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CASE REPORT

Fetus-In-Fetu or Well-Differentiated Teratoma- A Continued Controversy

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Abstract Fetus in fetu is an uncommon anomaly in pediatric age group. There have been controversies regarding its differentiation from well differentiated teratoma. While presence of axial skeleton has been considered mandatory for diagnosis, there are many reports of fetus in fetu without any vertebral column. It has been suggested that the two entities form a spectrum of lesions depending upon the timing of embryological insult. The present case is another example of a lesion that fits in this spectrum of interesting lesion.

Keywords Fetus in fetu · Retroperitoneal teratoma

Introduction

Fetus in fetu and retroperitoneal teratoma are uncommon entities in childhood. Recent literature questions the necessity of presence of axial skeleton for definitive diagnosis of fetus in fetu. We present a case that was diagnosed as fetus in fetu without the presence of vertebral column.

Case History

A 4 months old male child was brought to the hospital by his parents when they noticed a lump in the upper abdomen for last one week. The child was born at full term out of a non consanguineous marriage in young parents.

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There was no significant obstetric history. Antenatal ultrasonography had not been performed during gestation. There was no history of congenital malformations in other sibling. He had been taking feeds well and weight gain had been normal. There were no symptoms of obstruction either in gastrointestinal or genitourinary tract.

Infant appeared well nourished and healthy. There were no signs of dehydration, anemia or jaundice. Abdomen appeared slightly distended in the right upper quadrant. There was a mass extending across midline, firm, immobile, circumscribed about 15×12 cm palpable in the right upper and umbilical quadrant. There was no organomegaly or free fluid in the abdomen. Rectal examination was normal.

Laboratory investigations revealed hemoglobin of 12 gm/dl. The liver and renal function tests were normal. Alpha fetoprotein levels were 59.4 IU/mL (normal range: 0–8.1 IU/ mL) and β hCG levels were 0.90 IU/L.

The plain film of the abdomen showed right-sided calcifications adjacent to the spine at the third and fourth lumbar vertebrae. Abdominal ultrasonography showed a hypoechoic mass with hyperechoic structures inside that extended from the lower border of the liver to the umbilical region. An abdominal computed tomography (CT) scan (Fig. 1) showed a well defined mass that measured approximately 15×11 cm that consisted of solid, cystic, and calcified components. A vertebral axis was not visualized. The mass was displacing the transverse colon and small bowel downward.

Based on these imaging findings, a clinical diagnosis of retroperitoneal teratoma was made preoperatively.

At surgery, a well-encapsulated retroperitoneal mass behind the transverse mesocolon (Fig. 2). This mass had a pedicle that was connected to the superior mesenteric artery. Excision of the capsule revealed a yellowish fluid and an incompletely developed fetus covered by vernix caseosa (Fig. 3). A radiograph of the specimen showed areas of calcification but no vertebral column.

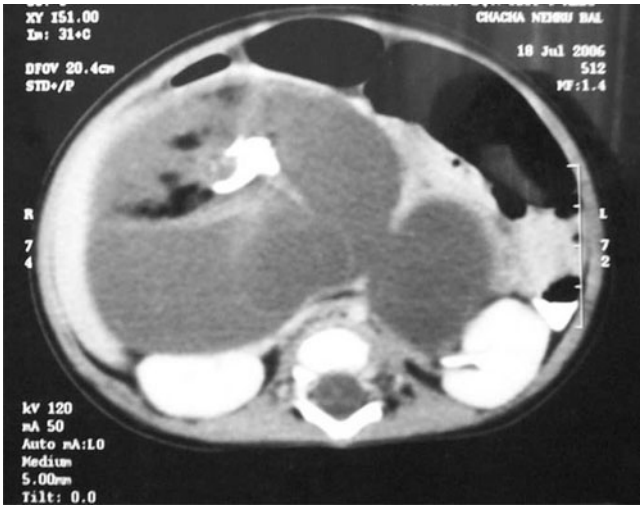

Fig. 1 Computed tomography showing cystic lesion with calcification

On the macroscopic pathologic examination, the mass measured 15×10×8 cm and was composed of a head with hair, a trunk, and rudimentary limb corresponding to an incompletely developed fetus. Microscopically, there was evidence of neural, intestinal and cartilaginous tissue (Fig. 4a, b, c). A diagnosis of fetus in fetu was given on the basis of histopathological findings.

The postoperative course was uneventful and the patient was discharged on the fifth postoperative day. At six months follow up, the child was well and alpha fetoprotein levels had come down to less than 1 IU/mL.

Discussion

Fetus in fetu is a malformed parasitic monozygotic diamniotic twin that is found inside the body of the living partner. The pathology is uncommon and the incidence is said to be about 1 in 500 000 births. About 90% cases are identified before 18 months of age with no specific sex predilection [1, 2].

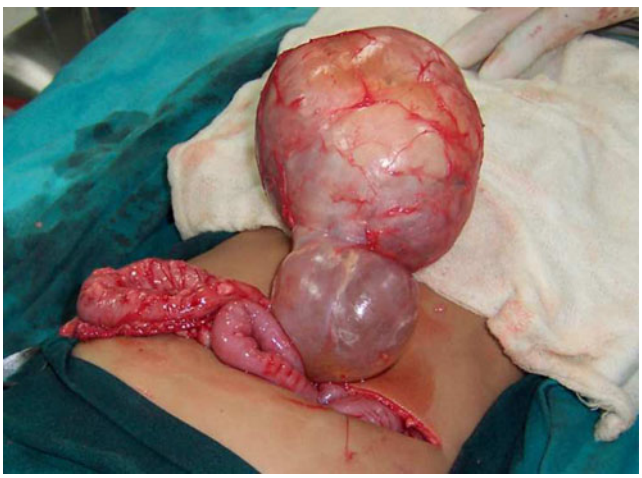

Fig. 2 Operative photograph showing cystic mass in relation to transverse colon

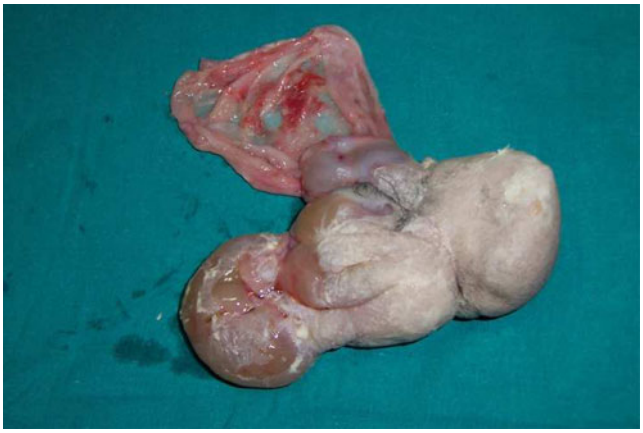

Fig. 3 Completely excised mass after opening the sac showing brain, intestine and attempt at limb formation

Although Meckel is said to have given the first description of fetus in fetu [FIF], over hundred cases have been reported in the literature. There has been continued controversy where the definition of FIF has been questioned. It has been suggested that the fetus in fetu should have a) separate spinal column and b) the organs should have developed in an organized manner with same degree of maturation [3]. On this basis, many cases reported in literature were termed retroperitoneal teratoma as they did not have evidence of axial skeleton considered mandatory for diagnosis of FIF.

Spencer has suggested that an FIF must have one or more of the following conditions: (1) be enclosed within a distinct sac; (2) be partially or completely covered by normal skin; (3) have grossly recognizable anatomic parts; (4) be attached to the host by only a few relatively large blood vessels; and (5) either be located immediately adjacent to one of the sites of attachment of conjoined twins or be associated with the neural tube or the gastrointestinal system [3]. A teratoma, on the other hand, shows lesser organization of microscopically identifiable tissues. Teratomas have a broader attachment site with multiple small blood vessels.

The differential diagnosis of a complex mass with calcific components in a fetus or neonate includes FIF, teratoma, meconium pseudocyst, and neuroblastoma [4].

Recently some cases of FIF with differentiation of tissues but no vertebral column have been described [5]. There are also many reports of sacrococcygeal teratomas and intracranial lesions that have been labelled FIF despite the absence of vertebral column [6, 7]. In a large review of all the cases reported, about 10% cases did not reveal vertebral column [2].

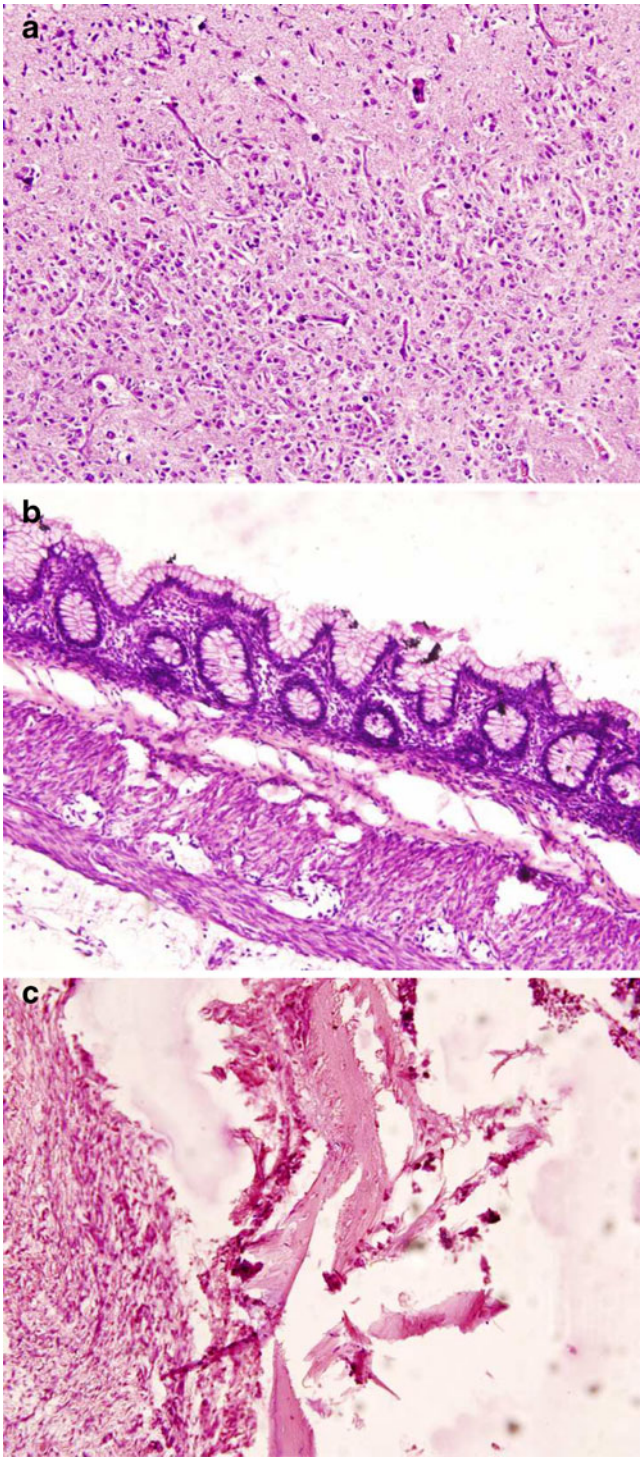

Fig. 4 **a**, **b**, **c** Microscopic examination showing **a**) glial tissue; **b**) intestinal tissue and **c**) cartilage (×10, Hemotoxylin and Eosin)

Spencer suggested that FIF and many teratomas are an aberration of monozygotic twinning [3]. His hypothesis points to a spectrum of anomalies ranging from conjoined twins to malformed external parasitic and acardiac twins to FIF to so called "fetaform" teratomas to well-differentiated teratomas. Depending on the degree of malformation and timing of the embryologic insult, interruptions of monozygotic twinning can lead to any of these observed phenomena. Fetus in fetu is thought to occur at a very early stage of development, before ventral fusion of the lateral body walls. It has also been suggested that there is a continuum between FIF and teratomas. Supporting this hypothesis are the observations that FIF and teratomas are increased in families with a history of twinning; FIF may contain multiple fetuses; FIF and teratomas can coexist; and FIF and teratomas are found in locations that are involved embryologically in conjoined twinning. While describing a case of sacrococcygeal teratoma with cardiac activity and differentiated intestinal tract de Lagausie et al9 state that absence of vertebral column does not rule out the possibility that none existed during earlier stages of development. On the basis of histopathological findings, Basu et al placed their two cases to be intermediate between FIF and highly differentiated teratoma [5]. The two entities have similar features on radiological investigations [4]. Medical imaging plays an important role in this diagnosis and it is important to be aware that nonvisualization of the vertebral axis on plain film of the abdomen or on CT scan does not exclude this diagnosis [2].

Treatment of the two entities includes complete excision of the mass. An important aspect is to keep the patients in follow up along with markers [8] like alpha fetoproteins so that recurrences with malignancies can be diagnosed at an early stage.

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