Giant retroperitoneal fetus-in-fetu: An unusual cause of respiratory distress

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ABSTRACT

Fetus-in-fetu (FIF) is a rare congenital condition in which a vertebrate fetus is incorporated within its host, a newborn or an infant. We report an interesting case of FIF presenting as an abdominal mass with respiratory distress in a 4-month-old girl, and review the existing literature. An ultrasound abdomen showed a right sided cystic mass containing multiple calcifications. Laparotomy revealed a well-encapsulated right retroperitoneal mass above the right kidney with two rudimentary limbs attached to an amniotic bag by an umbilical cord. Excision of the capsule revealed a yellowish fluid and an incompletely developed fetus covered by vernix caseosa. Pathologic examination showed an irregular fetiforme mass, weighing 1200 gm, with two rudimentary limbs, encephalus, stomach, duodenum, bowel, bone, cartilage, bone marrow, upper and lower respiratory tissue, and spleen. Complete excision of the FIF was performed. The postoperative period was uneventful and the patient was discharged on the seventh postoperative day.

KEY WORDS: Fetus-in-fetu, intra-abdominal mass, monozygotic diamniotic twinning, teratoma

INTRODUCTION

Fetus-in-fetu (FIF) is a very rare condition, with a reported incidence of 1 in 500,000 live births.[1] It most likely represents a monozygotic diamniotic twin that implants itself and grows within the body of its normal karyotypically identical sibling.[2] Although abdominal masses are commonly encountered in pediatric oncology, the finding of a giant FIF causing severe respiratory distress is certainly unexpected. We report such a case of a giant FIF and highlight important features of this rare disorder.

CASE REPORT

A 4-month-old girl presented in the emergency department with massive abdominal distention and respiratory distress. Her SpO2 was 82% on room air. Abdominal examination showed massive distention and a mass in the upper right abdomen. Abdominal ultrasonography showed a hypoechoic retroperitoneal mass containing multiple central irregular calcifications, which extended from the lower border of the liver to the hypogastric region. Based on these findings, teratoma was presumed to be the most likely diagnosis. Because of her deteriorating respiratory distress an emergency laparotomy was planned. On laparotomy, a well-encapsulated retroperitoneal mass behind the transverse mesocolon, measuring 20 cm × 10 cm was found [Figure 1]. This mass had a pedicle that was arising from the superior mesenteric artery. Excision of the...
capsule revealed a yellowish fluid and an incompletely developed fetus covered by vernix caseosa. Complete excision of the mass was performed. On macroscopic examination, the mass weighed 1200 gm, measured $20 \times 15 \times 8$ cm, was ovoid, smooth surface, with two rudimentary limbs, a head and a trunk, corresponding to an incompletely developed fetus. [Figures 2A and 2B]. A radiograph of the specimen showed presence of bone calcification; however, no vertebral axis was observed [Figure 3]. The postoperative course was uneventful and the patient was discharged on the tenth postoperative day. On microscopic pathologic examination, it showed mature embryonic tissue containing elements of the three germinative layers. It was possible to show the presence of skin, germinative buds of limbs, central nervous tissue, alimentary tissue, bone, cartilage, and choroids plexus [Figure 4]. Most of these cells were well-differentiated. The bony tissue was essentially composed of osteoblasts with little bone marrow. The nervous tissue was mostly formed by glial cells and choroids plexus. Even after an elaborate search, vertebral column was not seen.

**DISCUSSION**

FIF is a malformed parasitic monozygotic diamniotic twin that is found inside the body of the living child or adult.[2] It was first described by Meckel in 1800 and defined by Willis in 1953 as a mass containing a vertebral axis often associated with other organs or limbs around this axis.[3] It is a rare pathology, with an incidence of 1 in 500,000 births.[4] Majority of these cases occur in children younger than 18 months with the chief complaint of an abdominal mass.[4] As far as location is concerned, it is predominantly retroperitoneal in 80% of cases,[5] but could also be found in atypical locations, such as skull,[6] scrotum,[7] sacrum,[8] mouth,[9] and adrenal gland.[10] The fetus is usually single in 88% of the cases, although up to five fetuses have been found in the skull of a newborn.[11] Symptoms of FIF relate to its mass effect in the area of its occurrence and in its most common location cause abdominal distention, feeding difficulty, emesis, and rarely jaundice.[10,12] In our knowledge, this is the first reported case where FIF presented with massive abdominal distention leading to respiratory distress. Only in 16.7% of these cases it is possible to show a preoperative diagnosis of FIF, differential diagnosis usually being teratoma and meconium pseudocyst. Occasionally, FIF can be identified on a prenatal ultrasound.[13] Plain abdominal radiograph may be helpful in diagnosis, which may show presence of vertebral column and axial skeleton.[10] With the advent of computed tomography and MRI it is possible to confirm the diagnosis pre-operatively.[14-16] Early and complete resection of the fetus and associated membranes is expected to offer complete cure.[4] The fetus is typically suspended by a pedicle within a sac containing fluid or vernix caseosa. Definite vascular connections to the host are rarely seen and if present arise from superior mesenteric artery or inferior mesenteric artery.[1,10] The size of the FIF is related to its blood supply and can range from 13 to 2000 gm.[10,17] The vascular pedicle in our case...
was significantly large, which accounted the weight of 1200 gm. The distinction between FIF and teratoma has for long been the subject of controversy.[18] According to Willis, the distinction is classically based on the absence of an axial skeleton in a teratoma. However, the radiology and pathology findings in our case and supported by other reports, clearly suggest that FIF can exist even in the absence of a vertebral axis.[19] To resolve this controversy, Gonzalez-Crussi have proposed that the definition of FIF is applied to any structure in which the fetal form is either in a very high development of organogenesis, as in our case or there is presence of a vertebral axis.[19] Although the prognosis for FIF is more favorable than for cystic teratoma, the presence of immature elements warrants early and frequent postoperative surveillance by clinical examination, imaging studies, and alpha fetoprotein.[20]

REFERENCES


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