History

20 year old female G1P0 referred at 19 weeks gestation for further evaluation of abnormal fetal findings on routine ultrasound.

Findings

Serial ultrasound examinations were performed at 19, 23, and 33 weeks. At 33 weeks, obstetric MRI was requested to help in management decisions. On the initial examination, there was asymmetric ventriculomegaly which became progressively worse especially on the left. The asymmetry in ventricular enlargement became more obvious with advancing gestational age. Because of this asymmetry, the possibility of cerebral infarction on the left was entertained.

MRI was performed demonstrating absence of the corpus callosum and a very large left sided arachnoid cyst. The right lateral ventricle can be easily separated from this cyst. The left lateral ventricle is compressed laterally. More posteriorly, the occipital horn and cerebral cortex are markedly attenuated as the cyst enlarges.

Diagnosis

Arachnoid cyst resulting in asymmetric ventriculomegaly

Discussion

Arachnoid cysts are developmental or acquired collections of CSF located within the layers of the arachnoid membrane. Primary arachnoid cysts are congenital and develop secondary to abnormal CSF flow, leading to development of a diverticulum. Secondary arachnoid cysts result from post-inflammatory accumulation of CSF in the subarachnoid space following trauma, hemorrhage, or infection. Arachnoid cysts account for 1% of intracranial lesions and are most often supratentorial with 50% located in the middle cranial fossa.

Most arachnoid cysts are asymptomatic. However, they can cause hydrocephalus, and arachnoid cysts are frequently associated with other intracranial anomalies, such as agenesis of the corpus callosum and increased risk of mental retardation. The most common presenting symptoms postnatally are seizure, headache, and focal neurologic signs. Size and location of the cyst and the gestational age at diagnosis are the most important prognostic factors.

Prenatal ultrasound detection of intracranial fluid collections is challenging. Arachnoid cysts must be differentiated from noninterhemispheric fluid collections, vascular malformations, and other interhemispheric fluid collections. Interhemispheric fluid collections can be divided into two groups: cystlike lesions related to physiologic median structures and pathologic cystlike lesions.

Lesions related to physiologic median structures include enlargement of the cavum septum pellucidum or cavum vergae and cysts of the velum interpositum. Pathologic cystlike lesions include arachnoid cysts, neuroepithelial anomalies, and cystic tumors. Larger cysts, cysts that are stable or increasing in size, and the presence of other intracranial abnormalities favor a pathologic interhemispheric cystic lesion.

References


Midsagittal MRI Image

There is absence of the corpus callosum. The arachnoid cyst (arrow) displaces the cerebellum inferiorly (arrowhead).
History

Twenty-one year old primigravida with no prior prenatal care. Serial ultrasound examinations show multiple abnormalities that suggest a diagnosis. Please give only one diagnosis as your answer.

Discussion

Trisomy 18 is the second most common autosomal trisomy among liveborn children. It occurs once in every 6,000 – 8,000 live births. The phenotypic syndrome was first described in 1960. It is characterized by multiple congenital malformations and severe psychomotor and growth retardation.

The best estimates to date are that no more than 5% of trisomy 18 conceptions survive through birth. There is a prenatal selection against males. At the time of amniocentesis the sex prevalence is nearly even, but females constitute 80% of live births. Ninety-five percent are full trisomy 18. Roughly 5% are mosaic with variable phenotypic expression. A small fraction represent a translocation trisomy. Over 90% of the time the trisomy is caused by maternal nondisjunction, and therefore is more common with advancing maternal age.

Many of the trisomy 18 malformations are detectable on prenatal ultrasonography. The most prevalent [90%] and characteristic finding is the distinctive hand posture - clenched with the index finger overriding the middle finger and the fifth digit overriding the ring finger. Another is the characteristic cranial form of a microcephalic elongated skull with a narrow bifrontal diameter and prominent occiput.

In second trimester ultrasound examinations, approximately 40% will demonstrate cardiac defects, a two vessel umbilical cord and choroid plexus cysts. Less frequent but also commonly seen are IUGR, omphalocele, renal malformations and poly/oligohydramnios.

When Trisomy 18 is suspected on the basis of ultrasound, the referring physician should be notified immediately so patients can receive referrals for genetic counseling and chromosomal analysis.

For patients with a prior full trisomy 18 pregnancy, there is a less than 1% risk of recurrence. However, when a parent is a balanced carrier, the risk of recurrence in subsequent pregnancies can be high.

Findings

Axial image through the fetal chest shows a four chamber heart with a large ventricular septal defect (arrow).

Axial image of the umbilical cord shows a single umbilical artery.

Axial image through the abdomen at the level of the cord insertion shows a markedly distended urinary bladder.

Axial image of the fetal head through the level of the lateral ventricles demonstrates large bilateral choroid plexus cysts (arrows).

References

25 year old female with positive UPT and vague abdominal pain.

**Findings**

Abdominal pregnancies are rare, accounting for 1 percent of ectopic pregnancies with an incidence ranging from 1:3,500 to 1:15,000. Abdominal pregnancies are typically associated with high maternal and perinatal mortality. Maternal mortality in the United States is estimated to be 7.7 times higher than that due to a tubal pregnancy and 90 times higher than that due to an intrauterine pregnancy. The perinatal mortality rate is estimated to be 40-95 percent. With the incidence of ectopic pregnancy rising in the United States, the ability to accurately diagnose this condition becomes increasingly important.

The diagnosis of abdominal pregnancy continues to be a challenge, delaying diagnosis and management decisions. Clinical suspicion must be high. However, most symptoms are nonspecific. The most common presenting symptom is mild, vague abdominal pain. Other symptoms include vaginal bleeding, cessation of fetal movement, and abnormal fetal heart sounds.

Abdominal pregnancy is divided into two categories: primary and secondary. In the primary type, the ovum implants directly on the peritoneal surface instead of traveling within the fallopian tube. The fallopian tubes and ovaries must be normal, and there can be no evidence of uteroplacental fistula. Primary sites of implantation include the pouch of Douglas, posterior uterine wall, uterine fundus, liver, spleen, lesser sac, and diaphragm. The secondary type accounts for the majority of abdominal pregnancies and occurs after rupture of a tubal gestation with subsequent reimplantation on the peritoneal surface. Alteration of tubal anatomy or function, such as tubal surgery, prior ectopic pregnancy, or PID, predispose to the secondary type.

Criteria for the prenatal diagnosis of abdominal pregnancy include demonstration of the fetus outside an empty uterus, location of the placenta outside of the uterine cavity, oligohydramnios, close approximation of the fetus to the maternal abdominal wall, abnormal fetal lie, poor visualization of the placenta, failure to see myometrium surrounding the fetus, and failure to establish continuity between the cervical canal and amniotic cavity.

Ultrasound is the primary diagnostic tool of obstetric imaging allowing for early and accurate diagnosis of ectopic pregnancy. However, the diagnosis of abdominal pregnancy is often missed. It is imperative that the uterus be identified in relation to the fetus outside an empty uterus, location of the placenta outside of the uterine cavity, and visualized in the same image. Therefore, with its ability to image in multiple planes, MRI plays a role in diagnosing late abdominal pregnancy. Other advantages of MRI include excellent soft tissue differentiation, no exposure to ionizing radiation, and the elimination of overlying bowel gas.

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**Discussion**

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References


**Axial MRI images**

The cross-sectional image of the uterus shows the bright signal of the endometrium (long arrow). The fetus (short arrow) and placenta (arrowheads) are to the left of the uterus.
History

33 year old G3 P2 referred for evaluation of mass in the fetal head.

Findings

Ultrasound images obtained at 34 weeks and at term demonstrate a septated cystic mass posterior to the cerebellum. The cerebellum and cisterna magna appeared intact. There was concern for a calvarial defect at the location of the mass. Thus, the possibility of an occipital cephalocele was entertained. MRI was performed to confirm the diagnosis. MRI images demonstrate a multi lobulated cystic mass measuring 3 cm by 1.3 cm. The mass is shown to arise from the soft tissues of the cranio-cervical junction. There was clearly no connection between the brain structures in the posterior fossa and the mass. No calvarial defect is noted on any of the images.

Discussion

Cystic hygromas are congenital malformations of the lymphatic system caused by failure of the embryonic lymphatics to connect with the venous system during the sixth week of gestation. The incidence of cystic hygroma is not truly known as many fetuses miscarry or are electively terminated, but the prevalence is estimated at 1-30 per 10,000 births. There appears to have been an apparent increase in the number of cystic hygromas in the past 25 years, which is thought to be due to increased use of prenatal ultrasound and improved imaging techniques.

Cystic hygromas are frequently associated with chromosomal abnormalities, classically Turner syndrome, but also trisomies 13, 18, and 21. Cystic hygromas have also been associated with Noonan syndrome, Roberts syndrome, teratogen exposure, and cardiac and genitourinary anomalies. The risk for cystic hygroma increases with increasing maternal age, which may be related to increased risk of aneuploidy.

The most common location is the neck (75%), with twice as many occurring on the left. Other sites include the axilla (20%) and less commonly the mediastinum, abdomen, and retroperitoneum. Cystic hygromas typically develop in the late first trimester or early second trimester. The typical ultrasound appearance is a mass of anechoic cysts of variable size. The differential diagnosis for cystic hygroma includes cephalocele, meningocele, hemangioma, and teratoma.

The prognosis for fetuses with cystic hygroma is poor when associated with hydrops fetalis, which occurs in up to 75% of cases. Large size and the presence of septations are also associated with poor prognosis.

References

History
Twenty-eight year old primigravida being evaluated for a known twin gestation.

Discussion
A Twin Reversed Arterial Perfusion (TRAP) sequence is one of the abnormalities seen only in multiple gestations which share a placenta. First described grossly in scientific literature in the 16th century, the reversed arterial circulation was postulated in the 19th century. The TRAP sequence occurs in 1 out of 35,000 pregnancies, and in approximately 1% of monochorionic twin gestations.

In this condition, the “acardiac” twin is without a functioning circulatory pump, either from acardius development or an acephalic state. This twin receives its blood supply from its normal or “pump” twin. The pump twin accomplishes this by delivering retrograde flow through the umbilical artery of the acardiac twin [Figure 1]. The reversed circulatory circuit is completed by blood flow exiting through the umbilical vein of the acardiac twin.

The primary risk to the pump twin is cardiac decompensation caused by the additional circulatory demand. Thus, it is not surprising that the size of the acardiac twin is a predictor of pump twin survival. If the acardiac twin is at least 75% the mass of the pump twin, the prevalence of mortality for the pump twin is almost 90%. Even if the acardiac twin is one half its mass, the pump twin has an almost two-thirds incidence of death.

The diagnosis is made by ultrasound visualization of the acardia and Doppler flow demonstration of the reversed circulation. Echocardiography is utilized to assess the cardiac status of the pump twin. Even if the mass of the acardiac twin is not significant, heart failure may still be present, and intervention indicated.

In addition to documenting the requisite monochorionic condition of the gestation in a TRAP sequence, ultrasound evaluation of the mono or diamniotic status is important for selecting treatment options. Furthermore, the reported incidence of chromosomal abnormality is as high as 1:10 in the pump twin. Therefore, evaluation for a genetic anomaly is imperative when weighing a decision to intervene on behalf of the pump twin.

Interventions have consisted of cord occlusion techniques, such as embolization or laser coagulation, and methods of intrafetal ablation such as radiofrequency. The end result of all of these measures is termination of the circulation to the acardiac twin. If the gestation is monoamniotic, cord transection may also be considered to reduce the risk of subsequent cord entanglement and compromise of the normal twin’s blood supply.

Findings
Axial images through the uterus show markedly discrepant appearances between twins. Fetus A on the maternal left shows no structural abnormalities. Fetus B on the maternal right is markedly hydropic with no definable internal structures. No cardiac structures were seen, consistent with the acardiac twin in a TRAP sequence.

Axial and longitudinal images obtained to the right of midline show the head (arrow) of Twin B and cystic masses around the cervical spine (arrowheads). No other recognizable fetal structures can be appreciated.

References
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Diagram of TRAP Sequence vascular flow.