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Case Report

FETUS IN FETU WITH JAUNDICE- A RARE PRESENTATION OF A RARE DISEASE

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Abstract:

Fetus and fetu is a rare mysterious medical phenomenon in which an acardiac fetiform mass is commonly located in abdomen of a neonate or infant. We report on a case of a 7 month-old girl with a gradually enlarging right upper abdominal mass and progressive jaundice, whose plain abdominal radiograph, ultrasonography, and computed tomography scan revealed a mass in which the contents favor a fetus in fetu. Obstructive jaundice was caused by billiary obstruction by FIF .

Keywords: Fetus in fetu, Obstructive jaundice in infant, Liver cirrhosis in infant.

Introduction:

Fetus in fetu (FIF) is a rare congenital anomaly. It is a condition in which malformed and parasitic fetus is located in the body of its twin. The anomaly was first defined in early nineteenth century by Meckel. This rare congenital anomaly, which was reported around 69 times among pediatric age group since its first definition in the nineteenth century very rarely present with obstructive jaundice.

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Case report:

Our patient Sazia Jannat, weighing 7kg, 7 months old girl child of her nonconsanguinous parents hailing from Jamalpur admitted in DMCH under PSU-I on 13th March, 2015, with the complaints of –

- Abdominal lump for 3 months
- Yellowish discoloration of skin for 1 month
- Itching over whole body for 1 month

The abdominal lump was incidentally found 3 months back which gradually increased in size. She developed yellowish discoloration of skin and itching all over the body with no associated rash for last one month. She passed whitish stool. Her symptoms were not associated with fever, vomiting, shortness of breath. Her mother had not under regular antenatal check-up and undergone LUCS on 38th week of pregnancy due to less fetal movement.

The playful baby was mildly anemic and moderately icteric. The intra-abdominal, non-pulsatile lump with well defined margins, occupying the right side of umbilical region and part of right hypochondriac region was firm to hard in consistency, about 11 x 10 cm in size, globular in shape, non tender, smooth surfaced, did not move with respiration, slightly mobile from side to side and less in above downwards. The lump seemed to be fixed to the underlying structures but free from the overlying abdominal wall.

The lump is free from lower border of the liver. Liver was just palpable, about 1 cm from costal margin in mid clavicular line. Her breath sound was bilateral vesicular with ronchi in both lungs.

Reports of routine investigations were within normal limit. LFT revealed raised S. Billirubin and S. Alkaline phosphatase. Serial S. Billirubin was 8.3 gm/dl (14/03/15), 7.3 gm/dl (18/03/15), 6.4 gm/dl (24/03/15), 4.84 gm/dl (29/03/15) & 4.19 gm/dl (04/04/15).Repeated study of S. Alkaline Phosphatase was 604 U/L (18/03/15) & 496 U/L (29/03/15). SGPT, Prothrombin Time, S. Total protein was within normal limit. S. á Feto-Protein was 37.6 IU/ml and S. âhCG was <2.39 ng/ml, both were within normal value. Viral markers were negative. Urine RME and Urine C/S revealed no abnormality.

Chest X-ray was normal. X-ray Abdomen showed a large radio-opaque lesion in right hypochondrium.

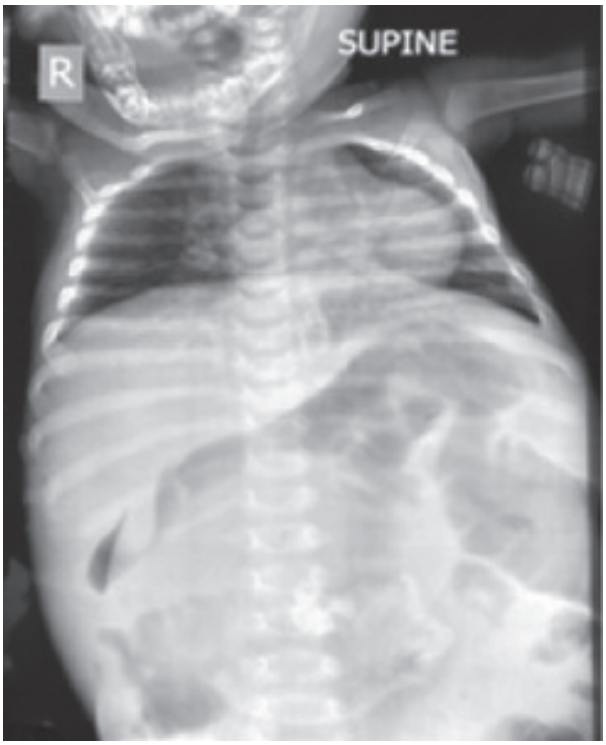

Fig.-1: *X-ray abdomen showing a large radio-opaque lesion.*

USG of whole abdomen revealed abdominal swelling corresponds to huge complex mass (about 11×10 cm) in the central abdomen & displacing the intestinal loops at the periphery. There is a deformed fetus within the cystic component, skull, feet & long bones could be identified & other structures could not be clearly delineated. Umbilical cord like structure was floating in the cystic component. BPD of fetoid mass was 22 mm, corresponds to about 12-13 wks of gestation, FL= 33 mm, corresponds to about 20 wks of gestation. Liver was normal in size (6cm vertically) with uniform parencymal echotexture; no focal lesion seen; hepatic veins are not congested. CBD is mildly dialated (7.4 mm in diameter) along with dialated intrahepatic billiary tree in both lobes.

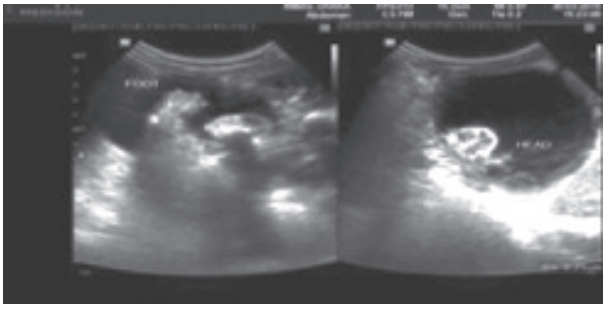

Fig.-2: *2D USG view of deformed foot and head of fetus in fetu.*

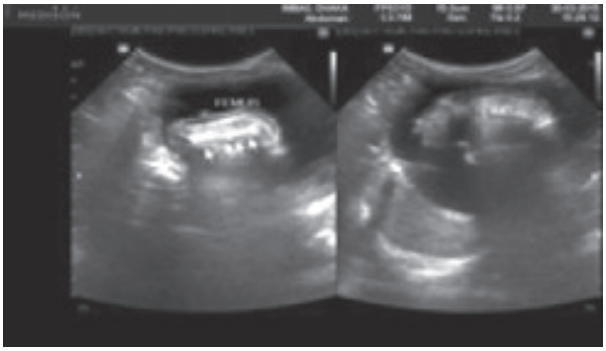

Fig.-3: *2D USG view of femur and limb of fetus in fetu*

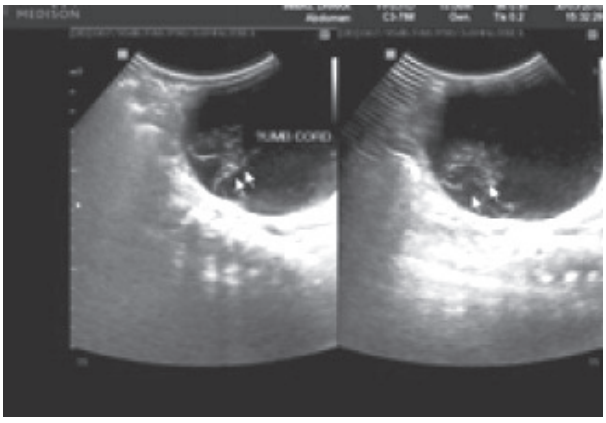

Fig.-4: *2D USG showing umbilical cord attached with the fetus in fetu*

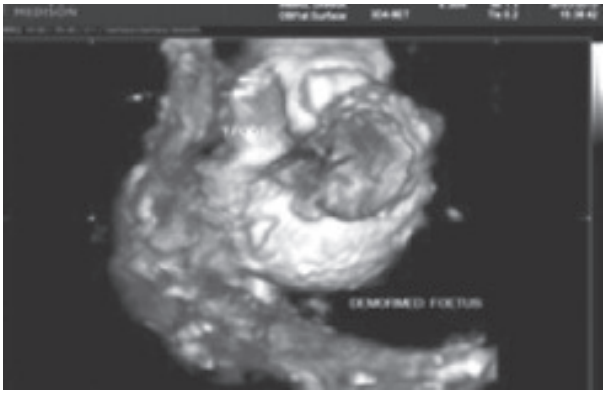

Fig.-5: *3D USG view showing deformed fetus.*

Contrast CT scan of whole abdomen revealed a large well defined cystic mass of about 11x9.5 cm having anencephalic, acardiac soft tissue mass with identifiable extremities and spine within it is noted in central part of abdomen extending up to pelvic cavity. The mass is separated from adjacent organ but compression over the liver resulting dilatation of intrahepatic billiary channels. The mass causing displacement of bowel loops upward, forwards and laterally. The mass also compressed upon both kidneys. No ascites of intra-abdominal lymphadenopathy is noted. Rest of the scanned portion is normal. Final comment by specialist radiologists was a very large abdominal mass suggestive of fetus in fetu and billiary obstruction.

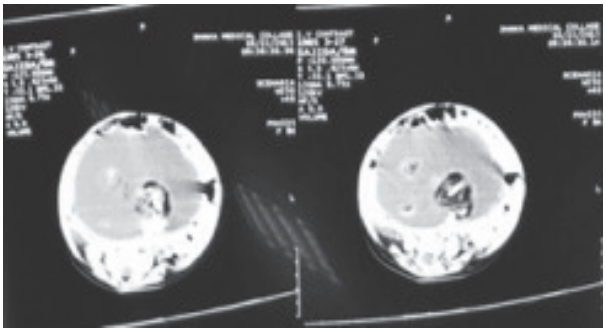

Fig.-6: *CT scan showing intra-abdominal fetiform mass*

Multidisciplinary medical board evaluated the baby and advised oral antibiotics for chest infection and proper hydration. Exploratory laparotomy followed by removal of deformed fetus with liver biopsy was the surgical plan. After optimization of the patient satisfactorily, surgery was scheduled on 15.04.2014.

After proper sign in and time out, right supra umbilical transverse incision with medial extension was made. Per-operative findings were deformed fetus presenting well formed lower limbs, one rudimentary upper limb, spina bifida and anencephaly. It was surrounded by a separate gestational sac about 22 wks size. Umbilical cord was communicated with the mesenteric vessels. Cyst contained yellowish fluid. Liver was shrunken & cirrhotic. Umbilical cord transfixed and separated. Deformed fetus was removed and gestational sac excised. Fetus weighed 415 gm. Liver biopsy was taken. A wide bore drain tube was kept in left hepatorenal pouch. 140 ml screened and matched fresh whole blood transfused per-operatively. Reverse from anaesthesia was smooth. Her immediate and early post-operative period was uneventful. Sips of plain water were started from 2nd POD and feeding was started from 3rd POD. Alternate stitches were removed on 8th POD and all other stitches on 9th POD.

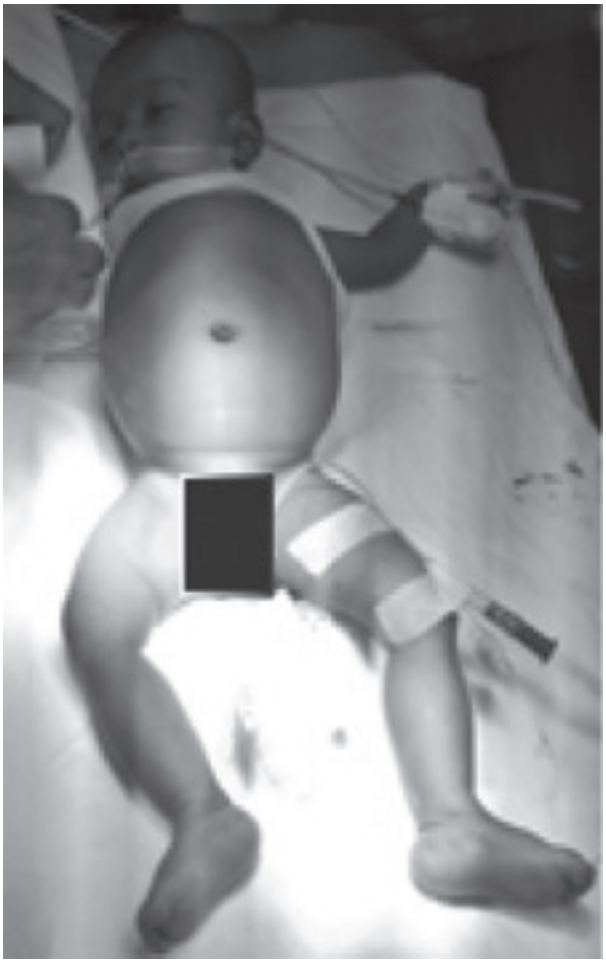

Fig.-7: *Pre-operative picture of the patient*

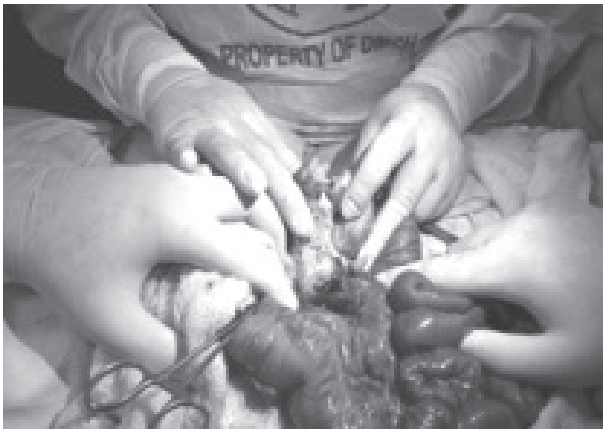

Fig.-8: *Per operative picture of Fetus in fetu, showing adhesion with the mesentery.*

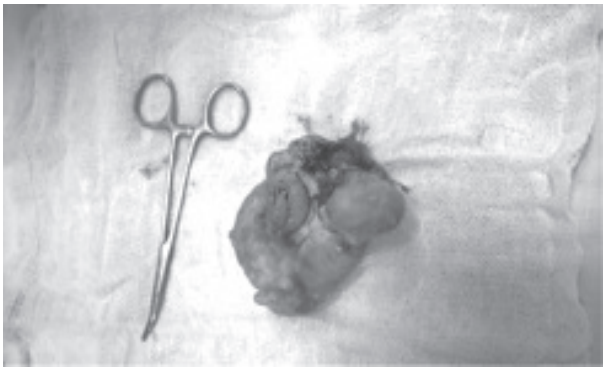

Fig.-9: *Specimen of fetus in fetu, arrow showing the cranial part & arrowhead showing the under-developed lower limb.*

Liver biopsy stated sections of submitted liver tissue show degenerating nodules encircles by fibrosis. It also reveals intrahepatic cholestasis & bile duct proliferation. Knodell scoring- Piecemeal necrosis-Absent, Necrosis, apoptosis & focal inflammation-Mild (score 1), Portal inflammation- Mild (score 1), Fibrosis- Mild (score 10), Total Histology Activity Index (HAI)- 03/22. Report of pathologist was Active Cirrhosis, Total Knodell score- 3/22.

Patient was discharged on 11th POD, with follow up advices. Liver function test, S. á Feto-Protein and USG of whole abdomen with special attention towards hepato-billiary system was planned for follow up. Unfortunately, patient did not come for follow up.

Discussion:

Fetus in fetu has been defined as the existence of a parasitic, monozygotic, diamniotic fetus in the body of its twin. Preceded by Willis in 1935, in 1954, Lord claimed the presence of a vertebral column and extremities and organs located at appropriate places around it as the basic diagnostic for FIF. These criteria are still, to a wide extent, valid today. However, there are those who claim this pathology to be a teratoma that is well-differentiated and highly organized[1]. Our patient had the basic diagnostic features of FIF.

Fetus in fetu most frequently (80%) inhabits the retroperitoneal region. However, there have been few reports of FIF location in the head, sacrum, scrotum and the mouth. Despite the requirement of the presence of a vertebral column for diagnosis, there are reports of the cases without a vertebral column[1]. FIF presented as features of obstructive jaundice was not yet reported. Our provisional diagnosis for this patient was Choledochal cyst and differential diagnoses were teratomas, omental cyst and right ovarian cyst.

Fetus in fetu is considered as a benign condition. Consequently, some researchers have stated that to facilitate the excision, it is possible to leave some sections of the capsule in its place. Nevertheless, in one case, the mass has been reported to recur as a yolk sac tumor after 4 months. This has been attributed to the presence of immature tissues in the small areas and the remnants of the capsule of the mass.

In summary, FIF is considered as a benign condition, while the potentially malignant characteristics of teratoma constitute the basis of the discussion. This argument may lead to differences in the follow-up and treatment of such cases. Although there is a consensus on FIF being a benign condition, considering the reports of malignant recurrences, as stated above, there is a need for the total removal of the mass including its capsule. In addition, we consider the evaluation of the postoperative tumor markers and periodical ultrasound examinations is an appropriate approach.

A. N. Gangopadhyay stated that the prognosis for FIF is more favorable than for cystic teratoma, the presence of immature elements nevertheless indicates the need for close clinical, radiological and serological (AFP) follow-up. Despite the AFP levels before and after surgery remaining at normal values,

a possible recurrence of a malignant teratoma after FIF resection must best be kept in mind. This was the reason why we continued to monitor the serial tumor marker levels of our patient, while also conducting cross-sectional imaging follow-up examinations[2].

Conclusion:

Fetus in fetu causing obstructive jaundice in its host is a rare presentation of this rare diagnosis.

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