Case Report

TWIN FETUS IN FETU - A VERY RARE ENTITY: A case report with review of literature

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ABSTRACT:
Objective: 1) To report a very rare condition of conjoined twining in fetus in fetu in an infant. From literature it appears to be the 4th case being reported.
2) To emphasize the importance of proper examination of the fetus in fetu grossly by doing extensive dissection, radiologically and detailed histological tissue examination from several sites.
Design: Detailed report of an unusual intra-abdominal mass in an infant.
Setting: Department of Pediatric Surgery, Children Hospital, PIMS, Islamabad.
Results: Successfully treated with surgical removal and interesting diagnosis emerged.
Conclusion: Twin fetus in fetu (FIF) is a very rare anomaly due to an abnormal embryogenesis of fetus during pregnancy. Malformed fetus grows in the body of co-twin and it is frequently detected as an abdominal mass. There is no clear distinction between the term of FIF and fetiform teratoma (FT). We are reporting a case in which a girl was referred as a case of nephroblastoma but at operation and on detailed examination of the specimen it turned out to be a case of twin fetus in fetu.

KEY WORDS: Twin fetus in fetu, postoperative diagnosis.

Pak J Med Sci October-December 2003 Vol. 19 No. 4 306-309 www.pjms.com.pk

INTRODUCTION

A fetus-in-fetu is an encapsulated, pedunculated vertebrate tumor. It represents a malformed monozygotic, monochorionic diamniotic parasitic twin included in a host (or autosite) twin.[1, 2]

Twin fetus in fetu is a very rare anomaly among fetus in fetu.[3] Most cases that have been reported in the literature occur within the upper retroperitoneum or within abdomen[4]. Unusual locations for fetus in fetu have included the scrotal sac[5], pelvis[6], iliac mesentery[7] and in the cranial cavity.

Characteristically the fetus-in-fetu complex will be composed of a fibrous membrane (equivalent to the chorioamniotic complex) that contains some fluid (equivalent to the amniotic fluid) and a fetus suspended by a cord or pedicle. The presence of a rudimentary spinal architecture is used to differentiate a fetus-in-fetu from a teratoma, since teratomas are not supposed to develop through the primitive streak stage (12-15 days). More recently, such criteria have been questioned[8]. The presence of an amnion or umbilical cord, tissue indigenous to an embryo/fetus, is also considered acceptable diagnostic criteria for fetus-in-fetu. CT scan demonstrating a tubular configuration of almost pure fat around a central body density (a leg or an arm) should suggest a

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* *Received for publication:* June 20, 2003

Accepted: July 25, 2003

diagnosis of fetus-in-fetu rather than a teratoma.[9,10]

CASE REPORT

A five months old female who was referred from another hospital with an abdominal mass which was producing a marked bulge in the left lumbar region which mother had noticed a few days earlier (Fig 1). It was non-tender and had considerable mobility. Antenatal examination was unremarkable. An ultrasound report, which she carried reported to be a left nephroblastoma. The other kidney was normal. Routine hematological investigations were within normal limits. At laparotomy, there was an encysted retroperitoneal mass to which was plastered the left kidney and it had been compressed out inferiorly and had become completely flat. But it could be conserved because its blood supply could be preserved and it was carefully separated from the mass. The mass had a cord like structure, which contained its artery and vein and these were seemed to be arising from abdominal aorta and vena cava. This feeding vascular pedicle was ligated and divided and the mass was easily removed.

On gross examination the entire mass was enclosed in a firm thin covering. On opening this covering there was a small amount of dirty fluid with sticky brownish coating the entire multi-structured mass. This substance had stark resemblance with vernix caseosa. On further examination one could identify several structures which included two heads, single heart just below these completely formed fused livers with gall bladder, irregularly formed limbs with bony structures, loops of intestine and several other rounded firm, solid and cystic structures which could not be grossly identified (Fig 2).

X-ray of the specimen revealed two vertebral columns with spinous processes, the skulls with well-developed bases like sella turcica. There was identifiable pelvis and rudimentary limbs skeleton (Fig 3).

On histological examination, sections revealed fragments of mature tissue from all three germ layers. Skin with appendages (Fig 4), pseudostratified columnar ciliated epithelium, salivary gland, serous acini and neural tissue etc (Fig 5). In addition, interesting findings were eyeball (Fig 6) and optic nerve with surrounding muscle and orbital cavity, fully formed intestinal mucous membrane, cartilaginous model of limb showing endochondral ossification and bone marrow formation in the central cavity (Fig 7). No immature or malignant tissue was found in these sections.

DISCUSSION

Fetus-in-fetu may not be a distinct entity, but actually a mature, highly organized embryonal dysgenesis, or even a teratoma that has developed into architecturally well-defined and vascularized organs. It usually occurs retroperitoneally, but occasionally may occur at other sites.[4,5] The antenatal sonographic detection of a fetal mass that resembles a skeleton or other fetal parts mandates a neonatal evaluation with ultrasound and CT imaging.[11]

When discovered in a newborn child during physical examination, the differential diagnosis includes all the common masses such as Wilm's tumor, hydronephrosis and neuroblastomas.[12] In our case preoperative diagnosis was nephroblastoma.

Prenatally, the main differential diagnosis is with teratoma. By differentiation and induction, they can achieve striking organization, with examples of several organs being well formed. However, teratomas do not have vertebral segmentation, craniocaudal and lateral differentiation, body coelom or systemic organogenesis. Thus the presence of a mass with a spinal organization and surrounded by fluid suggests the correct diagnosis as it was in our case. Yet, the coexistence of a fetus-in-fetu and a teratoma as well as the occurrence of a teratoma 14 years after removal of a twin fetus-in-fetu have been reported, supporting the older hypothesis of a continuum between twin and teratoma.[13]

Cases of fetus in fetu behave differently from retained fetuses in abdominal pregnancies (i.e.,

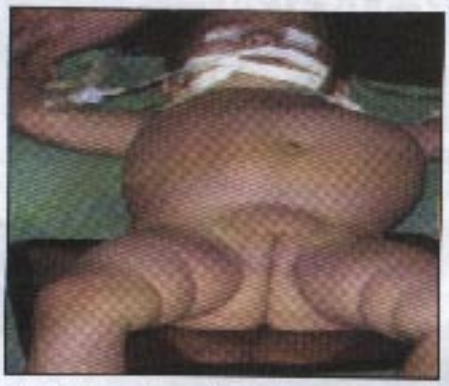

Fig 1: Asymptomatic flank mass which on USG, was reported to be a nephroblastoma

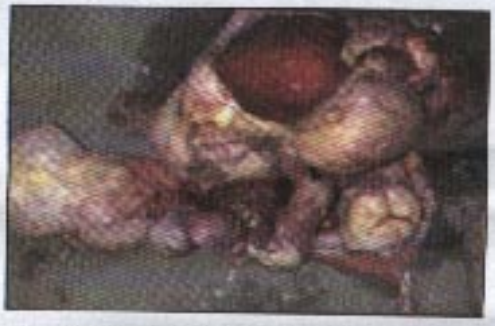

Fig 2: Gross appearance of mass after removal of it's covering. It was covered with vernix caseosa like material. Heart liver, two skulls and other distorted organs can be grossly identifed.

Fig 3: X-ray of specimen showing different skeletal structures.

Fig 4: Slide shows well developed skin and its appendages of fetus in fetu.

Fig 5: Microscopic appearance of brain tissue obtained from within the skull of one of the twins.

Fig 6: Low power Microscopic appearance of appearance of fully developed eyeball.

Fig 7: Photomicrograph of one of the long bones of the limbs.

the saponification and skeletonization of a longstanding abdominal pregnancy has not been described in cases of fetus in fetu).[10]

Surgical extirpation is the treatment of choice for fetus-in-fetu. We emphasize the need of proper prenatal investigations in suspected cases. Postnatally any intrabdominal mass should be subjected to thorough investigations and peroperative and postoperative confirmation of the mass with histopathology to verify the diagnosis as was done in this case, which helped us in arriving at a diagnosis of conjoined fetus in fetu with two well developed skulls, spines, heart and fused liver. It further improved our understanding by radiographic study of skeletons and detailed histopathological findings of different organs. We feel that several cases of fetus in fetu are not diagnosed because of lack of depth and detail of examination of specimen, skeletal survey and extensive histopathological examination of several areas of tissues from the mass.

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