OLGU SUNUMU/CASE REPORT

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Fetus-in-fetu: Disguising Lymphocyst

İntrauterin Lenfokist Olarak Takip Edilmiş bir Fetus-in-fetu Vakası

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ABSTRACT

Fetus in fetu (FIF) is a rare anomaly resulting from abnormal embryogenesis. Patients often present with an abdominal mass. We report a case of FIF initially diagnosed as a lymphocyst in fetal ultrasonography (USG) during prenatal screening.

Keywords: lymphocyst, fetus-in-fetu, abdominal mass

ÖZ

Fetus in fetu, anormal embriyogenez sonucu oluşan oldukça nadir bir hastalık grubudur. Hastalar genellikle abdominal kitle nedeni ile başvururlar. Bizde kliniğimizde intrauterin dönemde lenfokist olarak takip edilmiş olan ve sonrasında FIF tanısı alarak takip edilen bir vakayı sunmak istedik.

Anahtar Kelimeler: lenfokist, fetus-in-fetu, karında kitle

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BACKGROUND

The FIF is an uncommon pathology with an incidence of 1 in 500.000 births (1). There are many theories in the etiology. Most accepted one is, during embryogenesis, unequal cell division during the blastocyst stage causes a cellular mass to remain in the mature embryo. Thus, diamniotic monochorionic pregnancy is the development of the remaining cellular mass in the body of normal co-twin's body (1). Multiple pregnancy and a variant of mature teratoma are among other theories (2). Becauseof the risk of malignancy in mature teratomas, differential diagnosis with FIF should be performed. FIF can be distinguished from mature teratomas by the presence of vertebral structures. Most common presentation is a mass in the abdomen. In addition, the mass can be inthe cranium, madiastinum, sacrum, scrotum or in mouth. Some cases are diagnosed in prenatal period. In this report a case of FIF initially diagnosed as lymphocyst in prenatal USG was presented.

CASE REPORT

A female infant of 39 week's gestational age was born to a 33-year-old woman by cesarean section due to previous cesarean. Her birth weight was 4500 g, APGAR scores were 9 and 10 in 1th & 5th minutes respectively. Pregnancy was uneventful but an intraabdominal mass, supposed to be a lymphocyst, was detected by USG in the 28th week of gestation. It was reported as a 46x40x59 mm cystic mass without a vein and with a solid component of 12 mm in diameter. Due to its thin wall, it was primarily thought to be compatible with the lymphocyst. There was no consanguinity between the parents and prenatal history was otherwise unremarkable. She was followed in the neonatal intensive care unit after delivery. Physical examination revealed 2 cm palpable liver and palpable mass in the right lower quadrant. After delivery AFP was 69.533 IU/mL (Normal range:48.406±34.718 IU/mL). USG showed a mass approximately 5.3x2.9 cm midline in the abdomen, with lobulated contours, smooth cystic areas and microcalcifications (Figure 1). Computerized tomography (CT) revealed calcified and ossified lesions in the right upper and lower abdomen, the lower extremities and vertebral structures (Figure 2). The abdominal mass was operated and removed without ruptiring the amniotic membrane. Umblical cord, amniotic sac and extremity fragments were determined in the macroscopy of the material. Pathology was re- ported as FIF, umblical cord with two arteries and containing a vein (Figure 3). Macroscopic examination revealed no development of heart and brain (Figure 4). In the examination of

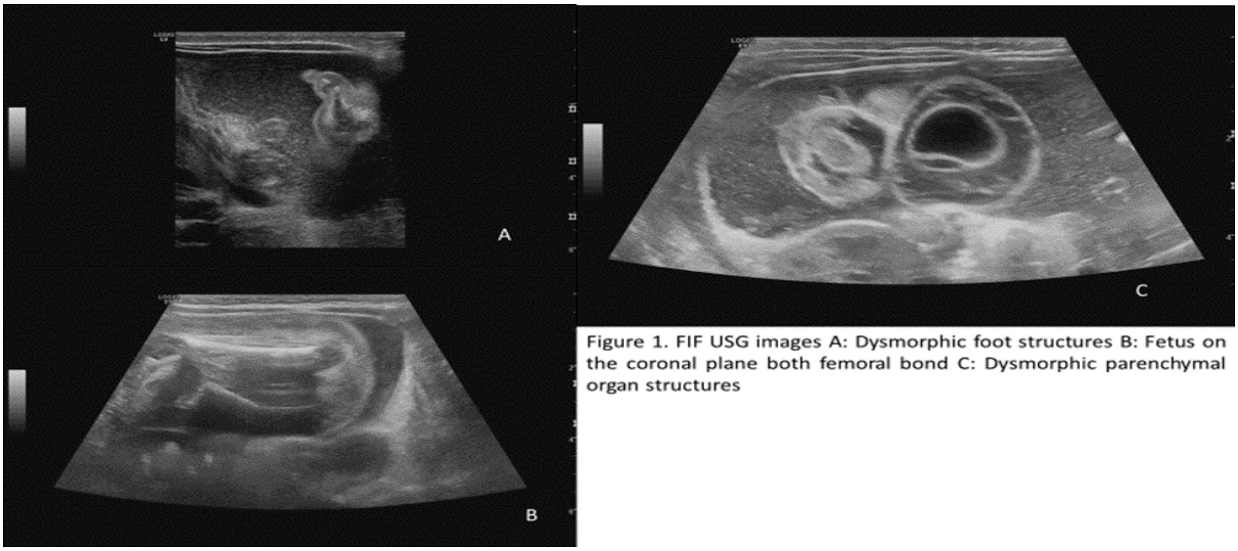

Figure 1. FIF USG images A: Dysmorphic foot structures B: Fetus on the coronal plane both femoral bond C: Dysmorphic parenchymal organ structures

microscopic samples, mature skin, central nervous tissue, intestinal epithelium, muscle, bone, cartilage, fat and bone marrow tissue were ob- served (Figure 5). The patient was found to have AFP:3490 IU/mL (2654±3080 IU/mL)
and beta-HCG:0.66 MIU/mL (0-0.5 mIU/mL) at 20 days postoperatively. There was no gross pathology in the postoperative USG of the patient who is currently 17 months old and the patient is under close follow up with AFP, beta-HCG and USG.

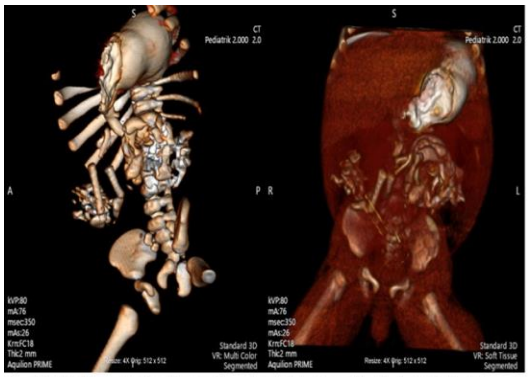

Figure 2. In the CT coronal reformat images, deformed skeletal structures of the fetus located in the abdomen of the infant are seen on the right.

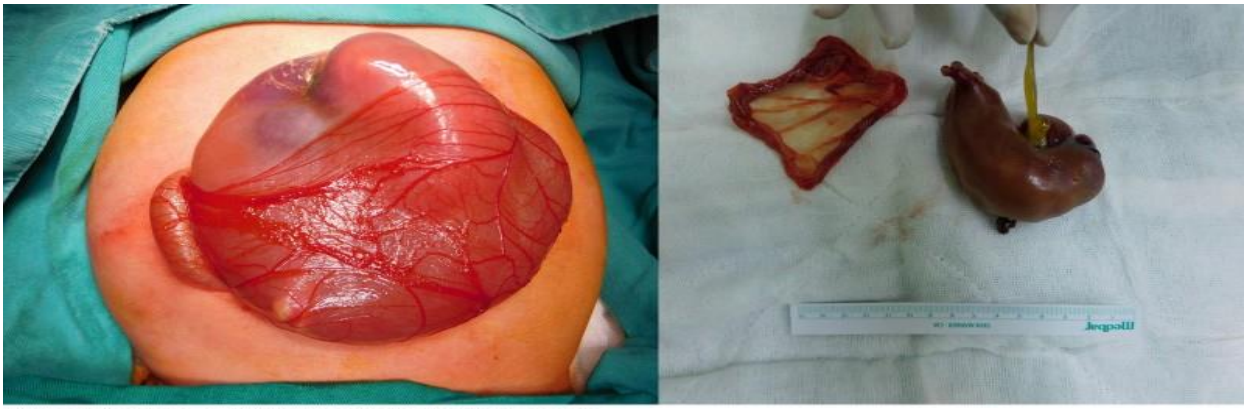

Figure 3. Amniotic sac, umblical cord and extremity fragments

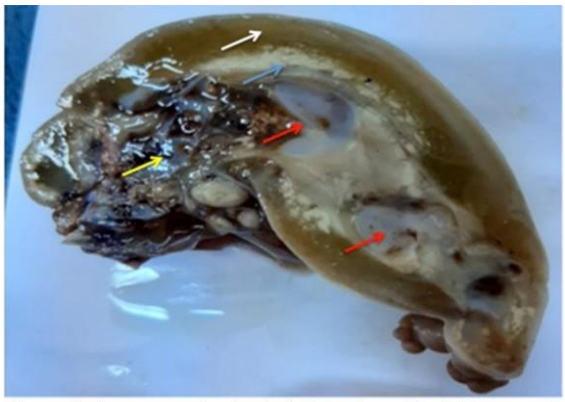

Figure 4. In macroscopic longitudinal sections skin (white arrow), subcutaneous fat tissue (blue arrow), cartilage tissue (red arrow) and occasionally cystic hemorrhagic areas (yellow arrow) were seen.

DISCUSSION

FIF was first descibed in the 18th century by Friedrich Meckel (3). FIF is seen at approximately 1 in every 500.000 births and is more common in males (4). Although mainly reported in infants and children younger than 18 months, there are several cases reported in adults (1).
The presence of all or a part of the vertebral column and the presence of axial or appendicular organs or bones are the prominent characteristics of FIF. Different tissues and organs can be seen in the fetus such as costa, central nervous system, gastrointestinal system and vascular structures. Conventionally, these fetuses are acardiac and anencephalic (5). In our patient, we haven't seen the development of heart and/or brain tissue, in macroscopic examination. The blood supply to FIF is usually derived from vascular structures in the abdominal wall, and the size and weight of FIF varies according to the blood supply. If it has a large and wide blood network, it is seen as a large and well-developed mass (1). Patients are often diagnosed as having an abdominal mass. The cli

nical symptoms are usually due to mass effect such as abdominal distension, constipation, dyspnea etc. Serum AFP and beta-HCG levels may be normal or elevated (5). In our case, preoperative AFP and postoperative AFP and beta-HCG were within normal limits.

Many authors classify the FIF completely seperate from the teratoma. The presence of vertebral structures and extremities allows the differentiation of FIF from the teratoma. During the embryogenesis, the vertebral column develops in the normal embryo in three weeks from the primitive line. Existence of a vertebral column in the mass is a clue for the diagnosis of FIF because it reflects the embryogenesis (6). However, although there is no vertebral column in FIF, there are case reports that can include tissue in the advanced stage of organogenesis (5). The teratoma may contain different tissues originating from three germ layers. Therefore, some authors consider FIF as a highly organized teratoma (1). Some authors evaluated FIF and teratoma as the same pathological disease in different maturation stages (5). The stage of tissue formation in our patient had completed the maturation. Imaging modalities, especially CT, are descriptive for vertebral column and other skeletal elements. X-ray, USG, CT and/ or magnetic resonance imaging can be used as imaging method. In our patient, due to its thin wall, it was primarily thought to be compatible with the lymphocyst. CT revealed calcified and ossified lesions in the right upper and lower abdomen, the lower extremities and vertebral structures and findings were compatible with a fetus

Surgical excision is recommended in the management of the FIF (6). In our case, surgical excision was performed and the pathological diagnosis was confirmed. The follow up of the patient is recommended with AFP, beta-HCG and USG and after 17 months she is well without a complaint. In differential diagnosis of lymphocyst in prenatal USG, FIF should be kept in mind.

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