 10.000 live births, this number may be underestimated due to stillbirths and elective pregnancy terminations.

Case Presentation: We present the case of a 34-year-old primigravida with an uneventful medical and family history, who attended our unit for the second trimester fetal ultrasound examination. The ultrasound scan showed a singleton live fetus with a gestational age of 23 weeks and normal growth parameters for the gestational age of the pregnancy. The sonographic evaluation of the fetal head revealed a posterior protruding sac-like structure, which appeared to originate from the right lambdoid suture. The mass measured 22.6 x 27 x 16 mm and did not appear to include brain tissue. MRI revealed the apparent elevation of the straight venous sinus, a pathognomonic feature of congenital atretic parietal cephaloceles.

Conclusion: Careful evaluation of the fetal head during the second trimester ultrasound is essential for the timely and accurate diagnosis of atretic cephaloceles. MRI is helpful to differentiate sculp lesions such as sinus pericranii, lipomas, teratomas, sarcomas and cephaloceles. Early prenatal detection of cephaloceles allows more time for delivery planning and parental counselling.

KEY WORDS Cephalocele; atretic parietal cephalocele; meningioma; congenital brain lesions; central nervous system abnormalities

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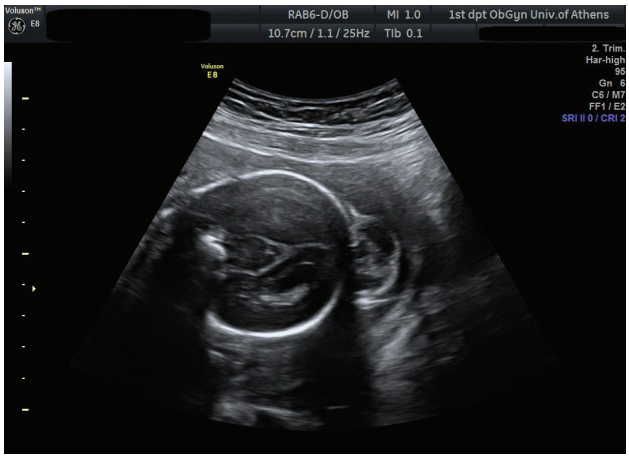


Figure 1

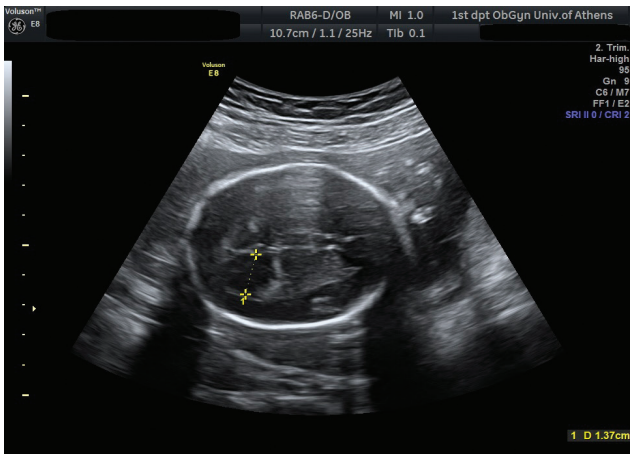
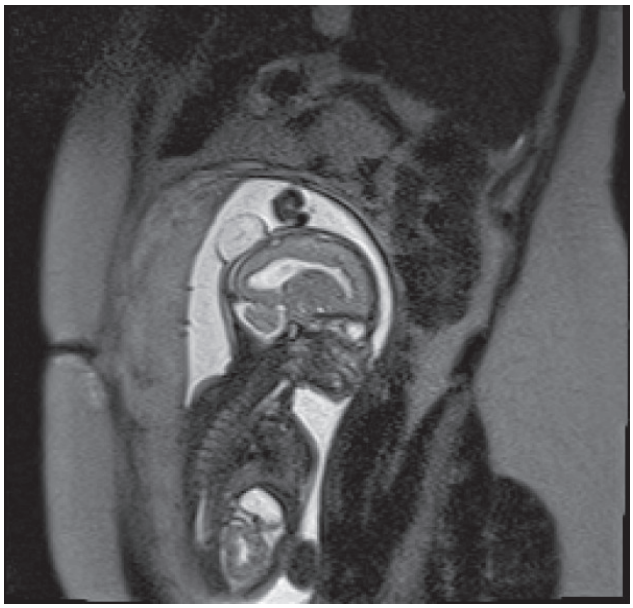


Figure 2

Figure 3



Introduction

Cephalocele is the herniation of intracranial structures through a fetal skull defect (1). The protruding mass consists of meningeal and vestigial tissues such as arachnoid, glial and central nervous system rests. When the lesion includes brain tissue the lesion is classified as encephalocele (1). Cephaloceles are categorized in Type I which consist mostly of arachnoid tissue and anomalous blood vessels and type II which have ectopic foci of neural and/or glial elements (2). Further classification separates cephaloceles in primary which are present at birth and secondary which are a result of surgery or trauma.

The incidence of cephaloceles is estimated at 0.8 - 4:10,000 live births (3). However, this number may be underestimated due to stillbirths and elective pregnancy termination. A female predisposition has also been reported in small case series (4). Atretic cephaloceles account for 4-17% of all cephaloceles and they occur more commonly near the lambdoid suture either parietally or occipitally. Parietal cephaloceles account for 37.5-50% (2,5).

Diagnosis of cephalocele is usually made postnatally, due to the detection of a scalp lesion or as part of congenital hydrocephalus evaluation. However, prenatal identification of cephaloceles has also been reported and it facilitates postnatal planning and treatment. We

report the case of an atretic parietal cephalocele that was identified during a routine antenatal visit in the second trimester of pregnancy.

Case description

A 34-year-old pregnant woman presented to the Obstetrics Ultrasound Department of Alexandra Maternity Hospital in Athens, Greece during the second trimester of her pregnancy. The woman was Gravida 1 Para 0 (G1P0). The gestational age of the pregnancy was 23 weeks and 0 days. The woman's past medical history was uneventful.

During her pregnancy, she was subjected to prenatal testing including a first trimester scan. Ultrasound examination revealed one fetus with normal growth parameters and amniotic fluid index for the gestational age of the pregnancy. Biparietal Diameter was 56.5mm, Head Circumference 197.2mm, Abdominal Circumference 186.6mm, Femur Length 43.6mm and the estimated fetal weight was 617gr. The sonographic evaluation of the fetal head revealed a posterior protruding sac-like structure (Figure 1). The mass appeared to originate from the right lambdoid suture and measured 22.6 x 27 x 16 mm. The lesion did not appear to include brain tissue. However, a small vascular structure was detected within the protruding mass. Further evaluation of the fetal head revealed ventriculomegaly, as the posterior horns of the lateral ventricles measured 13.7mm and 12mm respectively (Figure 2). The anomaly scan did not reveal any other congenital malformations. Considering the position of the lesion, a fetal brain Magnetic Resonance Imaging (MRI) scan was suggested. The MRI scan showed an apparent elevation of the straight venous sinus, the sagittal venous sinus and the cerebellar tentorium. The bone defect measured 5mm laterally to the right lambdoid suture (Figure 3a and 3b). Development of the cerebral cortex appeared pathological, as suggested by the presence of cerebral grooves which do not correspond to the normal brain development for the gestational age of the fetus. The subarachnoid space was diminished, and a small impression of the fetal cranium was noted. The posterior horns of the lateral ventricles appeared enlarged at 10.4mm and 10mm respectively. The cerebellar tonsils and the cerebellar vermis appeared normal. The corpus callosum also appeared normal. After careful evaluation of the ultrasound and MRI characteristics of the lesion, the diagnosis of cephalocele was suggested.

Discussion

There are many theories regarding the cause of atretic cephaloceles. A viable theory suggests that the origin of the cephalocele can be attributed to the persistence of neural crest remnants, while others have proposed the persistence of a fetal neural bleb to be the aetiologic factor of cephaloceles (5–7).

Abnormal presentation of the straight sinus, which is positioned vertically, is a common find in parietal cephaloceles (7). The straight sinus is positioned vertically during fetal cranial development until the third month of gestation when cerebral hemisphere expansion results in a more horizontal orientation (6). The embryonic positioning of the straight sinus could be a result of a fibrous strand connecting the tectum to the membranous cranium resulting in the interruption of the normal fetal cranial development (6).

Differential diagnosis of atretic cephaloceles includes sinus pericranii, lipomas, teratomas, sarcomas and other sculp lesions. In the majority of cases, the presence of a vertical straight sinus is sufficient to differentiate atretic cephaloceles from other lesions. Sinus pericranni can be differentiated by its relationship with the underlying Dural venous sinus (8).

The presence of atretic cephaloceles has been associated with a variety of other congenital anomalies. Occipital atretic cephaloceles have been associated with Meckel-Gruber syndrome and Walker-Warburg syndrome (5,9). Atretic parietal cephaloceles have also been associated with Dandy Walker Syndrome, Holoprosencephaly, Chiari type II malformations and corpus callosal agenesis (2).

Determination of the prognosis of patients with atretic cephalocele remains challenging given the rarity of the condition and the lack or relevant studies. It is generally accepted that the prognosis of infants with atretic cephaloceles varies depending highly on the presence or absence of other central nervous system abnormalities (10). Good prognosis has been reported for patients with no other central nervous system abnormalities (10). In any case, early prenatal detection allows more time for parental counselling and delivery planning. ■

Conflict of interest

The authors declare that they have no conflict of interest.

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