

## Fetus-in-Fetu: A Rare Entity

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### Abstract

**Background :** Fetus in fetu is a rare congenital anomaly in which a malformed fetus or parasitic fetus grows within the body of its twin. It is a parasitic twin of a Diamniotic, monozygotic twin. Fetus in fetu is often overlooked in the differential diagnosis of an abdominal mass in infants and children. Teratoma is one of the closest differentials and must be excluded. Unlike teratomas, fetus in fetu is a benign disorder. The objective of our study is to describe the sonographic findings of an extremely rare pathology. We therefore the present diagnosis, pathology, management of fetus-in-fetu.

**Case Presentation:** On 6th June 2021, a two years boy was brought by his parents to Chittagong Medical College Hospital (CMCH) for treatment with the complains of abdominal mass, occasional abdominal pain and loss of appetite. The patient was evaluated radiologically by plain radiographs, USG and pre & post contrast CT scan of the abdomen. All radiological modalities showed a mass in the left lumbar region having internal bony structures like-long bone, vertebra and teeth. CT scan played the vital role in clearly demonstrating the bones and confirming the presence of vertebra. Radiologically a diagnosis was made as fetus-in-fetu and the possibility of Teratoma was excluded by the presence of vertebra. The patient underwent surgery and the mass was completely excised. A deformed parasitic fetus enclosed in a complete capsule was removed from the patient's abdominal cavity. Autopsy of the specimen showed deformed limbs, presence of vertebra, long bone, other soft tissue, organogenesis around an axis and histopathology confirmed the presence of vertebra, bone and bone marrow, cardiac muscle, nerve tissue, intestine, renal tubules with glomerulus, lymphoid tissue, respiratory epithelium and skin. All findings were compatible with fetus-in-fetu.

**Conclusion :** Although fetus in fetu is a rare condition, correct diagnosis using imaging can be made before surgery. It should be considered as a differential diagnosis for lump abdomen especially in infant. Complete excision is curative.

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### Introduction

Fetus in fetu is a rare disorder. The incidence is 1 per 500,000 births.<sup>1</sup> with less than 200 cases reported in the world to the best of our knowledge.<sup>2</sup> The present study is an effort to present a rare case of fetus in fetu with review of the available literature.

We present a case of fetus-in-fetu in a 02 years old male child from the diagnosis to management. Its pathogenesis and differentiation from Teratoma have been well established. A fetus in fetu is monochorionic-diamniotic, monozygotic twin of its bearer. It is usually intraperitoneal or retroperitoneal but may also present in scrotum, or in cranial cavity.<sup>3</sup> It is usually surrounded by a membrane analogous to amniotic sac and supplied by a single feeding vessel. A true placenta is usually absent. Absence of an independent circulatory system explains the subsequent growth retardation. Developmentally speaking, it has gone through the stage of primitive streak which is why it has vertebral body and organ arranged around axis. This differentiates it from Teratoma. Fetus in fetu is usually malformed because of pressure exerted by the host organ.<sup>4</sup>



**Fig 1** Distension of abdomen & mass in left side of abdomen of the patient

### Case Presentation

A 2 years old male child, coming from CRB Chattogram was admitted into Pediatric Ward (Ward no- 09) Chittagong Medical College Hospital on the 6<sup>th</sup> of June, 2021, with the complaints of abdominal distension and feeling of a mass in the abdomen for last 6months. The child's parents also complained of loss of appetite of the child. On general examination the child's appearance, height, weight, pulse, BP, temperature, breath sounds, heart sounds were all normal. No jaundice, no oedema, no cyanosis was seen. Only mild anemia was seen clinically. On local examination of the abdomen a firm hard mass, was felt in the left side of abdomen, mainly occupying the left lumbar region and extending upwards and downwards into left hypochondrium and left iliac region respectively. Mobility restricted, no ascites. Clinically a provisional diagnosis was made as Nephroblastoma (Wilm's tumor). Among lab investigations, CBC showed Hb%- 10.8 g/dl, ESR- 10 mm in 1<sup>st</sup> hour, WBC- 14,800/ cm, Lymphocyte-40%, Monocyte-05%, Eosinophils- 25%, Platelet- 400000/ cmm. Blood film (Thick & thin)- No malaria parasite seen. PBF showed- Microcytic hypochromic anaemia with eosinophilia. S. Albumin- 4.6 gm, S. creatinine- 0.27 mg / dl, S. HCG- 0.01 mIU/ ml, Alfa feto protein- 1.84 ng/ml. With the provisional diagnosis of Nephroblastoma (Wilm's tumor) the child was sent to Radiology and Imaging Department of Chittagong Medical College for further evaluation. In the radiology and imaging Department of CMCH, we evaluated the patient by the following radiological investigations:

#### Plain X-ray abdomen

Multiple bones- long bones, vertebra and teeth like structures were seen in left side of abdomen in left lumbar region.



**Fig 2** Plain X-ray abdomen shows multiple bone structures and teeth like structures in left lumbar region

#### B-Mode and Color Doppler USG of abdomen

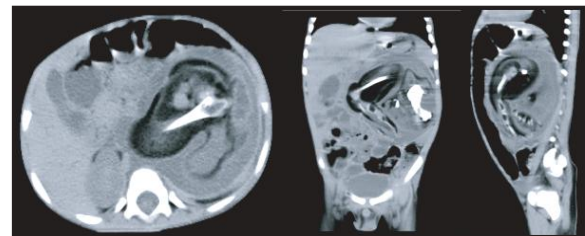
A fairly large, size about 12 x 11 cm soft tissue mass was seen in left side of abdomen. The mass showed internal high echogenic structures resembling bone or calcifications. The mass was separate from the spleen, stomach, left kidney and pelvic organs. The left kidney was displaced and compressed downwards. No significant blood flow was seen in the mass on color Doppler interrogation. There was no signs of any renal mass on USG, so the possibility of Nephroblastoma was ruled out.



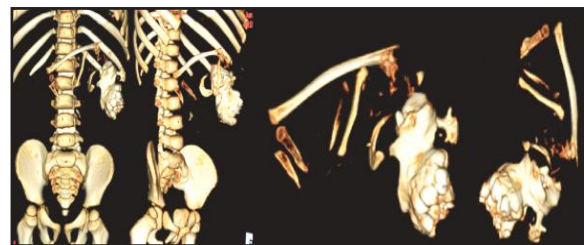
**Fig 3** B mode & color doppler USG of abdomen showing the mixed echogenic mass and calcified bony structures within the mass. Color doppler shows mild blood flow signal

#### Contrast CT scan of abdomen with 3D reconstructions

A large lobulated non-enhancing mixed density soft tissue mass measuring about 12 x 10 x 11 cm was seen in left side of abdomen. The mass had areas of soft tissue and fat density components. Multiple bony structures were identified within the mass- long bone, rib, pelvis and deformed lower vertebral column. All the elements were enclosed by a fluid containing sac.



**Fig 4** Axial, coronal and sagittal CT scan images of the mass. CT scan clearly demonstrate the long bones and vertebra within the mass. The mass shows mixed fat, fluid and soft tissue densities



**Fig 5** CT 3D VRT reconstruction images. Long bones, vertebral and a deformed pelvis are seen

### Per-operative findings

With the final diagnosis of Fetus-in-feto, the patient underwent surgery. Laparotomy was done by left supra-umbilical transverse incision over the swelling. A 12 x 10 x 11 cm mass was found occupying the epigastric, left hypochondriac and left lumbar region. The mass displaced the left kidney to pelvis sharing common covering. The mass was attached laterally with lateral body wall, medially with great vessels, superiorly with splenic flexure of colon and inferiorly with left kidney. The mass had a mixed consistency containing sebaceous secretion, hair, intestine, bone and limbs. The mass has a complete capsule. The capsule was medially closely adherent to abdominal aorta.



**Fig 6** Laperotomy and excision of the mass



**Fig 7** The excised specimen. A deformed & mal-developed fetus was found

### Autopsy of the specimen (Fetus in fetu)

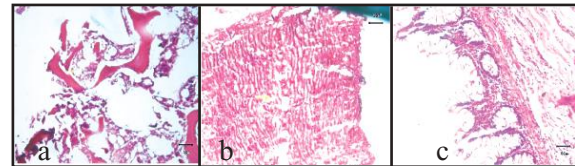
Deformed fetus with partially cut and attached sac, measuring about 15 x 9 x 7 cm. Covered by skin, areas of surface hair, a hand like structure, eyes like structures, two lower limbs each containing 07 rudimentary digits and nails and an umbilical cord was found. Vertical cut section showed fatty tissues, cartilages, bony tissue resembling vertebral column, cardiac chamber like cavity, intestine, multiple cystic areas containing serous fluid and sebaceous material.



**Fig 8** Gross specimen of fetus-in-fetu and vertical dissection of the specimen

### Histopathology findings

Sections made from the specimen showed fibro-collagenous and fibro-fatty tissue revealing presence of skin having stratified squamous epithelium, skin adnexal structures (Wheat gland, sebaceous glands and hair follicles) cartilages, bones with bone marrow, bony specules, axial arrangement of bony tissues resembling vertebral column, lymphoid structures, cardiac muscle, nerve bundle, salivary gland structures, intestinal wall with benign intestinal glands, ganglion cells, respiratory pseudo-stratified ciliated columnar epithelium, renal tubules with few glomerulus, foreign body type giant cells, chronic inflammatory cells infiltration and areas of necrosis was found. No granuloma or malignancy was seen. All findings were compatible with fetus-in-fetu.



**Fig 9** Histopathological slides showing bone and bone marrow (a) Cardiac muscle (b) Intestinal tissue (c)

### Discussion

Fetus-in-fetu is usually single, but rarely, more than 1 parasitic twin is observed in the host body. To our knowledge, the maximum number of fetus-in-fetu previously documented was 11.<sup>5</sup> Studies of genetic markers, such as blood group, sex chromosome constitution, protein polymorphisms, and DNA marker, suggested that host infants and their fetiform mass are genetically identical.<sup>6,7</sup> Aberration of monozygotic diamniotic twinning in which unequal division of the totipotent inner cell mass of the developing blastocyst leads to the inclusion of a smaller cell mass within a maturing sister embryo.<sup>7</sup>

Thakral et al reported equal male and female prevalence but Patankar et al and Federici et al noted a 2:1 male predominance. In our case the patient was a male 02 years old child.<sup>8-10</sup>

Most fetus-in-fetu are located retroperitoneally along the ventral midline, while other rare reported sites include the cerebral ventricles, liver, pelvis, scrotum, and mediastinum.<sup>11-16</sup> The common presentation of fetus-in-fetu is mass in the abdomen, almost 80% in the retroperitoneum.<sup>17</sup> The fetus in fetu produces symptoms due to mass effect

leading to distention, difficulty in feeding, vomiting, jaundice, urinary retention. In our case the patient had symptoms of abdominal distension, pain and feeling of a mass in the left side of the abdomen.

To qualify as an fetus-in-fetu, one of the following characteristics must be present: A mass enclosed within a distinct sac, partially or completely covered by skin, grossly recognizable anatomic features and attached to the host by a pedicle containing a few relatively large blood vessels.<sup>18</sup> The Fetus in fetu in our report fulfilled the criteria for being Fetus in fetu and not a teratoma. In our case, during the surgery, the fetus was found to be surrounded by a capsule that grossly resembled a fetal membrane. It contained a variable amount of sebaceous fluid and was attached to surrounding body wall and viscera. Engorged blood vessels were seen on the surface of the mass, the larger one arising from retro-peritoneum.

Preoperative diagnosis is possible radiologically, especially with computed tomography. Plain abdominal X-ray may be helpful in diagnosis. In half of the cases, X-ray shows the vertebral column and axial skeleton. Though a rare anomaly, fetus in fetu can be identified radiologically in the preoperative period.<sup>19</sup> Radiological differential diagnosis includes Teratoma and meconium pseudocyst.<sup>20</sup> Maternal and host serum alpha fetoprotein levels may also be raised. In our case the host child had normal alpha fetoprotein levels.

Pathological controversy arises during differentiation of fetus in fetu from a mature or well organized Teratoma. According to Willis, the presence of axial skeleton with vertebral axis and an appropriate arrangement of other limbs and organs goes more in favor of fetus in fetu.<sup>21</sup> Consistent with the theory of Willis, in our case, a deformed vertebral column was detected preoperatively by plain X-ray abdomen and CT scan of the abdomen and also in histopathology by the pathologist. However, review of literature showed that in about 9% of cases of fetus in fetu, no vertebral column was found even on pathological examination.<sup>22</sup> Therefore, Gonzalez-Crussi suggested fetus in fetu to be applied to any structure in which the fetal form has a highly developed organogenesis or to the presence of vertebral axis. In our case there was no great

confusion as CT scan clearly demonstrated the vertebral column within the mass. On the other hand, teratoma is an accumulation of pluripotent cells in which there is neither organogenesis nor vertebral segmentation.<sup>23</sup> Another important aspect of fetus in fetu is that they almost never become malignant whereas teratoma is known to become malignant. Until now, only one case of malignant fetus-in-fetu has been reported.<sup>24</sup>

Treatment of fetus in fetu is essentially surgical and excision gives complete recovery.<sup>8</sup> In our case, the mass was completely resected with no subsequent complications to the best of our knowledge. Postoperative follow-up by tumor markers  $\beta$ -HCG and AFP is often used and is further supported on the basis of malignant recurrence of fetus-in-fetu.

#### **Limitations**

MRI of the abdomen was not done due to lack of resources and financial constraints.

#### **Conclusion**

Fetus in fetu is a rare entity that typically presents as an abdominal mass in infancy or early childhood. It can be diagnosed in the preoperative period confidently by appropriate radiological investigations like CT scan of the abdomen. In our case, CT scan of the abdomen proved to be a real eye opener and savior in making a diagnosis. The identification of bones and vertebra within the mass, which was easily seen on CT scan, helped to make the diagnosis as Fetus-in-fetu. USG was able to demonstrate the mass and confirm its extra renal location and excluded Nephroblastoma but was not able to make a confident diagnosis. Complete excision of mass is curative and confirmatory.

#### **Recommendations**

Though a rare entity, fetus-in-fetu should be kept in mind as a differential diagnosis for abdominal mass in infancy and early childhood and should be well differentiated from Teratoma which is a common entity but has potential for malignant transformation. In the diagnostic work up CT scan of abdomen should also be done, because USG is sometimes not enough to make a concluding and confident diagnosis in rare abdominal masses.

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#### Contribution of authors

JC- Conception, design, acquisition of data, drafting and final approval.

SQ- Acquisition of data, data analysis, critical revision and final approval.

KC- Acquisition of data, interpretation of data, critical revision and final approval.

KMMU- Acquisition of data, data analysis, interpretation of data, critical revision and final approval.

#### Disclosure

All the authors declared no competing interest.

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