

Case Report

Hidden Fetus in Fetu in a Male Adult

Seyed Hassan Hosseini Hooshyar¹, Hamid Zeinalinejad¹, Mehran Mosallami¹,
Shahriar Dabiri², Jahanbanoo Shahryari², Moeinadin Safavi²

1. Dept. of Surgery, Kerman University of Medical Sciences, Kerman, Iran

2. Dept. of Pathology, Kerman University of Medical Sciences, Kerman, Iran

ABSTRACT

We present unusual retained remnants of hidden conjoined twin of possible abdominal pagus after 21 years who presented with abdomenpain and incidentally have found his forbidden conjoined twin. He presented for many years with a palpable epigastric mass, and recent abdominal pain, early satiety, nausea and vomiting. The mass was a like deformed fetus weighing 815 gr who was anencephalic and deprived of viscera but there was a distinct vascular connection through the right limb. Microscopic examination revealed a normal epidermis with melanocytes overlying dermis with mature adnexa. The fetus plain film after surgery showed a rather complete skeletal system. All above findings were in favor of fetus in fetus, a rare congenital entity in which a nonviable parasitic fetus grows within the body of its twin.

Keywords: Fetus in Fetu, Male, Adult, Iran

Introduction

Fetus in fetu is a rare congenital entity in which a nonviable parasitic fetus grows within the body of its twin. Historically it was first described by Meckel in early 19th century. It is a rare phenomenon occurring 1 in 500000 births. To our knowledge, nearly 100

cases have been reported so far (1).

Fetus in fetu complex mainly includes a fibrous membrane that encircles some fluid and a fetus suspended by a cord structure or a vascular pedicle (2). One characteristic is presence of vertebral axis associated with other organs or limbs around (3). At early intrauterine life stages,

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Address communications to: Dr. Jahanbanoo Shahryari, Department of Pathology, Kerman University Of Medical Sciences, Kerman Iran.

Email: Jshahryari@yahoo.com

growth of subordinate twin (fetus in fetu) is parallel to its twin, but stops suddenly because of either vascular dominance of its twin or its own inherent defects.

Fetus in fetu is the rare differential diagnosis of teratoma. However, its prognosis is better in comparison with teratoma. Thus, determination of criteria for discriminating these two entities seems to be necessary (4).

Although fetus in fetu is prevalent among infants and children, there have been few reports of this anomaly in older subjects due to its asymptomatic nature (5, 6). So we decided to share our experiences with other clinicians and paraclinicians about pathogenesis, diagnosis, complications and follow up of this rare phenomenon in an adult.

Case Report

The patient was a 21-year-old man with epigastric mass, which had been palpable since he was one year old with gradual growth and no significant symptoms. However, it grew faster from 3 years ago leading to abdominal pain, early satiety, nausea and post prandial vomiting especially from 3 months ago. Physical exam revealed a huge mass about 30×20 cm supraumbilically, which was mildly mobile with no significant tenderness. He was product of a triple pregnancy that was diagnosed by sonography and delivered by normal vaginal delivery. His live twin was a non-identical female one. However, they did not find the third mysterious one.

Abdominal CT scan was performed for more evaluation before surgery, which showed a fetus like mass with a rather complete axial skeleton in the upper to mid retroperitoneum separated from surrounding viscera by a distinct fibrous membrane compressing the adjacent viscera (Fig.1).

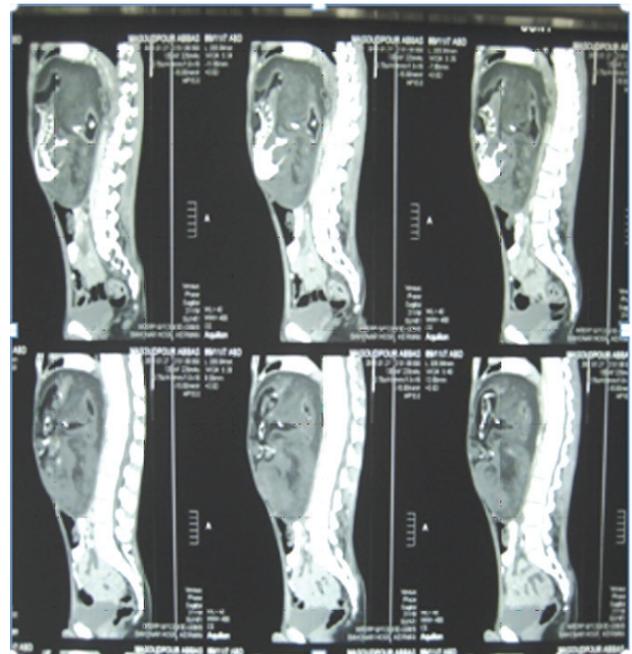


Fig. 1: Abdominal CT scan, sagittal view exhibited a heterodense mass containing fat, bone, liquid and soft tissue density in the upper to mid part of abdomen. Bony particles constitute a skeletal axis like structure. It also compressed abdominal viscera to the sides.

Then he was operated with the following procedure: After prep/drape under general anesthesia in supine position the peritoneal cavity was entered via midline incision, there was a huge mass peritoneally posterior-inferior to transverse colon mesentery after dividing the peritonea the mass was resected with meticulous sharp dissection. During the procedure, an active hemorrhage happened due to indeliberate cutting of the vascular pedicle connected to the superior mesenteric artery. After removal of mass, the cavity was irrigated with normal saline. There was no other pathology in exploration so the abdominal cavity was closed as routine surgical procedure.

The pathology report was a monster like fetus weighing 815 gr with the following measurements: crown rump length=21 cm, crown heel length=38 cm, head circumference=20 cm. It was anencephalic and its head was partly covered by long terminal hairs. The face members were

atretic including eyebrows, eyes, ears, nose and teeth. Right and left upper extremities of the fetus were deformed phocomelia like processes measuring 7 cm and 5 cm respectively. The upper right extremity seemed to be the feeding site through a vascular connection. Lower extremities were anomalous too, the right one possessed toe like processes, and the lower extremities measured 18 and 14 cm for the right and left one. The back of the fetus had a meningocele like lesion superior to the natal cleft. On the autopsy, the abdomen was opened but no viscera were detectable and the abdominal wall was directly lying on the vertebral column. The chest contained no lungs or heart (Fig. 2).



Fig. 2: Gross appearance resembled mummified miniature of a fetus. Additional autopsy revealed no viscera.

On microscopic examination, skin tissue was seen with a normal epidermis overlying dermis with mature adnexa containing sebaceous glands and hair follicles. Melanocytes were also detectable in the basal layer of epidermis (Fig.3). The biopsy of vascular connection site revealed a vascular network composed of dilated and congested vessels of different sizes (Fig. 3) The fetus X-ray Rontgography of specimen after surgery revealed a rather complete skeletal

system including skull vertebral column, shoulder, humerus, pelvis, femur, fingers and toes (Fig.4)

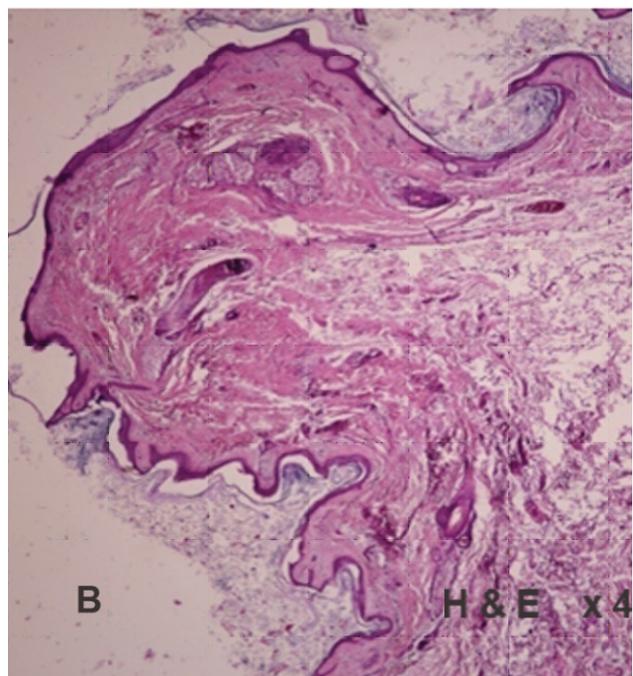
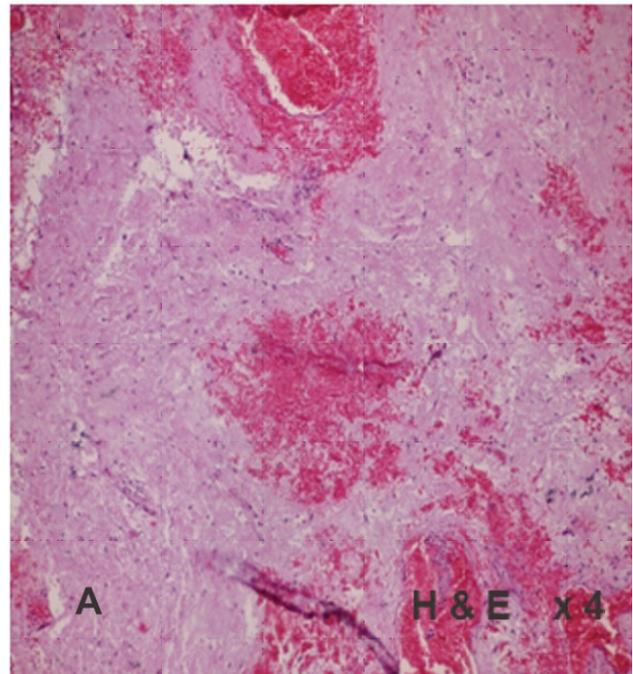


Fig.3: Microscopic appearance revealed (A) the vascular connection site with variable sized and congested vessels in a fibromyxoid stroma (Hematoxylin and Eosin $\times 40$) and(B) mature skin of fetus with its adnexa (Hematoxylin and Eosin $\times 40$).

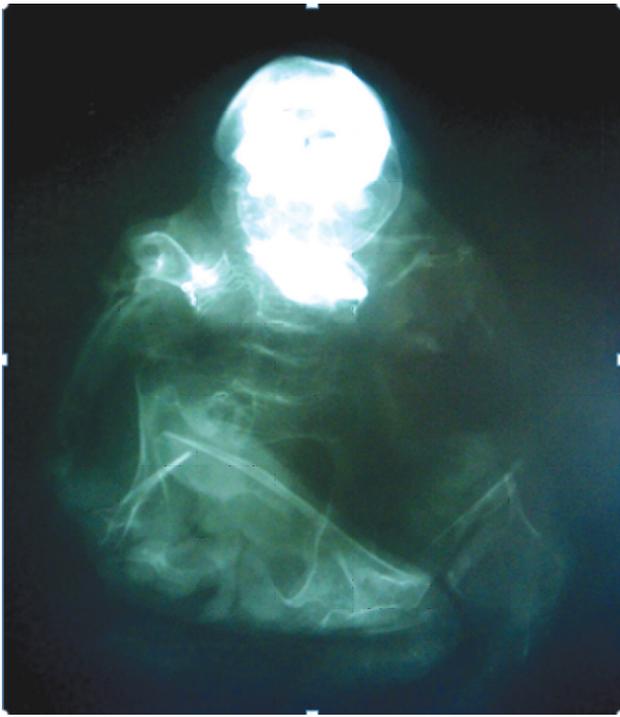


Fig.4: Postsurgical plain x-ray of the fetus revealed a rather complete skeletal system.

Discussion

Several theories have been suggested for clarifying pathogenesis of fetus in fetu. Two of them are more popular. The first one declares that fetus in fetu results from diamniotic-monozygotic twin pregnancy and subsequent unequal division of totipotent inner cell mass leading to a smaller cell mass formation within a maturing sister embryo. The second suggests that fetus in fetu may be a highly differentiated form of mature teratoma (7). Spencer set 5 criteria for fetus in fetu diagnosis including: 1- a distinct sac enclosing fetus, 2-normal skin coverage, 3-grossly obvious anatomic parts, 4-attachment to the host through a few relatively large vessels, 5-Association with gastrointestinal tract or neural tube (8). According to Spencer's opinion on criterion is enough for diagnosis of fetus in fetu. Our case met at least four of five criteria. Other clues in favor of parasitic twin rather than a well-developed teratoma are male sex, location of mass in the upper retroperitoneum and approximately thorough skeletal system.

On the other hand, teratomas are composed of two or three germ cell layers, having multiple tissue type. Most of them are sacrococcygeal. They can also commonly originate from ovary or testis. Other possible locations are thyroid, mediastinum, stomach and retroperitoneal tissue. Teratomas do not show organogenesis or vertebral column formation (9,10).

In contrast to our case, the majority of this cases present in infancy with the oldest case reported in a 47-year-old man (2). Eighty nine percent of fetuses in fetu cases are less than 18 months old (9). To the best of our knowledge only five cases have been reported in adulthood (older than 20 years) up to now and this case is the sixth one and the largest reported case in adults. Evaluation of other characteristics in adult type fetus in fetu such as location revealed that all of them just like our case were in upper retroperitoneum except two cases, which were placed in iliac fossa and right upper quadrat. This might be due to more potentially spacious retroperitoneum, which can accommodate parasitic twin with less or no symptom. The other common finding in adults was the male sex of subjects that constituted gender of almost all cases except one adult female. Therefore, although male sex is a contributing factor for differentiating fetus in fetu from teratoma but it is not absolute (11-15). As in this case, the most common site of fetus in fetu is the upper retro peritoneum (16). Rare cases of this anomaly have been reported in central nervous system, gastrointestinal tract or genitourinary tracts. It is even more rarely found in lungs, adrenal glands, pancreas spleen or lymph nodes (17).

Symptoms of fetus in fetu are related to its mass effect. As its most common site is upper retroperitoneum, feeding difficulty, vomiting, abdominal distention and rarely jaundice are the leading symptoms (2,18).

Definite vascular connection to the host is a very rare finding (2,19,20) but in this case a distinct vascular connection through the right upper

extremity was the route of nourishment for the fetus and it might be the cause of secondary visceral atresia.

According to previous reports, fetus in fetu weight ranges from 13 grams (2) to 2000 grams (21). The size of the fetus is likely related to its blood supply. Fetuses with distinct vascular connections to the host are relatively bigger with better-developed features. Our fetus weighed 815 gram which is due to a definite vascular connection

Preoperative imaging scan can be helpful for diagnosis before surgery. Plain abdominal x-rays confirm the diagnosis of approximately half of the cases by revealing vertebral column and an axial skeleton (19). The rest of cases may have an underdeveloped or dysplastic vertebral column (22). In such cases CT scan and MRI have been proven to be very practical in preoperative diagnosis (23). In the presented case abdominal CT scan had the pivotal role in preoperative diagnosis of this congenital anomaly.

Fetus in fetu can be treated by complete resection. Sometimes complete excision becomes impossible because of its adherence to the adjacent organs (24). In this case, the fetus and its surrounding sac were completely excised but a massive hemorrhage happened due to the vascular connection to the large vessels of abdomen. Thus, surgeons must consider this complication during operation. Successful preoperative detection of vascular connection has been reported once through three-dimensional CT scan (25). Therefore, we recommend this imaging modality especially in more developed fetus in fetu and adult subjects before operation to evaluate the risk of hemorrhage.

Prognosis of fetus in fetu is better compared with cystic teratoma. However, presence of immature elements require a close follow up by serial radiological and serological (alpha-fetoprotein) evaluation (6). As this case had completely mature organs, no extra follow up except post surgical imaging seemed essential.

Acknowledgement

The authors declare that there is no conflict of interest.

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