Fetus-in-fetu as a differential diagnosis of fetal abdominal tumors

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Abstract

Fetus-in-fetu (FIF) is a rare condition in which an abnormal parasitic twin grows inside the body of its co-twin. Fetal Medicine specialists should be suspicious of dealing with FIF when ultrasound imaging shows a heterogenous mass inside the fetal abdomen. We present a case of a 21-year-old pregnant woman who presented an abnormal ultrasound at 31 weeks gestation that revealed a fetus with an intra-abdominal mass. The fetal ultrasound evaluation was suggestive of FIF. The patient had a vaginal delivery after spontaneous onset of labour at the end of pregnancy. An exploratory laparotomy of the newborn was performed four days after birth and allowed removal of an incomplete fetus with distorted head and limbs enveloped by a semi-transparent sac-like capsule placed in retroperitoneum and fed by a small branch of the aorta. The autopsy confirmed the diagnosis of a FIF. The newborn was discharged healthy.

Background

Fetus-in-fetu (FIF) is a rare congenital anomaly with an incidence of 1 per 500,000 births [1]. The pathogenesis of FIF can be explained by the ‘Included - twin’ theory in which the FIF is a diamniotic, monochorionic, monozygotic twin that becomes incorporated into the body of the host twin during organogenesis [2]. The most common presentation is a mass in the abdomen located in the retroperitoneum (80%) [3]; however, FIF can be detected in other sites include intracranial, mediastinum, sacrum, scrotum, and mouth [4]. Due to the similar ultrasound features, FIF is frequently diagnosed as a teratoma, but it is differentiated from that by the vertebral organization with limb buds and other organ systems [5].

FIF are usually single, rarely multiple tumors, and are genetically identical to their host, as proven by karyotyping and DNA analysis [5, 6]. Diagnosis can be made via X-ray, ultrasound, CT scan and MRI, and some cases were diagnosed after prenatal screening as the case report presented here. After delivery, computerized tomography (CT) scan and magnetic resonance imaging (MRI) can be performed to minimize the differential diagnosis and evaluate the patients’ condition before the surgery [3,7].

Case Report

A 21-year-old pregnant woman, primigravidae, presented to our institution after an abnormal ultrasound at 31 weeks gestation that revealed a fetus with an intra-abdominal mass. She was asymptomatic. On the transverse and sagittal plane, the obstetrical ultrasound detected a heterogeneous echogenicity mass inside the fetal abdomen with solid, cystic, and calcified elements measuring 30 X 24 X 26mm and without significant internal blood flow (Figure 1). The mass was in the left hypochondriac region, pushing the stomach upwards and under the left lobe of the fetal liver. Components favoring a fetus seen within the mass include a deformed skull and long bones. The color Doppler showed a branch of the fetus’s abdominal aorta, which supplied blood to the mass. The prenatal diagnosis was FIF. There was an increase in the size of the mass, and the long bones became more evident at the 36 weeks+5 days scan (50 X 46 X 34mm) (Figure 2). At 38 weeks + 4 days of gestation, the patient had a vaginal delivery of an infant weighed 2825 grams and Apgar scores of 8/9 after spontaneous onset of labour.

The infant’s abdominal ultrasound revealed a heterogeneous and predominantly cystic mass in the epi/ mesogastric region with coarse calcifications (parts of a skull), one of them elongated as a long bone-like appearance. The mass measured 50 X 47 X 37mm and displaced the other adjacent abdominal organs (Figure 3). Doppler velocimetry showed mild vascularity supplying the wall of the mass. An exploratory laparotomy of the newborn was performed four days after birth. The mass’s gross description was like the prenatal and preoperative diagnosis: an incomplete fetus with distorted head and limbs enveloped by a semi-transparent sac-like capsule placed in retroperitoneum and fed by a small branch of the aorta (Fig-
The postoperative period was uneventful.

The autopsy confirmed the diagnosis of a FIF weighing 16.0 grams and measuring 9.0 cm (Figure 5). The FIF was composed of a remnant of the cranium, and thoracic cage, the right upper limb measuring 20 mm and the left lower limb measuring 65 mm.

Figure 1: A heterogeneous echogenicity mass inside the fetal abdomen with solid, cystic, and calcified elements was detected at the 31 weeks + 5 days fetal scan.

Figure 2: The FIF increased in size, and the long bones became more evident at the 36 weeks+5 days fetal scan.

Figure 3: The abdominal ultrasound evaluation of the two-day-old infant showed an image suggestive of FIF.

Figure 4: The postoperative specimen showed a FIF with a remnant of the skull, thoracic cage, the right upper limb, and the left lower limb. Part of the umbilical cord can be seen.

Figure 5: X-ray of the autopsy specimen showing a FIF with parts of a skull, two long bones (right humerus and left femur), and some bones of the metatarsal and toes. Definitive diagnosis: FIF.

Discussion

FIF is a sporadic disease in which a malformed fetus is inside of the body of its co-twin. There are some hypotheses of the pathogenesis mechanism; however, the diamniotic monochorionic twins’ hypothesis is the most accepted. It states that a FIF occurs at a very early stage, during the embryonic folding at the second and third week of gestation [8]. Taher et al. (2020) [9] demonstrated the combined experience of four sizeable tertiary referral centers with FIF. They showed that the diagnosis occurs during infancy in most cases by symptoms suggestive of abdominal distension and feeding or respiratory difficulties. Only a few reported cases were above the age of 2 years.

The advancement of ultrasound has enhanced the image quality and accuracy of diagnosis in a shorter time. It is the first-line tool in diagnosing FIF, particularly during fetal life. Prenatal ultrasound shows a fluid-filled mass with solid pieces and bone-shaped calcification inside [10]. Some authors highlight the presence of axial skeleton or long bone as an essential feature in diagnosing FIF. FIF may be confused with meconium peritonitis, teratoma, neuroblastoma, adrenal haemorrhage, and viral infection [5]. Fetal Medicine specialists and pediatric surgeons should be suspicious of dealing with FIF when preoperative imaging shows an axial skeleton, in the presence of multiple highly organized structures around it, when a glissening amniotic sac encases the tumor [9]. The treatment of choice for FIF is complete resection as there is still a controversy over whether FIF is an entity on its own or is a highly organized teratoma, the potential for malignant transformation or recurrence exists [11]. So, establishing the true nature of FIF is of great interesting since teratomas have more than a 10% malignancy rate. The removal of the FIF must involve careful planning as there is a high possibility of bleeding intraoperatively as the mass is highly vascularized. A preoperative CT angiography may help review the mass’s vascularity and identify the feeding vessel [7].
References


