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Case Report

Imaging findings in Fetus-in-fetu-misdiagnosed commonly as teratoma

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ABSTRACT

Fetus in fetu is a rare condition associated with abnormal embryogenesis in a diamniotic, monochorionic pregnancy, wherein one of the fetus is enclosed within the body of another normally developing fetus. It should be differentiated from a teratoma because of the later's malignant potential. Here we report a case of 2 months old girl child who presented with complaints of abdominal distension. USG showed a solid cystic retroperitoneal mass resembling an anencephalic fetus. Contrast enhanced Computed tomography (CT) showed similar findings with visualization of bones resembling femur, sacrum and vertebrae. Findings were correlated with MRI and post-op pathology. The preoperative diagnosis of FIF is based on the observation of vertebral column or limbs in a mass on imaging modalities and our case meets the required criteria.

Keywords: Fetus-in-fetu, teratoma, Ultrasonography, Computed tomography, Magnetic resonance imaging

INTRODUCTION

Fetus in fetu (FIF) is a rare condition associated with abnormal embryogenesis in a diamniotic monochorionic (DAMC) pregnancy, wherein one of the fetus is enclosed within the body of another normally developing fetus.[1] The incidence is estimated to be 1/500,000 births.[2] FIF is mostly anencephalic, but in almost all cases its vertebral column and limbs are present.[2] At the same time, its lower limbs are more developed than the upper limbs. Other possible mechanism for fetus in fetu is a highly differentiated teratoma. It should be differentiated from a teratoma because of the later's malignant potential. Preoperative diagnosis is based on radiologic findings.

Other reported locations include cranial cavity, mediastinum, ovary and scrotum.[1] There are reports on antenatal detection of FIF within the abdomen of foetuses using 3-dimensional ultrasound.[7] In most cases of FIF, only one fetus exists inside the baby. Only in extremely rare cases are multiple foetuses found.

Here we report a case of 2-month-old baby with abdominal distension due to a foetoid tumor like mass which was confirmed as FIF.

CASE REPORT

A 2-month-old baby girl with complaints of abdominal distension was referred to our department for computed tomography (CT) examination. Physical examination revealed a well-developed and well-nourished girl. No family history of twinning was present. There was no significant prenatal history. It was a normal term vaginal delivery and postnatal period was uneventful. The parents brought the baby to the hospital for excessive crying and a subtle abdominal distension. On local examination, abdomen was grossly distended with a bulge of left lateral abdominal wall. A non-tender mass of variable

consistency was palpated, measuring approximately the size of a cricket ball. No abnormal sounds were auscultated over the swelling. Ultrasound of the abdomen was performed, which showed a large, predominantly cystic lesion with central solid heterogeneous mass in the abdomen. Imaging resembled a rudimentary fetus with anencephaly and ill defined lower lumbar vertebrae/ sacrum and iliac bones. There were parallel, linear calcified areas resembling fetal long bones. Minimal vascularity was appreciated on color doppler with a vessel showing arterial flow at the periphery of the mass (Figure 1).

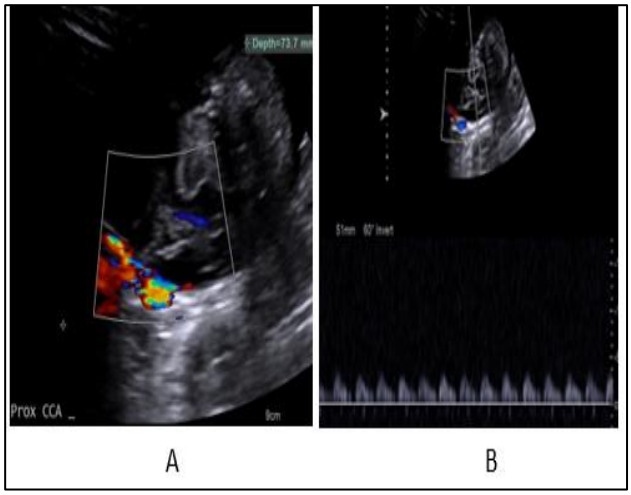

Figure 1: Colour doppler (A) showing a vessel within the mass which on pulse wave doppler (B) shows arterial flow.

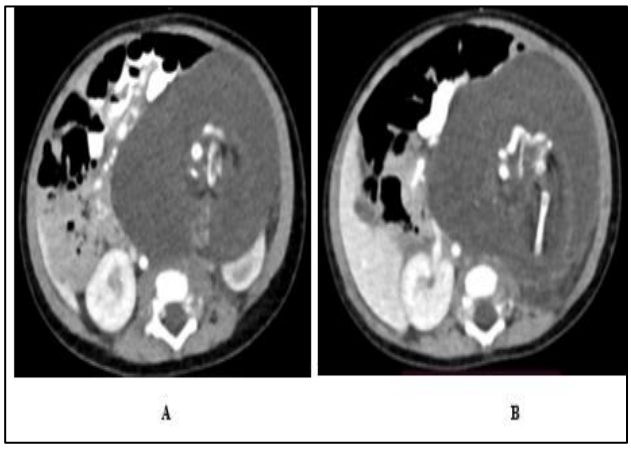

Figure 2: CECT axial section at the level of upper abdomen shows a well-defined predominantly cystic, retroperitoneal mass (A) containing rudimentary limb bones, sacrum and iliac bone within (B).

Plain and postcontrast CT scan of abdomen was performed on a 128 slice multiple detector CT scanner after intravenous sedation and non-ionic water soluble contrast. CT scan showed a 7.7×7.9 ×7.7 cm (AP×TRA×CC), well-circumscribed retroperitoneal mass lesion with bulk towards left side. The lesion contained fatty tissue surrounding a central bony structure having foetoid morphology (Figure 2). The bony structure resembled rudimentary spine, pelvis and femur bone. No bony structure resembling cranium was seen. It had cystic areas surrounding it, with few thin septations in the lower part. Blood supply to the lesion was from Superior Mesenteric artery (Figure 3). The lesion was producing mass effect on the left kidney and displacing it postero-inferiorly without any features of infiltration.

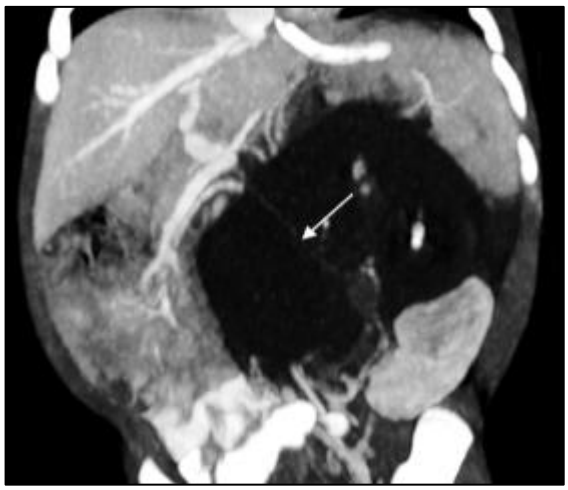

Figure 3: Coronal MIP image showing the artery (arrow) supplying the mass is arising from the SMA.

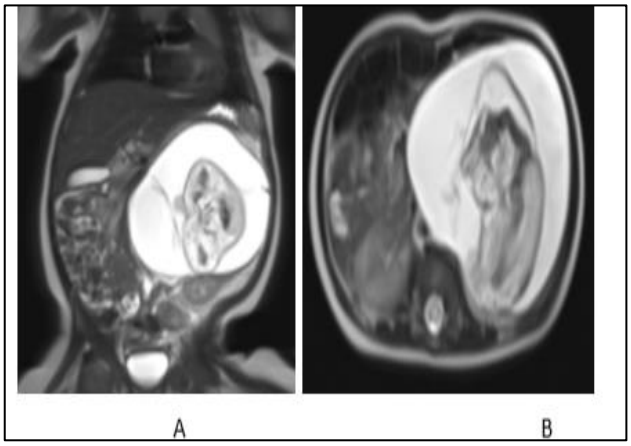

Figure 4: T2 weighted MRI in coronal (A) and axial (B) planes depicting the solid cystic mass with bony structures surrounded by soft tissue.

MRI of the patient was done which also revealed a solid cystic retroperitoneal mass lesion on left side with centrally placed bony structures surrounded by soft tissue (Figure 4).

The infant underwent open surgery and complete removal of the mass lesion. The gross specimen was a large cystic lesion covering a soft tissue by membranes. On removal of the membranes and fluid, it revealed a soft tissue component which resembled a thigh with pelvis and

rudimentary spine attached to the membranes. (Figure 5) The infant was discharged on day 10 with uneventful follow up.

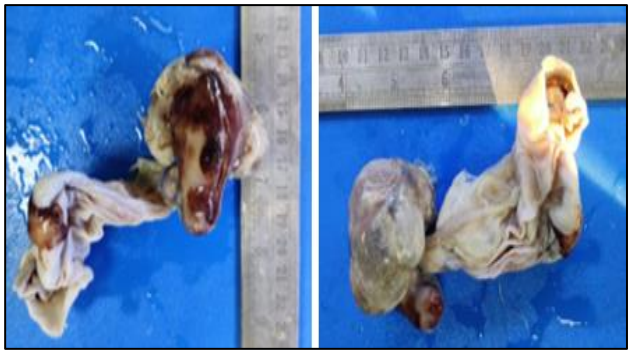

Figure 5: Post-op gross specimen showing the mass lesion resembling one thigh, pelvis and rudimentary spine attached to the membranes.

DISCUSSION

The term "fetus in fetu" was first described by Johann Friedrich Meckel during the late 18th century. Later Willis described it as a rare condition where a parasitic twin resides in the body of its host usually the abdominal cavity.[11] Exact embryogenesis is controversial. However, two theories have been proposed. One describes FIF as a variant in the spectrum of monozygotic twinning and the other labels it a highly differentiated teratoma.[1]

According to the first theory, FIF represents an aberration of monochorionic diamniotic twinning in which unequal division of the totipotent inner cell mass of the developing blastocyst leads to the inclusion of a smaller cell mass within a maturing sister embryo.[10] Miura S et al found that both the host and the FIF have the same genotype and said that two blastocysts are formed from a single zygote and one gets implanted into the other.[2] Identification of the vertebral column indicates that the fetal development of the included twin must have advanced at least to the primitive streak stage to develop a notochord, which is the precursor of the vertebral column.[11]

According to the second theory, FIF is described as a development of high organotypic type of teratoma with formation of vertebra and arrangement of tissues around it. A teratoma consists of pluripotent cells but organogenesis or vertebral segmentation is unusual. One observation pointing towards the teratoma theory is the occurrence of a mass in the expected locations of teratoma in an infant, like retroperitoneum and ovaries. Another observation holding against a twin fetus theory is the occasional teratoma formation after the FIF resection in the same surgical site.[5] Sometimes teratomas contain well-developed foetiform structures like brain and spinal cord. But the occurrence of vertebral segments is never reported. Features highlighted by Spencer to differentiate teratoma from FIF are- lesser organization of microscopically identifiable tissues in the former and broader attachment site with multiple smaller blood vessels.[1]

The usual presentation of FIF is as a solitary mass in infants and young children. Few cases of two FIF in single host have been reported.[9]

From a radiological and surgical perspective, making a preoperative diagnosis and differentiating it from the usual and more common congenital teratoma is important. This is because of the fact that there is an associated 10% chance of malignancy developing in a teratoma, whereas FIF is a benign condition. The presence of a complete or partial vertebral column plus other appropriately situated axial or appendicular bones or organs is the hallmark of FIF and differentiates it from teratoma. Visualization of the vertebral column and limbs forms the key differentiating feature to aid diagnosis. The Willis criterion stresses much emphasis on the development of axial skeleton and vertebral axis.[1]

Imaging plays a role in accurate preoperative diagnosis. Ultrasound also helps in early diagnosis of FIF and there are few cases diagnosed antenatally also by ultrasound.[7] CT scans especially with multiplanar and volume rendering capabilities can correctly identify the vertebral column and other skeletal elements. It is also useful in correctly detecting fat and soft tissues. Possible pressure effect on the adjacent organs as well as the feeding arteries and draining veins can be correctly identified. Usually, the tumor derives its blood supply from the rich capsular vascular network, usually derived from the plexus where the sac is attached to abdominal wall. A few cases had definite vascular connections from major vessels like in our case it is from the SMA. In our case, a correct diagnosis of FIF was possible due to the visualisation of fetal pelvis, long bone and vertebrae. The mass was removed in toto with surgical clipping of the supplying and draining vessels.

CONCLUSION

The preoperative diagnosis of FIF is based on the observation of vertebral column or limbs in a mass on imaging modalities which is present in our case. The treatment of choice for FIF is complete resection. Postsurgical follow-up imaging and serum AFP and HCG monitoring is the recommendation to rule out recurrence. In conclusion, the case presented in our report meets all the accepted criteria of an abdominal fetus-in-fetu.

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