Fetal Scalp Cysts
Challenge in Diagnosis and Counseling

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The prenatal detection of a cystic lesion in the fetal occipital region could be alarming, because it may represent a meningocele or an encephalocele. We report a case of fetal scalp cyst detected on prenatal ultrasonography. This case is reported because the definitive antenatal diagnosis could be very difficult, but a correct diagnosis is important to avoid unnecessary intervention, in particular, termination of pregnancy.

Case Report

A 26-year-old gravida 1 woman was referred to our unit because of the finding of a cystic lesion over the fetal occipital region during a routine second-trimester ultrasonographic examination. There was no family history of congenital malformation.

The patient was seen at 20 weeks of gestation. A 7.4 × 4 × 5-mm cystic lesion was detected over the fetal occipital region. It was located in the midline immediately above the level of the cerebellum and was completely covered by the scalp (Fig. 1). No bone defect could be identified. There was no demonstrable herniation of intracerebral content into the cyst. No significant blood flow within or going into the lesion was observed on color Doppler sonography. The head size was normal, but a “lemon sign” deformity of the frontal bone was noted (Fig. 2). The intracerebral structures appeared normal, except that the lateral ventricular size was marginally increased to 0.94 cm. There was neither spina bifida nor other fetal structural abnormalities. The couple was counseled carefully about the possibility of a small meningocele or subcutaneous soft tissue cyst. It was emphasised that even if the lesion were an encephalocele, its size and location were
In general associated with a better prognosis than usual for this condition.

Subsequent ultrasonographic examination at 22 weeks of gestation revealed similar findings. The size of the cyst and the lateral ventricles remained the same, but the lemon sign had disappeared. Again, no bone defect or intracerebral abnormality could be identified. It appeared that the lesion was most likely of cutaneous or subcutaneous origin. The couple decided to continue with the pregnancy. The lesion was no longer visible during subsequent transabdominal scans at 26 and 32 weeks, most probably because of its small size and the position of the fetal head in the maternal pelvis. Transvaginal scanning was not performed, because the results of that procedure would not have altered treatment. No hydrocephalus or ventriculomegaly was present.

The neonate was delivered at 41 weeks of gestation by emergency cesarean section because of suspected fetal distress. The neonate weighed 2655 kg. A small, subcutaneous soft tissue mass that measured 1 cm was detected at the occipital region with fluctuation. No skull defect or intracranial lesion was demonstrated on postnatal ultrasonography or skull radiography. No surgical intervention was required. Magnetic resonance imaging was performed, which revealed a subcutaneous nodule at the occipital region. No intracranial extension of this lesion or any sinus tract could be demonstrated.

**Discussion**

When a cystic lesion is detected over the fetal occipital region during an antenatal ultrasonographic examination, encephalocele is the primary differential diagnosis. Other conditions, such as cystic hygroma, hemangioma, and cutaneous tumors, could have occipital cystic lesions, although these are much more rare.1–4

Encephalocele may appear as a pure cystic lesion (i.e., meningocele), which is particularly difficult to differentiate from other lesions. The correct diagnosis is important because the most common condition, i.e., encephalocele, may be associated with significant neurologic disability, whereas mistaking an epidermal cyst as encephalocele may lead to unnecessary interventions, which in most cases may result in termination of pregnancy.1

The correct diagnosis of an encephalocele is most definitive if a bone defect is identified or if herniation of cerebral tissue into the lesion can be demonstrated. However, identification of a bone defect may be difficult if the defect is small or if the fetal position does not afford an optimal view. On the other hand, care should be taken not to misinterpret suture lines as bone defects. Acoustic artifacts may also result in images resembling bone defects; therefore, multiple planes at different angles are necessary to confirm a genuine bone defect. Herniation of intracerebral contents is usually not obvious unless the defect is large.
When a smaller occipital cystic lesion without obvious bone defects or cerebral herniation is detected, the presence of other features, such as ventriculomegaly, distortion of the cerebellum, obliteration of the cistern magnum, microcephaly, and an abnormal head shape, may assist in the diagnosis of encephalocele. However, these signs are not specific and therefore should be interpreted with caution. For example, the lemon sign is not specific for meningomyelocele and has been detected in 1% of fetuses with no structural abnormalities. In our case, the borderline ventricular size and “lemon-shaped” head were retrospectively innocuous. Color Doppler ultrasonography may be useful in demonstrating the presence of abnormal vascular connections through the bone defect of an encephalocele or the vascular nature of a hemangioma. In doubtful cases, measurement of the maternal or amniotic fluid α-fetal protein level may assist in reaching a correct diagnosis of an open neural tube defect. However, a normal α-fetal protein level does not exclude the possibility of a small neural tube lesion or a lesion with intact overlying skin or scalp.

Occasionally, especially with a small lesion, it may not be possible to confidently exclude the possibility of encephalocele. Under these circumstances, one should be extremely cautious about prenatal counseling. Although encephaloceles may be associated with significant cerebral and functional abnormalities, occipital meningoceles, small lesions, the absence of brain tissue in the herniation sac, and the absence of hydrocephalus are good prognostic signs, because they are associated with either no or mild physical or functional disability. Date et al found that patients with encephaloceles as large as 5 cm could survive without neurologic deficits, although the mean size of such defects without neurologic deficits was 2.88 cm. Therefore, the involvement of a neurosurgeon or other specialists should be considered to provide a more precise estimation of prognosis in the process of counseling.

In summary, the detection of an occipital cystic lesion requires careful consideration of differential diagnoses other than encephalocele. This is particularly important if a bone defect cannot be identified with certainty and there is no evidence of associated intracerebral abnormalities. Correct recognition and diagnosis of the benign nature of some conditions, such as the one in this report, are important to avoid unnecessary termination of pregnancy.

References