Is definition of “fetus in fetu” needs revision?: A case report and review of literature

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Abstract
Fetus in fetu is a rare clinical entity consisting of malformed fetus inside body of the living partner. It is a mass containing a vertebral axis and often other organs and/or limbs. Unlike other classical cases, this case was not having a vertebral axis but other clinico-pathological findings were suggestive of fetus in fetu. To include such types of cases, the traditional definition by Willis of fetus in fetu needs revision.

1. Case summary
A 12-year old male child presented with a mass over the lower back since birth, which was gradually increased in size. The mass was present in sacrococcygeal region, approx. 20 × 20 cm in size, non-tender and had variegated consistency. It was fixed with underlying structures and was covered with normal skin. The anal orifice was normally placed. On palpation of abdomen, another hard and fixed lump measuring approx. 8 × 7 cm in size was felt in suprapubic region. Plain X-ray of the abdomen and pelvis showed calcified mass pushing the bowel and bladder to one side. Ultrasonography revealed, heterogeneous mass in the pelvis and sacrococcygeal area. MRI showed calcified and ossified component within the elongated fatty tissue with large formed long bone of the limb (Fig. 1). Level of serum alpha fetoprotein was 2.3 ng/ml.

Laparotomy was done. Retroperitoneal tumor was mobilized and dissected up to the level of lower end of coccyx. Patient was turned into prone position, an inverted V incision was given over the tumor. The muscles above the tumor were preserved. Tumor was dissected out meticulously from the neighboring structures. On gross examination, tumor had both solid and cystic component with a covering membrane looking like skin around it. After opening the cystic sac, a well formed long bone and a finger with nail was found (Fig. 2). The whole tumor was taken out with coccyx. The entire covering membrane of the cystic cavity was stripped out. No feeding vessel was found supplying the tumor. Anorectal anatomy was preserved.
were described in clinical literature. The controversy about the recurrences and malignant transformation. Management is surgical resection. Follow up is done with ultrasound, plain radiographs, ultrasonography, CT scan & MRI. The chorionic gonadotropin may be normal or elevated. Diagnosis is systemic, dyspnea. The levels of serum alpha fetoprotein and human chorionic gonadotropin might be raised in older children because chances of malignancy are more in them. Unlike teratomas, fetus in fetu rarely has malignant transformation.

The essential component of FIF is the presence of vertebral column. One fourth case reports of FIF do not have vertebral column but have advanced stage of organogenesis. Gonzalez-Crussi and Spencer redefined the different components of definition of FIF. Prescher LM (2015) states that FIF and teratoma have same pathology but are detected at different stage of maturation. This hypothesis was supported by many case reports in which FIF into teratoma at the two ends of same disease spectrum.

The teratomas are an accumulation of pleuripotent cells lacking organogenesis or vertebral segmentation. Level of alpha fetoprotein might be raised in older children because chances of malignancy are more in them. Unlike teratomas, fetus in fetu rarely has malignant transformation.

The index case was not having vertebral axis but the other diagnostic features of fetus in fetu as proposed by Spencer were present. The definition of FIF proposed by Willis which includes vertebral axis as essential component needs to be modified as approximately 25% cases reported of FIF in literature have no vertebral axis. Also, many reports have shown changes of FIF into teratoma with malignant components, again suggesting FIF and teratoma at the two ends of same disease spectrum.

2. Discussion

FIF is a rare anomaly consisting of a malformed monozygotic, diamniotic twin found inside body of the living partner. The common sites of occurrence are abdominal cavity (retroperitoneum), posterior mediastinum, sacrococcygeal region, neck [4]. Usually a single parasitic twin is seen but maximum five fetuses had been reported in a single patient [5]. Common organs seen inside the fetus in fetu could be vertebral column, limbs, central nervous system, gastrointestinal tract, vessels, genitourinary tract. Classically these fetuses are acardiac and anencephalic. The important differential diagnoses are teratomas, meconium pseudocyst, neuroblastoma. The clinical symptoms are mainly due to the mass effect. These are abdominal distension, constipation, feeding difficulties, pressure effects on renal system, dyspnea. The levels of serum alpha fetoprotein and human chorionic gonadotropin may be normal or elevated. Diagnosis is done by plain radiographs, ultrasonography, CT scan & MRI. The management is surgical resection. Follow up is done with ultrasound of the local areas and serum alpha fetoprotein levels to know about recurrences and malignant transformation.

After Meckels description of fetus in fetu, almost 100 cases were described in clinical literature. The controversy about the definition of fetus in fetu is still continuing. According to Willis definition, a fetus in fetu should have (a) separate vertebral column, which demonstrates that the fetus has passed through a primary stage after gastrulation, involving formation of the neural tube, metamereization, and symmetrical development around its axis and (b) the organs should have developed in an organized manner with some degree of maturation [6]. Occasional cases have been reported where no spinal column was found on radiography as well as on pathological examination [7]. To address these cases, Gonzalez-Crussi (1982) proposed another definition: “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 vertebral axis” [8]. Later on Spencer (2001) had suggested that the fetus in fetu must have one or more of the following: (a) be enclosed within a distinct sac, (b) be partially or completely covered by normal skin, (c) have grossly recognizable anatomic parts, (d) be attached to the host by only a few relatively large blood vessels, and (e) either be located immediately adjacent to one of the sites of attachment of conjoint twins or be associated with the neural tube or the gastrointestinal system.

In this case, surgical findings and histopathology confirmed the diagnosis of fetus in fetu. The findings in favor of the fetus in fetu were following:

a) There was heterogenous mass in sacrococcygeal and pelvic area. This mass was a having both solid and cystic component and a large formed long bone of the limb in the cystic component.

b) The level of serum alpha fetoprotein was within normal limit.

c) There was well defined long bone with attached finger and nail.

d) There was also evidence of cartilage, hairy areas, covering skin. Microscopically, components of all three germ layers were present.

The teratomas are an accumulation of pleuripotent cells lacking organogenesis or vertebral segmentation. Level of alpha fetoprotein might be raised in older children because chances of malignancy are more in them. Unlike teratomas, fetus in fetu rarely has malignant transformation.

In the changed clinical setting Willis definition of FIF appears less relevant than Spencer proposed criteria of FIF.

3. Conclusion

The index case was not having vertebral axis but the other diagnostic features of fetus in fetu as proposed by Spencer were present. The definition of FIF proposed by Willis which includes vertebral axis as essential component needs to be modified as approximately 25% cases reported of FIF in literature have no vertebral axis. Also, many reports have shown changes of FIF into teratoma with malignant components, again suggesting FIF and teratoma at the two ends of same disease spectrum.

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