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Fetus-In-Fetu Mimicking Retroperitoneal Teratoma: A Series of Two Cases

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Abstract: *Fetus-n-fetu is a fascinating diagnosis explaining the development of 'baby within a baby'. It results out of a monozygotic diamniotic twin pregnancy in which a twin is absorbed into the body of its sibling. The surviving twin often presents with abdominal mass and the diagnosis is only confirmed by surgical exploration. It is often mimics retroperitoneal teratoma on imaging and poses a challenge for a physician in diagnosing pre-operatively. Surgical excision is often curative.*
Keywords*: fetus-in-fetu, mass, surgery, teratoma, twin*

I. Introduction

Fetus in fetu is a rare abdominal tumor of childhood. Its preoperative radiological diagnosis is made with surity in only a fraction of cases. It was first described by Meckel in the late 18th century[1]. Fetus in fetu , a term quoted by Willis[2] was first described as a rare condition in which a malformed parasitic twin was found inside the body of its partner usually in the abdominal cavity. It represents an aberration of monozygotic diamniotic twinning in which an 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.
This is a rare entity with an incidence of 1 per 500000 births[3], with fewer than 100 reported cases worldwide[4]. The majority of cases occur in infancy, with the oldest reported case being a 47 year old man[1]. Tharkral et al[5] reported equal male and female prevalence. In 70 % of cases, the chief presenting complaint was an abdominal mass[6]. The mass was predominantly retroperitoneal in 80% of cases[2], while reported uncommon sites include the oral cavity[1], sacrococcygeal region[7], and scrotum[4].

II. Case Profile

2.1 Case 1

A two year male child born through a normal delivery with birth weight of 2.8 kg presented to our outpatient department with a prediagnosis of abdominal mass,, felt by the mother on left side of abdomen. The clinical examination confirmed the presence of mass and further work-up was done. On X-ray abdomen, a large left sided soft tissue opacity with calcified osseous structures within, could be appreciated (Fig.1).

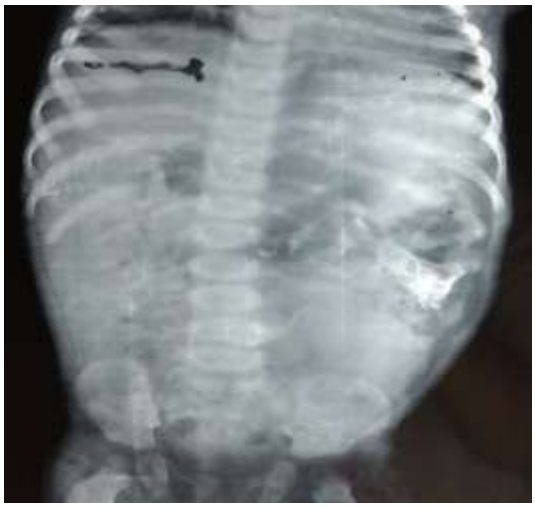

Figure1: Anteroposterior abdominal radiograph shows a large soft tissue mass in left hemiabdomen with calcified osseous-appearing densities of varying shapes and sizes

Abdominal USG could better define the lesion as a complex apprearing mass displacing left kidney and measring approximately 10.5x7x8 cm with cystic areas in it. Calcification within the lesion were identified and a provisional diagnosis of retroperitoneal teratoma was made. CECT Abdomen was carried out and a large

retroperitoneal mass with areas of varying densities, viz. fat, soft tissue and bone was noted (Fig.2).

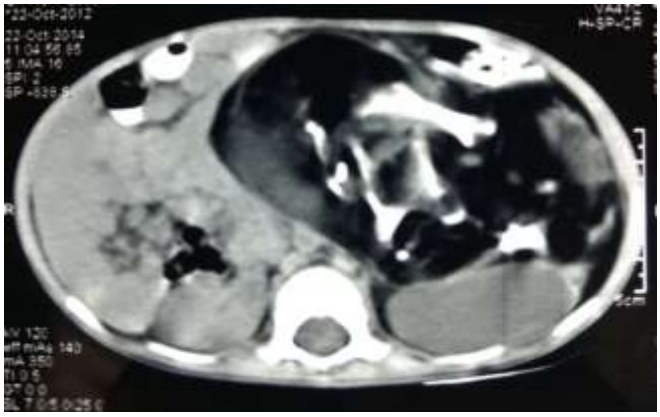

Figure 2: Computed Tomography scan of the patient's abdomen reveals a large retroperitoneal soft tissue mass. There are long hyperdense opacities that resemble fetal bones Surgical exploration revealed a large well encapsulated mass located superior to left kidney and encasing left renal vessels. Left kidney was sacrificed during the procedure. Gross pathological specimen measured 15x10x8 cm and spinal column could be seen upon dissection (Fig.3,4).

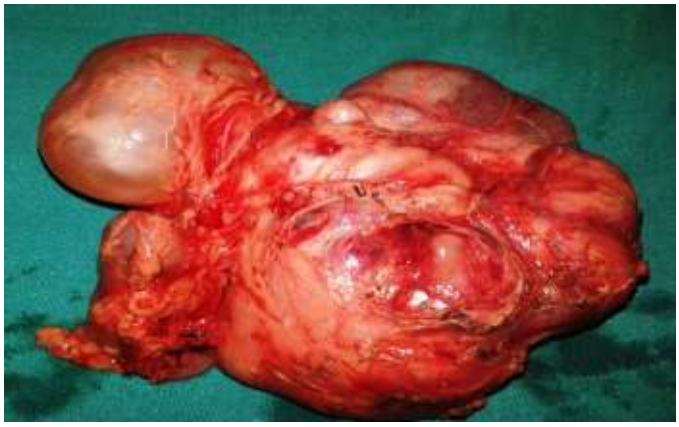

Figure 3: The postoperative specimen showing a lobulated mass lesion which is well encapsulated, alongwith excised left kidney.

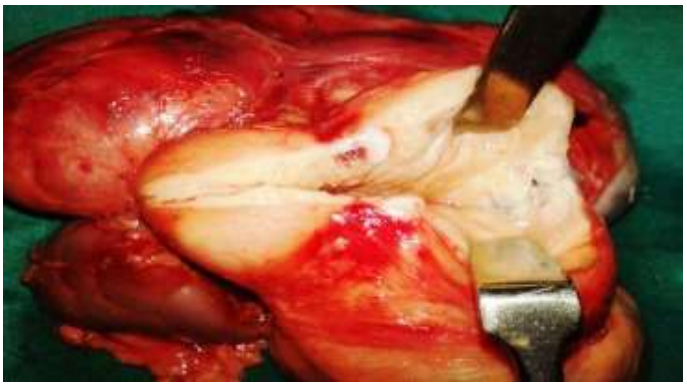

Figure 4: The dissected specimen showing opened spinal column of the fetus.

Histopathogy confirmed the diagnosis and revealed a variety of tissues including fibrocollegenous, fibroadipose tissue with cartilage, bony trabeculae, bone marrow elements, nerve bundles, neural elements, cystically dilated glands and muscle bundles with areas of hyalinization, necrosis and degeneration.

2.2 Case 2

A 2800-g, 8 day old female, term baby, born through spontaneous vaginal delivery to a mother of 24 years of age in the first pregnancy, presented to our hospital with abdominal lump that was noticed by her

mother incidentally just after birth. On physical examination, the physical appearance of the baby was normal. There was a semimobile mass of 6× 8 cm, which could only be noticed by deep palpation. α-FP levels, among other laboratory investigations were normal. CECT abdomen revealed a complex left sided, retroperitoneal mass lesion with scattered bony densities in it. Areas of fat attenuation and necrosis were also noted (Fig.5).

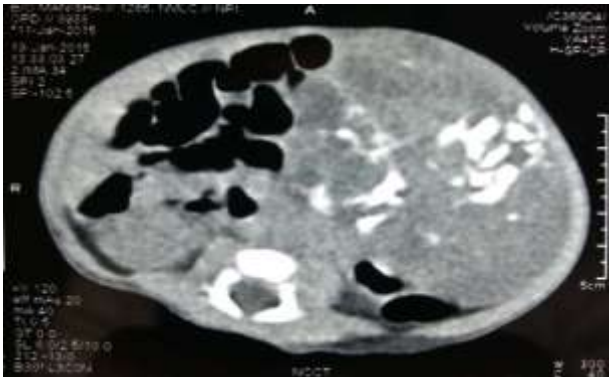

Figure 5: Computed Tomography scan of the patient's abdomen reveals a large retroperitoneal soft tissue mass with hyperdense opacities that resemble fetal bones Histopathological examination demonstrated skin, skin extensions, bone, bone marrow, fat tissue, venous structures, striated muscle and peripheral nerve sections. The postoperative period of the patient was uneventful.

III. Discussion

Fetus-in-fetu is an entity that is encountered on rare events. It most likely represents monozygotic diamniotic twin that implants itself and grows within the body of its karyotypically normal, identical sibling. Lord reviewed a number of similar cases and defined the lesion as a parasitic twin found within the abdomen of its sibling[8]. It is most commonly located in upper retroperitoneum or in the retroperitoneal portion of fetal abdomen. The fetus is usually single and is always anencephalic although upto five foetuses have been reported[9,10]. Most often, as Knox and Webb pointed out, the definitive diagnosis of fetus-in-fetu cannot be established pre-operatively, even though the radiological diagnosis retrospectively confirms it[6]. A crucial feature leading to the correct diagnosis is the presence of vertebral column. Willis stated that the embryologic development of an included twin must include the primitive streak stage, which invariably produces vertebral column[2]. Fetus-in-fetu is often misdiagnosed as retroperitoneal teratoma, owing to the similar radiologic features. Willis cleared the distinction and stressed that teratoma is a true neoplasm as opposed to the other entity[2]. Lewis also emphasized that teratoma is associated with malignant degeneration, rarely observed with fetus-in-fetu[11]. Although calcified densities are noted in both the cases, the arrangement of very low density fat surrounding a central bony density in fetus-in-fetu and interspersion of fat and bone in an helter-skelter fashion in teratoma aids in making an accurate diagnosis.

IV. Conclusion

Fetus-in-fetu is an important diagnosis to be differentiated from retroperitoneal teratoma preoperatively due to potentially malignant characteristics of the latter. Although the benignity of fetus-in-fetu is well established, there is always a consensus on complete excision of the lesion.

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