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Retroperitoneal fetus in fetu in a 65-year-old man

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ABSTRACT

Fetus in fetu (FIF) is a rare congenital anomaly in which a malformed fetus is incorporated within the body of its twin. It was first described in the late 18th century and has an incidence of 1:500,000 live births. In most cases, the diagnosis is made in infants or young adults. To date, the oldest patient reported in the literature was 47 years old. We describe the case of a 65-year-old patient with FIF, now the oldest reported in the literature. Our patient meets all the diagnostic criteria for FIF, including the presence of a limb in advanced formation inside the lesion. The treatment was surgical excision. FIF should be considered in the differential diagnosis of abdominal masses, typically recognized in infancy. Symptoms arise from mass effects. Surgical resection should be performed due to the potential for malignant transformation.

Keywords: *Fetus in fetu; Surgery; Teratoma*

INTRODUCTION

Fetus in fetu (FIF) is a rare condition that was first described by Johann Friedrich Meckel in the late 18th century[1], with a worldwide incidence of 1:500,000 live births[1-3]. The pathogenesis of FIF is still under debate because of its similarity to mature teratomas. The most common theory is the "included-twin" theory, in which a diamniotic, monochorionic, and monozygotic twin is incorporated within the body of the host twin[1,4], most commonly in the retroperitoneal cavity. The usual clinical presentation consists of a gradually-increasing abdominal mass[2]. By definition, FIF has a central axis with structures surrounding this axis, suggesting that it has undergone the initial stage of organogenesis[2,5].

Most cases were described in neonates and children, with only a few cases reported in adults. To the best of our knowledge, this is the oldest reported case of FIF in the literature.

CASE REPORT

A 65-year-old man was admitted with upper gastrointestinal bleeding secondary to rupture of esophageal varices, as well as self-reported weakness and anorexia. He had arterial hypertension, dyslipidemia, and chronic liver disease due to nonalcoholic steatohepatitis. Current medications included simvastatin, ramipril, hydrochlorothiazide, lactulose, pantoprazole, and propranolol. The patient underwent coronary angioplasty for acute myocardial infarction in 2011 and had no previous operations.

Abdominal ultrasonography revealed a large lesion in the retroperitoneum with irregular contours. Computed tomography (CT) and magnetic resonance imaging (MRI) revealed a lesion measuring 26 × 18 × 21 cm with a volume of 5,103 cm^3, foci of calcifications, a long bone structure with epiphysis, diaphysis, and metaphysis, and the presence of adipose tissue (Figure 1).

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The mass was anterior to the aorta, inferior vena cava, and portal vein. The mass compressed but did not infiltrate adjacent organs and structures such as the stomach, liver, and kidneys (Figure 2). The main diagnostic suspicion was FIF, and surgery was indicated.

A midline laparotomy was performed. Due to the volume of the mass and the difficulty in identifying adjacent vessels and structures, it was decided to open the mass and remove its contents. A large amount of fatty liquid, hair, well-formed bones, and teeth were removed. An inadvertent small lesion of the posterior wall of the superior mesenteric vein was appropriately sutured. After risk-benefit analysis, a small part of the cystic wall was left behind due to firm adherence to the portal vein and vena cava. After surgery, the patient was admitted to the intensive care unit (ICU).

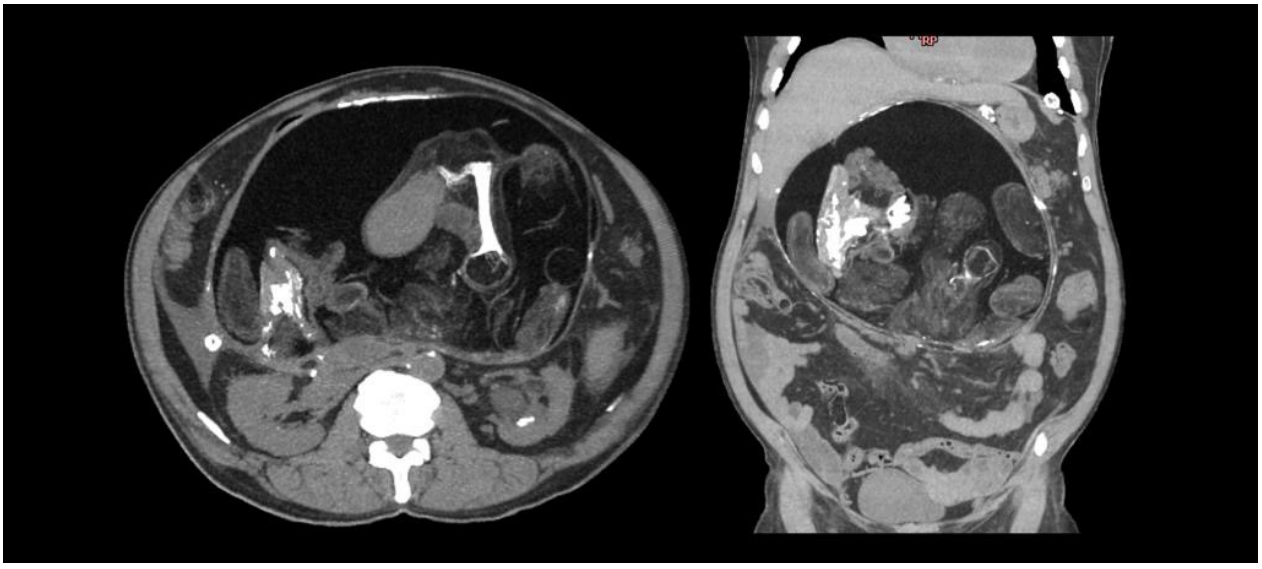

Figure 1: Computed tomography showing a large mass measuring 26 × 18 × 21 cm (volume of 5,103 cm^3) and containing a long bone structure with epiphysis, diaphysis, and metaphysis. A large adipose tissue is also documented.

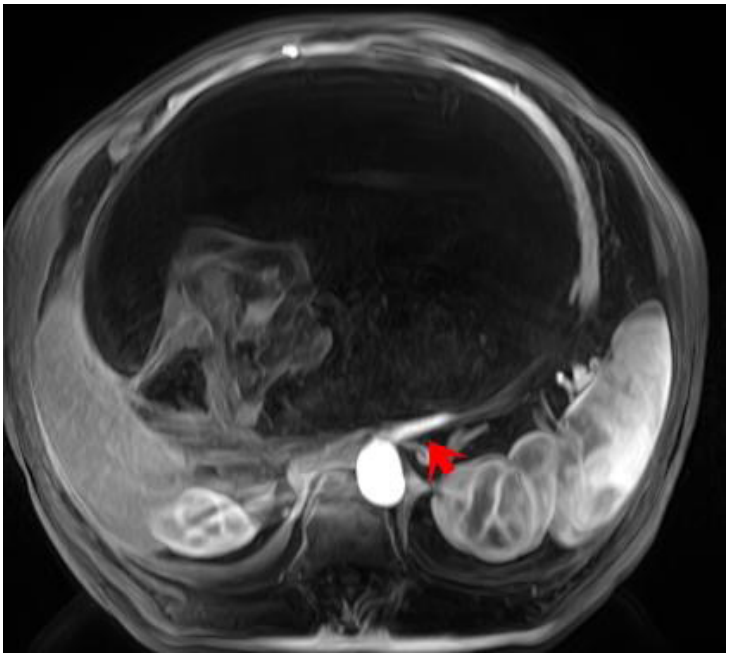

Figure 2: Computed tomography showing a mass anterior to the aorta (red arrow – celiac trunk), inferior vena cava, and portal vein. The mass is compressing adjacent structures such as the stomach and liver.

Postoperative recovery was slow, with complete norepinephrine withdrawal on postoperative day (POD) 7. The patient was extubated on POD 10. During this period, the patient received nasoenteric tube feeding. He was discharged from the ICU on POD 13. A total of 1,200 mL of serous fluid was percutaneously drained from the right abdominal flank under CT guidance. The patient subsequently improved and was discharged on POD 46.

DISCUSSION

Two theories may explain the development of FIF. The main one is the "included-twin" theory, in which a malformed, anencephalic twin (parasite) lacking several organs is incorporated within the body of the host twin[2]. The other theory defines FIF as a well-differentiated and highly organized teratoma[6].

Willis[7] defined FIF as a mass containing a vertebral axis with other organs or limbs surrounding this axis. In the absence of a vertebra, the presence of identifiable limbs and long, well-formed bones may also characterize FIF[5]. Recently, Kumar et al.[8] proposed a review of this concept and suggested that advanced organogenesis without the presence of a spinal column is enough to characterize FIF. This fetiform mass is usually surrounded by a yolk sac-like tissue with liquid or sebaceous content[2,5]. The presence of vertebrae indicates that FIF has undergone the gastrulation stage, given that the neural tube develops before the skeleton[5]. Conversely, teratomas are formed by the disorganized accumulation of pluripotent cells, without systemic organization or vertebral segmentation[1,9], clearly distinct from the case reported here.

FIF is a very rare congenital anomaly typically developed during infancy or early childhood. Most cases are diagnosed during the neonatal period or in infancy, with fewer than 20 cases reported in adults[8]. To date, the oldest patient reported in the literature was 47 years old[10]. Our patient was 65 years old. The diagnosis was established by CT findings of a large mass in the retroperitoneal space associated with a long bone structure with epiphysis, diaphysis, and metaphysis. The diagnosis of teratoma was ruled out by the presence of limb bones around the vertebral axis. A literature review reported the presence of spine in 76%-91% and limbs in 84% of cases[5,8]. Lower limbs are usually more developed than the upper limbs[3]. FIF is anencephalic and heartless[5]. Despite the diagnosis being established based on histopathological findings[1], criteria for adequate differentiation between teratoma and FIF are still lacking, considering that well-developed tissues are found in both conditions. In 80% of cases, FIF develops in the retroperitoneum, but it may occur in other regions, such as the mediastinum, cranial cavity, neck, back, sacrococcygeal region, scrotal sac, and oral cavity[1,3].

FIF symptoms result from mass effects, thus they vary according to the affected site. Even though FIF is a benign condition, the treatment of choice is complete excision, including the entire membrane[5]. Although rare, there is a risk of malignant transformation, thus the need to ensure the absence of remaining tissue[8]. Due to the presence of immature tissue, follow-up with measurement of serum levels of beta-human chorionic gonadotropin and alpha-fetoprotein plus CT or MRI is recommended[5]. In conclusion, our case meets the established criteria for retroperitoneal FIF in a 65-year-old patient, the oldest age ever reported in the literature. Preoperative diagnosis was based on CT findings, which showed the presence of a spine and limbs, revealing advanced organogenesis. The treatment of choice was surgical resection.

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Conflicts of interest

The authors declare no conflicts of interest.

Humans and animal rights

This article does not contain any study with animals performed by any of the authors.

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