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Case Report:

Fetus in Fetu: A Rare Presentation in an Adult Female

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ABSTRACT:

Herewith we reported a case of fetus in fetu, a pathologic condition resulting from abnormal embryogenesis in a diamniotic, monochrionic pregnancy. This condition usually presents in infancy and childhood. In this present case, a 38 year old patient misdiagnosed preoperatively as retroperitoneal teratoma is presented with acute abdominal pain with a lump and sepsis. CT scan, ultrasound and X-ray was performed. We would like to conclude that , FIF is a very rare clinical entity. It is expected to be seen in childhood. Presentation at the age of 38 years is very rare. Fetus in fetu should always be kept as differential diagnosis of retro peritoneal Teratomas and dermoid in adults.

Keywords : Fetus in fetu , CT scan

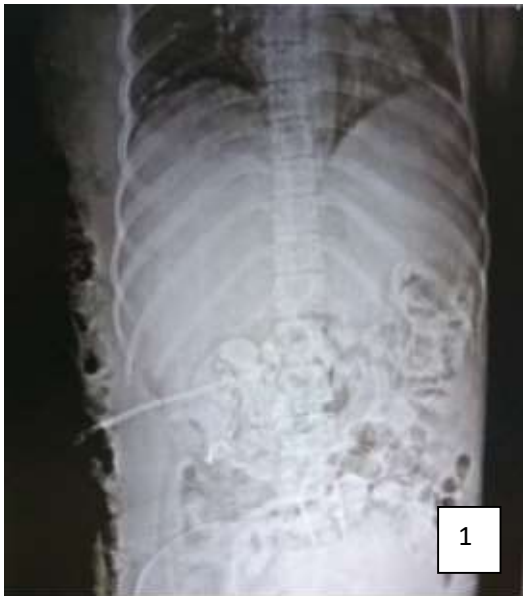
INTRODUCTION :

Fetus-in-fetu (FIF) is a rare condition in which a malformed parasitic twin is found within the body of a living child or adult. It is an encapsulated fetiform calcified mass which is present in the abdomen of its host. It is supposed to be a highly differentiated form of teratoma.[1] However, in view of the fact that body parts can be identified within it, there is a tendency to consider this condition as being distinct from a teratoma. It has been suggested that if spinal elements are absent, the lesion is a teratoma, whereas if they are present the tumor can be considered to be a FIF.[1] We describe the case of a FIF that was diagnosed on CT scan.

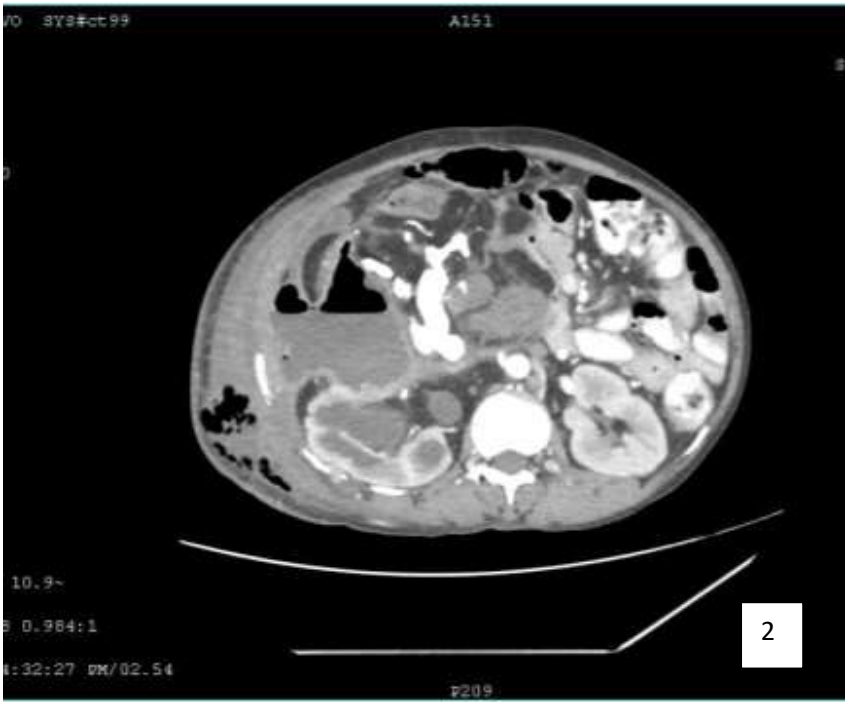
CASE REPORT:

A 38 year-old married female was admitted with a lump in the right side of the abdomen of 1 year duration. The lump was noticed immediately after pregnancy and it was gradually increasing in size. The patient had not conceived after the appearance of the lump. There were no gastrointestinal or genitourinary complaints. There was also no history of twinning or Teratoma in the family. A large 14cms x 15cms mass of firm consistency was palpable in the right hypochondriac and lumbar region.

Conventional Radiography of the abdomen showed irregular radio dense lesion with calcification. Abdominal USG showed a complex mass in the lower abdomen, containing nodular soft tissue components and multiple echogenic areas with post-acoustic shadows. Based on these findings a provisional diagnosis of retroperitoneal teratoma was made. Alpha fetoprotein and beta-human gonadotropin hormone assays were not obtained.



Abdominal CT scan was performed for further evaluation and this showed a lobulated complex mass in the retroperitoneum in the midline; the mass contained a set of spinal elements, a set of rudimentary pelvis, sacra, extremity bones, phalanges, and other osseous elements surrounded by fat, all enclosed in a common sac. Free fluid was also seen in the abdomen, the density of which was high . And the mass was invading the right kidney, causing resultant hydronephrosis and ballooning with thinned out cortex. With this constellation of findings, a diagnosis of ruptured d fetus-in-fetu (FIF) was made and exploratory laparotomy was performed.



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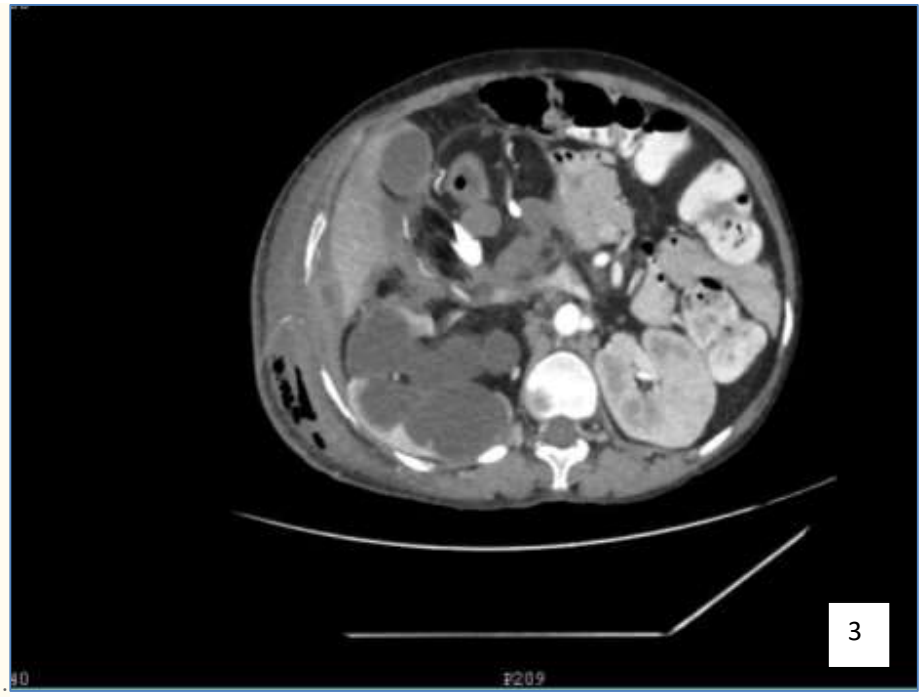
Axial contrast-enhanced CT scan shows the spine of the bigger fetus surrounded by fat and enclosed in a capsule.. Ascites (*) is also seen



On laparotomy, a 15-cm encapsulated retroperitoneal mass with a broad vascular pedicle (attached to the retroperitonium below the aortic bifurcation) was found. The mass was resected *en bloc*; however, the capsule was found ruptured at places. On removing the capsular covering of the resected surgical specimen, it was found covered with hairy skin and a cheesy, pultaceous material; there were rudimentary limbs showing nails and teeth The excised specimen was cut open, and removal of the overlying fat and soft tissue material revealed an anencephalic malformed fetus within the mass, fetus showing many matured bony elements, including the spine (the most defining element) and extremities; flipper-like rudimentary lower limbs. The diagnosis was also confirmed on histopathology

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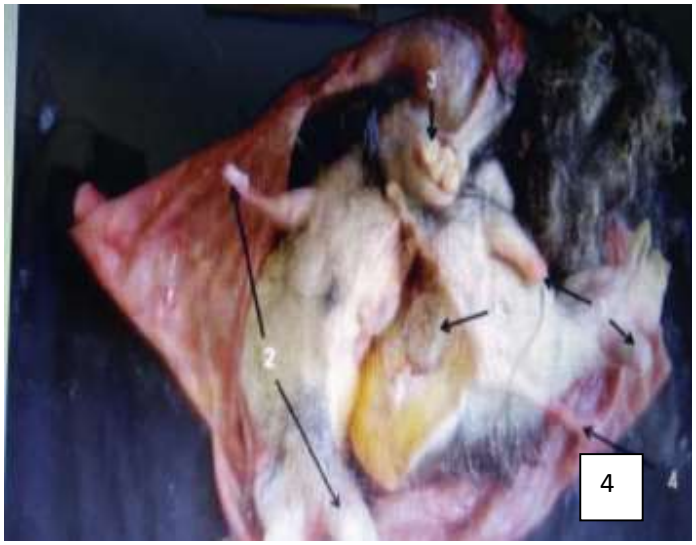
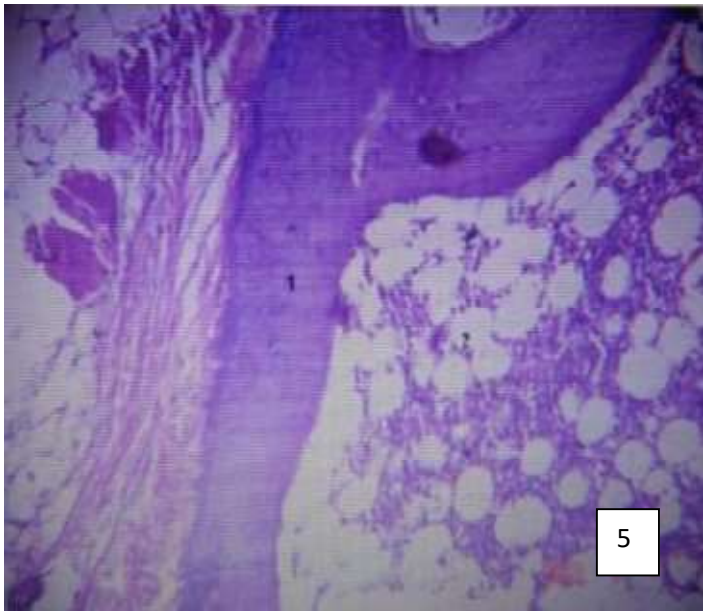


Specimen showing- upperlimb buds, lower limb buds, teeth, vertebral coloumn with fat.



Histopathology shows vertebra and nerve roots.

DISCUSSION:

FIF is a rare congenital entity in which a nonviable parasitic fetus grows within the body of its twin. This term was first coined by Meckel in the 18th century and the first case was reported by Young in 1809. The incidence is about 1 in 500,000 births and only about 100 cases of FIF have been reported to date.[2]

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There are many theories that have been postulated to explain the exact pathogenesis of FIF. The most commonly accepted theory is that it results from an abnormal diamniotic-monozygotic twin pregnancy in which a smaller cell mass is included within a maturing sister embryo due to unequal division of the totipotent inner cell mass of the developing blastocyst. According to the next most popular theory, it may be a highly differentiated form of dermoid cyst, which itself is a highly differentiated form of a mature teratoma.[3]

The condition usually presents in childhood. FIF is usually single, though as many as five FIFs have been reported by Kimmel *et al*., who described a case of a newborn with a cerebral tumor containing five human fetuses. Although, an FIF may increase in size and cause local mass effect and hemorrhage, rupture is not common.[3,4]

FIF is usually seen as a well-defined complex mass enclosed within a capsule, which is usually an amnion-like membrane. Identifiable bones and fat may be seen on plain films, but these findings are more vividly demonstrated with CT scan or MR studies. In addition, the vascular pedicle may be detected on CT scan.[5] On USG, it is seen as a mass with a complex echo pattern, with fluid, soft tissue, and calcification all being often identified within it.[6]

FIF usually presents as a fetiform osseous / calcified mass, often in the abdomen of its host, with the retroperitoneum being the most common site to be affected; however, the cranial cavity, pelvis, scrotal sac, sacrococcygeal region, mesentery, right iliac fossa, and oral cavity are also affected rarely. The condition usually presents in childhood. FIF is usually single, though as many as five FIFs have been reported by Kimmel *et al*., who described a case of a newborn with a cerebral tumor containing five human fetuses. Although, an FIF may increase in size and cause local mass effect and hemorrhage, rupture is not common.[3,4]

The contents of the mass may vary; for example, it may contain extremity bones, pelvic bones, ribs, thoracic and abdominal organs, eyes, ears, mouth, skin, hair, nails, etc. Willis has emphasized that the identification of the vertebral column secures the diagnosis of FIF and differentiates this entity from teratoma.[7] In addition to the presence of spinal elements with limb buds, other features distinguishing this condition from teratoma are its embryological origin, its invariable benignity, and the underdeveloped (though at times well-developed) organ systems within it. [3,7,8]

On, antenatal USG, FIF usually presents as a complex mass in the fetal abdomen. The general appearance is of a well-delineated, encapsulated, echogenic mass suspended in or partially surrounded by fluid. Occasionally, the diagnosis may be easily made if the rudimentary spine is recognized.[8]

CONCLUSION:

In the end, we would like to conclude that , FIF is a very rare clinical entity. It is expected to be seen in childhood. Presentation at the age of 38 years is very rare. Fetus in fetu should always be kept as differential diagnosis of retro peritoneal Teratomas and dermoid in adults.

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