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Case Report

Intra-abdominal retroperitoneal fetus in fetu: A case report ☆

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ABSTRACT

Fetus in fetu (FIF) is a rare abnormality where a vertebrate parasitic fetus develops inside the body of another normally developing fetus. It is distinct from teratomas, tumors composed of cells from multiple germ layers and have malignant potential. Symptoms of FIF arise from the mass effect, causing abdominal distension, feeding difficulties, and pressure effects on organs. FIF is commonly found in the retroperitoneal region but can also occur in other locations. It often includes certain organs such as the vertebral column, limbs, central nervous system, gastrointestinal tract, vessels, and genitourinary tract. Early diagnosis of FIF by ultrasound, computed tomography, and magnetic resonance imaging can improve patient outcomes. Surgical resection is the primary treatment approach, aiming to alleviate symptoms, and molecular analysis helps differentiate FIF from malignant teratomas. Regular follow-up is necessary due to the potential recurrence of teratomas.

Introduction

Fetus in fetu (FIF), derived from the Latin term "fetus within the fetus," is a rare congenital anomaly characterized by a vertebrate fetus developing inside the body of its twin sibling [1]. This condition arises due to abnormal embryogenesis [2]. The phenomenon of the FIF was initially documented by Meckel in 1800, and later defined by Willis in 1953 as a mass containing a vertebral canal and often accompanied by other solid organs or upper and lower limbs [3]. The reported cases of FIF remain few and far between no more than 200 to date [4]. There appears to be a male predominance, with a ratio of approximately 2:1 [5].

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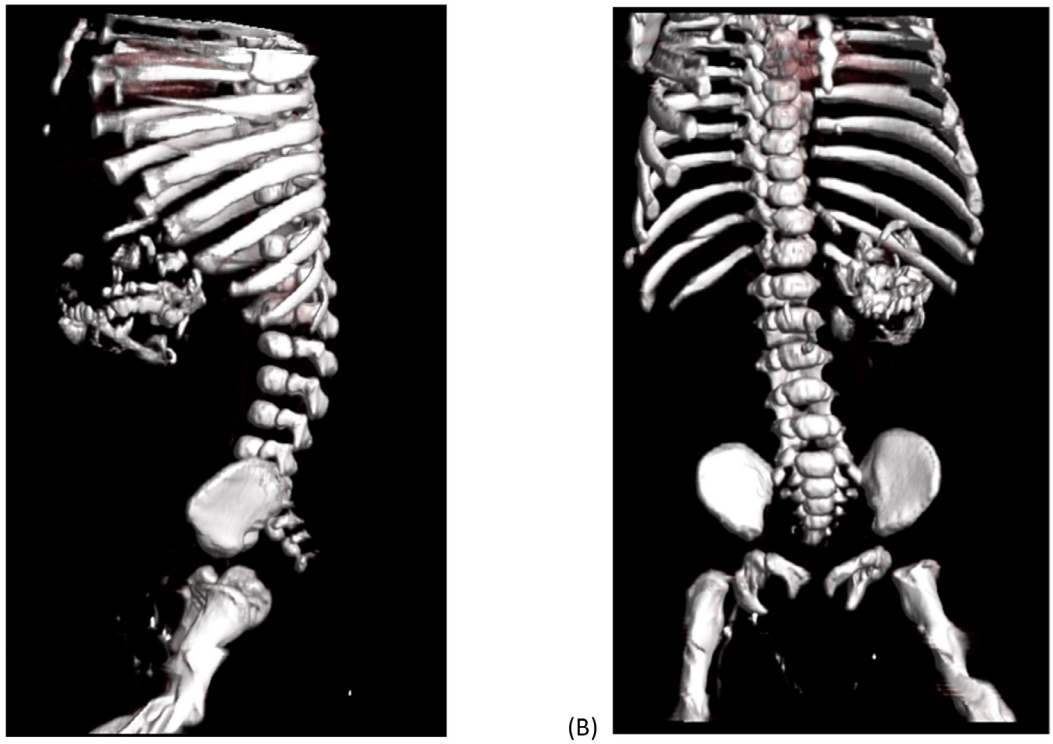

Fig. 1 – (A and B) of 3D VRT (volume rendering technique) images demonstrate multiple bony fragments as well as spine, ribs, upper, and lower limbs of the parasitic fetus.

Case presentation

A 2-month-old male infant, born through spontaneous vaginal delivery presented to the outpatient department of the hospital with the complaint of a lump on the left upper quadrant of the abdomen. Physical examination showed approximately 8 × 12 cm solid and semi-mobile mass, firm to hard consistency with no tenderness on the left hypochondrium area. The swelling extended into the left lumbar and epigastric regions. Ultrasonography revealed abdominal mass. A CT scan was done for the patient in the radiology department. The contrast-enhanced abdomen CT scan demonstrated a well-defined heterogeneous lesion in the left upper quadrant, with resultant displacement of bowel loops as well as major abdominal vessels. The lesion had soft tissue, fat, fluid and bone components predominantly with apparent spine, ribs, facial bones, upper and lower limbs, scapula (Fig. 1A, B and Fig. 2A-C).

After a thorough assessment and stabilization, the patient underwent surgery under general anesthesia. The surgery involved a left-side transverse supraumbilical muscle-cutting incision. A mass was discovered in the retroperitoneum, which was adhered to surrounding structures and the pancreas. The mass was contained within a complete membranous sac and was supplied by major vessels originating from the aorta. It was successfully separated from the surrounding structures and excised (Fig. 3A). The abdomen was closed in layers, and an abdominal drain was inserted. The patient's postoperative recovery was normal.

Upon opening the sac, an incompletely formed fetus was observed. The fetus exhibited a rudimentary head with hair (anencephaly), a well-developed spine, upper limbs with fingers, and lower limbs with feet and toes (Figs. 3B and C). The patient is currently following up and (is doing well with no complaints.)

Discussion

FIF is a rare abnormality associated with abnormal embryogenesis in a diamniotic, monochorionic pregnancy, in which a vertebrate fetus is enclosed within the body of a normally developing fetus [6].

Despite the 2 pathogenetic controversies regarding whether FIF represents a distinct pathogenetic entity, known as the parasitic-twin theory, or is part of the spectrum of fetiform teratomas, referred to as the teratoma-spectrum theory, it is crucial to differentiate FIF from teratomas. This differentiation holds significant clinical importance due to the potential malignancy associated with teratomas. Even mature cystic teratomas carry a risk of malignancy ranging from 3.5% to as high as 6.67%, while immature teratomas should be considered malignant [7,8].

Pathologically, FIF is characterized by highly differentiated tissue surrounding a vertebral skeleton. In contrast, teratomas are composed of discordant aggregations of pluripotential cells lacking systemic organization, incorporating differenti-

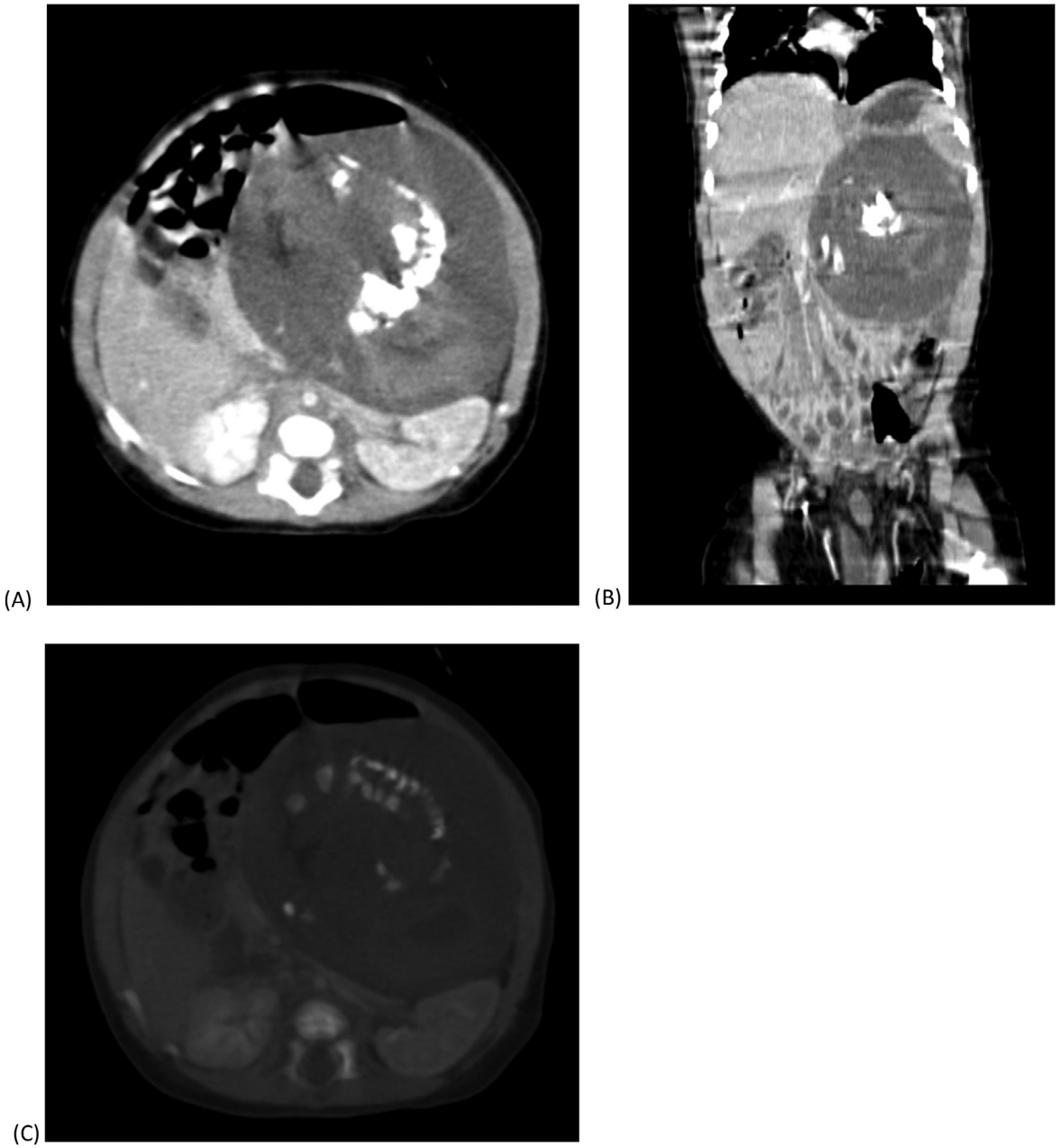

Fig. 2 – (A-C) Selected axial, coronal soft tissue window, and axial bone window images demonstrate well-defined heterogeneous lesion in the left upper quadrant, resulting in the displacement of bowel loops as well as major abdominal vessels.

ated cells from multiple germ layers. Consequently, teratomas can develop into fully mature tissue and possess malignant potential [9]. The growth of the FIF initially mirrors that of the host, continuing as long as there is sufficient blood supply. However, beyond a certain size, further growth comes to a halt [10].

The typical presentation of FIF is observed before the age of 18 months, although the oldest reported case involved a 47-year-old adult. Symptoms associated with FIF primarily arise from its mass effect, leading to manifestations such as abdominal distension, feeding difficulties, pressure effects on the renal system, and dyspnea [11].

As in our case, the patient initially presented with abdominal swelling and a palpable mass. Following physical examination, the pediatric surgeon raised suspicions of either a Wilms tumor or neuroblastoma as potential underlying causes for the observed symptoms.

The retroperitoneal region is the most frequently observed location for FIF, accounting for approximately 80% of cases. However, FIF has also been reported in other locations, including the scrotum, nuchal region, and intracranial area [12–15].

In our case the mass was identified in the left retroperitoneal region, leading to displacement of the abdominal vessels and bowel loops towards the right side.

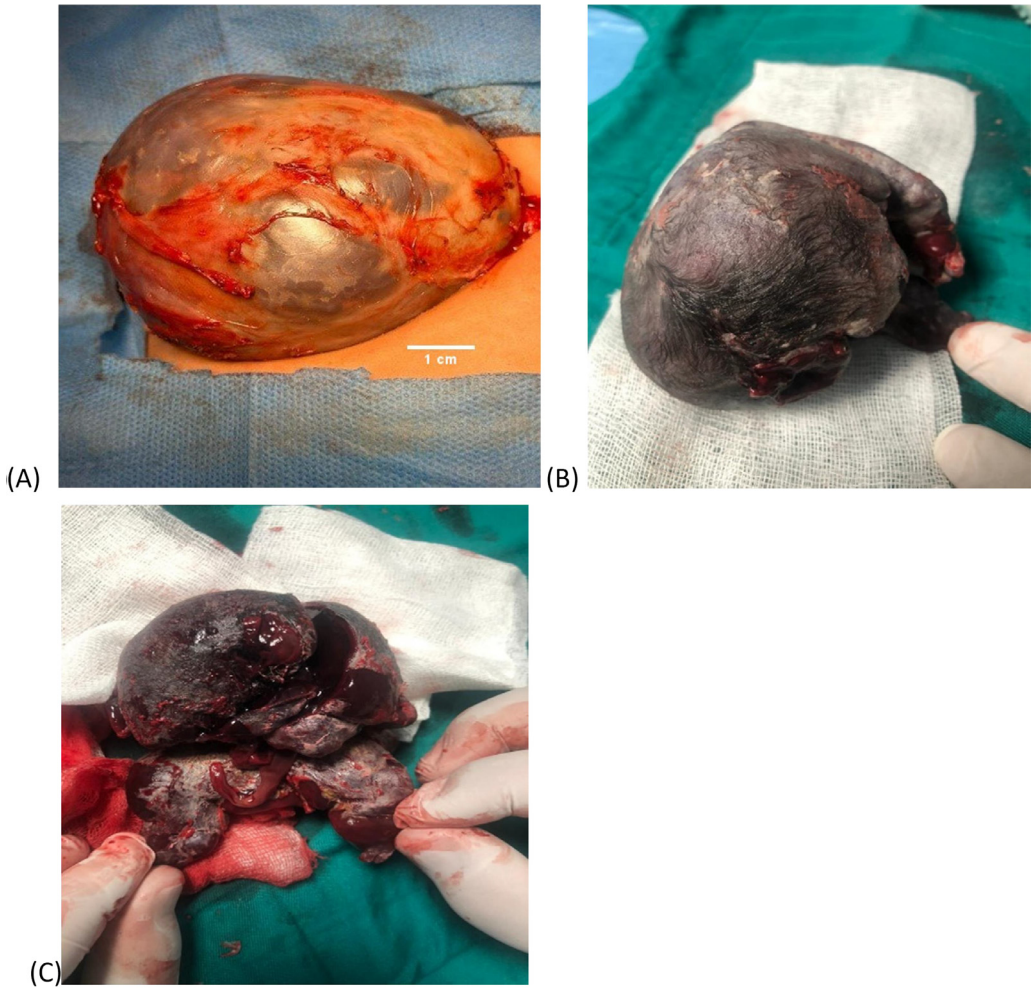

Fig. 3 – (A-C) Selected images were taken during the surgery, showing the pre-excised mass within its membranous sac (A). Postexcised images revealed a mass in which an incompletely formed fetus was observed. The fetus exhibited a rudimentary head with hair (anencephaly), a well-developed spine, upper limbs with fingers, and lower limbs with feet and toes (B and C).

FIF commonly exhibits the inclusion of certain organs, with reported frequencies as follows: vertebral column (91%), limbs (82.5%), central nervous system (always anencephalic) (55.8%), gastrointestinal tract (45%), vessels (40%), and genitourinary tract (26.5%). Notably, the presence of the heart in FIF has been rarely documented, with only 1 to 2 reported cases to our knowledge [16].

With the advancements in ultrasonography, early detection of FIF has been associated with enhanced patient outcomes. However, ultrasound findings may not always be conclusive, often leading to a broad range of differential diagnoses. Fortunately, the increased utilization of CT and MRI has significantly improved the capacity to narrow down to a specific diagnosis. Upon reviewing the available radiologic literature, only 1 case involving 3-D reconstruction demonstrating the rare phenomenon of FIF has been reported. The use of multidetector CT technology in FIF can argue for an accurate preoperative diagnosis, facilitating the required surgical planning for such complex cases. Advanced imaging techniques offer improved resolution and unique insights into the spatial relationship of the mass with its surrounding structures. This, in turn, enhances the surgeon's ability to develop a more informed operative plan [17–19]. Plain abdominal X-ray can be a useful tool in the diagnosis of FIF. Approximately half of the cases exhibit findings on X-ray that reveal the presence of the vertebral column and axial skeleton [16,20].

The primary treatment approach for FIF is complete surgical resection, except in cases where the mass is adherent to the host's organs. The main objective of resection is to alleviate or prevent symptoms associated with an intra-abdominal mass [21,22]. Molecular analysis employing informative genetic markers of chromosomes 14 and 15 reveals genetic concordance between the host infant and the fetiform mass. This supports the diagnosis and aids in distinguishing FIF from malignant teratomas. While the prognosis for FIF is generally more favorable than that of cystic teratomas, the presence of immature elements necessitates diligent clinical, radiological, and serological (alpha-fetoprotein) follow-up. Pre- and post-

operative alpha-fetoprotein levels remain within the normal range. However, considering the potential recurrence of malignant teratomas following FIF resection, the patient undergoes regular monitoring of tumor marker levels and cross-sectional imaging as part of the follow-up protocol [23].

Conclusion

We presented a rare case of FIF, accentuating the significance of accurate diagnosis and surgical intervention. This anomaly remains rare, and our case highlights the successful surgical excision of a well-defined heterogeneous lesion representing the parasitic fetus. Differential diagnosis of the FIF from teratomas is crucial, and advanced imaging techniques aid in preoperative planning. Complete surgical resection, molecular analysis for genetic concordance, and long-term follow-up are essential for optimal management. Further research and reporting are needed to enhance understanding and improve patient outcomes.

Patient consent

Written informed consent for publication of their case was obtained from the patient parents.

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