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Correspondence to:
Sandip Kumar Rahul
Email: sandeep.rahul65@gmail.com
ORCID: 0000-0002-9620-9305

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Case Report

Fetus-in-Fetu Presenting as a Symptomatic Mass in an Infant: A Case Report

Sandip Kumar Rahul[1], Rupesh Keshri[1], Digamber Chaubey[2], Md Asjad Karim Bakhteyar[3]

1 Pediatric Surgeon, Department of Paediatric Surgery, All India Institute of Medical Sciences (AIIMS), Deoghar, Jharkhand, India
2 Associate Professor/Pediatric Surgeon, Department of Paediatric Surgery, All India Institute of Medical Sciences (AIIMS), Patna, Bihar, India
3 Pediatric Surgeon, Department of Paediatric Surgery, Indira Gandhi Institute of Medical Sciences (IGIMS), Patna, Bihar, India

ABSTRACT

Fetus-in-fetu represents an extremely unusual condition where a malformed parasitized twin with an organized vertebral column is present within the body of its host, leading to a variety of symptoms due to its mass effect. Its pathogenesis and features differ from both teratoma and fetiform teratoma. Detailed radiological evaluation helps in the assessment of the local anatomy and in planning the surgical procedure. The clinical features of a 9-month-old female with progressive abdominal distension and feeding difficulties are described. A firm, non-tender left-sided abdominal mass on palpation was found to be a well-defined encapsulated retroperitoneal lesion with bony structures resembling a vertebral axis on abdominal sonography and computed tomography. Surgical excision revealed an encapsulated mass containing rudimentary limb buds, hair, and partially developed vertebral elements. Histopathology confirmed the diagnosis of fetus in fetu. The postoperative course was uneventful, and the patient remained asymptomatic on follow-up.

Key words: Fetus-in-fetu, vertebral column, teratoma, fetiform teratoma

INTRODUCTION

Fetus-in-fetu (FIF) is a rare congenital anomaly characterized by the presence of a malformed parasitic twin having organized vertebral and limb structures. [1,2] Retroperitoneum is the most common site of FIF, although it may also occur in the mediastinum, cranial cavity, and Sacrococcygeal region. [1] The rarity of the condition and its resemblance to teratoma and fetiform teratoma make the clinical diagnosis challenging. Differentiation from a teratoma and a fetiform teratoma is important. We present a case of an infant with FIF where complete surgical excision was curative and histopathology confirmed the diagnosis.

CASE REPORT

A 9-month-old female presented with abdominal distension associated with repeated non-bilious vomiting but without any other bowel or bladder symptoms. There was a progressively enlarging painless left-sided upper abdominal mass noted for the last 2 months.

On examination, the abdomen was distended and a large heterogeneous non-tender palpable lesion occupying the left hypochondrium, epigastrium, left lumbar and umbilical regions (**Figure 1**) was palpable and confirmed to abut the stomach, small

bowel, pancreas, left kidney and colon with calcification in its lower portion on ultrasound and contrast-enhanced computed tomography scan (**Figure 2**). Bone appeared to assume the shape of a vertebral column. Clinically, it was not possible to differentiate whether it was an intraperitoneal or retroperitoneal mass due to its large size, but imaging (abdominal ultrasound and computed tomography scans) conclusively described its retroperitoneal location. Additionally, there was a well-defined, approximately 15 cm × 12 cm fluid-filled sac containing this mass. Serum alpha-fetoprotein level, routine hematological, and biochemical parameters were normal.

Excision of the lesion was performed under general anesthesia. Intra-operatively, a well-formed fluid-filled sac containing solid components with variable consistency was found. It measured around 15 cm × 12 cm and had polarity with the presence of organized bones in addition to soft tissue content. Poorly formed head and body parts were present (**Figure 3**).

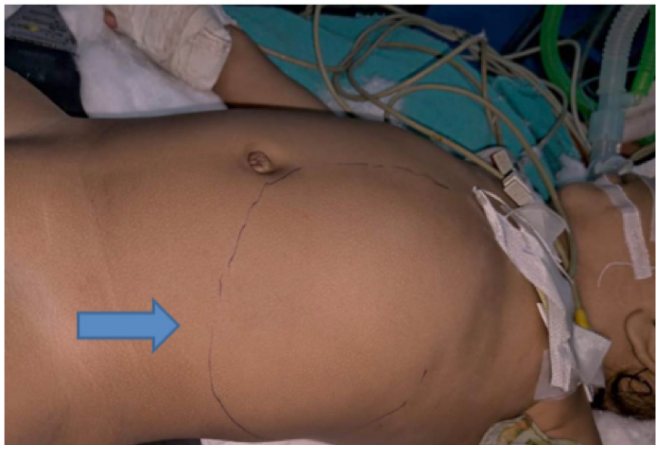

Figure 1: The marked area shows the span of the mass lesion.

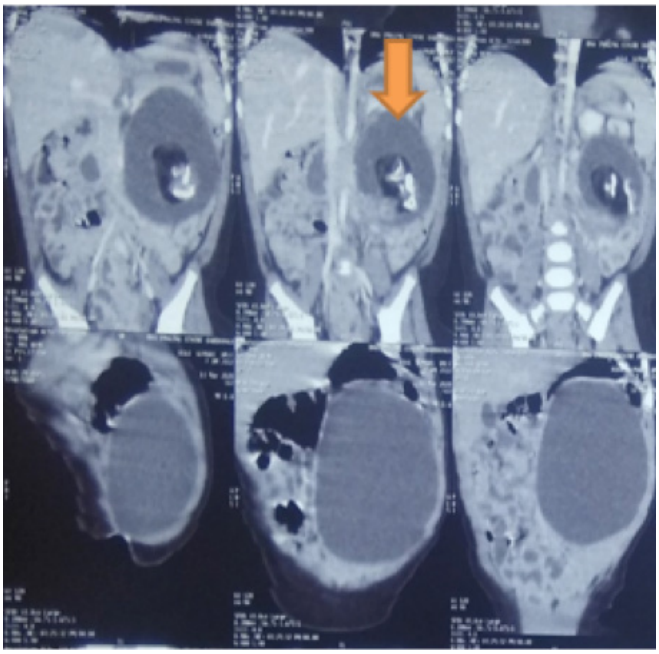

Figure 2: Computed tomography scan pictures showing the presence of organized bone in the cystic mass.

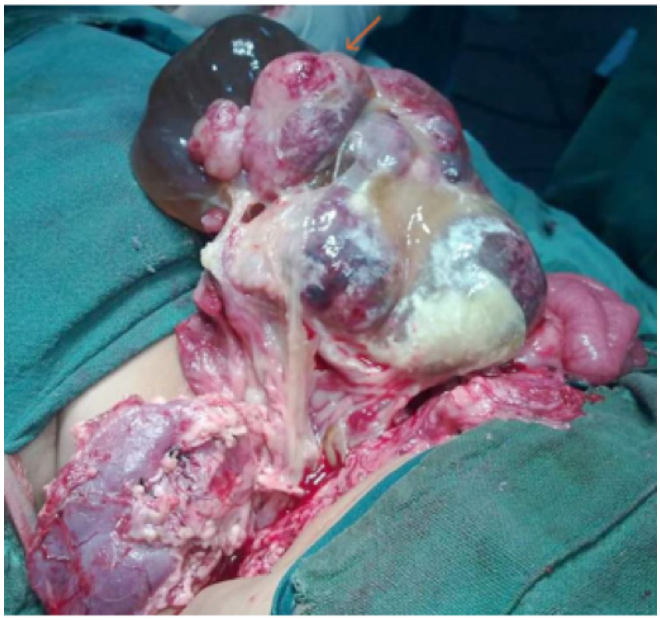

Figure 3: Fetus-in-fetu characterized by a head end, vertebral column, and inconspicuous other organs (brown arrow marks the head end).

An ill-formed cord-like structure connected it to the sac. The vascular pedicle was from the local retroperitoneal region. Rudimentary limb buds were poorly appreciated. The lesion was excised and sent for histopathology, which confirmed the presence of bones, cartilage, skin, and a cyst with cuboidal epithelium and evidence of FIF. There was no evidence of any immature element in the histopathology report. The patient has been asymptomatic on follow-ups in the last 3 years.

DISCUSSION

FIF is exceedingly rare, with an incidence of 1:500,000 births. [1] Growth of an FIF initially parallels that of its twin, but stops abruptly because of either the vascular dominance of the host twin or an inherent defect in the parasitic twin. The parasite lags in growth and persists as a mass lesion. On examining the mass, a fibrous membrane (equivalent to the chorio-amniotic complex) that contains some fluids (equivalent to the amniotic fluid) and a Fetus suspended by a cord or pedicle are both obvious. This was also evident in our case. Literature suggests the possibility of the presence of various tissues in different proportions: vertebral column, limbs, Central Nervous System (CNS) , Gastro Intestinal Tract (GIT), vessels, and genitourinary. [2] We could appreciate an ill-formed head and vertebral column, but other parts were inconspicuous.

A teratoma is a type of germ cell tumor that may contain several different types of tissue, such as hair, muscle, and bone. A fetiform teratoma (*homunculus*) is a rare pathology and manifests as an organized teratoma mimicking a malformed fetus. While differentiating a fetiform teratoma and FIF, it is important to note that while the former is the result of neoplastic processes of germ cells, the latter occurs due to aberrant embryogenesis, specifically the inclusion of a parasitic monozygotic twin. Vertebral columns and long bones are usually not seen in fetiform teratomas. [3,4] FIF is the most organized and differentiated of the three lesions and consists

Table 1: Differentiating fetus-in-fetu (FIF), teratoma, and fetiform teratoma.

Feature	FIF	Teratoma	Fetiform teratoma (highly organized teratoma)
Origin	Monozygotic twin (parasitic fetus)	Germ cell tumor (totipotent cells)	Teratoma with extreme organization
Vertebral column	Present (diagnostic)	Absent	Absent
Organ development	Partial organs (limbs, bones, etc.)	Disorganized tissues (hair, teeth, cartilage)	Mimics fetal parts, but no true organ systems
Malignancy potential	Benign	Can be benign or malignant	Usually, benign
Blood supply	Host-dependent vessels	Tumor vasculature	Tumor vasculature
Imaging (CT/MRI)	Fetal-like structures, spine visible	Heterogeneous mass, calcifications	It may resemble a fetus, but lacks a spine
Treatment	Surgical removal	Surgical removal ± chemo/radio	Surgical removal

of a parasitic monozygotic twin usually found inside the body of a newborn or infant. [3] **Table 1** summarizes the differences between FIF and fetiform teratoma. According to Spencer, FIF is the result of abnormal monozygotic diamniotic twinning with developmental arrest of a parasitic twin within the host. [3] This contrasts with the earlier Willis theory and teratoma theory. [5] The pathogenesis of parasitic twins found externally has also been similarly described. [3,6] The externally attached parasitic twins have now been known to develop by both abnormal fission and fusion mechanisms. [3,6,7]

Our case had all the characteristics of FIF. Excision, as in this case, provides a cure and, along with histopathology, confirms the diagnosis. Osama et al. described a similar case in an 11-month-old female, and excision led to a complete cure. [1] Tiwari et al. described two cases of FIF in 1-month-old infants; both had similar management with successful outcomes. [8]

CONCLUSIONS

FIF is rare but should be suspected in infants presenting with a well-encapsulated abdominal mass containing calcified structures. It needs to be differentiated from teratoma and fetiform teratoma clinically, radiologically, and histologically. Complete surgical excision is the treatment of choice, and an excellent prognosis is usually seen, as in our case.

PATIENT CONSENT

Written informed consent was obtained from parents for the publication of this case report and all associated images.

AUTHORS' CONTRIBUTION

All authors have significantly contributed to the work, whether by following the case at the bedside, conducting literature searches, drafting, revising, or critically reviewing the article. They have given their final approval of the version to be published, have agreed with the journal to which the article has been submitted, and agree to be accountable for all aspects of the work.

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CONFLICT OF INTEREST

None.

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