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Fetus in Fetu: CT Appearance— Report of Two Cases¹

Fetus in fetu is a rare abnormality secondary to the abnormal embryogenesis in a diamniotic, monochorionic pregnancy. It is an unusual condition in which a vertebrate fetus is enclosed within the abdomen of a normally developing fetus. Presented are the computed tomographic findings in two cases in which imaging findings were diagnostic of this entity.

To the best of our knowledge, fetus in fetu originally was described by Meckel in the late 18th century (1). It is a rare pathologic condition, with 79 cases reported in the literature (2).

The exact embryogenesis of fetus in fetu is controversial. Some investigators propose that fetus in fetu occurs from the anomalous embryogenesis in a diamniotic monochorionic twin pregnancy in which a malformed monozygotic twin lies within the body of its fellow twin (1). Others consider it to represent a highly organized teratoma.

This condition has a 2:1 male predominance, with most patients presenting with an abdominal mass in the 1st year of life. Other reported locations include the cranial cavity and the scrotum (3,4). The number of fetuses in fetu is usually single; however, multiple fetuses in fetu have been reported (5). The intent of this case report is to describe the characteristic computed tomographic (CT) findings of this unusual entity.

I Case Reports

Case 1

A 16-year-old male patient presented with a several-year history of a slowly growing mass in the upper abdomen and periumbilical region. At physical examination, the mass was firm, smooth, and nontender. Laboratory test findings were

unremarkable. A conventional radiograph of the abdomen showed a large, partially calcified soft-tissue mass in the upper abdomen. Ultrasonography revealed a large, hyperechoic, inhomogeneous intra-abdominal mass that appeared to contain areas of calcification. Some of these calcifications on the longitudinal scans were suggestive of bones.

CT scans of the abdomen and pelvis obtained before and after the administration of contrast material revealed a large heterogeneous mass in the upper abdomen. The mass predominantly contained areas of fat that surrounded a central bony structure (Fig 1). The bony components of the mass resembled the remnants of a vertebral column.

Surgical exploration revealed an encapsulated, large retroperitoneal mass that was adherent to the mesenteric root. There was an anomalous blood supply to the mass that arose directly from the abdominal aorta. The mass was resected en bloc.

Gross examination revealed the mass to be an anencephalic fetus in fetu that weighed approximately 2 kg and was 30 cm in length. Both extremities were well developed, and the entire skin was covered with hair. The gluteal cleft and genital tubercle were identified. No anal dimple was found. Chromosomal analysis was not performed.

Case 2

A 3-year-old girl presented with a slightly painful mass of 2 weeks duration in the left upper abdomen. At physical examination, a smooth, tender left flank mass was present. The remainder of the physical examination was unremarkable.

A conventional radiograph of the abdomen revealed a mass that contained numerous calcifications that resembled bony structures. A CT scan showed a large heterogeneous mass in the retroperitoneum that displaced the left kidney posteriorly. The mass consisted predominantly of fat that contained multiple bony structures. The bony components resembled vertebral bodies and long bones (Fig 2).



Figure 1. Case 1. Transverse CT scan of the upper abdomen reveals a large retroperitoneal soft-tissue mass (arrowheads) that displaces the stomach and the liver anteriorly. The mass consists of fat (solid arrows) that surrounds a central bony structure that resembles the vertebral column (open arrows).

Surgical exploration revealed a large retroperitoneal mass between the left kidney and the spleen. The mass was enclosed in a purplish-pink capsule that ruptured during its removal. No major vascular connections were identified.

Gross examination showed an anencephalic fetus in fetu that weighed approximately ½ kg and had skin that covered its entire body. The lower extremities were formed, with flipperlike feet. The trunk was formed incompletely, and an immature external genital ridge was present. No anal dimple, scrotal sac, or testicles were seen. No glial tissue or other evidence of organogenesis was seen. Chromosomal analysis was not performed.

Discussion

Fetus in fetu is thought to result from the unequal division of the totipotent inner cell mass of the developing blastocyst. This results in the inclusion of a small cell mass within a maturing sister embryo. The result is an included vestigial remnant of a diamniotic monochorionic twin that is located within the body of the otherwise normally developed twin (6).

There is controversy as to whether fetus in fetu is a distinct entity or represents a highly organized teratoma. A teratoma is defined as a neoplasm with a slight potential for malignancy that is composed of multiple tissues foreign to the part in which they are located (7). It often is difficult to make a distinction between teratomas and vestigial remnants that result from abortive attempts at twinning. As a result, some authors think that fetus in fetu is within the spectrum of abnormalities that can result from the

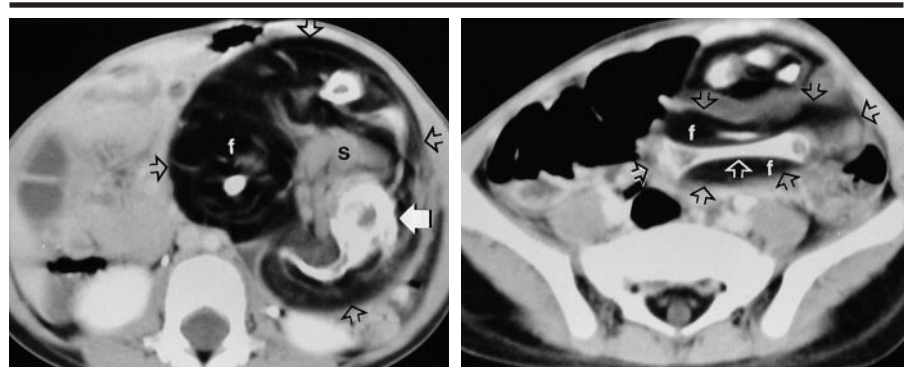


Figure 2. Case 2. Fetus in fetu. (a) Transverse CT scan of the upper abdomen reveals a heterogeneous mass (open arrows) that has fat (*f*), soft tissue (*s*), and bony components. The bony components (solid arrow) resemble a vertebral body in cross-section. (b) Transverse CT scan at the level of the pelvis demonstrates a soft-tissue mass (black arrows) that consists of an ossific component that resembles a femur (white arrow) surrounded by fat (*f*).

anomalous embryogenesis in a diamniotic monochorionic pregnancy. The spectrum includes conjoined symmetric twins; parasitic fetuses; embryonic vestigial fetal inclusions; and organized teratoma (7). Thus, some authors claim that fetus in fetu is only a well-differentiated highly organized teratoma (7).

However, many other investigators suggest that fetus in fetu is a pathologic entity that is distinct from teratoma for several reasons (8). Malignant degeneration associated with fetus in fetu is extremely rare, with only one reported case to our knowledge (8). Two-year follow-up has shown no evidence of recurrence in either of our two patients. Fetus in fetu occurs most commonly in the upper retroperitoneum, while teratoma usually occurs in the lower abdomen, ovaries, or sacrococcygeal region (9). Both of our patients' masses were located in the retroperitoneum.

A final important feature that has been used to distinguish between fetus in fetu and teratoma is the presence of a vertebral column. Willis (6) emphasized that the identification of a vertebral column secures the diagnosis of fetus in fetu and differentiates this entity from teratoma. Identification of the vertebral column indicates that fetal development of the included twin must have advanced at least to the primitive streak stage to develop a notochord, which is the precursor of the vertebral column (6). Occasional cases have been reported in which the spinal column could not be identified at imaging (10). These cases probably were due to an underdeveloped and markedly dysplastic spinal column that prevented the identification of vertebral bodies at imaging (11).

The fetus in fetu may increase in size and cause local mass effect and hemorrhage (12). Surgical excision is the recommended treatment. The intraabdominal fetus in fetu is usually contained in a complete sac, without any major vascular connections to the host. A few instances of definite vascular connections have been reported, but the predominant blood supply appears to be derived from the plexus where the fetus in fetu and the sac are attached to the abdominal wall (4). One of the masses was attached to the aorta by a vascular pedicle just below the superior mesenteric artery. This may explain the large size of the fetus in fetu and the delayed manifestation, as the fetus may have grown in the abdominal cavity.

Imaging played an important role in our ability to correctly diagnose both cases in a prospective fashion. The diagnosis of fetus in fetu can be made with abdominal conventional radiographs, by identifying a vertebral column and/or specific bony structures (10). Nocera et al (11) initially described the CT appearance of fetus in fetu. The CT findings were those of a mass that consisted of a round or tubular collection of fat that surrounded a central bony structure. The identification of vertebrae or long bones is essential for establishing this diagnosis prospectively. Our findings were similar to those reported and allowed us to correctly diagnose this rare lesion at the time of mass identification. CT also was helpful in determining the relationship of the mass to the other abdominal structures.

In summary, fetus in fetu is a pathologic condition that occurs from the abnormal embryogenesis in a diamniotic, monochorionic pregnancy. The issue is

still debatable as to whether it represents a highly organized teratoma. However, its characteristic CT appearance allows correct prospective diagnosis of this very rare disease entity.

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