

Fetus in Fetu

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Fetus in fetu is a congenital abnormality in which a nonviable, parasitic fetus grows within its twin. It is a rare cause of retroperitoneal abdominal mass in infants and children. The authors report a recent case of a six-month-old girl who presented with unexplained abdominal distention. A sonogram and a magnetic resonance imaging examination showed a multiloculated, complex cystic mass with calcified and soft tissue components. A definitive diagnosis and discrimination from a teratoma was difficult to make because of the absence of a distinctive criterion, the presence of a vertebral column. Pathologic examination showed a complex mass consisting of well-formed bowel and upper respiratory tract segments as well as mature neuroglial tissue, skeletal muscle tissue fibers, and bone tissue that contained bone marrow, supporting the diagnosis of fetus in fetu. Therefore, the nonvisualization of a vertebral column on imaging should not exclude fetus in fetu from the differential diagnosis.

Key words: fetus in fetu, teratoma, sonography, vertebral column, monozygotic diamniotic twinning

Fetus in fetu is an extremely rare developmental abnormality where mature tissue from all three germinal layers is enveloped inside a viable embryo. There is some controversy as to what this abnormality represents: a mature teratoma or an accident of monozygotic, diamniotic twinning. It is usually discovered in a newborn or infant but has been described in all age groups. Seventy-five percent have been documented to occur in the abdomen or retroperitoneum.¹ Although criteria for the diagnosis of fetus in fetu have been established by

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Willis,² the diagnosis of this entity has proven to be difficult and often depends on the pathological examination.³

Case Report

A six-month-old girl presented to our affiliated institution with symptoms of increasing abdominal girth. Except for the unexplained increase in abdominal size, her medical history and laboratory findings were unremarkable. A sonographic examination of the abdomen was ordered. The sonogram was performed using an ATL 5000 sonographic instrument (Bothel, Washington), with an 8-MHz curved-array transducer. The examination revealed a large, multiloculated, complex cystic mass with calcified and soft tissue components directly inferior to the liver and filling most of the abdomen (Figures 1 and 2). The mass mildly displaced the abdominal organs superiorly and was immediately adjacent to the fundus of the uterus (Figure 3). The mass reached both sides of the abdomen and did not appear to arise from either kidney (Figures 2 and 4). The precise origin could not be determined, but it was suspected to be retroperitoneal. The combination of these findings led to the suspicion of fetus in fetu and cystic teratoma. Correlation with magnetic resonance imaging (MRI) was recommended.

The MRI was performed seven days later, and the findings correlated well with the sonogram. The MRI revealed a very large mass measuring approximately 11 cm craniocaudally, 8 cm anteroposteriorly, and 15 cm transversely (Figures 5 and 6). The mass was found to have a large cystic component with septations that contained a large fatty component (Figure 7). In addition, the mass was found to have no interaction with the sacral area and contained echogenic foci that were suspected to represent calcifications. Because of the complexity and diagnostic variability of the mass, exploratory laparotomy was performed after the parent's consent was obtained. Surgical excision of the mass was performed, and the infant's recovery was uneventful.

Pathological examination revealed a 16 × 12 × 5-cm complex mass with cystic and solid components. The cystic components were lined with

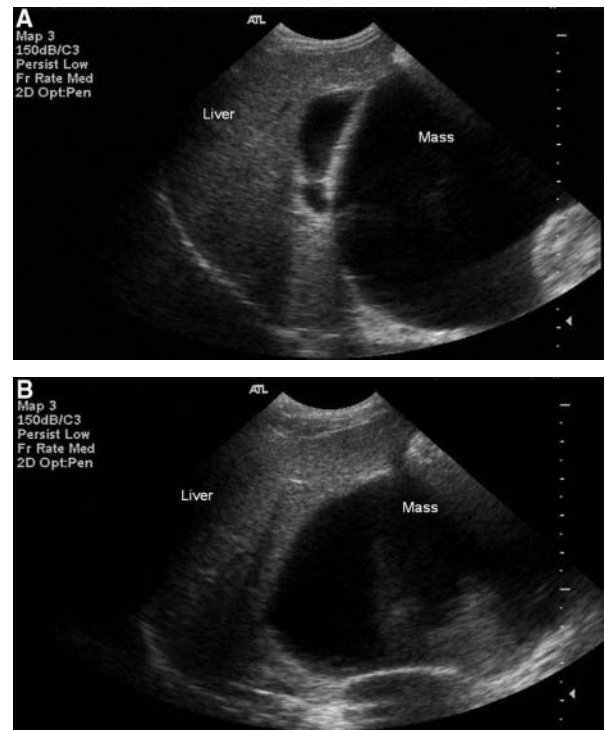


FIGURE 1. (A) A sagittal image of the right upper quadrant demonstrating the relationship of the mass to the liver. The mass contained both cystic and solid components. (B) A transverse image of the right upper quadrant demonstrating the relationship of the mass to the liver. The mass contained both cystic and solid components.



FIGURE 2. A sagittal image of the right upper quadrant showing the relationship of the mass to the liver and right kidney. Calcified elements producing acoustic shadowing were seen (white arrow).

respiratory-type epithelium, squamous epithelium, or mucous epithelium. Underneath the epithelial layer, structures such as hair follicles and sebaceous



FIGURE 3. A sagittal image of the pelvis showing the mass adjacent to the fundus of the uterus and the dome of the urinary bladder.

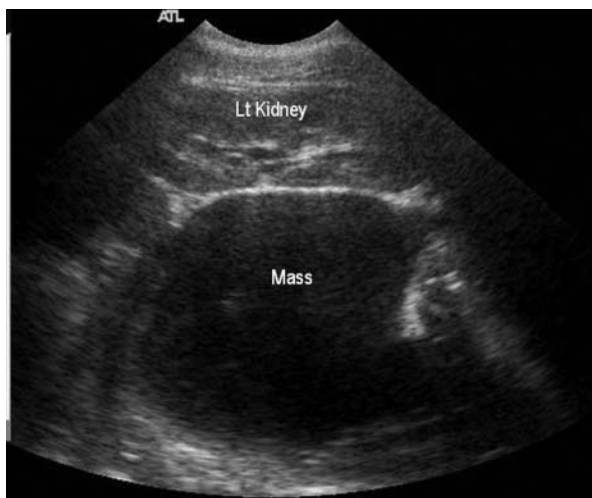


FIGURE 4. A sagittal image of the left lower quadrant demonstrating the complex mass abutting the left kidney and not arising from it.

and sweat glands were found. The fluid in the cystic areas was described as slightly gelatinous and clear. The solid area contained two tubular structures measuring approximately 3 cm in diameter. One was a section of well-formed bowel containing ectopic gastric mucosa. The other resembled a segment of an upper respiratory tract complete with a cartilage rim. The rest of the solid area contained mature neuroglial tissue, fibers of skeletal muscle tissue, cartilage, and bone tissue that contained bone marrow. There were no immature cells

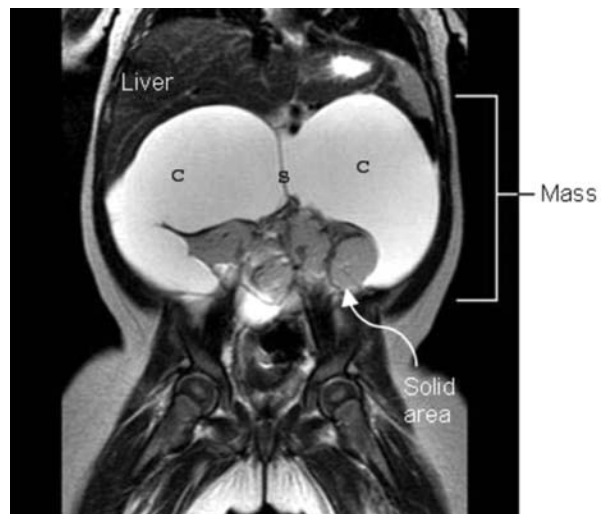


FIGURE 5. A coronal T2-weighted image of the abdomen showing the complexity of the mass. The mass had a cystic component (c), a septation (s), and a solid component (white arrow).

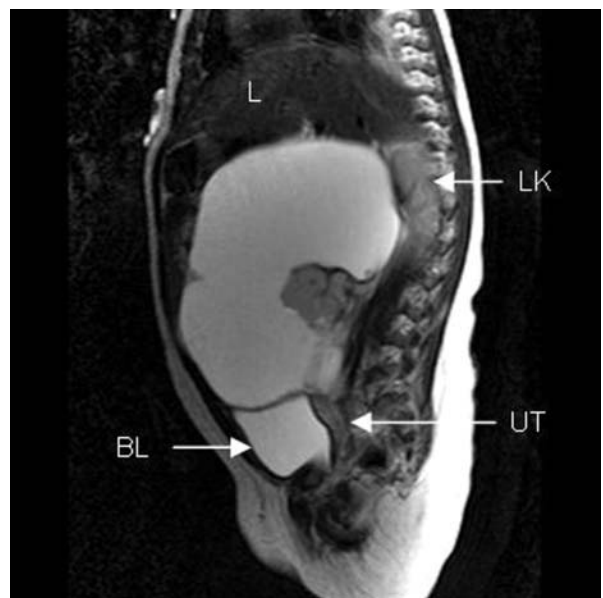


FIGURE 6. A sagittal T2-weighted image showing the relationship of the mass to the liver (L), left kidney (LK), urinary bladder (BL), and uterus (UT).

seen, and no evidence of malignant transformation was found. The pathologic findings were consistent with fetus in fetu.

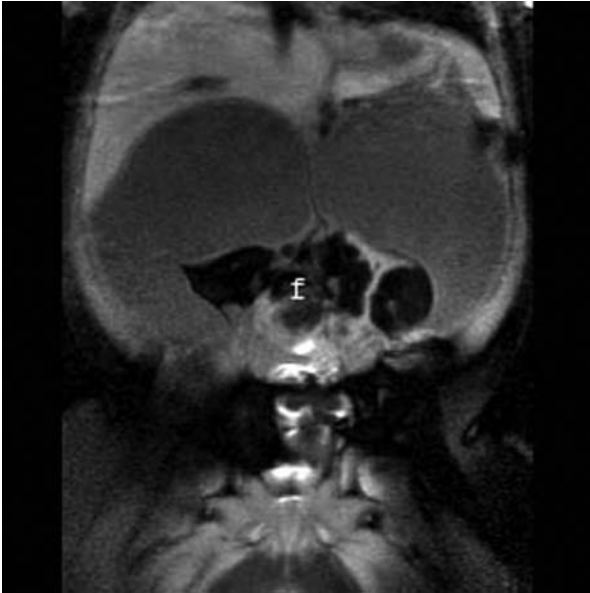


FIGURE 7. A T1-weighted image with fat suppression showed that the solid area is mostly composed of fatty tissue (f).

Discussion

Fetus in fetu is an extremely rare abnormality that occurs secondary to a developmental aberration of monozygotic diamniotic twinning. Fewer than 100 cases have been reported in the literature.^{4,5} Its incidence is reported to be 1 in 500,000 births.^{1,6,7} It affects males twice as often as females, and most patients present with symptoms during the first year of life.⁸ Fetus in fetu has also been documented in different age groups.⁶ Seventy percent of cases present with an abdominal mass as the chief complaint.⁹ Symptoms are usually secondary to compression of the mass on internal structures. In 80% of the cases, the mass is located in the retroperitoneal cavity.¹⁰ Other locations such as the skull, sacrum, scrotum, mouth, and neck have been reported.^{11–15}

The exact nature or embryogenesis of this abnormality is controversial. Some researchers consider fetus in fetu to represent one end of a teratoma continuum that ranges from immature to mature.^{16,17} Others suggest that fetus in fetu represents a monozygotic diamniotic twinning abnormality where one twin in a monochorionic, diamniotic pregnancy becomes enclosed inside its

karyotypically identical co-twin.⁷ The distinction between a teratoma and a twinning abnormality is often difficult.⁸ Despite the difficulty in distinguishing between a teratoma and fetus in fetu, Willis² reported that clinical and pathological criteria can be integral in making the diagnosis. He stated that teratomas do not have the ability to develop through the stage of primitive streak, which provides the fetus with its vertebrate pattern. Furthermore, teratomas are located extraperitoneally¹⁸ and possess a malignant potential.¹⁹ On the other hand, Willis believes that any highly differentiated teratoma that contains a vertebral axis around which were organs or limbs constitutes a fetus in fetu.^{20,21} According to Willis,²¹ the presence of a vertebral column can be considered the best finding in distinguishing between a teratoma and a fetus in fetu.

The use of imaging modalities such as sonography, computed tomography, and MRI has enhanced the preoperative diagnosis of fetus in fetu.^{22,23} Although diagnostic imaging has improved the diagnosis of fetus in fetu, a definitive diagnosis can be uncertain in those cases where the vertebral column is dysplastic and underdeveloped. In reviewing the literature up to the mid-1990s, Hoeffel et al⁶ found that 16.7% of the cases of fetus in fetu were diagnosed preoperatively. In addition, it was indicated that the differential diagnoses were teratoma and meconium pseudocyst.²⁴ More recently and according to Spencer,²⁵ fetus in fetu is usually encapsulated and enclosed in a well-circumscribed sac without major attachments to the host via large blood vessels.

The treatment of choice for fetus in fetu is surgical. The surgical resection of the mass is necessary because fetus in fetu tends to grow proportionally with the host, eventually causing abdominal distention, pain, jaundice, feeding difficulty, and compression and disruption of organ systems.²⁶

Conclusion

The discrimination and differentiation of fetus in fetu from a teratoma is often difficult. The diagnosis of fetus in fetu is straightforward on imaging when a vertebral column is present. But the nonvisualization of a vertebral column on imaging, especially sonography, should not exclude the diagnosis of fetus in

fetu. In our case, the suspicion of fetus in fetu was made preoperatively and confirmed by pathology. Therefore, in the absence of a vertebral column and the presence of a well-circumscribed, fluid-filled sac enclosing a solid mass with calcifications in sonography, fetus in fetu should be suspected.

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