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Case Report

Fetus in fetu presenting as suprarenal mass: a case report

Md Nawad Azam[1]*, Baikuntha Narayan Mishra[2], Subrat Kumar Majhi[1], Ranjit Kumar Joshi[1], Prabin Prakash Pahi[1]

[1]Department of Pediatrics and Neonatology, AMRI Hospital, Bhubaneswar, Odisha, India
[2]Department of Pediatric Surgery, AMRI Hospital, Bhubaneswar, Odisha, India

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***Correspondence:**
Dr. Md Nawad Azam,
E-mail: dr.nawed2007@gmail.com

ABSTRACT

Fetus in fetu (FIF) is a rare condition in which baby harbors its monozygotic malformed twin fetus inside its own body. This is even rarer in supra renal location, mostly asymptomatic but may present as a palpable abdominal lump. Here in we describe one newborn male child with antenatal diagnosis of mass abdomen. Post-natal ultra-sonogram of abdomen was suggestive of left supra renal mass. Operative findings and histopathology were consistent with fetus in fetu.

Keywords: FIF, Newborn, Suprarenal mass, Teratoma

INTRODUCTION

Fetus in fetu (FIF) is an uncommon condition due to defect during embryogenesis leading to development of abnormal smaller monozygotic twin which get encased into normally developing twin fetus. The development of abnormal fetus ceases after a while and presents as a lump postnataly and can also be diagnosed by antenatal imaging. In 19th century Meckel reported the first case and less than 200 cases have been reported so far.[1,2] It has an incidence of 1 in 500,000 births.[3] Common age of presentation is early infancy although oldest case reported was 47 yrs.[4] It has almost equal incidence in both male and females.[5] Usual presentation is intra abdominal mass, mostly in retro peritoneum.[6] X-ray, USG followed by CECT/MRI abdomen are the usual investigations. Intra-abdominal FIF should be differentiated from teratoma, though it is not well established. Axial differentiation and metamerisation is an important finding in FIF.[7] Teratomas have higher malignant potential and needs long term follow up. Complete surgical excision is the treatment of choice.

CASE REPORT

A 28 year old mother delivered a term 3.04 kg male child by LSCS in our hospital with antenatal ultrasound suggestive of intra-abdominal mass in fetus. Baby was admitted to NICU and initial examination showed no dysmorphic features. Baby's vitals were normal, abdominal cystic mass of size 5 X4 cm, was palpable in left flank. Initial post-natal ultrasonography confirms a well-defined solid and cystic lesion of size 58 X 46mm with well-formed bones within seen over left supra renal location. Plain X-ray abdomen showed irregular calcifications along with a soft tissue shadow in left renal area. CT scan of the abdomen was suggestive of a cystic lesion of the size 69 X 67 X 71 mm (AP x ML x CC) with a large solid component inside. Multiple bony elements were noted in the solid component. The spleen was displaced anteriorly and left kidney was displaced inferiorly due to mass effect (Figure 1). Serum α-feto

protein level and beta HCG levels were normal. Exploratory laporatomy was performed through trans peritoneal approach and a cystic lesion was found (Figure 2) in the supra renal location displacing the left kidney down ward, spleen anterior and upward. The cyst was closely adherent to left supra renal gland. Cyst contained straw colored fluid with a fleshy mass and the cyst wall resembled amniotic membrane (Figure 3).The mass had a stalk appearing like umbilical cord with single vessel (Figure 4) .No placental tissues identified. The cut section showed bony elements (vertebra) inside the mass (Figure 5).

The entire mass was dissected free from adrenal tissues and excised completely. Baby had an uneventful recovery and discharged home on 7th post operative day. Histopathology was consistent with FIF. He is on regular follow up.

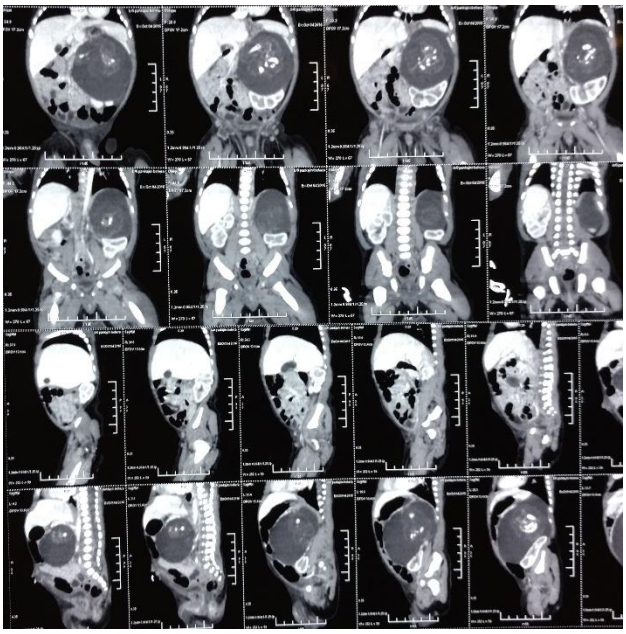

Figure 1: CECT abdomen showing suprarenal mass displacing spleen anteriorly and kidney inferiorly.

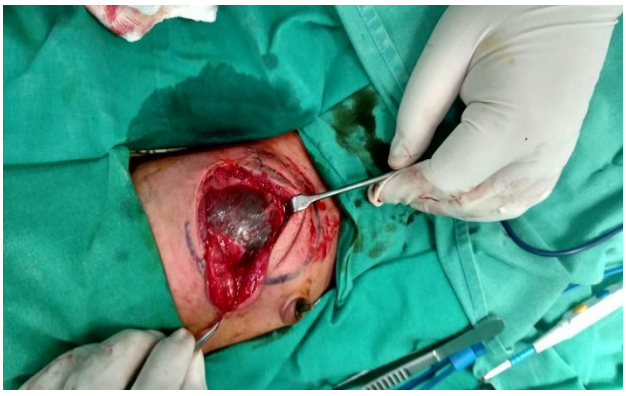

Figure 2: Exploratory laporatomy showing cystic mass.

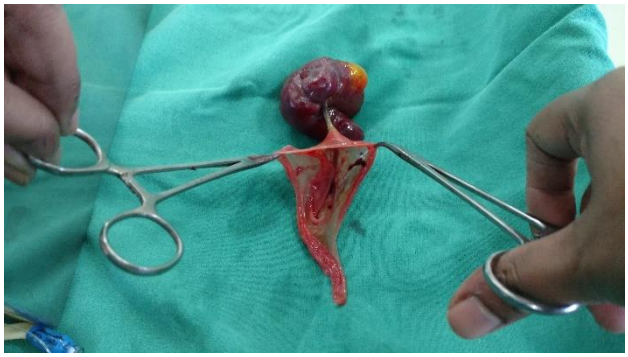

Figure 3: Dissected mass showing cyst wall and a membrane resembling amniotic membrane.

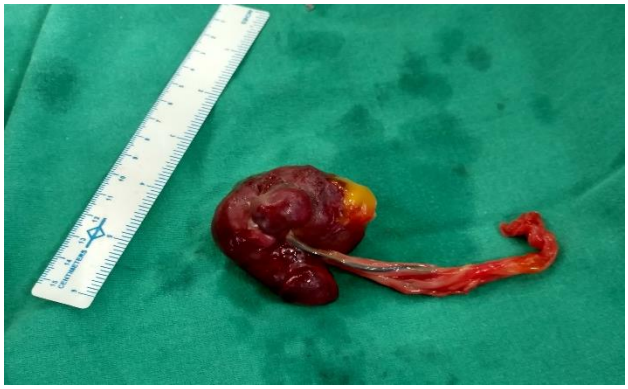

Figure 4: The mass had a stalk appearing like umbilical cord with single vessel.

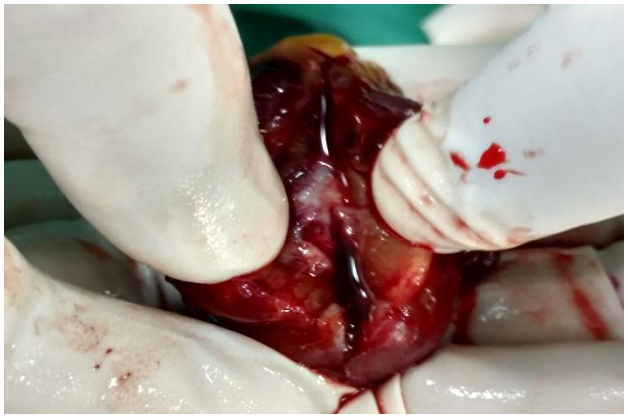

Figure 5: Cut section of mass showing bony elements (vertebrae).

DISCUSSION

Fetus in fetu is an extremely rare congenital anomaly, resulting from an abnormal embryogenesis in a diamniotic monochoroinic twin pregnancy. Antenatal ultrasound can detect fetus in fetu but a cystic mass in abdomen is the most common presentation. It may remain asymptomatic for a long period. Sometimes it may present as feeding intolerance, jaundice, vomiting, hydronephrosis and intestinal obstruction due to pressure

effect.[5,6] In our case mass abdomen was detected antenatally and no other pressure symptoms were noted.

Fetus in fetu is mostly retroperitoneal in location.[6,8] Other atypical sites include intracranial, oropharynx, neck, adrenal gland, mediastinum, lungs, ovary and sacro-coccygeal region.[5,8-13] Post-natal ultra sonogram and CECT abdomen were suggestive of a left supra renal cystic mass in our patient.

Ultrasound remains the first line of investigation to detect fetus in fetu. CT scan/MRI imaging adds to diagnosis of FIF. According to Knox and Webb, final diagnosis of FIF is made only after excision and histopathological examination.[14]

Suprarenal mass in infancy has varied diagnosis ranging from benign adrenal hemorrhage, intra-abdominal pulmonary sequestration, teratoma to malignant lesions like neurblastoma. Fetus in fetu is a rare entity with only few cases are ever reported as supra renal mass.

FIF is surrounded by a membrane resembling amniotic sac, float in a cystic fluid and supplied by a single feeding artery, Placenta like structures are not usually present.[7] We had similar findings.

The development of aberrant fetus has passed through initial stages of gastrulation, symmetrical development around neural axis and sometimes metamerisation before being halted due to lack of independent circulatory system.[15,16] The vertebral column and limbs were present in 91% and 82.5% respectively.[17] Cut sections of the specimen in our case showed vertebral bodies, no long bones detected.

There is a controversy, whether FIF is a distinct entity or a mature teratoma. Teratoma is a neoplasm, having malignant potential; while FIF is benign. Though differentiation is difficult it is important to distinguish FIF from teratoma. Raised level of Alpha feto protein and beta HCG are useful marker for teratoma. Willis et al suggested that identification of vertebral column is pathognomic in differentiating FIF.[7] But recent studies by Lagausie et al found that absence of vertebral column does not rule out the possibility of FIF.[18] Infact in 9% of cases of FIF, no vertebral column was identified. In such case, high degree of organogenesis has to be taken into account, as suggested by Gonzalez-Crussi.[19]

Complete excision is the treatment of choice with careful dissection to avoid injury to surrounding tissue. Excised mass is usually sent for Histopathological study to confirm fetal tissue. Histopathology was consistent with FIF in our case. USG abdomen of the baby was found to be normal on follow ups.

FIF in supra renal location is rare, complete excision is the treatment of choice. Close follow up needed to detect recurrence in remnant tissue, if any.

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