Original Research Paper



Paediatrics

FETUS IN FETU

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ABSTRACT Fetus-in-fetu (FIF) is an uncommon pathology with an incidence of 1 in 500000 births. Less than 200 cases have been reported worldwide. Multiple theories are proposed regarding embryogenesis ¹². The anomaly can be antenatally diagnosed or present at later age as abdominal lump or complications due to pressure symptoms. We report a case of antenatally diagnosed fetus in fetu which was removed in neonatal age.

KEYWORDS: Fetus in fetu

Case report

27 year old lady G3P2 was referred to us at 33 weeks of gestation with USG report suggestive of a well defined, mixed echogenic lesion in the fetal abdominal cavity. Multiple calcifications and osseous tissue resembling fetal bones were seen. Fetal MRI revealed a large cystic mass in fetal abdomen measuring 5x5x7 cm showing multiple soft tissue and bony parts suggestive of fetus in fetu. She delivered a girl at 37 weeks via LSCS with birth weight of 2.8 kg. Baby's vitals were stable. Per abdomen was distended with diffuse swelling, firm in consistency. CT scan suggests- a heterogenous mass with bony and soft tissue component measuring 10x8x8 compressing the IVC and abutting the aorta, lying in close proximity to the celiac trunk(Fig 1). As the child was getting distended we decided to operate. Upper abdominal transverse incision was taken. Duodenum was kocherised and retroperitoneum approached. The lump was enclosed in an amniotic sac like structure and was removed en mass taking care to preserve the vascularity and avoid damage to the surrounding structure (Fig 2). Surgery was uneventful. On opening the sac an anencephalic fetus with 4 limbs could be distinguished. (Fig 3)

Discussion

FIF was defined by Willis' in 1935 as "a mass containing a vertebral axis often associated with other organs or limbs around this axis". Since then, the presence of a vertebral axis in a fetiform mass has been considered pathognomonic of FIF

Besides the retroperitoneum which is the most common site of occurrence, it has also been described in the cranial cavity, oral cavity, mediastinum, lung, sacrococcygeal region, kidneys, intra-abdominal and scrotum ^{3,4}. It can present as an incidentally detected mass or with symptoms due to compression of adjoining structures in the abdominal, thoracic or cranial cavities. Demonstration of metameric segmentation of its spinal axis on imaging modalities usually clinches the diagnosis.

Prenatal diagnosis of FIF is possible in about 15% of reported cases 4 .Prenatal MRI can also be useful. Serum markers like AFP and β hCG may be normal or marginally elevated $^{5.6}$. The most important differential diagnoses to be considered are teratoma and meconium pseudocyst; both of which will also show calcifications on plain radiography 7 .

Detailed and meticulous gross as well as histopathological examination of the excised specimen is the gold standard for diagnosing FIF. Complete surgical excision is the treatment of choice as malignant recurrence has been reported when the membrane was left behind ⁸. For the same reasons, these patients should be kept under close follow up with serial ultrasounds and serum tumour markers.

Theories of origin of fetus in fetu included twin theory which regards FIF as a rare form of monozygotic twinning whereby an aberrant asymmetric twin becomes internalized in the other twin thus acting endoparasitically. Beaudoin's 5 theory of defective implantation during the second week of development resulting in the invasion of a second embryo (that becomes a homunculus) into the extra-embryonic mesenchyme of the host foetus or autosite, instead of the uterine wall, appears plausible.

The absence of normal umbilical vessels and a definite vascular connection explain the growth retardation and arrest of organ differentiation in almost all cases of FIF ¹.

The presence of an axial skeleton implies development of the included fetus past the primitive streak stage when the notochord is formed, a stage thought to be too developed for formation of a teratoma ⁴

Fetiform teratoma theory: Willis nurtured the theory that teratomas were derived from embryonic pleuripotential cells associated with the primitive streak, which escaped organizer influence to form a true neoplasm, which may later exhibit benign or malignant characteristics

To conclude, with improvement in imaging modalities, more cases of FIF are being diagnosed preoperatively and even prenatally. This has led to successful treatment and better survival

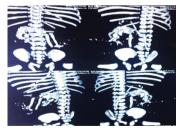


Fig 1: CT scan showing fetal skeleton



Fig 2: Retroperitoneal mass



Fig 3: Fetus in fetu

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