



Two cases of fetus in fetu

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Abstract Fetus in fetu (FIF) is a rare cause of abdominal mass in children. One of the malformed monozygotic diamniotic twins is located in the body of other twin. It is differentiated from teratoma by the presence of vertebral organization with limb buds and other organ systems. Diagnosis is based on radiologic findings. Surgical excision is the treatment of choice, leading to the complete removal of the mass. To our knowledge, less than 200 cases have been described in the literature. Herein, we report 2 cases of FIF, a newborn who was diagnosed antenatally and a three-and-half-year-old boy diagnosed with mediastinal FIF after admission for recurrent respiratory tract infections.

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Fetus in fetu (FIF) is a result of abnormal embryogenesis in a monozygotic diamniotic pregnancy in which a vertebrated fetus (the parasite) is located in the body of its normally developed twin (the autosite). The exact pathogenesis is still unknown; but during the ventral folding of trilaminar embryonic cyst, diamniotic monozygotic twin is included within its host. At the beginning of pregnancy, the parasitic twin grows parallel to its host twin; but later, this growth stops because of either its own inherent defects or dominant vasculature of normal twin [1]. In most cases, parasitic twin is anencephalic and usually contains a vertebral column and budding limbs. The upper limbs are less developed than lower limbs.

Fetus in fetu is usually located in the abdomen of the host twin [2]; but locations in gastrointestinal tract, genitourinary tract, vessels, and central nervous system have been rarely reported. Other even more rare locations are pancreas, adrenal glands, lungs, mediastinum, lymph nodes, and spleen [3]. Usually, there is a single parasitic twin; but rarely, more than 1 parasitic twin is observed in the host body [4].

Diagnosis can be made by ultrasonography (USG), plain radiography, computed tomography (CT), and magnetic resonance imaging (MRI). It should be differentiated from a teratoma by the presence of a vertebral column. Both conditions may have a calcified mass in imaging studies. Operative and histopathologic findings confirm the diagnosis. The treatment of FIF is complete excision, which is followed by a good outcome.

We report 2 cases of FIF, one located in abdomen and the second located in the mediastinum, which is a very rare location.

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1. Case 1

A male infant was delivered at 38-week and 4-day gestation via elective caesarean delivery. The infant had Apgar scores 8 and 9 at 1 and 5 minutes, respectively. There was no history of maternal illness, exposure to radiation, or drug intake during pregnancy nor consanguinity between parents. An intra-abdominal mass, resembling a FIF, was detected at 32 weeks of gestation by antenatal ultrasonography. At birth, the baby's weight was 4060 g (90-97 percentile), his length was 52 cm (50-75 percentile), and the head circumference was 36 cm (50-75 percentile). Physical examination revealed a 6 × 6 cm mass in the right lower quadrant. Complete blood count and kidney-liver function tests were within reference ranges. Serum β human chorionic gonadotropin (β -HCG) was 1 mIU/mL (normal, <5 mIU/mL) and serum α -fetoprotein (AFP) was 11 040 ng/mL (normal, 9452 ± 12610 ng/mL). Plain abdominal radiographs revealed a calcified right abdominal mass (Fig. 1A). Abdominal ultrasonography confirmed the presence of a malformed fetus within its sac posterior to the

urinary bladder. Magnetic resonance imaging of the abdomen revealed a heterogeneous well-encapsulated cystic mass 87×72 mm in size, displacing the right kidney superiorly and bladder anteriorly. There was a semisolid nodular mass with calcified and lipomatous components within the cyst resembling a teratoma (Fig. 1B).

On the fourth day of life, elective laparotomy was performed, showing a 10×10 cm tense, retroperitoneal cystic mass, pushing the bowel anteriorly. The sac contained clear yellow fluid, which ruptured during the dissection. The fetus in the sac was approximately 70 mm in vertical length that was appropriate for 11th gestational week and had visible limb buds (Fig. 1C). The cystic component was decompressed, and the mass was dissected off the retroperitoneum with ligation of the feeding vessels. Histopathologic examination revealed neuroglial tissue with ventricular system including choroidal plexus, skin with adnexae, a vertebral column with cartilage, bone and bone marrow elements, striated muscle, lymphoid tissue, sympathetic ganglia, and adipose tissue. The specimen was surrounded by 1 layer epithelium resembling to amniotic

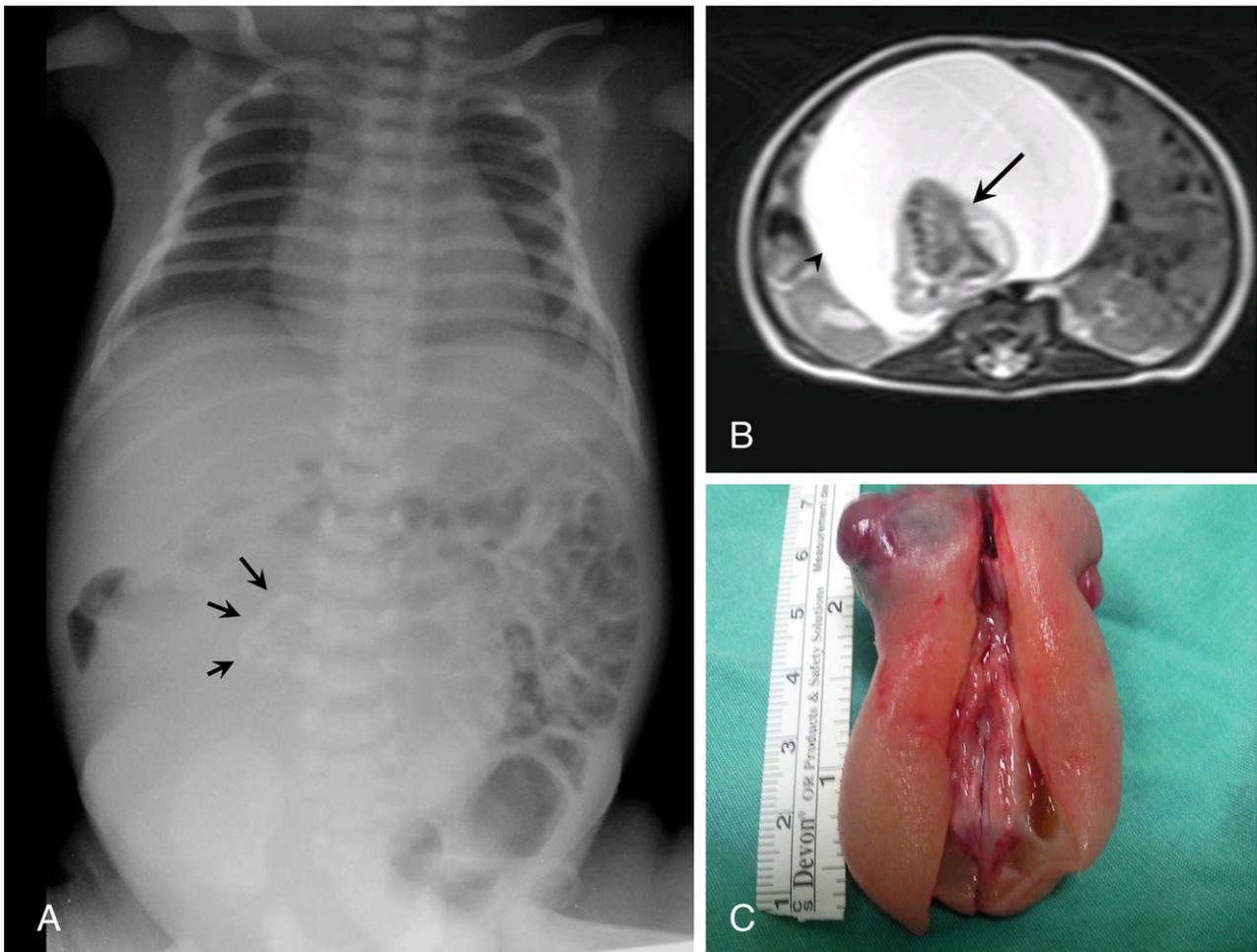


Fig. 1 A, Plain radiograph of the abdomen shows a calcified mass in the right abdomen (see black arrows). B, Abdominal MRI shows a semisolid nodular mass with calcified and lipomatous components within the cyst. Arrow head shows the amniotic sac; and black arrow, the vertebral mass. C, Macroscopic view of FIF with vertebral bodies and limb buds.

membrane. The infant had an uneventful recovery and was discharged on the seventh postoperative day. He was 3 months old at the last follow-up visit with no complaints and the serum levels of β -HCG and AFP were less than 1 mIU/mL and 81 ng/mL, respectively.

2. Case 2

A three-and-a-half-year-old boy presented to the pediatric outpatient clinic with fever, cough, respiratory distress, and a history of recurrent respiratory tract infections. His clinical examination revealed mild respiratory distress with a respiratory rate of 46 per minute. Laboratory studies showed normal blood glucose, liver, and kidney function tests. The serum AFP level was 1.5 ng/mL (<6.5 ng/mL), and serum β -HCG was 2.5

ng/mL (<5 ng/mL). His chest radiograph revealed a mass in the left upper hemithorax (Fig. 2A). Computed tomography demonstrated a 8-cm-long mass with lipid and calcified components, extending from the apex of the left lung to the inferior hilus and obstructing the mediastinal structures, left main bronchus, and hilar structures (Fig. 2B). Elective thoracotomy showed that the mass was located in the upper mediastinum, very close to the aortic arch. Blunt surgical dissection was performed, and the mass was separated from the aorta and pericardium, allowing its complete removal. During the dissection, the capsule of the mass ruptured, and muddy fluid drained (Fig. 2C). The lesion appeared to be a fetus with clear facial structures. The plain radiographic examination of the mass revealed several bones suggestive of vertebra (Fig. 2D). Histopathologic examination revealed bone, cartilage, mucous glands, neural, and pancreatic tissue. Postoperative

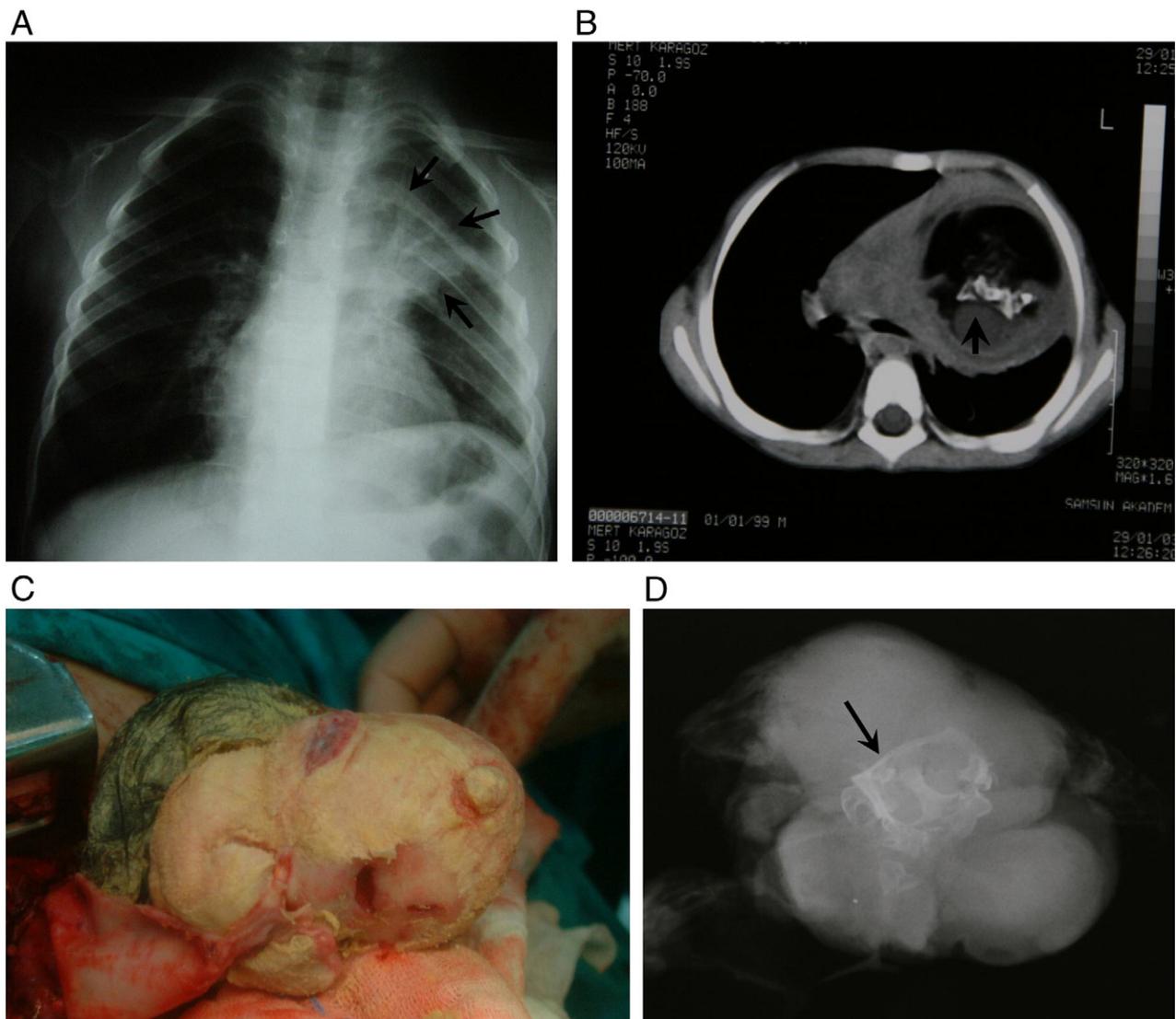


Fig. 2 A, Chest radiograph showing a mass in upper hemithorax (see black arrows). B, Computed tomographic scan showing heterogeneous lesion with lipid and calcified components in the mediastinum. Calcified portion shown with black arrow. C, Macroscopic view of material retrieved at operation. D, Plain radiograph of FIF showing vertebral organization (see black arrow).

course was uneventful, and he was discharged on the seventh postoperative day. He remained well 2 ½ years after surgery.

3. Discussion

Fetus in fetu is a rare pathology with an incidence of 1 per 500,000 births [5], in which a malformed fetus is located in the body of its twin, usually in the abdomen. It was first described by Johann Friedrich Meckel and is the result of an abnormality of monozygotic diamniotic twinning. Smaller cell mass in a developing normal fetus results from an unequal division of totipotent inner cell mass of the developing blastocyst. This term was first cited by Lewis [6].

Most of the reported FIF cases were in infancy [7], and the most frequent presentation is an abdominal mass, especially in upper retroperitoneum. Skull, scrotum, sacrum, lung, and mouth are other less frequently reported locations [3,8-10]. Three intrathoracic FIF cases were reported in the literature [3,11,12]. Two of them were located in the lower mediastinum [11,12], and the other one was in the lung parenchyma [3]. In our first patient, the mass was located in the abdomen. In the second patient, the mass was in the upper part of the mediastinum; and to the best of our knowledge, this case is the first reported case of FIF in the upper mediastinum.

Fetus in fetu can be differentiated from teratoma by the presence of vertebral bodies. If vertebral bodies exist, this means that the fetus passed the primary stage of gastrulation. Conversely, teratoma consists of pluripotent cells, without vertebral segmentation or organogenesis. Pathologic examination of the masses showed vertebral bodies in both our patients.

Fetus in fetu is a benign condition; however, the mass may compress the surrounding organs and tissues. In the second patient, the mass was very close to the aortic arch and pericardium. Compression of the lung by the mass may explain recurrent lung infections in this patient. After the surgical procedure, the child had no further lung infections and did well.

Plain radiographs, USG, CT, and MRI can be used to achieve a diagnosis of FIF. Our first patient was diagnosed antenatally by USG; postnatally, abdominal plain radiographs, MRI, and surgery confirmed the diagnosis. In our

second patient, a chest radiograph revealed a heterogenous calcification in the mass suggesting a teratoma-like structure. Computed tomography, MRI, and surgery confirmed the diagnosis of FIF.

Almost all of the FIF masses are acardiac and anencephalic. Intestinal, neural, pulmonary, gonadal, pancreatic, and adrenal tissue may be present. In our first case, the pathologic material consisted of peripheral nerves, sympathetic ganglia, striated muscle, choroid plexus, and fat surrounded by a layer of epithelium resembling an amniotic membrane. In our second patient, the mass consisted of neural and pancreatic tissues, which was covered by skin.

In conclusion, FIF is a rare anomaly that typically presents in infancy. Current imaging modalities enable accurate diagnosis before surgery. Enucleation is generally curative and allows confirmation of the diagnosis. In children with recurrent respiratory tract infections and a mediastinal mass, FIF should be considered in the differential diagnosis.

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