Fetus-in-Fetu


Fetus-in-fetu is a rare abdominal tumor of infancy and childhood. In some cases an accurate preoperative diagnosis can be established, while in others diagnosis is not accomplished until laparotomy. Often, the tumor is confused with a teratoma, but with adherence to certain diagnostic criteria, along with a computed tomographic (CT) finding noted in our patient, distinction between the two tumors should be possible before surgical intervention. Our case is presented because the tumor is rare and because it is the first reported case in which CT could suggest the diagnosis preoperatively.

Case Report

A 9-month-old girl was taken to an outpatient clinic because of irritability and colicky abdominal pain that had lasted 1 day. On physical examination, the family’s physician found a left-sided flank mass and performed excretory urography. This study revealed a calcified mass displacing the left kidney downward. Calcifications in the mass were well formed and suggested bony structures, but one could not identify a specific bone such as a radius, ulna, femur, or vertebral column (figs. 1A and 1B). Further, slight radiolucency of the mass suggested that it might contain fat. The patient was referred to the University of Texas Medical Branch at Galveston and further investigation was undertaken.

Transverse computed tomographic (CT) scans of the abdomen were obtained from the xiphoid process to the pelvis in 10 mm increments. A large mass was identified in the left upper quadrant extending to the lateral and posterior aspects of the abdomen. Several areas of markedly increased attenuation consistent with bone were noted; in addition, there were large areas of low attenuation within the mass (fig. 1C). Statistical manipulation demonstrated that the lower attenuation areas were in the range of fat, and the lesion was diagnosed as a large teratoma.

Medical history revealed that the mother had had a normal pregnancy and an uncomplicated spontaneous vaginal delivery. Birth weight was 3.2 kg. On admission the baby was well nourished, apparently healthy, and in no acute distress. Physical examination revealed the previously noted left flank mass. On admission, the following laboratory data were obtained: white blood cells, 8,000/mm³; hemoglobin, 11.1 mg/dl; hematocrit, 30.30/0; platelets, 267,000/mm³; prothrombin time, 10.5/12; and partial thromboplastin time, 33/32.

Surgical exploration of the mass was undertaken through a left flank transverse incision. The mass was exposed by retroperitoneal dissection. A mobile, well encapsulated mass was located superior to the left kidney and could be separated from it without difficulty. Lateral attachments contained enlarged blood vessels suggestive of a parasitic blood supply. The main blood supply seemed to arise from the adrenal vessels. The mass was removed intact without difficulty.

Gross pathologic inspection of the surgical specimen demonstrated a mass measuring 10.0 × 6.5 × 5.5 cm. A thin, shiny, purple-pink capsule, suggestive of an amniotic membrane, was stripped away from the ‘tumor.’ Attached to the capsule was a segment of skeletal muscle with several ligated vessels representing the tumor pedicle. Grossly, the ‘tumor’ was covered by skin, black hair, and superficial sebaceous material. There were bilateral lower extremity appendages, jointed and with flipperlike feet (fig. 2A). Between these ‘legs’ was an immature external genital ridge, and over the top of the poorly formed trunk was a shiny, pink fibromuscular membrane, which on opening revealed a segment of blind-ending bowel that contained meconiumlike debris.

Postoperative radiography of the specimen after removal of the capsule demonstrated shortened deformed long bones of the lower extremities and a markedly hypoplastic trunk (fig. 2B). No clear-cut vertebral column could be identified but a clump of deformed bone was present in the area where the spine should have developed.

On microscopic examination, there was evidence of a variety of organs and tissues including adrenal cortex, skin, adipose tissue, respiratory epithelium, choroid plexus, genitai ridge, corpus cavernosa with central lumen, lymph nodes, ganglia, large intestine with interspersed ganglion cells, transonsal cell mucosa, bone and bone marrow, and cartilage. Neither vertebrae or spinal cord was identified.

Discussion

Fetus-in-fetu is a term originally coined by Meckel around 1800. Lord [1] reviewed a number of cases of this rare lesion and defined the entity as being a parasitic twin found within the abdomen of its sibling. Willis [2] stressed the distinct difference between fetus-in-fetu and abdominal teratoma. Although not all subscribe to complete separation of these two entities, according to Willis, fetus-in-fetu represents an aborted monozygotic twin while a teratoma is a

Received October 9, 1981; accepted after revision December 15, 1981.
1 Department of Radiology, Child Health Center, The University of Texas Medical Branch, Galveston, TX 77550. Address reprint requests to C. K. Hayden, Jr., Division of Pediatric Radiology.
2 Division of Pediatric Surgery, The University of Texas Medical Branch, Galveston, TX 77550.

AJR 138:762-764, April 1982 0361-803X/82/1384-0762 $00.00 © American Roentgen Ray Society
true neoplasm. Lewis [3] also emphasized this distinction and further stressed that teratomas are associated with malignant degeneration while fetus-in-fetu is not. Another difference between the two tumors is that fetus-in-fetu occurs more commonly in the upper retroperitoneum, while teratomas usually arise in the lower abdomen, in the ovaries, or in the sacrococcygeal region [4]. When they do occur retroperitoneally, they are usually located in the upper para-vertebral gutters [5].

A final point in distinguishing fetus-in-fetu from teratoma lies in the presence or absence of a vertebral column. Willis [2] stressed that a vertebral column should be present to secure the diagnosis of fetus-in-fetu. He reasoned that the embryologic development of an included twin must involve the primitive streak stage, which invariably produces a vertebral column. In those cases where the vertebral column is very dysplastic and underdeveloped, it may not be identifiable on a gross radiologic basis. Further, even with histologic section it may be difficult to demonstrate its presence. This may have been the situation with our patient, for the
lower extremities and pelvis were so well developed that it
would seem unlikely that the lesion could represent any
form of teratoma. Consequently, although Willis most likely
is correct in the vast majority of cases, there may be those
cases where a spinal column is so vestigial that it cannot be
identified with certainty, and yet other features of the tumor
would strongly suggest that it is a fetus-in-fetu. It is in this
type of patient that the potentially diagnostic CT finding we
decribe might be of assistance. As an added note, con-
cerning the presence of a vertebral column, one of the
cases of so called retroperitoneal teratoma described by
Griscom [6] may well have been a fetus-in-fetu. In this case,
advanced organogenesis and a clearly identifiable vertebral
column were present. Nonetheless, disagreement regarding
just how to classify fetus-in-fetu and teratoma will persist,
but since there is a complete difference in malignant poten-
tial, there does seem reason to try to differentiate the two
conditions.

Most often, as Knox and Webb [4] pointed out, fetus-in-
fetu is not diagnosed preoperatively, even though the radio-
graphic features retrospectively are usually quite typical. In
our case, misdiagnosis of a teratoma was made; the main
reason was that a vertebral column was not identified.
Further, although formed bony elements were seen, no
specific bones such as the radius, ulna, or femur could be
identified. In retrospect, the most clearly visible bone within
the tumor must have bone one of the femurs, but it did not
look exactly like a femur on the abdominal radiographs.
Thereafter, when the CT scan demonstrated fat in the lesion,
the diagnosis of teratoma seemed even more plausible, but
in retrospect, we believe that one feature on the CT scan
should have alerted us to the possibility of the mass being
a fetus-in-fetu. This feature, for the most part, consisted of
a round or tubular collection of very low density fat surround-
ing a central bony structure (probably one of the legs). In
teratoma, fat and bone, along with fibrous tissue, are inter-
spersed in a helter-skelter fashion, and thus we believe that
the demonstration of almost pure fat in a round tubular
configuration around a central bony density should suggest
the diagnosis of fetus-in-fetu. This may prove incorrect, but
at the moment the finding does seem to have some potential
for differentiating fetus-in-fetu from teratoma.

REFERENCES
1. Lord JM. Intraabdominal foetus in foetu. J Pathol
   1956;72: 627–641
2. Willis RA. The borderline of embryology and pathology. Lon-
   don: Butterworth, 1958;147
3. Lewis RH. Fetus-in-fetu and retroperitoneal teratoma. Arch Dis
   Child 1961;36:220–226
4. Knox JS, Webb AJ. The clinical features and treatment of fetus
   in fetu. Two case reports and review of the literature. J Pediatr
   Surg 1975;10:483–489
5. Partlow WF, Taybi H. Teratomas in infants and children. AJR
   1971;112:155–166
6. Griscom NT. The roentgenology of neonatal abdominal
   masses. AJR 1965;93:447–463
This article has been cited by:


5. Drew A. Torigian, Parvati Ramchandani. Retroperitoneum 1953-2040. [Crossref]


