FETUS IN FETU: TWO CASES AND LITERATURE REVIEW

INTRODUCTION
Fetus in fetu is an uncommon pathological entity which results due to abnormal embryogenesis in a diamniotic monochorionic twin. The most commonly accepted theory states that unequal division of the blastocyst results in monoyzotic diamniotic monochorionic twins of unequal sizes [1]. The smaller twin then encases into the normally developing twin by unknown mechanism followed by the arrest of further growth of the encased fetus due to improper blood supply or inherent defects of the encased twin [1]. The true etiology is unclear [2]. The theory of dernised multiple pregnancy has gained support recently [2]. We report two cases of retroperitoneal fetus in fetu.

CASE SERIES

Case 1: A one month baby presented with a lump in the lower abdomen. A 10x10cms well defined, non-tender, ovoid, firm lump with a smooth surface was palpable arising from the pelvis. The lower border of mass was palpable on digital rectal examination. Abdominal ultrasound showed a 10x10.5cm cyst with a fat fluid level & calcific foci. Contrast Enhanced Computed Tomography (CECT) was suggestive of a large teratoma (fig. 1). The tumor markers (Alpha-Fetoprotein and Beta Human Chorionic Gonadotropin) were within normal limits. At exploration, there was a large 12x10x6cm retro peritoneal lump compressing left ureter, ovary & pushing the uterus and bladder anteriorly. The tubo-ovarian structures and uterus were normal. The tumor was mobilized all around and excised completely (fig. 2). The x-ray of the excised specimen was suggestive of a bony stalk like structure. Histopathology showed cartilage, smooth muscle, gastrointestinal epithelium (colonic mucosa with muscularis propria), adipose tissue and bone with marrow. All tissues were mature and arranged in an organoid form suggestive of fetus in fetu (fig. 3).

Case 2: A one-month female child presented with abdominal distention and intermittent non-biliious vomiting. There was a huge lump of variegated consistency involving almost whole of the abdomen of size 12x8 cm. The blood investigations and tumour markers were normal.
Abdominal ultrasound and CECT Scan was suggestive of retroperitoneal teratoma (fig. 4). Intraoperatively, there was a huge retroperitoneal solid cystic mass pushing the colon, right kidney and the right ureter (fig. 5). It was adherent to the inferior vena cava. The mass was completely excised. Limb bud could be seen in the gross specimen. Histopathology showed cartilage, bowel wall and bone with marrow arranged in an organoid form suggestive of fetus in fetu (fig. 6).

**DISCUSSION**

Fetus in fetu is an unusual cause of retroperitoneal mass in infants [3, 4]. The incidence is 1 in 500 000 live births [3] with a slight male preponderance [5]. It was
first described by Johann Friedrich Meckel in 1880 [6] as a malformed or a parasitic monozygotic diamniotic twin which is found inside the body of a living child or sometimes an adult, usually in the abdominal cavity [6]. It was defined by Willis in 1953 as a mass containing a calcified vertebral axis often associated with other organs or limbs around this central axis [6]. As per Gonzalez-Crussi, the term fetus in fetu is applied to any structure in which the fetal form is in a very high development of organogenesis and to the presence of a vertebral axis [6]. Teratoma is just the accumulation of pleuripotent cells without organogenesis or vertebral segmentation [6]. The identification of a vertebral column in fetus in fetu indicates development of the parasitic twin at least up to the stage of notochord [7].

There are two theories of its development:

a. Parasitic Twinning Theory/ Included Twin Theory: Most commonly accepted, it states that fetus in fetu is a rare form of monozygotic parasitic twin fetus growing within host twin [4]. Monozygosity can be confirmed by the presence of identical karyotype, histocompatibility types and blood groups [7]. Spencer’s observations also support this theory [5]. Beaudoin’s theory states that there is defective implantation during the second week of development resulting in the invasion of a second embryo into the extra-embryonic mesenchyme of the host fetus or autosite, instead of the uterine wall [8].

b. FetalformTeratoma Theory: Put forward by Willis, it states that fetus in fetu is a well differentiated, highly-organized form of a mature teratoma [9].

The pathogenesis is unknown. It is suggested that during the ventral folding of trilaminar embryo at second and third weeks of development, the diamniotic monochorionic twin is included within its host [10] because of a persistent anastomosis of the vitelline circulation during development [10]. It is also thought to result from an unequal division of totipotent inner cell mass of the developing blastocyst, causing a small cell mass within the maturing embryo [10].

The clinical presentation is usually as an abdominal mass during infancy. However, fetus in fetu has also been reported in adults. The differential diagnosis of an abdominal mass in an infant includes neuroblastoma, teratoma, meconium pseudocyst and fetus in fetu [10]. Teratoma is more common than fetus in fetu but rarely arises in retroperitoneum [10, 11]. Fetus in fetu is a highly differentiated tissue with or without a vertebral column, whereas teratoma is a discordant mass of pleuripotent cells with no systemic organization [10, 12]. Besides vertebral column (91%), other tissues like limb buds (82.5%), central nervous system (55.8%), gastrointestinal tract (45%), vessels (40%) and genitourinary system (26.5%) are commonly present in a fetus in fetu [6]. Gonads, adrenals, heart and primitive respiratory units are less commonly seen [10].

Though abdomen is the most frequent site of presentation, fetus in fetu has also been described in cranial cavity, mediastinum, lung, sacrococcygeal region, kidneys and scrotum [4]. Fetus in fetu is usually single, but up to five fetuses have also been reported [6]. The symptoms of fetus in fetu are those related to its site of presentation and its mass effects. In the abdomen, the symptoms are those of abdominal distention, feeding difficulties, vomiting, jaundice and pressure over the gastrointestinal and genitourinary tracts. It leads to dyspnea and respiratory difficulties in the thorax and dysphagia and cleft palate in the oral cavity. It causes neurological symptoms in the cranial cavity [6].

There is ongoing controversy about the differentiation of fetus in fetu from mature teratomas [10] because of pathologic overlap. Both of these entities may represent different degrees of spinal dysgenesis or residual posterior enteric remnants secondary to early fetal disturbance of endodermal-ectodermal differentiation [10, 13].

Abdominal USG usually gives a list of differential diagnosis like retroperitoneal teratoma, neuroblastoma and fetus in fetu. CECT and Magnetic Resonance Imaging (MRI) are helpful tools for diagnosis. There is only one case in literature with pre-operative radiological diagnosis of a fetus in fetu by 3-D CECT reconstruction [10]. Prenatal diagnosis is also now being done by USG and MRI where the main differential diagnosis is retroperitoneal teratoma [14].

Though considered benign, there have been isolated cases of malignancy following resection of fetus in fetu [10, 15]. This has prompted some surgeons to recommend complete resection followed by post-operative surveillance by tumour markers (Alpha-feto protein and Beta Human Chorionic Gonadotrophin) for two years [10, 15].

Surgical excision provides complete cure in almost all patients.

REFERENCES


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