Case Report

Fetus-in-fetu

Imaging and pathology

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ABSTRACT

جنين داخل جنين، والمعروف أيضاً باسم التوأم الطفيلي الداخلي هو شكل من أشكال الازدواجية الجنينية غير منتظمة الأجهزة والتي ينمو فيها الجنين غير الطبيعي متطفلاً على التوأم الطبيعي عن طريق الارتباط به داخلياً. نسجل في هذا التقرير حالة جنين داخل جنين والتي ظهرت ككتلة كيسية داخل البطن خلال الفحص بالأشعة فوق الصوتية للجنين في الأسبوع 29 من الحمل. وفي الأسبوع 34.5 من الحمل ولدت أم سعودية عمرها 32 عاماً ولادة تلقائية مولود ذكر. أظهرت الأشعة السينية والتصوير المقطعي للمولود بوضوح وجود أطراف وعمود فقري وأوعية دموية شاذة تغذي الكتلة. بعد الاستخراج الجراحي أُجريت للكتلة داخل البطن فحص مرضي تفصيلي. وأظهرت الدراسات المجهرية الإضافية وجود أنسجة دماغية، وتراكيب تشبه الأمعاء وأصابع وأطراف. وأخيراً تم تأكيد التشخيص للكتلة بأنه حالة جنين داخل جنين.

Fetus-in-fetu (FIF), also known as endoparasitic twin, is a form of asymmetric fetal duplication in which the abnormal developing embryo parasitizes the normal co-twin by attaching internally. Here, we report a case of FIF presented as an intra-abdominal cystic mass, which was first detected during an antenatal ultrasound examination of a 32-year-old Saudi mother. At 34 weeks and 4 days of gestation, she had spontaneous labor and delivered a baby boy. The x-ray and CT examination of the baby boy clearly suggested the presence of limbs, vertebral column, and anomalous blood supply to the mass. After laparotomy and surgical removal, the intra-abdominal mass was subjected to detailed pathological examination. Microscopic studies further showed the presence of brain tissue, gut-like structures, fingers, and limbs. The mass was finally confirmed as FIF.

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Fetus-in-fetu (FIF) is a rare abnormality where a malformed fetus is found within the body of a normally developing fetus.[1-3] To date, less than 200 cases of FIF have been reported in literature worldwide.[1,2] Over 90% of all the published cases were reported from Asia, Europe, and North America. Although it is known that the majority of FIF cases are intra-abdominal in location, exact distribution of the remaining cases is not well documented. According to recent analysis, FIF most likely represents a monozygotic diamniotic twin that implants itself and grows within the body of its normal karyotypically identical sibling.[4]

Fetus-in-fetu is an encapsulated pedunculated vertebrate fetiform mass. According to Lewis,[1] the term FIF (cryptodidymus) was coined by Meckel and is reserved, by consensus, for those formations that meet Nicholson's[2] and Willis'[3] criteria of a degree of morphologic development and structural organization that exceeds that which can occur in a teratoma. Embryonic development beyond the primitive streak stage is assumed by the presence of an axial skeleton and spinal column that imply metameric segmentation. The extreme degree of fetiformity generally includes encapsulation by a fluid-filled amniotic cavity and suspension of the mass from the inner surface of the capsule by a vascular pedicle in the manner of an umbilical cord.

In the uterus, the growth of an FIF initially parallels that of its twin, but stops abruptly because of either the vascular dominance of the host twin or an inherent defect in the parasitic twin.[2] Fetus-in-fetu is mostly anencephalic, but in almost all cases its vertebral column (92%) and limbs are present (82.5%).

Case Report. A routine ultrasound scan of an obese (110 kg) Saudi mother at 29 weeks of gestation showed a singleton fetus with an intra-abdominal heterogeneous mass (**Figure 1**). The mass was initially thought to represent either a well differentiated teratoma or a FIF. The baby boy was spontaneously delivered at 34 weeks and 4 days gestation. The 32-year-old mother had 5 previous pregnancies, of which 4 were live births and one still birth. There was no history of consanguinity

or multiple pregnancies. Besides being obese, she had a history of chronic asthma and hypothyroidism and received regular treatment with thyroxin, ventoline, fluticasone-seretide, and ranitidine. During pregnancy, the mother developed gestational diabetes which was controlled by diet. At birth, the baby weighed 2.89 kg with undescended testes and an APGAR score of 9 both at one and 5 minutes. The level of beta human chronic gonadotrophin (ß-HCG) was 5.0 IU/L and alpha-fetoprotein level was 157,750 ug/L.

Postnatally, this baby boy was subjected to a variety of radiological investigations including abdominal x-ray, ultrasound, and CT scan. Abdominal x-ray examination of the baby confirmed the presence of large mass arising from pelvis and extending in the upper abdomen (**Figures 2a & 2b**). The mass consisted of well differentiated elongated bones like femur located in its most anterior aspect. Abdominal and pelvis ultrasound revealed heterogeneous soft tissue mass with calcification and enhances vascularity associated with ascitic fluid (**Figures 3a & 3b**). The stomach and bowel were mainly collapsed and displaced superiorly, all other visceral organs including kidney, adrenal gland, and liver appeared grossly normal. Urinary bladder appeared compressed by the mass and contained small amount of urine. The CT abdomen and pelvis further confirmed the presence of large retroperitoneal mass filling the abdominal and pelvic cavities measuring 10.4 x 9.2 x 7.2 cm in maximum craniocaudal, transverse, and anteroposterior diameters respectively. Anomalous blood supply to the mass arose from the right femoral artery (**Figures 4a & 4b**). The mass was consisted of a large amount of fluid and a partially enhancing disorganized soft tissue component with fatty attenuation and areas of calcifications. Some of these calcifications showed differentiation between cortex and medulla and they simulated bony structures. Alignment of multiple small

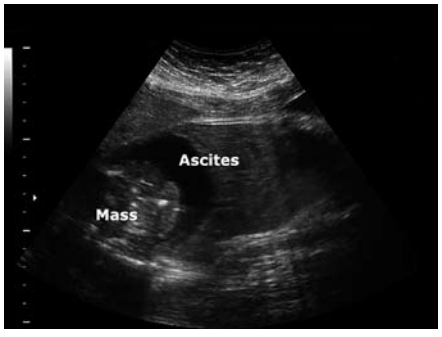

Figure 1 - Longitudinal section of antenatal ultrasound shows a fetal abdomen and thorax with a heterogenous mass at the lower fetal abdomen surrounded by ascetic fluid.

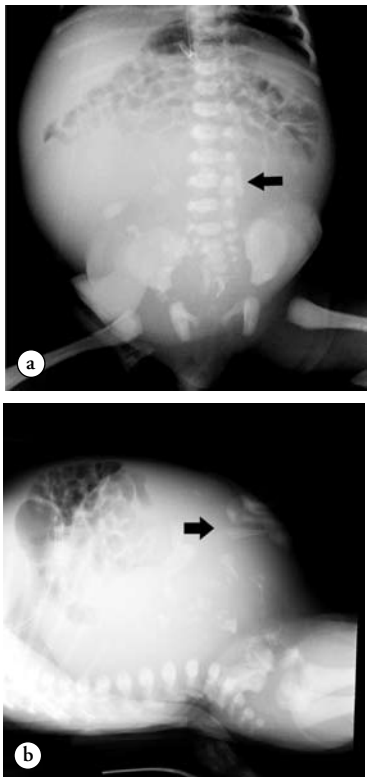

Figure 2 - Abdominal x-ray. **a**) Supine and **b**) cross table lateral showing the abdominal pelvic soft tissue mass with a well differentiated elongated bone likely representing the femur (arrows).

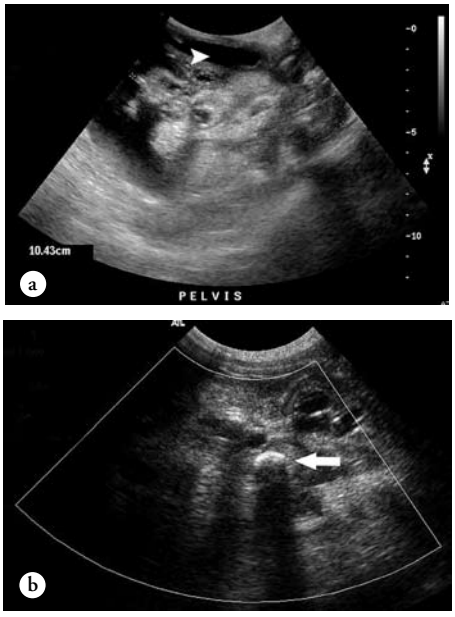

Figure 3 - Postnatal ultrasound of **a**) abdomen and **b**) pelvis showing complex mass and posterior acoustic shadow produced by the bones (arrow) associated with ascites (arrow head).

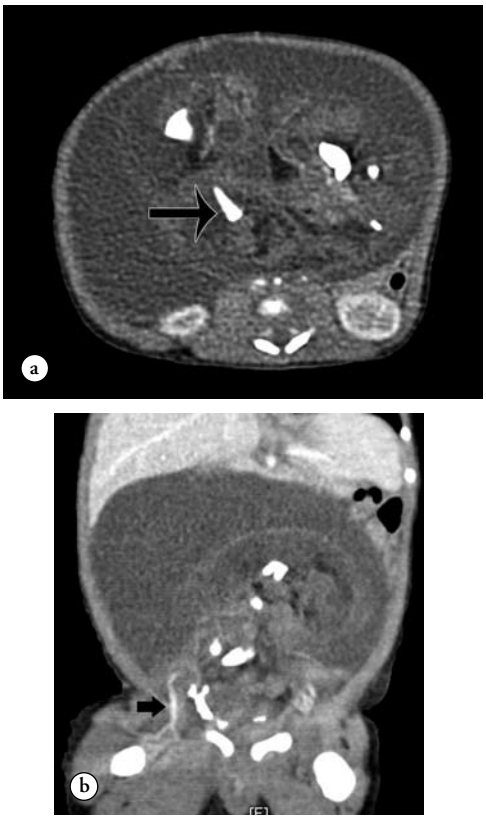

Figure 4 - Computed tomography scan of the abdomen showing **a)** axial cut at the level of the kidneys reveals a large abdomenopelvic mass consisting of a partially enhancing soft tissue component, fat component, and well differentiated elongated bones unlike mature teratoma (arrow). **b)** Coronal reformatted image reveals anomalous blood supply to the mass from the right femoral artery (arrow).

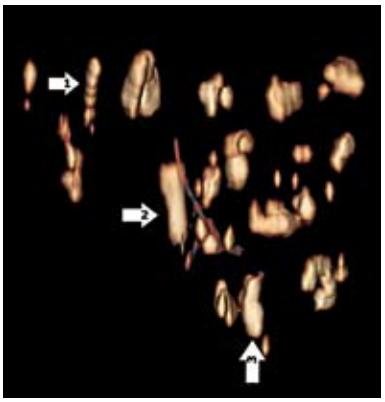

Figure 5 - Volume-rendered 3 dimensional CT demonstrates the dysplastic skeleton of the mass. Arrow 1 shows the vertebral column. Arrow 2 points to the femur. Arrow 3 illustrates the scapula.

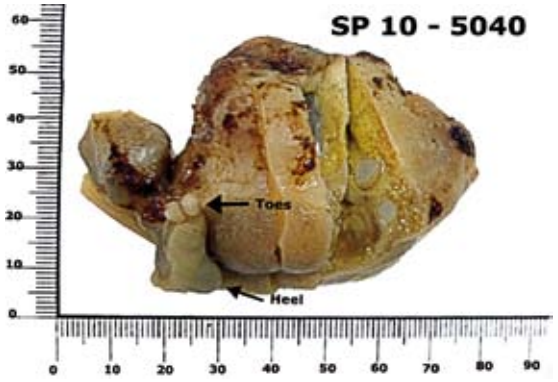

Figure 6 - The mass after pathological dissection shows skin and cartilage. Part of the foot can be seen with 2 toes and heel (arrows).

bones resembled a vertebral column while a V-shape bone resembled a scapula. An elongated bone resembled a femur (**Figure 5**).

The surgical removal of the intra-abdominal mass was carried out on the 7 days old male baby at Riyadh Military Hospital, Riyadh, Saudi Arabia by a supraumbilical transverse incision. The subcutaneous tissues including scarpa's fascia, muscle, and sheath were divided by diathermy and the peritoneal cavity was explored. A large mass which was encapsulated retroperitoneally was pushing both testes, vas, vessel, and urinary bladder anteriorly. With careful dissection, the mass was completely removed. After restoration of hemostasis and washing with warm normal saline, the wound was closed in layers. The mass was weighed 460 grams. Morphologically, the mass looked like a fetus containing upper and lower limb with skin and hair over it. The mass was sent to histopathology for further macroscopic and microscopic examination.

According to microscopic histological examination report, the excised specimen measured 11 x 9 x 7 cm and weighed 460 gms. The outer surfaced was covered by skin and showed a finger-like projection (**Figure 6**). Bisection of the mass showed cystic and solid areas with hard bony and cartilaginous tissue. Microscopically, the finger-like structure showed digital bones with cartilage on each end along with articular spaces between adjacent digits (**Figures 7a & 7b**). In other areas derivatives of 3 germinal layers, including stratified squamous and ciliated columnar epithelia along with mature brain tissue, cartilage and bone are identified (**Figures 8a & 8b**). Gut-like structures were also observed; they showed 2 layers of muscularis propria with myenetric plexuses in between. The lining mucosa showed a mixture of ciliated and non-ciliated columnar epithelium (**Figures 9a & 9b**).

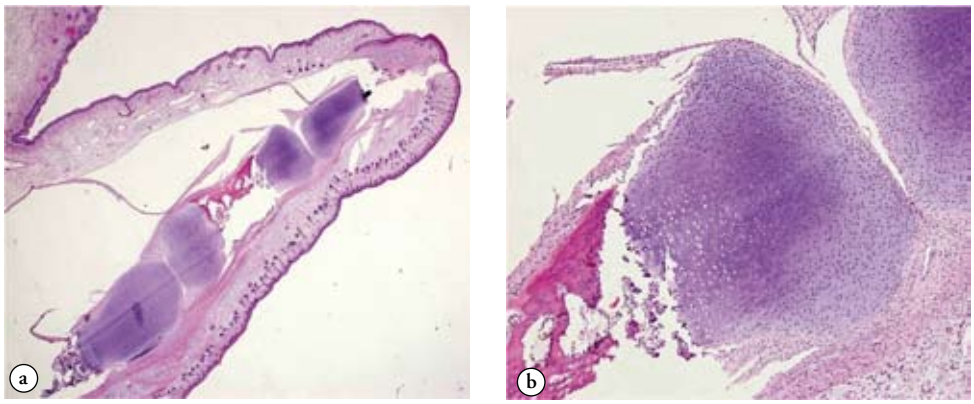

Figure 7 - Microscopically, **a**) the finger-like structure showed digital bones with cartilage on each end along with articular spaces between adjacent digits. **b**) High power showing the articular space.

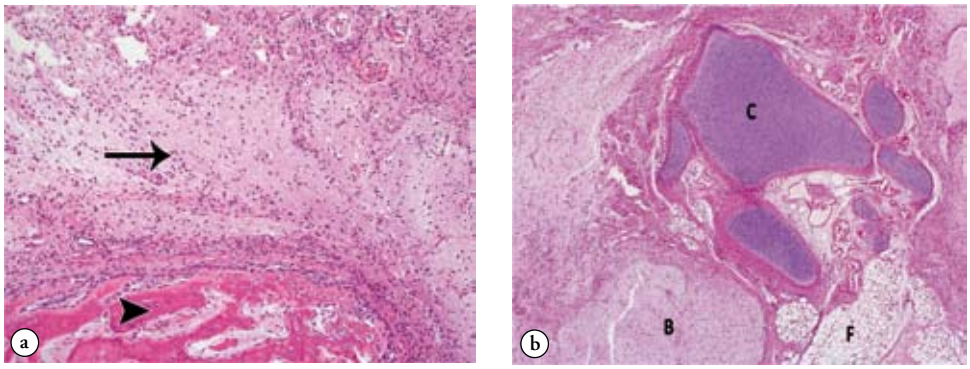

Figure 8 - Histology section showing **a**) brain tissue (arrow) and bone (arrow head) and **b**) the disorganized tissue composed of brain (B), mature cartilage (C) and fat (F).

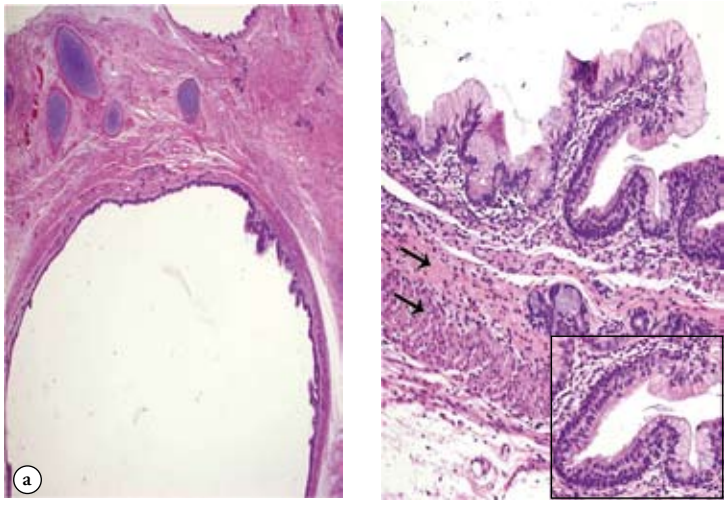

Figure 9 - Histology showing **a**) gut-like structures and **b**) high power of the wall shows 2 layers of muscularis propria (arrows). In situ: closer view shows ciliated and non-ciliated columnar epithelium.

A diagnosis of incorporated twin (FIF) was further confirmed.

Discussion. The diagnosis of the intra-abdominal mass in our case from both radiologic and pathological standpoints was confirmed to be FIF. Fetus-in-fetu or included twin is a descriptive term given to a fetus like structure found within the body of its bearer identified antenatally at birth or postnatally. Although FIF was apparently described as early as 1800 AD (cited in Lewis[5] and Majhi et al[6]), its distinction from a retroperitoneal teratoma was not emphasized until the late 1950's, when Willis stressed the advanced development of the mass as the *sine qua non* for the diagnosis of FIF. He proposed that the degree of structural organization at certain point in fetal development exceeds that which can occur in teratomas. According to Willis[7] the development of an axial skeleton and spinal column, with its implication of segmentation and associated musculoskeletal development and proof of embryonic development is beyond the primitive streak stage. Subsequently, reported cases of FIF have generally adhered to the Willis's criteria. However, these criteria have not been universally accepted.[3]

There are several problems with Willis's criteria to distinguish between teratoma and FIF. In fact, there is a considerable gray zone, exemplified by Majhi et al[6] in which the mass exhibits were more than organizational than the usual teratoma, but has no unequivocally identifiable vertebral column. Other investigations have also noted that potential difficulty in applying Willis criteria.[7] Kimmel et al's[8] reported 5 fetiform masses found in the brain of a 19-year-old hydrocephalic infant. Although only 2 out of 5 meet the criteria of Willis (presence of vertebral column), Kimmel et al[8] reported all the 5 masses as FIF as the basis of the presence of extremities in all the 5 masses.

Besides the presence of bony structures resembling the vertebral column, our imaging studies also showed other bones resembling femur and scapula. On the other hand, morphological studies on the removed mass showed the presence of limb skin and hair. Further detailed pathological examination of the mass supports our diagnosis of FIF which resembled the previously described cases showing the gut-like structure that has been the only visceral organ most readily identified whereas cardiac structures have never been seen.[9] Surprisingly, histopathological examination also showed the presence of mature brain tissue unlike majority of anencephalic fetuses indicating either inhibition of growth beyond Henson's node in utero or regression of craniofacial structure.

In this study, we observed that anomalous supply of the mass arose from femoral artery that is different from an earlier report in which investigators noted blood supply through the vessels arising from the superior mesenteric artery.[10] However, in majority of reported cases of FIF, investigators did not identify the source of vascular supply. Collectively, our imaging studies, morphological observation on extracted mass and histopathological examination confirmed the presence of FIF.

In conclusion, we describe a case of FIF presented as an intra-abdominal mass which was detected during prenatal ultrasound examination at 29 weeks of gestation. The x-ray and CT studies of the baby showed presence of limb, vertebra column, and anomalous blood supply. Post surgical removal, the pathological examination of the intra-abdominal mass showed presence of brain, gut-like structure, and limbs. Summation of pathological and imaging data clearly helped in diagnosis of FIF. It is important to report such cases to add to the knowledge of this rare field of fetal medicine.

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