Fetus-in-fetu presenting as a cryptorchid testis and abdominal mass: A report of a case and review of the literature

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Fetus-in-fetu (FIF) is a rare congenital mass containing fetal tissue thought to arise from a parasitic monozygotic twin within the host twin. This rare lesion often presents as an abdominal mass, and most often presents in young infancy.

1. Methods

We present the case of a 3-week old term neonate found to have an abdominal mass concerning for FIF. He was born at term via Cesarean section, and on routine physical exam, was found to have a right-sided abdominal mass and undescended right testicle. On workup, calcifications thought to correspond to an axial skeleton were seen on ultrasound (Fig. 1). This prompted additional imaging and on magnetic resonance imaging (MRI) (Fig. 2) an organized mass containing two femurs and a vertebral column was identified within a cystic structure. There was associated mass effect on the IVC, right kidney and liver. The pre-operative alpha-fetoprotein (AFP) level was 18,423 ng/mL.

At laparotomy, a large, circumscribed abdominal mass was present in the right upper quadrant (Fig. 3). It was removed from the abdominal cavity and was found to be associated with the vas deferens and gonadal vessels, appearing to be closely associated with the right-sided cryptorchid testis. Pathology revealed an intratesticular fetus-in-fetu. Fetus-in-fetu is a rare condition where a well-developed mass believed to arise from a monozygotic, parasitic twin, is found within its sibling. First reported by Dr. Meckel in 1800, this is thought to occur in 1 in 500,000 live births with less than 100 cases reported in the literature [1,2]. FIF is differentiated from mature teratoma by the presence of an axial skeleton with metameric segmentation and well-differentiated tissues. FIF is a benign disease, however surgical excision continues to be the treatment of choice, in order to confirm diagnosis by pathology and to exclude malignant teratoma. Fetus-in-fetu is a rare of congenital lesion that can present as an abdominal mass. Due to the high prevalence of retroperitoneal location, a FIF should remain on the differential for an infant with an abdominal mass and ipsilateral cryptorchid testis.

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mature teratoma by the presence of an axial skeleton with metameric segmentation and well-differentiated tissues [2–4]. Based on Hoeffel et al.’s review of reported cases, 91% have a vertebral column, limb development is present in 82.5%, and 55% will have an anencephalic central nervous system [5]. The vast majority, 80%, are located within the retroperitoneum [5]. Patients are diagnosed in infancy, with 89% of cases occurring prior to 18 months [5]. Abdominal pain was the chief complaint on presentation in 70% of cases [5].

Differentiating these lesions from a teratoma is paramount, as retroperitoneal teratomas can harbor a malignancy rate as high as 10%. On pathology, mature teratomas are tumors with a disorganized array of pleuripotent cells representing all three germ layers [6,7]. In FIF, the tissue growth is more organized, and must show an axial skeleton with metameric segmentation. If limb buds in appropriate arrangement are present, this supports the diagnosis. The karyotype of the mass should be identical to that of the host [7]. The presence of an axial skeleton is an important component in diagnosis; in order for this to be present, the fetus must develop past the primitive streak stage, a characteristic not present in mature teratoma [4,8,9]. The level of organization of the mass can be so advanced, that a morphology showing a homunculus can be seen; spontaneous movement of the FIF in utero has been documented in rare reports [4].

The maturation of a FIF is an area with controversy, especially with regards to its origins and development, with several proposed theories. The most accepted theory is the “monozygotic twin theory,” where the FIF is a diamniotic, monochorionic, monozygotic twin that becomes enveloped by the host twin during development [10,11]. Another proposed mechanism involves one blastocyst implanting on another blastocyst, subsequently developing as an inclusion body [10]. Kaufmann et al. propose that this process occurs as a result of twin–twin transfusion syndrome, with the demise of the non-dominant twin resulting in the envelopment of deceased twin during the third week of development [6]. Whatever the development, it is evident that fetal demise and arrest of growth occurs during its development, still another feature distinguishing it from the continuous growth potential of mature teratomas [11].

FIF is a benign disease, however surgical excision continues to be the treatment of choice, in order to confirm diagnosis by pathology and to exclude malignant teratoma. Only one case of malignant recurrence of FIF is present in the literature. The authors suggest that this occurrence of malignancy resulted from the FIF surrounding membrane being left in situ. As a result, Hopkins et al.

**Fig. 1.** Ultrasound image demonstrating axial skeleton.

**Fig. 2.** MRI images demonstrating calcified lesion with mass effect. a) coronal images; b) cross sectional imaging with arrow demonstrating fetal structure (the arrow is pointing at the thigh and femur within the cystic mass); c) cross sectional imaging with arrow demonstrating axial skeleton (the arrow is pointing at the fetal vertebral column within the cystic mass).
suggest, monthly monitoring of AFP and HCG for one year to ensure return to baseline [12].

3. Conclusion

FIF is a rare congenital anomaly that can present as an abdominal mass. Although many theories exist regarding the development of this lesion, treatment remains surgical excision in order to exclude malignancy. Pathologic differentiation may be difficult, but rests on the presence of organized development around an axial skeleton. Due to the high prevalence of retroperitoneal location, a FIF should remain on the differential for an infant with an abdominal mass and ipsilateral cryptorchid testis.

References