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Case Report

A case report of hepatic fetus-in-fetu: Approach to diagnosis ☆,☆☆,★

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ABSTRACT

Multiple theories have been proposed about the pathophysiology of Fetus-in-fetu (FIF). The most widely accepted theory is abnormal embryogenesis in diamniotic monochorionic pregnancies, in which a malformed parasitic fetus is found within the body of a twin host. Hepatic FIF has been reported in almost 1% of FIF cases, with only 2 case reports being published in the literature. This article presents the third case report of intrahepatic FIF. Additionally, we review the role of radiology in diagnosing these cases and guiding their proper management. This case report supports the monozygotic twin theory of FIF and the diagnostic dilemma of FIF vs. teratoma can be solved through collaborative work between radiologists and pathologists.

Introduction

Fetus-in-fetu (FIF) is a rare congenital condition characterized by a mass containing fetal parts located in various sites, including the intracranial, mediastinum, retroperitoneum, and sacrococcygeal regions. Multiple theories have been proposed about the pathophysiology and origin of these masses. Among them, the most widely accepted theory is abnormal embryogenesis in diamniotic monochorionic pregnancies, in which a malformed parasitic fetus is found within the body of a normally growing twin host [1]. Contrastingly, other authors have suggested that this mass results from a highly differentiated form of teratoma [2].

Worldwide, <200 cases of FIF have been reported, with an incidence of 1 per 500,000 live births and a 2:1 male-to-female ratio [1]. Most cases have been reported during infancy, with 1 case being reported in a 39-year-old patient [3]. These masses

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were commonly located in the retroperitoneum (80%). Other reported locations include the sacrococcygeal region, mediastinum, oral cavity, intracranial space, scrotum, and liver [3]. Hepatic FIF has been reported in almost 1% of FIF cases, with only a few case reports being published in the literature [4,5]. This article presents the third case report of intrahepatic FIF. Additionally, we review the role of radiology in diagnosing these cases and guiding their proper management.

Case report

An 8-year-old boy presented to the emergency department with abdominal pain and a visible abdominal mass. The patient's history was difficult to obtain due to the language barrier; however, his mother stated that she did not undergo prenatal follow-up during her pregnancy and that the abdominal mass had been present in the child since birth and had gradually grown. The patient had previously undergone assessments at another institution 4 years prior, including computed tomography (CT); however, we could not access the results or images. The patient did not undergo follow-ups due to limited access to medical care. Upon examination, a visible mass was observed on the right abdominal side, with mild tenderness on palpation. Routine laboratory findings were unremarkable. Initial abdominal radiography revealed well-defined, ovoid, and dysmorphic ossified structures overlying the hepatic shadow. Initial ultrasonography showed a large heterogeneous solid mass that was predominantly hyperechoic (fat) with a cystic component; further, a deformed hip joint was identified. Upon admission, previously obtained images were reviewed, and the patient was diagnosed with intrahepatic FIF. Follow-up magnetic resonance imaging (MRI) and CT showed a large, well-defined, encapsulated intrahepatic mass measuring 16 × 12 × 14 cm in segments (IV and VIII); moreover, the cystic component was characterized by macroscopic fat, an axial skeleton, pelvis, deformed limbs, teeth, and an anencephalic calvarium. Additionally, minimal enhancement was noted, with no major arterial supply or venous drainage (Fig. 1).

Given the continuous growth of the mass, segmental resection was planned and performed uneventfully through a transverse subcostal incision and midline incision. The mass was covered by a smooth capsule with bony components. First, the hepatic pedicle was identified, and the right hepatic pedicle was isolated. Complete liver mobilization and isolation of the right hepatic vein were achieved. The mass was dissected along its capsule, completely excised from the liver as 1 piece (Fig. 2), and sent for imaging (Fig. 3). There were no vascular pedicles or bleeding from the mass; additionally, the capsule was uninjured.

A mass measuring 18 × 15 × 14 cm with a smooth external surface was sent for pathological examination. Subsequently, obtained sections showed solid adipose tissue and large cystic areas containing mucoid materials. The upper segment of the mass contained hair- and tooth-like structures. The middle segment showed a possible vertebral column, cartilage, and gray-white solid areas. The lower segment contained limb-like bony structures with solid adipose tissue. Microscopic examination revealed a mixture of various well-formed organs, including the spinal cord and vertebral column (Fig. 4A), skin and subcutaneous tissues (Fig. 4B), prostate (Fig. 4C), and testicular tissue (Fig. 4D). The presence of these well-formed tissues was a diagnostic indicator of a fetus rather than a teratoma.

Early in the morning of postoperative day 1, the patient became critically ill with hemodynamic instability, significantly elevated liver enzymes, and acidosis. Upon returning to the operating room, the patient showed a right lobe (segment VII) infarction and a completely viable left liver lobe. Accordingly, a right lobe resection was performed. The patient was trans-

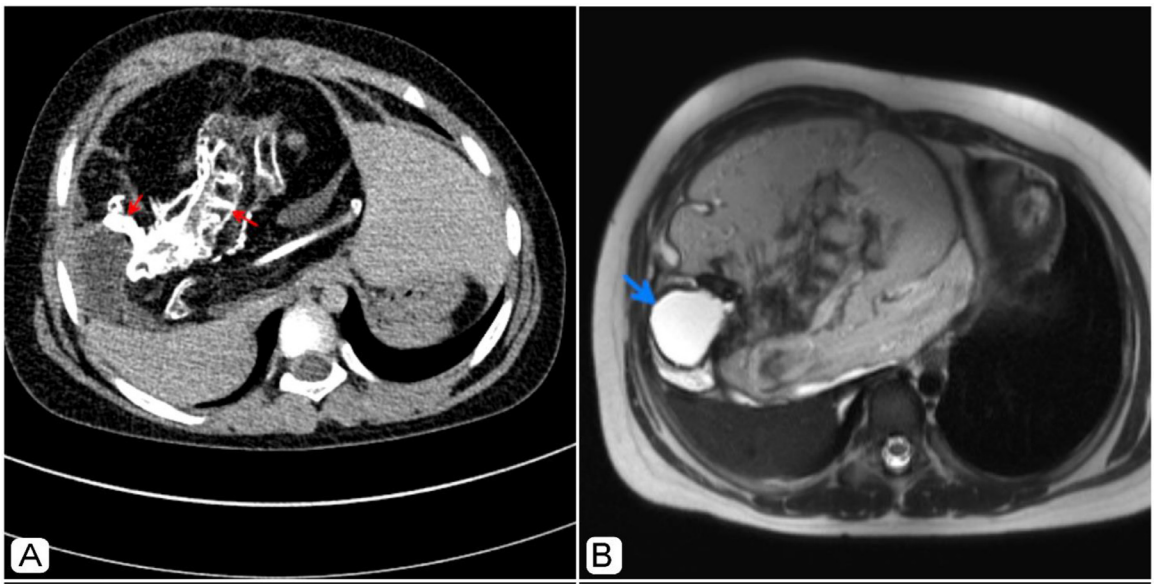

Fig. 1 – A CT scan showing a vertebral column and a skull base (red arrows) (A). Axial T2Wt MRI demonstrating a cystic component (blue arrow) laying above the cranium (B).

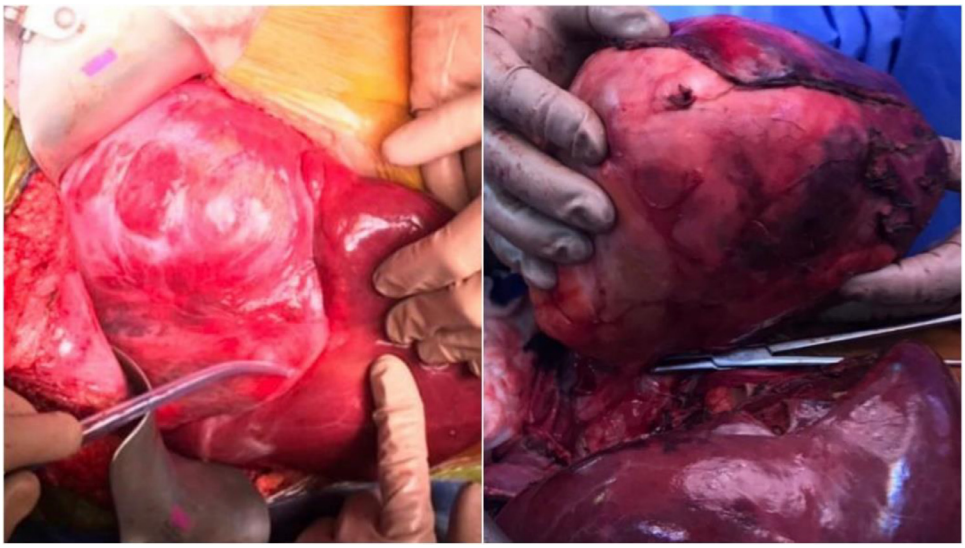

Fig. 2 – Gross photograph of the hepatic mass before the resection (left), and after the resection (right).

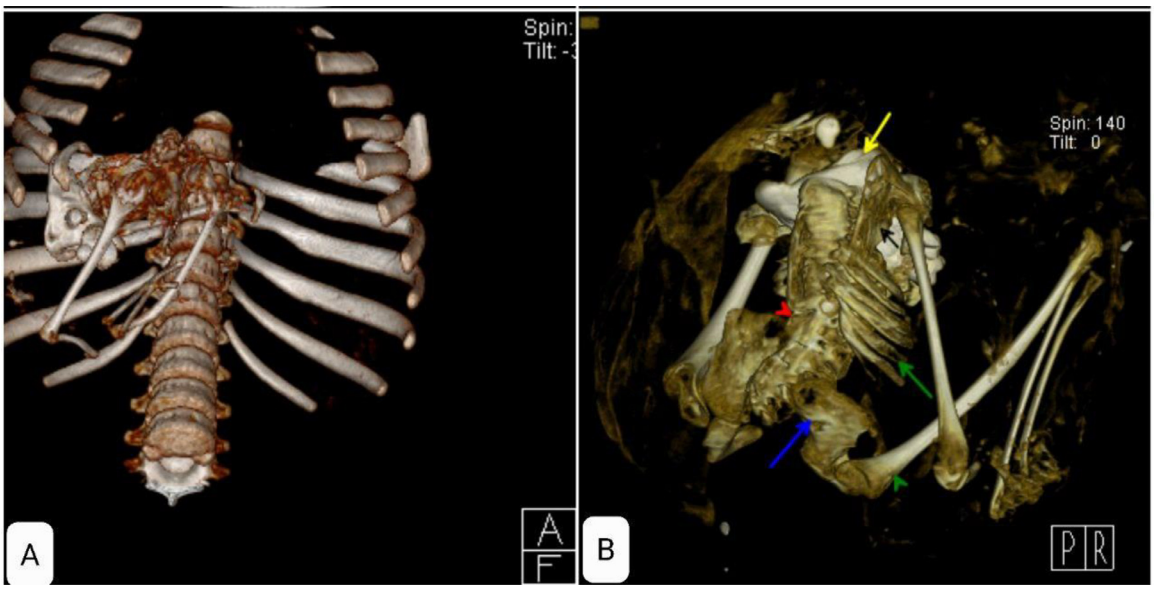

Fig. 3 – CT with 3D reformats showing an axial and perpendicular skeleton (A); further, a 3D specimen CT scan showing a vertebral column (red arrow), skull base (yellow arrow), scapula (black arrow), pelvis (blue arrow), ribs, and femur (green arrows) (B).

ferred to the intensive care unit and died shortly after surgery due to multisystem failure.

Discussion

The pathogenesis of FIF remains unclear; however, 2 theories have been proposed, i.e., the diamniotic monochorionic twins and teratoma theories. Willis et al. initially proposed that the diagnosis of FIF should be restricted to cases involving parts of the axial skeleton [1]. The presence of a spinal column and axial skeleton indicates that the embryonic development passed the primitive streak stage, which is consistent with the monozygotic twin theory [1,6]. Another study suggested diagnostic criteria for FIF, which are consistent with most commonly reported criteria of FIF, including (1) enclosed fetus within a distinct sac; (2) full or partial coverage of normal skin; (3) well-formed axial skeleton with a mature vertebral column; (4) grossly recognizable anatomical parts with systemic organization, including the upper and lower limbs, skin, prostate, and testicles; and (5) attachment to the host through a few relatively large blood vessels [6].

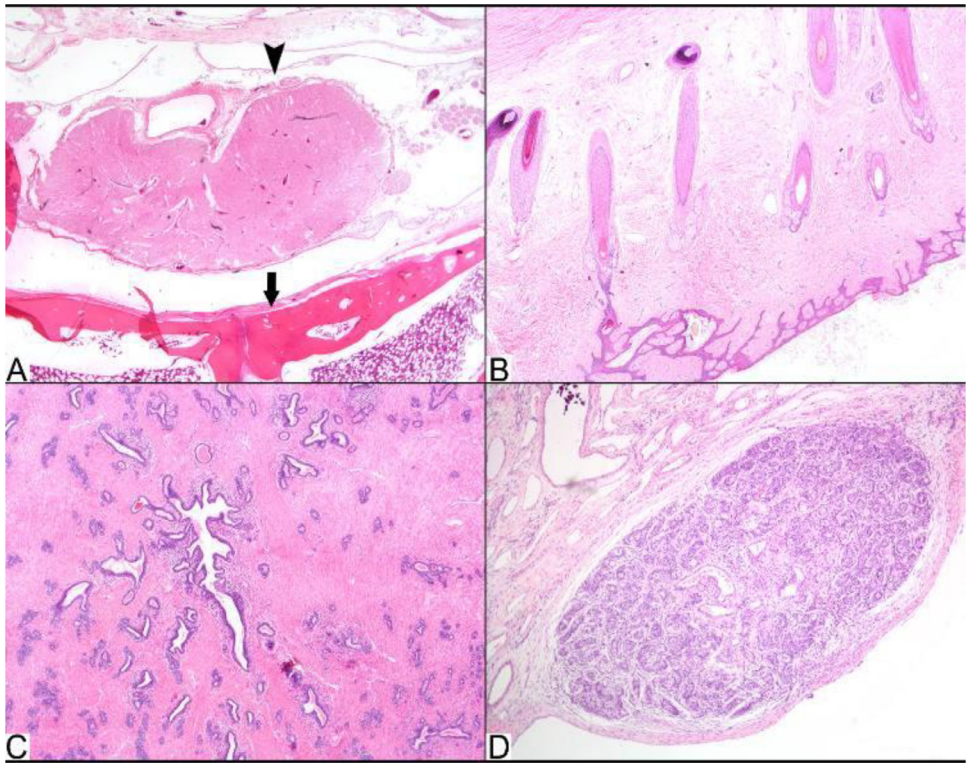

Fig. 4 – Histopathology showing a spinal cord and vertebral column (A), skin and subcutaneous tissues (B), prostate (C), and testicular tissue (D).

The teratoma theory suggests that FIF is an extreme form of highly organized teratoma with well-differentiated and mature organs [2]. Teratomas have a relatively lower degree of tissue organization without systemic organization regardless of the maturity of the contained organs.

Most cases of FIF are anencephalic, acardic, and have a vertebral column; however, 9%–24% of cases of FIF lack a vertebral column [5]. Moreover, 55% and 36% of cases involve central nervous system and genitourinary tissues, respectively, with cases presented with other tissues like gastrointestinal, pulmonary, adrenal, and pancreatic tissues [3].

In our case, all the criteria for FIF were met except for a large vascular supply and attachment to the host by a large vascular pedicle; however, Spencer suggested that only one or more features were required to consider FIF [6]. The presence of vascular pedicle has been suggested as a criterion since some cases of FIF are connected by both small and large vascular pedicles; contrastingly, teratomas are connected by multiple small vessels [6]. Although some cases of FIF have presented blood vessel-like structures, only 1 report describe specific vasculature and an aorta-like structure on CT [1].

Most cases of FIF are diagnosed during infancy. However, as observed in our case, limited access to medical care can contribute to delayed presentation. Generally, the most commonly reported symptoms of intra-abdominal FIF include abdominal distension with a palpable mass, vomiting, poor feeding, jaundice, and shortness of breath [3]. Specifically, the most frequent presentations of hepatic FIF, include palpable liver, failure to thrive, emesis, abdominal distention, and dyspnea [4,5]. Table 1 shows a comparison of all reported cases of hepatic FIF. Another case of FIF was found in the anterior abdominal wall but not intrahepatic. Interestingly, its liver tissue remained fused to the patient's own liver parenchyma [7]. All reported cases of FIF with a delayed diagnosis, as in our case, presented with progressive mass growth and a gradual increase in size. However, there was 1 case that showed a stable course without growth and was incidentally diagnosed in a 39-year-old patient [3]. Most cases of FIF have reported a single fetus in the mass, with only a few reports describing multiple fetuses [4].

In conclusion, this is the third reported case of hepatic FIF. Although FIF is rare, it should be considered as a differential diagnosis of growing abdominal masses in pediatric patients. Although an early diagnosis is usually established through routine antenatal imaging, many cases, including ours, are diagnosed late due to limited access to medical care. A multidisciplinary approach, including radiology, pediatric surgery, and pathology, is recommended for proper diagnosis and management. It is crucial to distinguish FIF from teratomas since teratomas have the potential to become malignant, and thus require close surveillance and monitoring of tumor markers. This case report supports the monozygotic twin theory of FIF and indicates that the diagnostic dilemma of FIF vs. teratoma can be solved through collaborative work between radiologists and pathologists since the suggested criteria have radiological and pathological aspects.

Table 1 – Comparison of reported cases of hepatic FIF.

	Our case	Magnus KG et al [4]	Al-Baghdadi R [5]
Gender	Male	Female	Male
Age of the diagnosis	8 y old	5 d old	4 mo old
Presentation	Growing abdominal mass with pain	Liver abnormality on prenatal US during the second trimester	Dyspnea, vomiting, and abdominal distention
Number of fetuses in the liver	1	2	1
Size of fetus	18 × 15 × 14 cm	Both are 3 cm	7 × 6 × 5 cm
Vertebral column presence	+	+	+
Outcome	Death on day 3 after surgery due to liver infarction and multisystem failure	Good after surgery	Not reported

Patient consent

Consent for the publication of the patient medical information was obtained.

Ethics approval and consent to participate

Consent for the participation and ethical approval was obtained.

Authors' contributions

SF, RA, and GH contributed in the patient's radiological reporting and manuscript writing. MT performed the histological examination of the specimen and participated in the manuscript writing, NP and MH contributed in the surgical part and manuscript writing. All authors read and approved the final manuscript.

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