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Case Report

Fetus in fetu: a rare entity

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ABSTRACT

Fetus in fetu is a rare congenital anomaly in which a malformed fetus grows within the body of its twin. It is detected as an abdominal mass in infancy. It is a heterophagus parasite of a diamniotic, monozygotic twin. It should be differentiated from teratoma which has no axial arrangement and has got malignant potential. Although fetus in fetu is a rare condition, diagnosis using imaging can be made before surgery. It should be considered as a differential diagnosis for lump abdomen in infants. Complete excision is the treatment of choice. We present a case of a 9 years old female with fetus in fetu who was successfully managed by surgical excision.

Keywords: Fetus in fetu, Teratoma, Heteropagus twins

INTRODUCTION

Heteropagus or parasitic twin is a grossly defective fetus, or fetal parts, attached externally, with or without internal connections, to a relatively normal twin (the autosite) in one of the same eight areas in which symmetrical twins are united.[1] They are usually composed of externally attached supernumerary limbs but may also contain viscera or visceral parts and only rarely a beating heart or intact brain. It is found usually in the abdominal cavity, rarely within the brain, with grossly recognizable fetal parts including an axial skeleton, attached to the autosite by a pedicle containing a few large blood vessels. Its growth rate is similar to the host within which it is discovered.[2]

A fetus in fetu is a monochorionic-diamniotic, monozygotic twin of its bearer. It is surrounded by a membrane analogous to the amniotic sac and supplied by a single feeding vessel. A true placenta is absent. Since there is absence of an independent circulatory system there is subsequent growth retardation. Developmentally, it has gone through the stage of the primitive streak so it has a vertebral body and organ arranged around the axis. This differentiates it from fetiform teratoma. The fetus in fetu is malformed because of pressure exerted by the host. It is rare and the incidence is 1 per 500,000 live births.[3]

The estimated incidence of heteropagus twins is much less than for symmetrical twins—1 per 1 million births or less.[4] The female preponderance seen in symmetrical twins is not evident in heteropagus twins where there is an equal distribution of the sexes.

It is postulated that heteropagus twins originated from symmetrical twins, one of which suffered secondary damage as a consequence of vascular compromise.[5] The affected "twin" would then have to rely on collateral blood supply from the autosite while ischemic damage occurred in various parts of the affected "twin." In support of this theory, hypoplastic umbilical vessels have been found in the heteropagus twin and vascular connections from the autosite to the heteropagus twin may be found during surgical separation. Others have disputed this theory because it cannot account for all the

abnormalities found in these twins and particularly cannot explain the acephalic occurrence. It is generally accepted that the heteropagus twin is genetically identical to the autosite, and this has been substantiated by DNA analysi.[6]

CASE REPORT

9 years old female presented with lump in abdomen since birth gradually increasing in size. There was no other gastrointestinal or genitourinary complaint. At clinical examination, a bulky, firm, rounded mass of 10×8×8 cm was palpated in the right hypochondrium extending upto right iliac fossa. The lump was seen to cross midline and palpated in left hypochondrium. Abdominal computed tomography revealed a large mass of 14×10.5×8 cm with cystic and solid calcified components in the retroperitoneum differential diagnosis being teratoma. Serum a- fetoprotein levels and b-hcg were within normal limits.

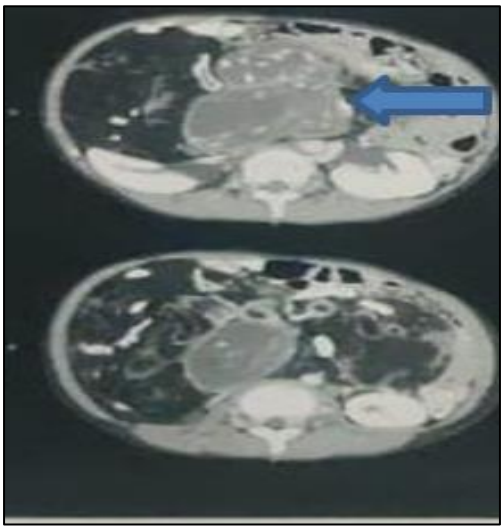

Figure 1: CT scan showing retroperitoneal tumour.

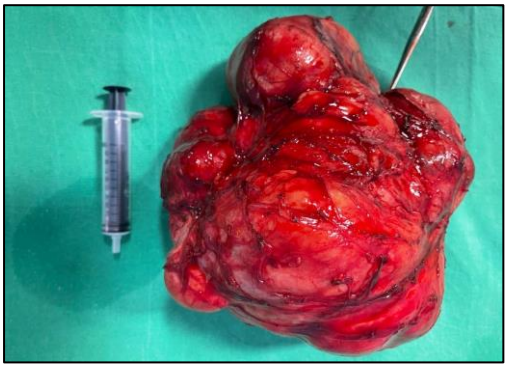

Figure 2: Gross specimen of tumour (fetus in fetu) weighing 980 gms.

Laparotomy was performed through a right transverse supraumbilical incision. At surgery, a thick walled cystic mass was found in the retroperitoneal region which compressed the inferior vena cava and medially displaced the duodenum. The tumor was encapsulated. The tumour seemed to receive blood supply from abdominal aorta. By opening the capsule, a fetiform structure floating in yellowish fluid was observed. Fetal head like structure with rudimentary vertebral column could be seen. Fetal limb like structures were also seen

Histopathology report stated-in view of presence of well developed limb bones, pelvic bones, presence of mature brain tissue, well developed respiratory system and colon, favour the diagnosis of fetus in fetu. No tissue showed immature, malignant cells.

The size of fetus was 18×12×9 cm weighing about 980 gms. The histopathology showed areas of mature adipose tissue, skeletal muscle and nerve bundles. It also showed presence of cartilage, bone and bone marrow.

Taking histopathological embryological development into account the gestational age of fetus was around 18 to 20 weeks.

On follow up after a month patient was asymptomatic and had not developed any complications.

Patient will be followed up yearly with Serum a-fetoprotein levels and b-hcg levels.

DISCUSSION

Fetus-in-fetu is a multiple aberrations of monozygotic diamniotic twinning with unequal division of the totipotent inner cell mass of the developing blastocyst leads to the inclusion of a smaller cell mass inside a maturing other embryo. The common presentation of fetus in fetu is abdominal mass 80% in the retroperitoneum. The fetus in fetu produces symptoms of mass effect leading to distention, difficulty in feeding, vomiting, jaundice, urinary retention.[7]

In this case during the surgery, the fetus was found to be encapsulated resembling a fetal membrane. It contained straw-colored sticky fluid and was connected by a peduncle to a vascular structure of the host. Preoperative diagnosis is possible with computed tomography and pre- or postnatal ultrasonography. A plain abdominal X-ray may be helpful in diagnosis. Though a rare anomaly, fetus in fetu can be identified in the preoperative period by radiological investigation. Radiological differential diagnosis includes teratoma and meconium pseudocyst. Maternal and host serum alpha-fetoprotein levels may be raised.[8]

Pathological controversy arises in differentiating a fetus in fetu from a mature well-organized teratoma. According to Willis, the presence of axial skeleton with the vertebral axis with limbs and organs goes more in favor of fetus in fetu.[9] On the other hand, teratoma is an accumulation of pluripotent cells in which there is neither organogenesis

nor vertebral segmentation. Another important aspect of fetus in fetu is that they never become malignant whereas teratomas are known to become malignant.[10]

Treatment of fetus in fetu is surgical and excision gives complete recovery. In our case, we completely resected the mass with no subsequent complication. It is necessary to keep the child in follow-up and surveillance as malignant recurrence has also been described.

CONCLUSION

Fetus in fetu is a rare and interesting entity that presents as an abdominal mass in infancy or early childhood. It can be diagnosed in the preoperative period with radiological investigations and complete excision of mass is curative and confirmatory. Though a rare entity, it should always be kept in mind as a differential diagnosis for lump abdomen in infancy and early childhood and should be differentiated from teratoma which is a common variant.

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