CASE REPORT

Fetus in fetu

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Fetus in fetu (FIF) is a very rare condition, with a reported incidence of one in 500,000 live births. It most likely represents a monozygotic diamniotic twin that implants itself and grows within the body of its normal karyotypically identical sibling, which typically manifests as a fetiform abdominal mass in a newborn or infant. The mass is located in the retroperitoneum in most cases, including our example, and is commonly surrounded by encapsulated fluid. However, FIF has been reported to occur in other locations, such as within the cranium, the scrotum, and the oral cavity. Usually only one fetus is present but very rarely multiple fetuses may also be present. Five fetuses in the cranium of a 1-day-old female infant with hydrocephalus (although three of these fetuses were composed of extremities only) had been reported. The fetus itself is incomplete, containing a variable number of identifiable organs. The lung, the liver, the adrenal gland, the pancreas, and the genital organs may be seen in the more complex specimens. The presence of a head with eyes, hair, and teeth has been reported, but most of these fetuses are anencephalic. We are presenting a FIF mimicking a solid and cystic renal mass in a 6-month-old boy.

Key Words: fetus, abnormalities, newborn, retroperitoneal space, twins, teratoma

Introduction

Fetus in fetu (FIF) originally was described by Meckel in the late 18th century[1] and over 100 cases have been reported in the literature.[2] FIF is an extremely rare developmental abnormality secondary to abnormal embryogenesis in a monozygotic diamniotic pregnancy. It occurs when a vertebrate fetus is enclosed within the abdomen on any site of a normally developed fetus that is attached to the host chorionic circulation by a vascular anastomosis.[3] FIF is defined as a fetiform mass in which organ-like tissues are arranged around a vertebral axis. We report a case of retroperitoneal FIF in a boy who was referred with the diagnosis of cystic Wilms' tumor.

Case report

A 6-month-old boy was referred to our center with an abdominal mass. The examination was normal except for the presence of a mass in the left upper abdomen. The plain film of abdomen showed left sided calcification adjacent to the eleventh rib. The calcifications had the appearance of extremities rather than vertebral bodies. Abdominal ultrasonography showed a well-circumscribed mixed cystic and solid mass measuring 80 mm in diameter and located in the left hemi abdomen. It contained calcified solid elements suggestive of fetal bone structures. Also, left renal parenchymal was found attached to the main structural mass at the pelvic region. Abdominal computed tomography scan confirmed the previous findings. The diagnosis of FIF was considered. A 99 m Tc dimercaptosuccinic acid (DMSA) scan confirmed downward displacement of the left kidney, Figure 1. The surgery was performed through a left supraumbilical transverse incision. The tumor was dissected free from the left retroperitoneal space which was delineated by a thin, translucent membrane. The mass was fed by two arteries, arising from the left renal artery. The left kidney was relocated immediately into the normal position following resection of the retroperitoneal mass.

Pathologic findings revealed a fetiform structure weighing 500 gr and surrounded by a yellow liquid. The fetiform mass has had skin covered with hair, while the four limb buds were only slightly developed.

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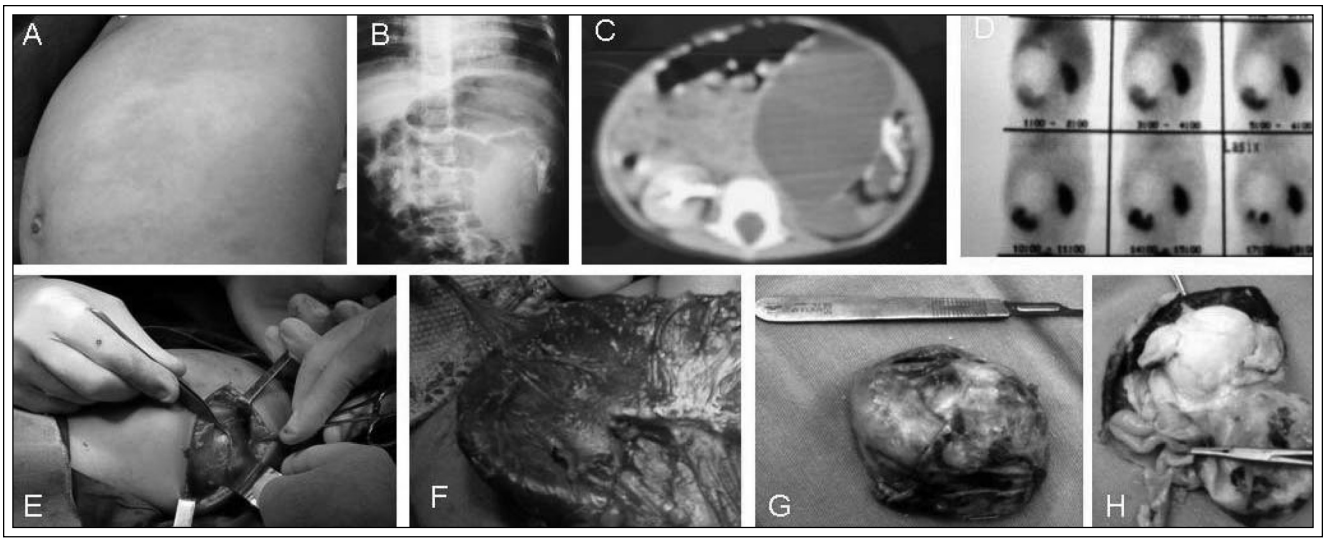

Figure 1. (a) left abdominal mass, (b) abdominal plain film showing the fetal bone structures, (c) axial CT scans of the mid abdomen level. The fetiform mass in the left retoperitonum and demonstration of fatty tissue surrounding the skeletal elements, (d) DMSA scan show a large left retroperitoneal mass displacing the left kidney downward, (e) a vascular pedicle, (f) vascular pedicle originated from the left kidney, (g) the mass with the covering amniotic membrane, (h) abnormal cerebral structures with four limb buds, intestine and appendix.

Dissection of the mass showed an extended cavity containing an intestinal structure connected to the appendix and vertebral column. The child had an uneventful postoperative course and 3 years follow-up evaluation showed no evidence of recurrence.

Discussion

FIF is a rare abnormality also defined as endoparazitic twin which has a 2 to 1 male predominance and is usually evident in first year of life.[4] It is an unusual condition consisting of a vertebrate fetus enclosed in the abdomen of a normally developing fetus. The incidence is 1 per 500,000 live births.[1]

Aberration in monozygotic twinning may rarely present as FIF. The entity was distinguished from teratoma by the presence of vertebral axis with limb buds. The pathogenesis of FIF is not fully understood, and two contrasting theories have been put forward. The theory of Willis[5] considers the FIF as an inclusion of a monozygotic diamniotic twin within the bearer. However, some investigators have proposed that FIF may represent a well-differentiated, highly organized teratoma.[6]

In some cases the non-calcified vertebral column invisible on the radiographs was identified by the pathologist; therefore, the non-visualization of the vertebral axis on radiography or on computed tomography scan does not exclude the diagnosis of FIF.

We believe that, this entity is not rare and some benign retoperitoneal mixed solid and cystic masses with bony calcification in children which are classified as teratoma could be a case of FIF. If the pathologist is aware about this condition and looks for fetal vertebrae, the diagnosis my convert to FIF and no further treatment is needed in this situation. Management of the FIF consists of complete surgical ablation. The prognosis is good, but given that the tumor is potentially malignant, clinical and radiological follow-up is indicated. □

References

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