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Case Series

Fetus in Fetu: Report of Two Cases and Literature Review

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Abstract

Fetus in Fetu (FIF) is a rare congenital anomaly in which a malformed parasitic fetus grows in the body of its twin. It is a rare pathological condition and about 200 cases have been reported in the literatures. Prevalence more in infant and children but may present in any age, the oldest reported case was 47 years old. Here we are reporting 2 cases of FIF, first one is 3 month 10 days old male child and second one is 7 month old female child. Our first case presented with an abdominal lump and the second case presented with huge abdominal distention with lump, respiratory distress and anaemia. After proper imaging studies elective surgery was performed and pathology confirmed the diagnosis in both cases. Here we discuss about FIF, difference between FIF and teratoma, chance of malignancy and post-operative follow-up.

Keywords: *Fetus in Fetu; Teratoma; Abdominal Mass*

Introduction

Fetus in fetu is a rare congenital anomaly in which a malformed parasitic twin is found within host [1,2]. The majority of cases appear in infancy with an incidence reported at 1 in 500,000 births [3]. The anomaly was first defined in early nineteenth century by Meckel [4]. About 200 cases have been reported in literatures. Multiple theories are proposed regarding embryogenesis. One is the teratoma theory where FIF is regarded as a highly differentiated form of mature teratoma [5-7]. The other is the parasitic twin theory, according to which FIF may be a parasitic twin fetus growing within its host twin.

Most FIF are anencephalic, but vertebral column and limb buds are present in almost all cases. During embryogenesis, one of the fetuses is enclosed in its host and it got arterial supply by superior mesenteric artery. As the superior mesenteric artery develops from the vitelline circulation, this explains its most common retroperitoneal location. Other reported sites are abdomen, scrotum, kidneys, adrenals, cranium, mediastinum and lymph nodes. A slight male preponderance has been reported. The usual presenting complaint is a palpable abdominal mass. Mass effects include intestinal obstruction, jaundice, hydronephrosis, meconium peritonitis, respiratory distress and vomiting [8-10]. Diagnosis is often made preoperatively with ultrasonography, plain radiography, computed tomography (CT) or magnetic resonance imaging (MRI). Histopathologic findings confirm the diagnosis, and the recommended treatment is complete excision and long term follow up.

Case Reports

Case 1

We report a case of 3 month 10 days old boy, from a rural area admitted in Division of Pediatric Surgery, Dhaka Shishu (children) Hospital, Bangladesh with the complaints of lump in left side of abdomen. Mother suddenly noticed the lump during bathing her child. On examination, patient was well alert, average built, not anaemic, non-icteric and not dehydrated. His pulse was 120/min, BP 70/50 mm hg and temperature was normal. Abdominal examination revealed a swelling in left side of abdomen without any visible pulsation or peristalsis. On palpation there was a large non tender intra-abdominal lump in left lumbar region measuring approximately 12 cm X 11 cm with an ill-defined margin. It was of variable consistency, had irregular surface and slightly moved with respiration. Insinuation was possible above the lump underneath the costal cartilage. There was no ascites or organomegaly. Bowel sound was present. Other systems appeared to be normal. Several laboratory investigations and imaging were performed during his stay in the hospital. Abdominal Ultrasound revealed - A large cystic mass with solid component within it. Total volume of mass is 306 ml (9.5 × 8.5 × 7.3 c.m). Suspected Fetus in Fetu or Dermoid.

CXR P/A - normal

CT scan of abdomen (Figure 1A) showed- Large retroperitoneal mass measuring about 12 × 11 cm is seen occupying most of the left side of the abdomen, cross midline towards right. The mass having fatty and calcified internal components within. Calcified component resembling fetal parts. The mass displace left kidney superiorly, adjacent bowel loops superiorly and towards right side. The mass causing compression over urinary bladder and left ureter resulting left sided moderate hydronephrosis. Possible diagnosis is Fetus in Fetu, may be Teratoma.

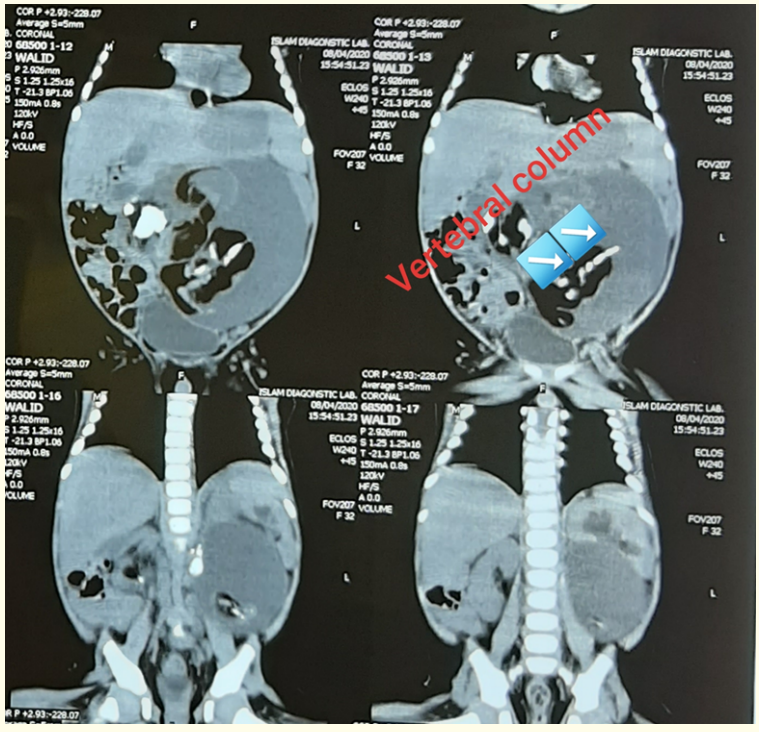

Figure 1A: *CT scan of Abdomen. Arrow shows vertebral column of FIF.*

Serum α-feto protein and S.LDH was normal. Other hematological tests, Urinalysis, Renal function test, Liver function tests were found normal. After clinical, laboratory evaluation and imaging the mass was thought to represent either a fetus in fetu or a dermoid cyst. Parents were counseled for surgery. Supra umbilical transverse incision was made and a large retroperitoneal, well capsulated mass was excised and the left kidney was found to be pushed superiorly. On gross eye appearance, the mass measured 13 × 9 × 7 cm, capsule was reasonably thick containing huge amount of sebaceous material and a well-developed fetiform mass with grossly visible four limbs (Figure 1B), well developed back, vertebral bone, anencephalic with fine hair over whole of the skin surface (Figure 1C). Weight of the fetus was 700 grams.

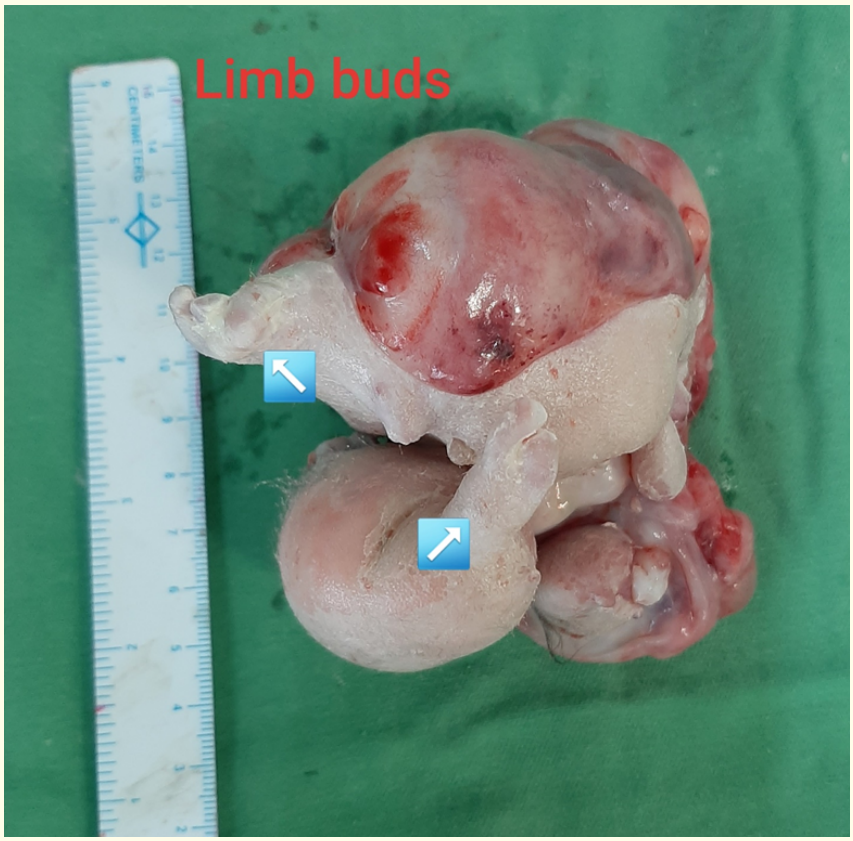

Figure 1B: *Arrow showing limb buds of FIF.*

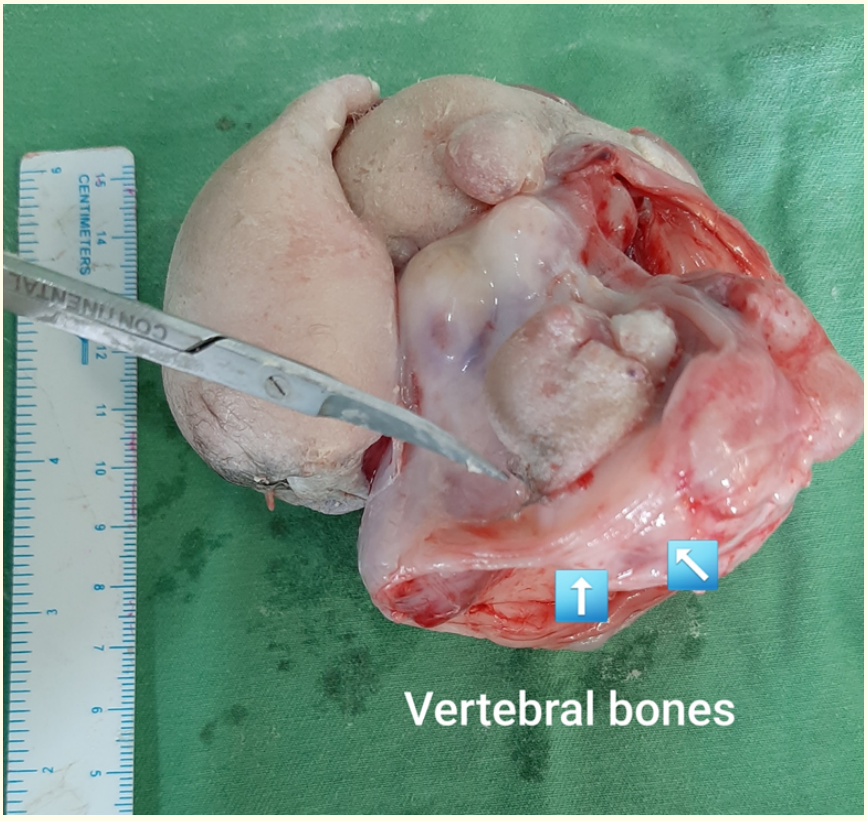

Figure 1C: *Showing buttock, fine hair over different part of the body and arrow showing vertebral bones.*

The child had an uneventful recovery and was discharged home on the third postoperative day. afp 1 month post-surgery was normal according to age. Histopathological examination showed a composition of mature benign tissues. A final diagnosis of a mature cystic teratoma was made.

Case 2

A 7 months old female child presented with gradual distention of abdomen for last month. Patient was mildly anemic, no palpable lymph node present but mechanical respiratory distress due to abdominal distention. Her pulse was 110/min, BP 80/50 mm hg and temperature was normal. On physical examination abdomen was Hugely distended more marked on left side. There was a palpable mass measuring about 20 × 15 cm, non-tender firm in consistency, smooth surface with well-defined margin. No visible peristalsis but engorged skin vein present, umbilicus protruded. No organomegaly or palpable abdominal lymph node present. Other systemic examinations revealed normal findings.

Abdominal ultrasound reveled - A hypoechoic mass which could not measured is seen along with left side of abdomen. Pushed bowel loops downward and right side, suggestive of neuroblastoma.

CT scan of abdomen reveled: Huge mixed density mass lesion cystic, solid, calcification and fatty component is noted occupying almost whole of abdomen (mainly on left side) measuring 15 cm x 16 cm. The mass causes compression and displacement of other abdominal organs. The mass causes significant compression and displacement of left kidney posteriorly and inferiorly. Suggestive of Teratodermoid/ Adrenal Neoplasm (Figure 2A).

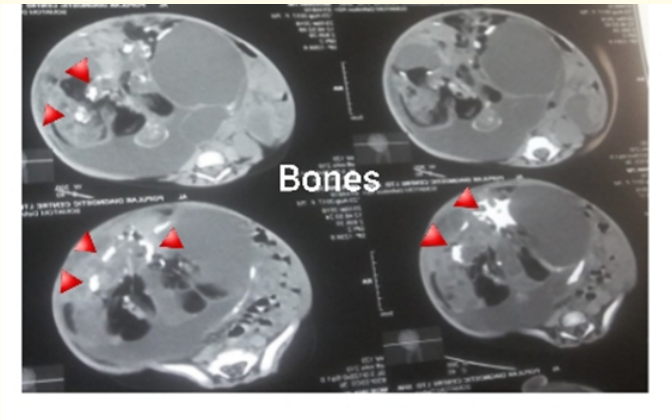

Figure 2A: *CT scan of Abdomen shows mixed density mass lesion cystic , solid, calcification and fatty component is seen.*

Serum α-feto protein, 24 hours urinary VMA and S.LDH was normal. Other hematological tests, Urinalysis, Renal function test, Liver function tests were found normal. After clinical, laboratory evaluation and imaging we thought that it may be a case of teratoma. Parents were counseled for surgery. Operative treatment was planned and a large retroperitoneal, well capsulated mass was excised. The left kidney was found to be pushed down and posteriorly. On gross eye appearance, the mass measured 18 × 16 cm, capsule was reasonably thick containing huge amount of sebaceous material and a well-developed fetiform mass with grossly visible four limbs, well developed buttock and back, developing genitalia, anencephalic with tufts of hair attached at back of the neck and fine hair over whole of the skin surface (Figure 2B and 2C). Weight of the fetus was 1300 grams.

Discussion

Fetus in fetu, malformed or parasitic monozygotic diamniotic twin that is found inside the body of a living child or sometimes in an adult, is first described by Johann Friedrich Meckel [11]. This abnormality occurs in 1 in 500,000 live births and male seems to be affected more than female [3]. The pathogenesis is still largely unknown and multiple theories are proposed regarding embryogenesis. One is the teratoma theory and the other is the parasitic twin theory. In teratoma theory where FIF is regarded as a highly differentiated form of mature teratoma [5-7]. In parasitic twin theory, according to which FIF may be a parasitic twin fetus growing within its host. FIF results

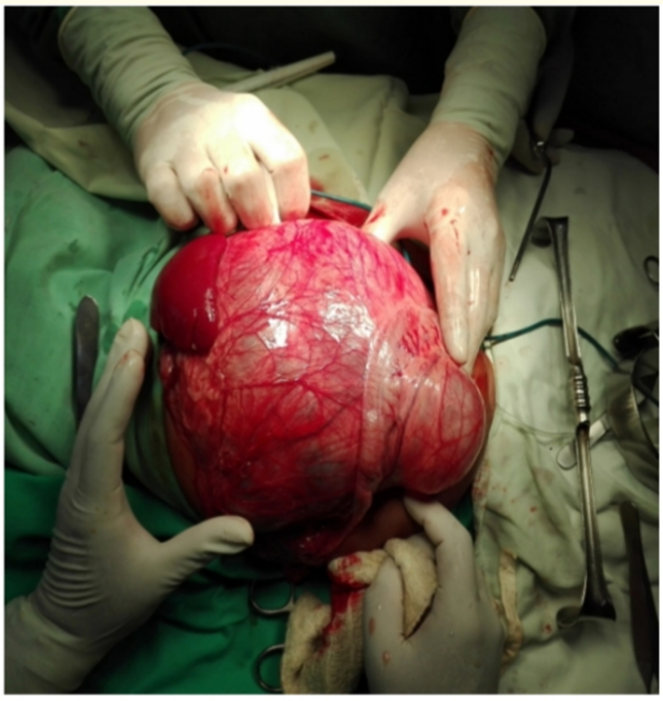

Figure 2B: *During operation.*

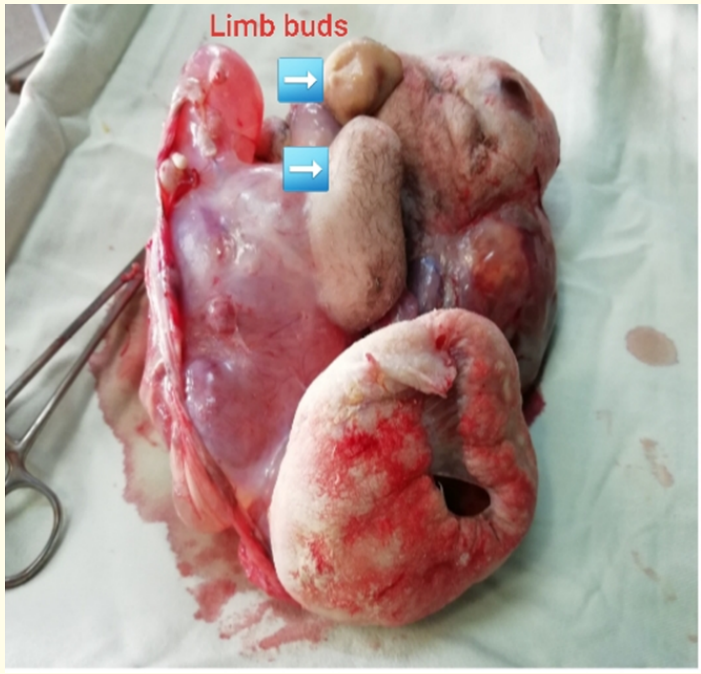

Figure 2C: *Pathological specimen showing limb bud, fine hair different part of the body and bony component.*

from abnormal embryogenesis in a diamniotic monochorionic twin pregnancy. This is followed by arrest of further growth of the encased fetus due to improper blood supply or inherent defects of the encased twin.

During ventral folding of the trilaminar embryonic disc, one of the fetuses is enclosed in its host. The most common reported sites are retroperitoneal location. Other reported sites are abdomen, scrotum, cranium, kidneys, adrenals, mediastinum and lymph nodes. Most FIF are anencephalic, but in almost all cases vertebral column (91%) and limb buds (82.5%) are present [8-10].

To know about differential diagnosis, Clinical and pathologic features should be discussed. In an infant with an abdominal mass showing diffuse calcifications or ossifications on ultrasonography, the differential diagnoses include neuroblastoma, FIF, meconium pseudocyst and teratoma [12]. Congenital neuroblastoma, the most common neoplasm in the neonate, usually presents with further involvement of skin, liver or bone. Teratomas rarely arise in the retroperitoneum, comprising less than 5% of retroperitoneal masses, compared to FIF, which are most often observed in the retroperitoneal location [13]. Teratoma are more common than FIF. Pathologically, FIF is highly differentiated tissue about a vertebral skeleton, whereas teratomas are discordant congregations of pluripotential cells (from more than 1 germ layer) without systemic organization [14]. In general, intra-abdominal fetus in fetu is usually suspended by a single pedicle within a complete sac containing fluid. Besides the vertebral column, structures commonly found in fetus-in-fetu include dermal, limb, gastrointestinal, and portions of the central nervous system [15]. Less commonly noted are the gonads, adrenal glands, heart, and a primitive respiratory unit.

With the advances in ultrasonography, early diagnosis is possible with improved patient outcomes [12]. But ultrasound is not confirmatory in all cases and which leads to a wide differential diagnosis. With the increased use of CT and MRI, the capability of narrowing to a single diagnosis has greatly improved [16].

According to Lindsey., *et al*. who reviewed 95 cases of FIF reported, 58 (63%) were male and 34 (37%) were female with 3 cases in which the sex could not be identified. A single fetus was seen in 89% percent of the case reports with the most common location being the retroperitoneum (72%), and one unusual case where 11 fetuses where identified in a single patient. Less frequent sites were intracranial, chest, mouth or neck. A vertebral column was identified in 76%, with 1 case revealing only a notochord. Based on literature review, most FIF masses are acardiac and anencephalic. However intestinal, neural, pulmonary, gonadal, pancreatic, and adrenal tissue are often present. In their review they found 40 (55%) fetuses had evidence of central nervous tissue, 61% had gastrointestinal tissue, and 36% had evidence of genitourinary development. A substantial number of fetuses (35%) were identified on prenatal imaging, with 50% identified within the first month of life and 75% within the first 2 years of life. Five cases involved an associated teratoma found at time of diagnosis or operation, and one case coincided with a yolk sac carcinoma [1].

As previously stated, most cases present as an abdominal mass during the first year of life. However, some reports of FIF in adults as old as 36 years of age have been reported [17].

Surgical treatment for fetus-in-fetu is curative as it is a benign disease. The main indication for resection is to prevent or palliate symptoms of an intra-abdominal mass. The most frequent symptoms reported are distension, palpable mass, emesis, poor feeding, jaundice, and/or dyspnea [18]. However, there have been isolated cases of malignancy following resection of a FIF, prompting some surgeons to recommend complete resection on a more urgent basis followed by postoperative surveillance of tumor markers for 2 years [19]. With the recent literature reviewed and our findings of coinciding tumors, we raise the argument that treatment guidelines should be identical for both conditions. Treatment of choice would therefore involve complete resection of the tumor and monitoring of alpha-fetoprotein levels because of the risk of malignant recurrence. The occurrence of a subsequent teratoma is not unprecedented.

Conclusion

Fetus in Fetu is considered as a benign condition but the potentially malignant characteristics of teratoma constitute the basis of the discussion. Some author belief that FIF may be malignant and chance of recurrence. Normal anatomy and physiology can be restored with surgical excision. With the ability of tumor marker surveillance and advances in radiographic technology physicians should consider the potential for a minimum of 2 year follow up to avoid a missed malignancy.

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