A 25 year old non-gravid female presents with severe abdominal pain one day after having a Copper-T IUD placed. A two part diagnosis must be given.

**Diagnosis**

- **Aberrant location of intrauterine device and right ovarian mature teratoma**

**Findings**

On the day of IUD placement, no string was visualized after insertion. During the physical exam on the following day, probing of the cervical canal demonstrated no evidence of an IUD or string.

A transvaginal ultrasound of the pelvis was performed. It demonstrated a uterus measuring 7.1cm X 5.1cm X 5.8cm. No intraovarian device was identified in the endometrial cavity. Instead a linear echogenic structure corresponding to the lost IUD was seen in the right adnexa. In addition, an ill-defined mixed echogenic mass measuring 4.6cm X 4.2cm X 6.4cm was also visualized. The mass had a lobulated contour with both solid and cystic components, highly suggestive of a dermoid. The right ovary itself had normal echotexture with multiple follicles.

Immediate surgery was performed, and the operative findings confirmed what was suspected on the ultrasound. Uterine perforation was noted in the posterior wall.

The IUD was found mildly adherent to the right ovary by filmy parietal adhesions. The ovary itself was intact and there was no blood in the pelvis. A 5 cm cystic right ovarian mass was also found. Hair and sebaceous fluid were seen upon rupture of the mass. Pathology confirmed this to be a mature teratoma.

Two interesting but unrelated findings are seen in this case: uterine perforation of an intrauterine device and mature ovarian teratoma. Uterine perforation is one of the most serious but uncommon complications associated with an intrauterine device. Other complications include infection, ectopic pregnancy, abortion, premature delivery, and maternal death. Complaints of pain at the time of insertion, expulsion, or menstruation are also associated with sterility. Torsion, infection, rupture, and malignant transformation are all possible complications of teratomas.

- **Torsion**
- **Infection**
- **Rupture**
- **Malignant transformation**

Although often used interchangeably teratomas and dermoid tumors are actually different entities. Teratomas contain tissue arising from the ectoderm, mesoderm, and endoderm (all three germ layers). Yet, it is rarely documented that the sign is caused by a very echogenic anterior component mass with posterior shadowing which causes the more posterior aspect of the mass to be obscured. Often a fat-fluid level may also be seen. The fluid portion is usually in the dependent aspect while the fat portion appears above or below. The echogenicity may vary slightly depending on the components of the tumor. In 10% of the cases these tumors are bilateral.

**References**

Hypochondrogenesis.

canals most prominent within the vertebral bodies and femora. These hypercellularity, decreased cartilaginous matrix, and large vascular cartilage also seen (Fig 2i-k). Microscopic evaluation of the bones revealed chondrocyte pubis, vertebrae, and calvarium. Short limbs with metaphyseal widening were severely micromelia, markedly shortened limbs, and a distended abdomen (Fig 26 menstrual weeks and confirmed the presence of short limbs. Additionally there was increased vertebral spine ossification; however, the sacrum and coccyx remained unossified. Approximately one week following the second ultrasound the mother was induced with pitocin and she delivered the fetus that died shortly after birth. An autopsy was subsequently performed. Postmortem pictures demonstrated an enlarged cranium and narrow thorax.

Figure 2a. Coronal US.

Figure 2b. Sagittal US.

Figure 2c. Coronal US of the thoracic spine.

Figure 2d. Transverse US of the thoracic spine.

Microscopic evaluation of the bones revealed chondrocyte hypercellularity, decreased cartilaginous matrix, and large vascular cartilage canals most prominent within the vertebral bodies and femora. These findings are most consistent with a diagnosis of achondrogenesis type II or hypochondrogenesis.

The physical features common to both include a flat nasal bridge, flat face, small thorax, short trunk, distorted abdomen, and short extremities.1-6 The radiographic pattern of deficient ossification along with the characteristic shape of the bones of the extremities, vertebrae and pelvis is diagnostic. In achondrogenesis the vertebral bodies are frequently unossified, particularly caudally. The sacral, pubic, ischial, calcaneus and talus bones are either absent or extremely delayed in ossification. The femora are short and broad, and the acetabulae have flat roofs. Additionally, the tubular bones of the extremities are severely shortened with metaphyseal flaring. One feature that helps distinguish achondrogenesis from other skeletal dysplasias is that the long bones are not bowed.1,3

Microscopic evaluation of the bones demonstrates disorganized endochondral ossification with columnization of cartilage and lack of normal bony architecture. There is chondrocyte hypercellularity as well as increased vascularity.1,4 Type I achondrogenesis is the most severe form of the disorder and is subdivided into type 1A (Houston-Harris) and type 1B (Faccaro), both of which are autosomal recessive in inheritance. Features characteristic of type 1A achondrogenesis include rib fractures, partial ischial ossification, and a poorly ossified skull. Type 1B neonates do not have rib fractures and have more extensive calvarial ossification.1,5

Type II achondrogenesis (Langer-Saldino) is likely of autosomal recessive inheritance and is thought to be caused by mutations of the COL2A1 gene which codes for type II collagen. Type II achondrogenesis is less severe than type I and is also caused by a defect of the type II collagen gene. No clear radiographic differences are seen between achondrogenesis type II and hypochondrogenesis. Given the similarity of radiographic findings, these two entities likely represent a spectrum of the same disorder.5,6

Hypochondrogenesis is another described skeletal dysplasia with similar findings as achondrogenesis type II, but is less severe. Hypochondrogenesis is of autosomal recessive inheritance and is also caused by a defect of the type II collagen gene. No clear radiographic differences are seen between achondrogenesis type II and hypochondrogenesis. Given the similarity of radiographic findings, these two entities likely represent a spectrum of the same disorder.5,6

Thanatophoric dwarfism is another lethal skeletal dysplasia, and is often confused with achondrogenesis. In contrast to achondrogenesis, the radiographic features of thanatophoric dwarfism include ossified vertebral bodies, sacrum, pubic and ischial bones, markedly flattened vertebral bodies with narrowed midpoints, short pubic bones without delayed ossification, and short bowed tubular bones with flaring of the metaphyses.3,5

References


20 year old female (G4P2) presents to the Emergency Room with bleeding, pelvic pain, and a positive urine pregnancy test.

An initial transabdominal ultrasound revealed a single intrauterine pregnancy with a crown rump length of 38.1 mm corresponding to a ten week 5 day gestation. The placenta demonstrates normal echogenicity and appearance. However, to the left of the gestational sac, the uterus shows multiple small cystic spaces within what was thought to be a mass. The ultrasound was interpreted as single intrauterine pregnancy with probable coexistent molar pregnancy. Clinical correlation with BHCG was recommended. A follow-up ultrasound was performed 6 days after the initial examination and shows similar findings.

An MRI of the pelvis was obtained for further evaluation. This demonstrated a single intrauterine pregnancy with no discrete mass to correlate with ultrasound findings. The myometrium was thickened and displayed enlarged vessels bilaterally. A follow-up ultrasound was performed a month later and shows normal vascularity.

Discussion

Small cystic spaces in the wall of the pregnant uterus can produce a confusing picture. Hydatidiform mole with coexistent fetus, degenerating fibroids, cystic degeneration of the placenta, and abruptio placentae are all differential diagnoses that are considered when these changes in the uterine wall are seen. Of these, hydatidiform mole with coexistent fetus is the least common, with degenerating fibroids being the most common.

It is crucial to distinguish whether the cystic spaces are within the uterine wall, the amniotic cavity, or placenta. This determination along with BHCG correlation, is helpful in differentiating this normal condition from the other differential diagnoses, particularly the hydatidiform mole. Hepatic increases in serum B HCG concentrations parallel the proliferation of trophoblasts. Abnormally rapid increases in this hormone indicate the presence of a hydatidiform mole. However, the presence of a normal pregnancy is against the diagnosis of a molar pregnancy.

Hypervascularity of the normal pregnant uterus

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Follow-up US approximately one week later continues to demonstrate a hypervascular uterus.

It is important to diagnose this entity because profuse bleeding may occur during invasive procedures in a hypervascular uterus. It is therefore imperative, that these ultrasound findings be communicated to the obstetricians prior to Cesarean section, amniocentesis or fetoscopy.

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A 19-year-old pregnant female with history of elevated maternal serum alpha-fetoprotein levels presented at approximately 28 menstrual weeks for a prenatal ultrasound.

Diagnosis

Pulmonary sequestration is a rare congenital abnormality characterized by nonfunctioning pulmonary parenchyma with no pulmonary connection to the tracheobronchial tree, which receives its blood supply from a systemic artery. Pulmonary sequestration likely results from early embryological malformation of the primitive foregut. Sequestrations are divided into two types: intralobar, in which the sequestration is located within the parenchyma of the normal lung and does not have a separate visceral pleura, and extralobar, in which the sequestration is surrounded by its own visceral pleura. Both types are composed of normal lung elements, which are arranged in a disorderly manner.

Extralobar sequestration accounts for 25% of pulmonary sequestration cases and has a 4:1 male to female predominance. Most often presents within the first year of life with symptoms of respiratory distress, cyanosis, and feeding difficulties. Extralobar sequestration is found on the left side in 90% of cases and is usually related to the left hemidiaphragm. It may be located between the lower lobe and the diaphragm, within the diaphragm, in the mediastinum, as well as in the peritoneal or pericardial cavity. The arterial blood supply in extralobar sequestration arises from the thoracic or abdominal aorta in approximately 80% of cases; however, other sources include subclavian, phrenic, gastric, intercostal, internal mammary, or pulmonary arteries. The venous drainage is usually via the inferior vena cava, axillary or hemiazygos veins, or the portal vein. Less common venous drainage includes the intercostal, esophageal, subclavian, and adrenal veins. The systemic drainage causes a left-to-right shunt. Congenital anomalies are associated with extralobar sequestration in approximately 50% of cases. Ipsilateral congenital diaphragmatic hernia is the most common associated defect occurring in 30% of cases. Other less commonly associated anomalies include pectus excavatum, congenital heart defects, pericardial cyst, vertebral malformations, esophageal communication, and colonic duplication.

Intralobar sequestration accounts for 75% of pulmonary sequestrations. Most patients are asymptomatic until recurrent localized pneumonia develops which frequently occurs in late adolescence. In approximately two-thirds of cases, the sequestration is located in the left lower lobe near the posterobasal segment. The sequestrated tissue typically derives its arterial supply from the descending aorta and the venous drainage via the pulmonary venous system. Intralobar sequestration is uncommonly associated with congenital anomalies.

Pulmonary sequestrations can be evaluated using several different imaging studies. It is often difficult to distinguish between intralobar and extralobar sequestration using plain radiographs since both typically appear as a homogeneously dense mass or cystic structure in the lower lobe. Ultrasonography can be a useful imaging modality, particularly if the sequestration is subdiaphragmatic or near the chest wall. The definitive diagnosis can be made by using angiography to demonstrate both the arterial supply to the sequestration as well as the venous system. CT and MRI are both helpful in diagnosing pulmonary sequestration; however, both are less reliable than angiography in identifying the vascular supply. The usual treatment for both types of sequestrations is surgical excision. Surgery for intralobar sequestration requires lobectomy since the sequestration cannot be separated from normal lung tissue.
An 18 year old primigravida presents for obstetric ultrasound because of uncertain dates.

**History**

The fetus was in cephalic presentation at the time of examination. Fetal biometry revealed an average sonographic age of 34 weeks. It was noted during examination that the fetal organs in the chest and abdomen were reversed in position. The orientation of the cervix with the fetus in cephalic presentation. Fetal biometry demonstrated reversal in position of the stomach and liver.

**Diagnosis**

Situs inversus totalis

**Findings**

The rapidity of detection is largely determined by the presence or absence of other associated anomalies. Transposition of the thoracic or abdominal viscera can be complete or partial. In complete situs, known as situs inversus totalis, transposition of both the thoracic and abdominal viscera occur. Both the thoracic and abdominal viscera are in a reverse position like a mirror image. On the other hand, situs inversus partialis may be confined to the thoracic viscera involving dextrocardia only without abdominal visceral involvement. Transposition of the abdominal viscera without other abnormality has also been reported.

**Discussion**

Situs inversus, also known as transposition of the viscera, is a diagnosis that often remains undetected until adulthood because it is usually asymptomatic. The rapidity of detection is largely determined by the presence or absence of other associated anomalies. Transposition of the thoracic or abdominal viscera can be complete or partial. In complete situs, known as situs inversus totalis, transposition of both the thoracic and abdominal viscera occur. Both the thoracic and abdominal viscera are in a reverse position like a mirror image. On the other hand, situs inversus partialis may be confined to the thoracic viscera involving dextrocardia only without abdominal visceral involvement. Transposition of the abdominal viscera without other abnormality has also been reported. The disease is thought to be genetically linked through an autosomal recessive gene.

One in ten thousand patients have situs inversus with dextrocardia. These patients are usually asymptomatic. However, situs inversus partialis is usually associated with more morbidity especially when the abdominal viscera is involved with a normally positioned heart. Situs inversus partialis is almost always associated with severe congenital heart disease. Renal anomalies are also more common with this form of situs. Situs inversus totalis can, however, be associated with other anomalies as well. Kartagener's syndrome is an example. This syndrome includes the triad of bronchectasis, sinusitis and or nasal polyps, and situs inversus totalis. Anomalies of the spleen, skeleton, pulmonary, and genitourinary system have also been reported.

A concrete grasp of the fetal orientation and position is crucial to making the diagnosis of situs inversus by ultrasound. A fetus with normal abdominal situs, when imaged in the cephalic position, should have its spine, stomach, and umbilical vein imaged in a clockwise manner on a transverse image. Counterclockwise imaging of these structures should occur in the breech position. This can become quite a challenge to determine when the fetus is not well imaged or is in an indeterminate or transverse lie. In these difficult cases determining where the cardiac apex is in relation to the stomach on transverse view may be helpful. Angling the transducer toward the fetal caudal structures should show the stomach and the cardiac apex on the same side. This at least excludes situs inversus partialis which is the more worrisome of the two disease processes. However, the diagnosis of situs inversus totalis would not be excluded by this method.

The cardiac and other morbidities associated with situs inversus partialis is why it is so important to make this diagnosis as early as possible. Prenatal diagnosis allows immediate postnatal management which facilitates early treatment. This in turn decreases the morbidity and even mortality that can be associated with this anomaly.

**References**