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Review Article

Fetus in Fetu: A Review of Rare Malformation with Imaging Findings

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ABSTRACT

Fetus in fetu is an extremely rare (1/500,000 live births) congenital anomaly and is defined as a malformed fetus which grows within the body of its twin. It is often overlooked in the differential diagnosis of an abdominal mass in infants or children. Unlike teratomas, fetus in fetu is a benign condition and has no malignant potential. Fetus in fetu is a rare condition with less than 200 cases reported in the world to the best of our knowledge. Although rare, fetus in fetu should be considered in differential diagnosis of paediatric and infantile abdominal masses and in presence of typical Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) findings, appropriate diagnosis can be made. The present article is an effort to review the epidemiology, etiopathogenesis and imaging findings in fetus in fetu.

Keywords: Computed Tomography; Fetus; Congenital; Malformed; Mass

INTRODUCTION

Fetus in fetu is a rare congenital anomaly and typically presents in infancy and early childhood. [1] It is a rare cause of abdominal mass in children. The term Fetus in fetu was first coined by Meckel and defined by Willis in 1935 as a fetiform mass in which organ-like tissues are arranged around a vertebral axis. [2,3] Fetus in fetu arises from an inclusion of a monozygotic diamniotic twin within the bearer. Presence of vertebral axis and appropriate arrangement of other organs or limbs with respect to the vertebral axis are the diagnostic features of fetus in fetu and help in differentiating this entity from the highly differentiated teratoma. [4] Majority of cases is detected before 18 months of age and present as an asymptomatic slow growing abdominal mass. [5]

Majority of cases consist of pedunculated vertebrate masses within a capsule containing fluid and with an umbilical cord composed of only 2 vessels. [6] Majority of them are seen in the retroperitoneal space. [6] Rare locations include skull, liver, pelvis, scrotum, sacro-coccygeal region, mouth, adrenal gland, mesentery and right iliac fossa. [6,7] Normally a fetus-in-fetu is single in number, but a few cases of more than one have also been reported. [7] Fetus in Fetu represents a diamniotic, monochorionic, monozygotic twin that becomes incorporated into the body of the host twin after anastomosis of the vitelline circulation. [8]

Pathogenesis:

Fetus-in-fetu is a rare entity and results from abnormal embryogenesis in diamniotic monochorionic twins. It is a malformed or parasitic monozygotic

diamniotic twin inside the body of a living child or sometimes in an adult. [9]

The pathogenesis is no exactly known but is believed to be due to inclusion of the diamniotic monochorionic twin within its host during the ventral folding of trilaminar embryonic cyst. [10] The inclusion in the sister embryo is thought to be due to persistent anastomosis of the vitelline circulation during development. [11] It is thought to result from unequal division of the totipotent inner cell mass of the developing blastocyst, causing a small cell mass within a maturing sister embryo. This ultimately results in a vestigial remnant, or fetus in fetu. [12] Superior mesenteric artery develops from vitelline circulation and thereby explaining the retroperitoneal location of the lesion [13] and absence of independent blood supply may account for fetal growth restriction in all cases.

Clinical Presentation:

Fetus in fetu is mostly detected before 18 months of age and present as an asymptomatic slow-growing abdominal mass with abdominal distension. [6] However age of presentation varies from ante natal period upto as old as 47 years. [14] When extremely large, patient may present with dull aching abdominal pain. Vomiting and other symptoms due to pressure effect have also been documented.

Imaging Findings:

Increasing use of obstetric ultrasound has made prenatal diagnosis of fetus in fetu more common. [15] In the antenatal period, the diagnosis may be suggested when a defined encapsulated cystic mass with solid and calcified components is seen. The mass often increases in volume throughout the gestation period and the calcified components take on the appearance of fetal skeletal bones. [16]

Post natal imaging modalities include X-rays, Ultrasonography (USG), CT and MRI. X-rays reveal an amorphous soft tissue density mass with numerous calcifications resembling bony structures. On USG, fetus in fetu appears as a large heterogeneous complex solid cystic mass with multiple dense calcifications and presence of fatty areas. Displacement of adjacent structures by the mass is also seen. On both X-rays and USG, findings are non specific and differentiation from teratoma is not possible.

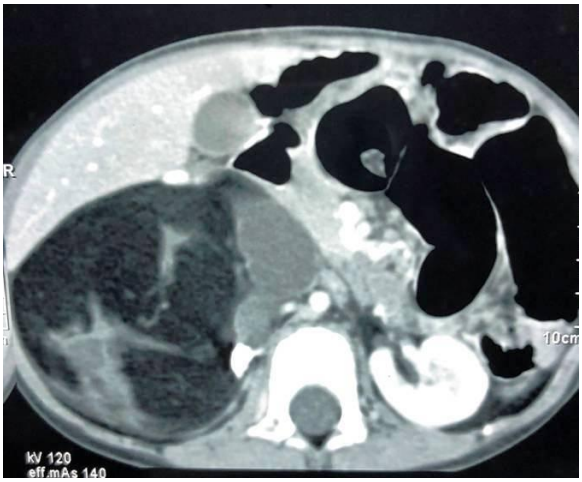

Figure 1: Axial CT image in a pathologically proven case of Fetus in Fetu showing presence of large right retroperitoneal mass with presence of fatty areas and cystic component. Small associated calcific density also seen at the medial aspect

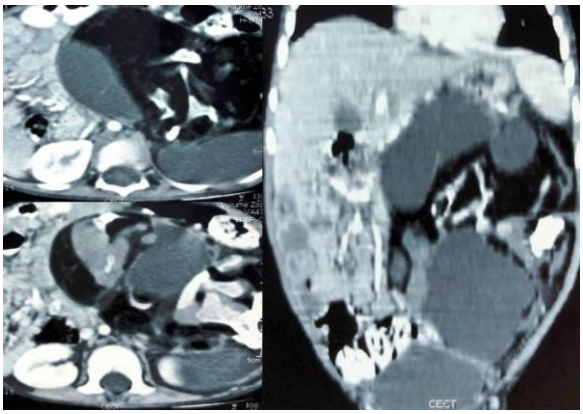

Figure 2: Axial and Coronal CT images in another pathologically proven case of Fetu in Fetu showing presence of large left retroperitoneal mass with presence of large cystic and fatty mass causing displacement of adjacent bowel loops. Multiple irregular vertebral bodies, rib like structures and long bones also seen in the mass.

CT findings are highly diagnostic for fetus in fetu. CT reveals a large well defined mass, commonly in the retroperitoneal region (Figure 1) with presence of cystic abd fatty components. Stomach and adjacent bowel loops are commonly displaced due to significantly large size of the mass. Few or multiple bony structures can be seen in the mass, predominantly irregular vertebral bodies, rib like bones and a well formed

femur like long bones (Figure 2). Facial and calvarial bones are uncommonly seen. Fat plane between the mass and surrounding structures were well maintained. The presence of bones is a significant finding which differentiates the lesion from teratoma. However, non visualization of vertebral column on CT does not exclude the diagnosis as stated by Hoeffel et al [17] (Figure 3). Spinal axis in fetus in fetu may be markedly dysplastic and underdeveloped and so may not be visualised on CT scan. So pathological examination of the specimen is always recommended to establish the final diagnosis.

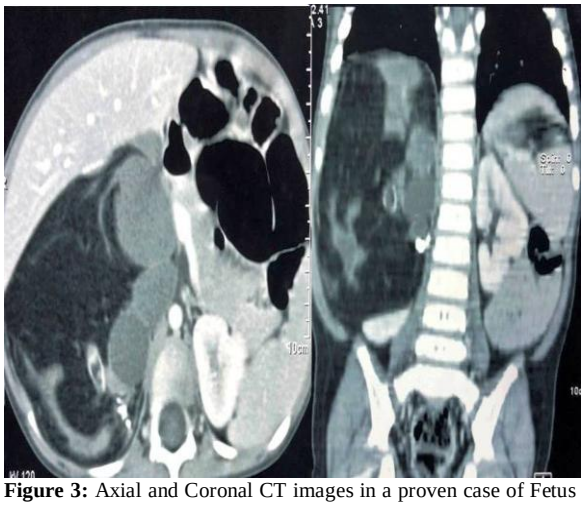

Figure 3: Axial and Coronal CT images in a proven case of Fetus in Fetu showing presence of a large right retroperitoneal mass with fatty and cystic areas. Small irregular long bone like structure and few calcific foci are also seen. Despite lack of spinal axis, pathological diagnosis of Fetus in Fetu was made.

Few case reports have advocated the use of MRI in diagnosis of Fetus in Fetu. [17-19] MRI has many advantages like high contrast resolution, multiplanar capability and choice of various pulse sequences, thus making the diagnosis accurately. MRI has distinct advantage over CT in evaluation of Fetus in Fetu: (1) lack of ionizing radiation, (2) it does not depend on detection of tissue calcification, this helps in identification of insufficiently calcified vertebral column, [17] (3) absence of bony artifacts, (4) no need of iodine based contrast agents. MRI may reveal a large fetiform retroperitoneal mass with mixed high, intermediate and low intensities and bony structures in the form of crumpled vertebral bodies, rib like bones and well formed long bones suggesting remnants of a second fetus. [20] Limb buds can be seen with primitive phalanges and an orbital socket. [21] MRI has emerged as an excellent alternate safe technique in evaluation of the pathology and can be used in antenatal period also as there is no known adverse biological effect on the fetus in utero.

Pathology:

Grossly, the fetus in fetu is surrounded by thin fibrous sac containing straw colored fluid along with an anencephalic fetus (100%) with limb buds (83%) and vertebral column. [20] Pathological diagnosis is confirmed by the presence of vertebral column. Another diagnostic criterion includes presence of a membrane covering fetal vasculature connections. [6] Rounded or tubular collection of very low density fat surrounding amorphous soft tissue is characteristic. [20] Other structures include skin, skin extensions, bone, bone marrow, venous structures, skeletal muscles and peripheral nerves. Uncommonly teeth, tongue, salivary glands, lymphatics, trachea, thyroid, parathyroid, lower respiratory tract, pancreas, spleen, kidney, gonads, testis, ovaries, urinary bladder, digits and nails are present in FIF. [14,21,22] By virtue of Karyotyping, serological markers and restriction, fetus in fetu represents a diamniotic monochorionic twin of the host, thus confirming a separate etiology as compared to teratoma. [6] No placenta or chorionic villi are found.

Management:

Treatment of Fetus in Fetu involves complete resection of the lesion along with the surrounding sac as it may cause symptoms because of mass effect over intra abdominal organs. [20] Associated risks include chances of infection, bleeding and pleura-peritoneal inflammation due to leak of sac contents. [20] Though benign, single case report of fetus in fetu with malignant degeneration has been described in literature

in which recurrence occurred after 4 month as a yolk sac tumour. [23] Recurrence has been attributed to the presence of immature tissues in the small areas and remnants of the surrounding capsule of the mass. [24]

SUMMARY AND CONCLUSION

Fetus in fetu is a rare cause of abdominal mass in children. It could be considered as a rare differential diagnosis of teratoma in an infant or young child presenting with progressively increasing abdominal swelling and vomiting. Characteristic finding of axial skeleton distinguishes Fetus in fetu from teratoma. Both CT and MRI findings are useful for its characterisation and diagnosis. MRI provides excellent definition of the salient anatomy without ionizing radiation and should be considered the ideal imaging modality. But MRI is expensive modality and less readily available than CT, especially in developing countries. Further, we emphasize that CT and MRI are important diagnostic valuable adjunctive tool to sonography for definitive preoperative diagnosis of FIF and surgical planning. Further non visualization of vertebral column on CT or MRI does not completely exclude the diagnosis and histopatholgical confirmation is necessary. The treatment of choice for FIF is complete resection and further research efforts are required to establish the true nature of Fetus in fetu.

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