Fetus in Fetu: Case Report of a 17 Years Old Male and Literature Review

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Abstract: Background: Fetus in fetu is an uncommon anomaly in pediatric age group. There have been controversies regarding its differentiation from well differentiated teratoma. It is defined as a monzygotic twin incorporated into abdomen of sibling during development. These kids are often overlooked in the differential diagnosis of an abdominal mass. Very much similar to teratoma, it’s a rare benign condition. Case presentation: Described here is a clinical presentation of a 17 years old male with an abdominal mass without any symptom came for his own concern regarding the mass in medicine outpatient department. His ultrasound was done but mass was so calcified that image could not be appreciated so underwent abdominal computerised tomography and magnetic resonance imaging which favoured the diagnosis of fetus in fetu. Conclusion: Fetus in fetu is a rare entity that typically presents in infancy and early childhood. It should be differentiated from a teratoma because of the teratoma’s malignant potential. Preoperative diagnosis is based on radiologic findings. The treatment is operative to relieve obstruction, prevent further compression and possible complications. Complete excision allows confirmation of the diagnosis and lowers the risk of recurrence.

1. Introduction

Fetus in fetu (or foetus in foetu) is a developmental anomaly where a mass of tissue resembling a fetus develop inside the body. The earliest example of this phenomenon was first described in 1808 by Geogre William Young.[1] Various theories of origin have been proposed concerning “fetus in fetu”. One theory opines that the mass begins as a normal fetus but becomes enveloped inside its counterpart twin.[2] The other theory finds this mass a highly differentiated form of dermoid cyst, itself a highly differentiated form of mature teratoma.[4] Parasitic twin theory regards a parasitic twin foetus growing within its host twin very early in a monzygotic twin pregnancy, in which both foetuses share a common placenta, one foetus wraps around and envelops the other. The enveloped twin becomes a parasite, in that its survival depends on the survival of the host twin, by drawing on the host twin's blood supply. The parasitic twin is anencephalic and lacks some internal organs, and as such is unable to survive on its own. As the host twin has to "feed" the enveloped twin from the nutrients received over a single umbilical cord, they usually die before birth.

2. Case Report

17 years old male presented to our hospital in medicine outpatient department with complain of abdominal mass. The boy was 5 feet 2 inch height, lean, afibrile, non-icteric, non-pale, acyanotic, no clubbing or oedema. Vitals were normal with the peripheral pulses well felt. Chest including cardiovascular examination was normal. There were no complaints as such except concern for abdominal mass. Abdominal examination revealed diarrihiva of recti and everted umbilicus. A non tender, central mass of firm to hard consistency about 10 X 8 cm, oval with regular well circumscribed margins extending from xiphisternum to umbilicus felt. Movement with respiration, no impulse on coughing but visible peristalsis could be seen. No associated hepatic or splenic enlargement or extension to internal organs found with no compressive signs. Detailed history taking revealed the patient noticing this mass since age of 4-5 years of age but with no difficulty in activities of daily living. Patient’s mother gave history of undue abdominal enlargement as compared to her other 2 pregnancies when this patient was in utero. She also states that while this patient was new born, the mass was bigger as compared to abdominal girth the protuberance of which reduced gradually to this size. The patient had normal sexual development for the age. All other siblings were completely normal with no family history of twin pregnancy or any similar symptoms. Thus, the patient was completely doing well and was himself curious about his lump.

Pertinent lab investigations done, the positive findings being 1) eosinophil 12.7% (N:1-6) 2) raised CEA value of 9.29ng/ml (N:5-6.5)

His abdominal X ray was suggestive of calcified mass with ill-defined long processes arising from the mass suggesting bony growth. The ultrasound whole abdomen revealed gross calcification. The NCCT abdomen and MRI suggested “a well-defined round fairly large midline hypointense lesion (18.0x14.8 cm) showing hypo intensity with hyperintense background on T2W, and loss of signal intensity on T1W with areas of low signal intensity s/o calcification in abdominal cavity causing compression and displacement of adjacent organs. Sagital and axial images corroborated with CT revealing calcification/bone taking a special pattern in form of spine and limbs, S/O FETUS IN FETU.
The x-ray abdomen erect posture suggestive of calcified intraabdominal structure of heterogenous morphology taking shape of poorly developed limbs, suggesting fetus in fetu

Contrast enhanced CT sagital and axial images corroborated with CT showing calcification/bone taking a special pattern in form of spine and limbs, S/O FETUS IN FETU.

3D recall MRI image clearly showing the fetus in fetu

3. Discussion

Fetus in fetu (FIF) is extremely rare pathology (1/500,000 live births) [3] in which a malformed foetus is located in the body of its twin. The liberal definition of FIF was proposed by Gonzalez-Crussi [13], who defined FIF as ‘high organotypic development and presence of a vertebral axis with arrangement of tissue around this axis’. In most cases, there is a single parasitic twin. But rarely, more than 1 parasitic twins can be observed in the host body. Studies of genetic markers, such as blood group, sex chromosome constitution, protein polymorphisms, and DNA marker, suggest that host infants and their fetiform mass are genetically identical. Eighty-nine percent of FIF lesions are noted before 18 months of age [14]. Most FIF are located retroperitoneally along the ventral midline, while other rare reported sites include the cerebral ventricles, liver, pelvis, scrotum, and mediastinum. Although FIF is a benign condition, the mass may compress the surrounding organs

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and tissue. Therefore, symptoms of FIF are primarily due to its mass effect such as abdominal distension, feeding difficulty, emesis, jaundice or pressure effects on the renal or respiratory system.

To qualify as FIF, at least one of the following characteristics must be present: a) mass enclosed within a distinct sac, b) partially or completely covered by skin, c) grossly recognizable anatomical features and d) attached to the host by a pedicle containing a few relatively large blood vessels. [18] Our patient also had an abdominal mass completely covered by skin under anterior abdominal wall, easily seen and felt thus fulfilling the criteria for being FIF. Ultrasonography and plain radiography can be used to achieve a diagnosis of FIF. Computed tomography scan and magnetic resonance imaging give a more accurate diagnosis and defines the relation of the FIF with the other intra-abdominal structures. FIF is usually overlooked in the differential diagnosis of a newborn abdominal calcification. In some cases, FIF may be confused with meconium peritonitis, which is commonly associated with calcifications. [19] Other causes of calcification include neuroblastoma, adrenal hemorrhage, and viral infection. In the above case, various lab investigations did not support any infective background. It is also important to differentiate between a retroperitoneal teratoma and a retroperitoneal FIF because the former have more than 10% malignancy rate. Until now, only one case of malignant FIF has been reported [16]. Clinically, FIF can be differentiated from teratoma by the presence of vertebral bodies and limbs which was seen in our case as having evidence of calcification/bone taking a special pattern in form of spine & limbs.

As described above, many authors agree that FIF corresponds to a monochorionic, monzygotic twin contained with the host. In the study of Miura et al [27] the investigators demonstrated that host infants and their foetus shared the same genotypes, further supporting the monzygotic theory. These findings also confirmed a separate aetiology for FIF as compared to teratoma. However, our knowledge of early molecular and genetic events that regulate embryo development and organogenesis is rudimentary. The possible association between FIF and highly differentiated teratoma is still controversial. Some investigators hypothesized that FIF represents a well-differentiated and highly organized teratoma [19]. In other words, FIF and teratoma may share a causal/pathogenetic mechanism. There are several observable phenomena support the teratoma theory. First, FIF are observed in the same sites as teratomas, including retroperitoneum and ovaries. Second, FIF can be associated with a teratoma. Retroperitoneal teratoma formation after FIF resection has also been reported. [19] A simple monozygotic (monochorionic, diamniotic) twin theory may be difficult to explain these phenomena. Third, there have been many reports of invertebrate teratomas containing well-developed fettiform structures, including brain-like tissue with ependymal- lined ‘ventricles’ and spinal cord with a central canal. [22] As there are many similarities at the histological level, and considerable overlap between FIF and teratomas, establishing the true nature of FIF is of great interesting.

The recommended treatment for FIF is surgical excision. Because the final diagnosis of FIF is not made until pathological analysis, all parts of the mass should be removed to prevent malignant recurrence. Postoperative follow-up with screening for the tumor markers β-HCG and AFP is often used and is further supported on the basis of malignant recurrence of FIF. The detection of raised CEA levels generally indicates advanced malignant disease. Therefore, the raised CEA level in our first patient is of great concern. Whether this association of abnormal CEA level is a manifestation of the FIF or an incidental finding is unclear based on our case. Further studies are needed to establish the significant of this phenomenon.

4. Conclusions

In conclusions, the cases presented in our report met all the accepted criteria of an abdominal FIF. The pre-operative diagnosis of FIF is based on the observation of vertebral column or limbs in a mass on imaging modalities. The treatment of choice for FIF is complete resection. Future research efforts should be made to establish the true nature of FIF. Further studies to determine the possible association between FIF and highly differentiated teratoma are also warranted.

References


