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Case Report

Twin Fetus in Fetu with Immature Teratoma: A Case Report and Review of the Literature

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Fetus in fetu is an extremely rare condition in which a fetus or fetus-like structure with a vertebral axis is seen in the body of its twin. This paper presents a case of fetus in fetu in a two-day-old female newborn who was referred for an abdominal mass, biliary vomiting, and feeding intolerance. After plain abdominal X-ray and ultrasonography, the patient underwent abdominal surgery with the primary diagnosis of teratoma or fetus in fetu. We found a retroperitoneal mass that consisted of double fetus in fetu and a separate undetermined mass. The pathologic examination confirmed double fetus in fetu and revealed a separate immature teratoma. She was discharged from the hospital after seven days in a healthy and normal condition.The level of serum alpha-fetoprotein was normal after three months of follow-up.

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Introduction

Fetus in fetu (FIF) was first described by Meckel in 1800 and defined by Willis in 1953 as a mass containing a calcified vertebral axis often associated with other organs or limbs around this central axis.[1] This congenital anomaly is a monozygotic diamniotic, parasitic twin, attached by a vascular anastomosis to its host chorionic circulation, which is usually incorporated into the body of its most endoparasitic twin. FIF is a rare anomaly which resembles to retroperitoneal teratoma. However, in contrast to teratoma, this fetus- like structure has spinal axis and shows variable degrees of organogenesis.[1]

Case Report

A two-day-old female newborn of healthy, not relative parents was referred for upper abdominal mass, biliary vomiting, and feeding intolerance. The patient was the seventh child of a 40-year-old female who had a history of stillbirth in her first conception. Except one of her children who died in a car accident, all others were healthy and normal. She had no history of prenatal illness including diabetes mellitus, taking teratogenic drugs, and radiation exposure. Her obstetric examination and ultrasonography were normal. She had a full-term delivery with cesarean section. The newborn's weight was 3250 g and had normal Apgar scores.

On admission, she had feeding intolerance and biliary vomiting, but she had expelled meconium. The abdomen was distended and a tense mass was seen in the right upper part. Plain abdominal x-ray revealed displacement of abdominal viscera to the left lower part and a spinal axis- like calcification in the right area. Ultrasonography showed a solid cystic mass structurally similar to vertebral column. According to imaging findings, the diagnosis of retroperitoneal teratoma or FIF was made and the patient underwent abdominal surgery. No tumor markers were evaluated prior to the surgery; however, alpha-fetoprotein (AFP) and beta human chorionic gonadotropin (βhCG) levels,

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immediately after the surgery were 2600 IU/mL and 1.3 IU/L, respectively. Because she had an emergency condition, MRI or CT scan could not be performed before the surgery.

The surgery revealed a well-defined retroperitoneal encapsulated vascular mass which caused displacement of the inferior vena cava (IVC) and abdominal viscera (Figures 1 and 2). During dissection, it ruptured and a yellowish clear fluid came out and subsequently two fetiform masses were seen. There was also another mass without specific diagnostic feature. All of them and the most part of the main capsule (except those parts with adhesion to main vessels) were resected (Figure 3). Two days after the surgery, oral feeding was started and the newborn was discharged five days later.

Pathologic description

Macroscopic examination revealed three masses consisting of:

- A soft tissue mass resembling acardiac fetus, weighing 150 g, and measured 20×5×3 cm. There were two malformed upper limbs: the right measured 20 mm in length with three malformed fingers and the left measured 12 mm in length without finger. There were also two rudimentary lower limbs. Both right and left measured 20 mm in length and had three and five fingers, respectively. Sections revealed a spinal cord housed in the complete spinal column. The chest cavity had mostly composed of myxoid material and in the abdominal cavity a tubular-branched structure was seen. No other internal organ was identified.
- The second was a malformed premature anencephaled fetus with an incomplete face which weighed 100 g and measured 20 mm in head circumference and 16 cm in crown-heel length. The fetus attached to an amniotic bag by umbilical cord of 4×0.2 cm. There were two rudimentary upper limbs. The right measured 15 mm in length with four fingers and the left measured 17 mm in length with three fingers. The right lower limb with four fingers measured 20 mm in length and the left one with five fingers was 15 mm in length. Sections revealed spinal column, a tubular- branched structure in the abdominal cavity with a proximal end near the cervical region, and the distal end reaching the distal end of the fetus.
- The third mass was an irregular spherical tissue with bosselated external surface, measuring 7×6×3 cm. In sections, it had solid cystic appearance with heterogeneous consistency.

Microscopic examination of both fetuses confirmed the presence of spinal cord with occasional ganglia as well as presence of a highly-

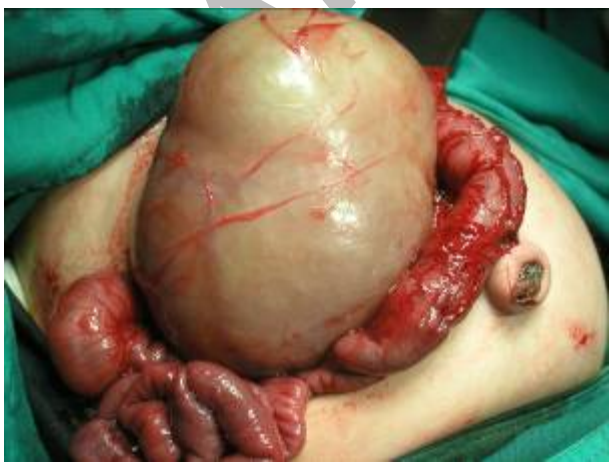

Figure 1. Intraoperative view of the well-capsulated mass.

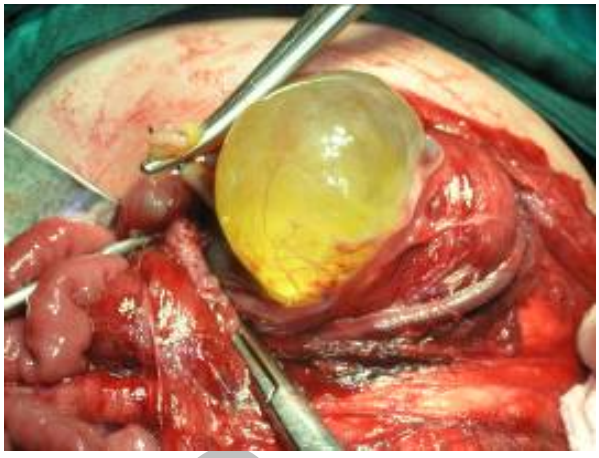

Figure 2. Amniotic sac with displacement of the inferior vena cava.

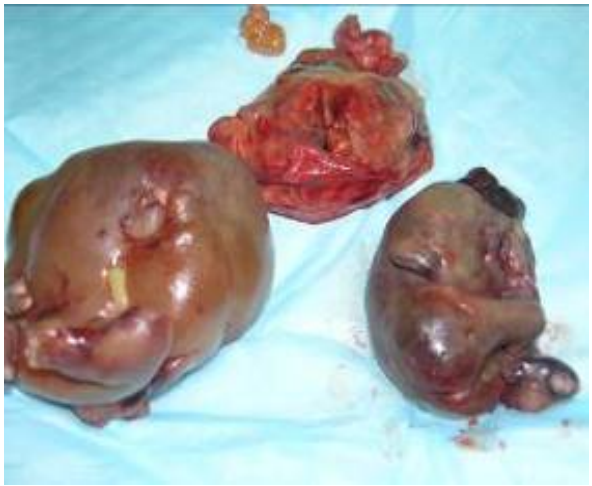

Figure 3. Twin fetus in fetu with immature teratoma.

differentiated dermal epithelium and subcutaneous tissue, cartilage, and gastrointestinal tract. In the second fetus, portions of respiratory tract, large vessels, and cerebroid tissue were also identified. Sections from third mass showed features of immature teratoma.

Discussion

FIF is a very rare congenital condition that has an incidence of one in 500,000 births with fewer than 100 reported cases worldwide.[2] It is a parasitic monozygotic diamniotic twin that is seen in the body cavity of newborns, infants, or rarely of adults. It was first described by Meckel as a rare condition in which a malformed parasitic twin was found inside the body of its partner, usually in the abdominal cavity.[1] To separate it from teratoma, Willis in 1953 set the criteria that FIF contains a vertebral axis with organs and limbs arranged around it.[1] However, review of the literature showed that in about 9% of cases of FIF, there was no vertebral column, even on pathologic examination.[3] This has led to another definition of FIF by Gonzalez-Crussi: "FIF is applied to any structure in which the fetal form is in a very high development of organogenesis and to the presence of a vertebral axis."[3] On the contrary, teratoma is an accumulation of pluripotential cells in which there is neither organogenesis nor vertebral segmentation.[4] Although the hypothesis regarding the origin of the FIF was widely accepted as the included twin theory, which was postulated as a diamniotic monochorionic monozygotic twin, some supporters of the teratoma theory have suggested that the FIF mass represents a well-differentiated, highly-organized teratoma.[5]

The term FIF is used to point out an unequal division of totipotential cells of a blastocyst where the result is the inclusion of a small cellular mass in the more mature embryo. This is a form of monozygotic diamniotic twin pregnancy where the parasitic twin installs and grows in the body of its partner.[6] This is a rare malformation that has some similarities with the retroperitoneal teratoma, but it is different from the latter by its fetiform aspect and the metameric segmentation of its spinal axis. A capsule covering this formation and a vascular pedicle are frequently encountered.[7]

The majority of cases present in infancy, with the oldest reported case occurring in a 47-year-old man.[8] Sex ratio was different in the reports; however, it seems that males are predominant.[9]

The most common site of presentation is abdominal cavity. It is also described in cranial cavity, oral cavity, mediastinum, lung, sacrococygeal region, kidneys, and scrotum. In contrast, teratomas usually arise in the lower retroperitoneum. FIF is usually single but two fetuses and up to five fetuses in the skull are also reported.[8–11] The size and weight of fetus varied from 4 cm to 24.5 cm, and from 1.2 g to 1.8 kg, respectively in the literatures.[12–15] According to Hing A et al., 89% of cases were less than 18 months.[16] The presenting symptom was abdominal mass in 70% of cases, which were retroperitoneal in 80% of them.[10] Different organs can be seen in these fetuses; vertebral column is seen in 91%, limbs in 82.5%, CNS in 55.8%, gastrointestinal tract in 45%, vessels in 40%, and genitourinary tract in 26.5%.[2]

Symptoms of FIF related mainly to its mass effect and the site of presentation and they present gradually. In the abdomen, it causes abdominal distention, feeding difficulty, emesis, jaundice, and pressure to genitourinary and gastrointestinal tracts. In the thorax, it causes dyspnea and respiratory symptoms; in the oral cavity, dysphagia and cleft palate,[25] and in the cranial cavity, neurogenic symptoms. The most reports up to 1980 showed that the preoperative diagnosis of FIF was made only in 16.7% of cases and up to the mid-1990, fewer than a quarter of the cases were diagnosed. The use of CT scans and MRI have since enhanced the accuracy of preoperative diagnosis.[9,2] The ability to diagnosis FIF on prenatal ultrasonography was first reported by Nicolini et al.[17] AFP and βhCG may be normal or elevated.[18–20] Maternal AFP level also is high in some cases,[21] and they will be normalized postoperatively. The sustained high levels could be the sign of malignancy.

The fetus is typically suspended by a pedicle within a complete sac containing fluid or sebaceous material. The umbilical cord-like structure may consist of only two vessels.[1,8,18,19] Definite vascular connection to the host is rarely described.[8,11,2] The absence of umbilical vessels and a definite vascular connection explain the growth retardation and arrest of organ differentiation in almost all cases of FIF. They are usually acardiac and anencephalic and the lower limbs are more developed compared with the upper limbs.[1,2,8,9,18] The karyotype is usually normal and similar to the host.

All the cases reported so far have kept the

asymmetric monozygotic twinning theory. For instance, the fetuses and their hosts have consistently belonged to the same sex. It has also been shown that the blood group and karyotype of the leukocytes of the FIF were exactly the same as that of the host.[22] Lee et al. reported a case that both FIF and the host had trisomy 21.[23]

Complete excision of FIF with its all surrounding membrane is the choice treatment because of the possibility of malignant transformation in the residue of FIF.[8,21] Although the prognosis for FIF is more favorable than for cystic teratoma, the presence of immature elements mandates close clinical, radiologic, and serologic follow-up,[24] and the possibility of recurrence of malignant teratoma after FIF resection should always be kept in mind.

The occurrence of a subsequent teratoma is not unprecedented. Hopkins et al.[20] reported on a five-day-old boy who was found to have a retroperitoneal FIF and who developed a right abdominal mass which proved to be a teratoma with malignant components requiring chemotherapy. Also Gilbert-Barness E et al.[26] reported the third known instance of FIF associated with a benign teratoma at age one.

In this report, we presented a relatively rare case of FIF in a two-day-old newborn girl. We found two FIF with separated immature teratoma. To the best of our knowledge, cases of concomitant FIF with immature teratoma have not been reported till now. Complete resection was performed for this patient as recommended for such patients. However, the presence of concomitant immature teratoma made it difficult to choose whether we should use chemotherapy or not?

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