Case Report

Retained Foetus-in-Fetu or Fetiform Teratoma – A Diagnostic Dilemma and Operative Challenge

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Abstract

Foetus-in-fetu (FIF) is a condition where one of the twin foetuses is found developing inside the other. Differentiating this condition from fetiform teratoma (FT), in which the teratoma becomes highly differentiated to take a foetus-like appearance, often becomes arduous. Unfamiliarity with the situation and the presence of bone and hair within such lesions can potentially mislead the surgeon to diagnose it as a dermoid cyst erroneously. Suboptimal surgery can complicate things further when it comes to redoing operations and the final histopathological reporting. We report this case of a partially removed cystic retroperitoneal mass in a 14-year-old male, posing a significant surgical challenge due to dense adhesions to the vital retroperitoneal structures, such as common iliac vessels and ureter. We also faced similar diagnostic dilemmas during histopathology due to the tissues removed during the previous surgery. Based on gross and microscopic features, the possibility of a FIF or FT was rendered.

Keywords: Dermoid cyst, fetiform teratoma, foetus, foetus-in-fetu, teratoma

Introduction

Foetus-in-fetu (FIF) is an extremely rare condition.[1] The presence of an axial skeleton along with other tissues, is pathognomonic. Fetiform teratoma (FT) is an even more exceptional condition in which a high degree of differentiation leads to a foetus-like appearance, thus making it difficult to distinguish it from the former.[2] The rarity of both conditions makes them less entertained diagnoses. The presence of hair and bone in a well-encapsulated cyst often misdirects the surgeon into believing the condition to be a simple dermoid cyst as it had happened with us. Although the operative procedures for these conditions can often be challenging, published works of literature dwell little on the difficulties faced during surgical extirpation.

The case we are reporting had already been operated prior, for the same condition, before presenting to us, where debridement of the abdominal lesion was done instead of complete removal due to morbid adhesions. The case is being reported to highlight the importance of adequate pre-operative work up to rule out the other differentials and to condemn any suboptimal surgery such as a marsupialization, which would not only make the redo surgery more technically demanding but also pose a big problem for a definite histopathological diagnosis when some tissues have been lost to debridement. The description of both surgical and diagnostic challenges of this case will be an example to learn from for future surgeons dealing with such rare conditions.

Case Report

A 14-year-old boy had a painless, slowly progressive lump in the lower abdomen since childhood, for which he underwent a laparotomy 1 year before, at a tertiary care hospital. Operative records revealed a large retroperitoneal cystic mass measuring 15 cm × 10 cm in size, densely adherent to the surrounding structures and including bowel and major blood vessels. Intraoperatively, dense adherence of the mass forbade complete resection of the mass, and the surgeon removed approximately 40% of the cyst wall, with the evacuation of

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its content and cauterisation of the inner wall of the remaining mass.

A year later, he presented to our institute with a well-defined, firm and non-tender lump in the hypogastrium measuring 10 cm × 10 cm and a fleshy protrusion in the midline over the scar from the index surgery about 2 cm below the umbilicus for the past 6 months. On examination, the mass was soft, non-pulsatile, non-compressible and non-reducible, and measuring about 1 cm × 1 cm in size, with some black hair at the periphery and mucopurulent discharge [Figure 1]. The rest of the abdomen, general and systemic examinations were normal. There were no signs of any neurovascular deficits distally.

On abdominal ultrasonography, a well-circumscribed mass measuring 10 cm × 15 cm with multiple hyperdense structures resembling bone was seen in the lower abdomen and pelvis. The contrast-enhanced computed tomography (CECT) scan revealed both the common iliac vessels densely adherent and thromboses at the site of adhesion [Figure 2]. Flow in the external iliac arteries was intact bilaterally. The right ureter was compressed by the mass, causing right-sided hydroureteronephrosis. Magnetic resonance of the abdomen did not show any intervertebral extension of the mass. Exploratory laparotomy for complete removal of the mass was planned with a diagnosis of a partially excised dermoid cyst.

Intra-operative findings

As a result of the previous surgery and possibly the cauterisation of the inner wall, there were dense adhesions with the surrounding structures, including the small bowel, common iliac vessels and right ureter. A plane between the outermost fibrous tissue layer and the outer wall of the mass was entered to attempt a complete removal of the mass without injuring the adjacent vital structures. The adherent umbilicus, skin and overlying part of the rectus muscle were removed with the lump [Figure 3]. The common iliac vessels on both sides appeared cord-like, without pulsation. Dissection was continued medially to the vessels leaving behind a thin layer of fibrous tissue with the vessels to avoid injury to the vessels. This manoeuvre further helped to preserve the ureters by dissecting them off to the retroperitoneum. The dissection was relatively more straightforward in the posterior and the caudal part of the mass, probably because the planes were not violated in the previous surgery. Despite the difficulties due to morbid adhesions and difficult planes of dissection, the entire mass, which resembled an undeveloped foetus bearing hair on the scalp with three rudimentary limbs, was successfully removed. The post-operative period was uneventful without any vascular or neurological deficits.

Histopathological examination

Gross examination showed a partially skin-covered, irregular mass, measuring 15 cm × 10 cm × 8 cm, with three limb-like rudimentary structures. The cut surface revealed yellowish fatty areas and a few hard bony areas. However, no definite vertebral column or cranium was identified. Few pearly white soft areas were also visible [Figure 4a and b]. Microscopic examination of two of the appendageal structures revealed the presence of bones with marrow and a surrounding layer of skeletal muscle focally with skin and subcutaneous fat covering



Figure 1: Fleshy mass, measuring 1 cm × 1 cm in size, with some black hair at the periphery, with mucopurulent discharge



Figure 2: CT scan showing common iliac vessels densely adherent and thrombosed at the site of adhesion. CT: Computed tomography

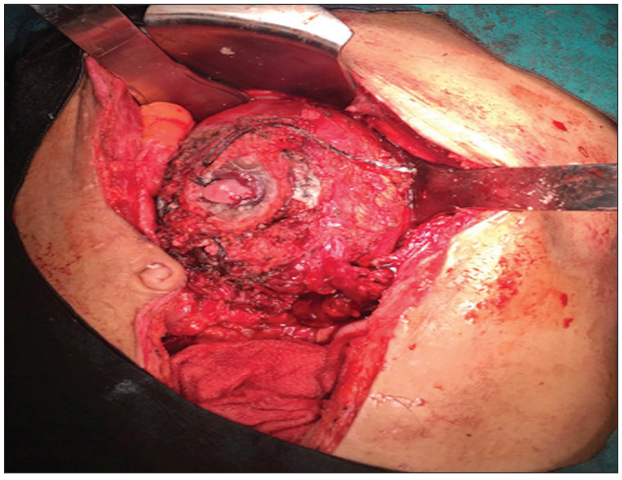


Figure 3: Intra-operative picture showing lump with adjacent tissues

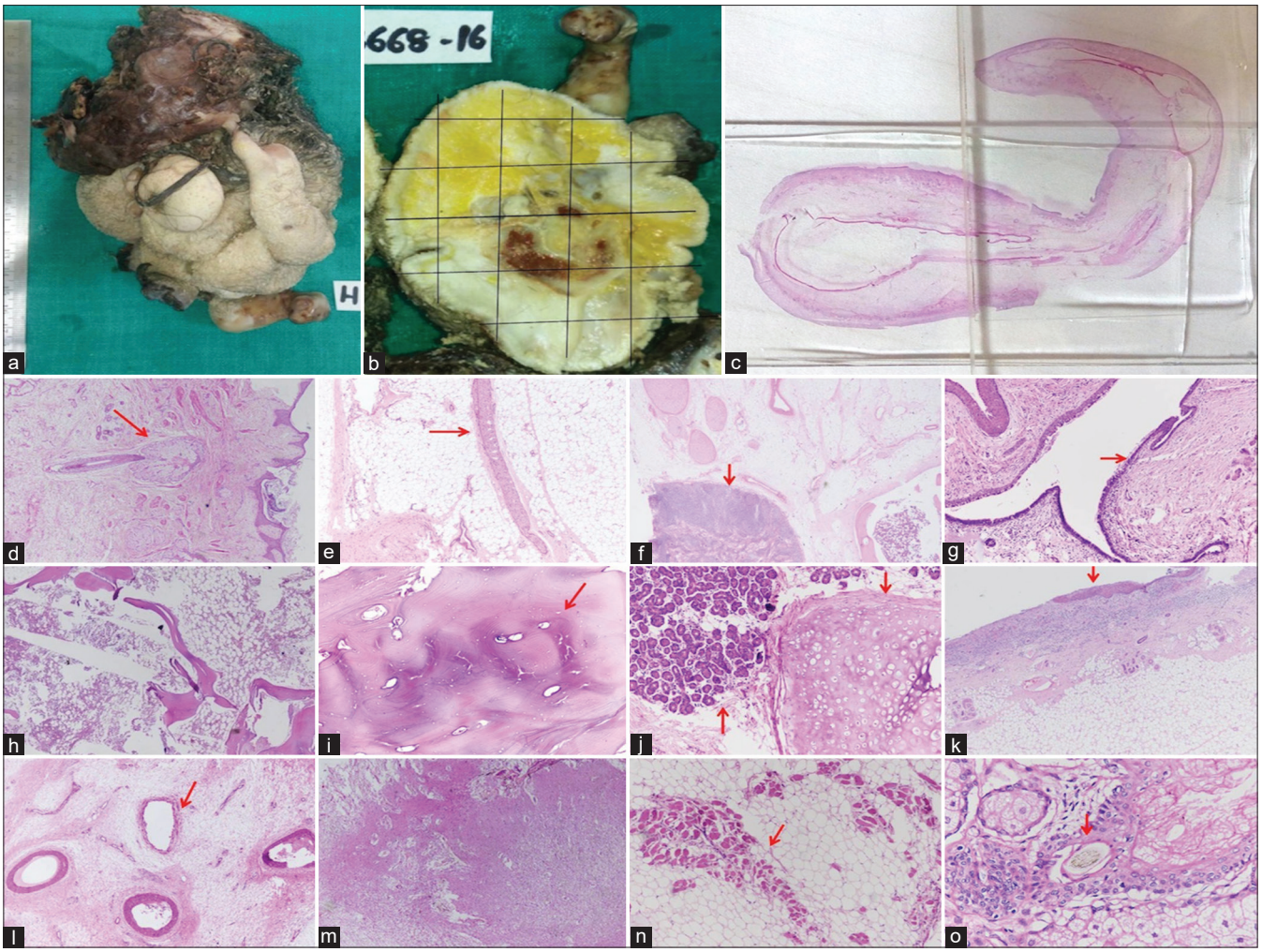


Figure 4: (a) Gross picture showing mass with a cauterised irregular area and a main lesion comprises a skin covered structure and presence of hair, limb-like structures, (b) The cut surface of the specimen shows mainly yellowish fatty areas and few hard areas of bone and cartilage, (c) The microscopic picture of limb-like structure (H and E), (d-o) The grid section of the entire specimen shows different areas such as the skin (d), nerve (e), lymph nodes (f), respiratory epithelium (g), bone marrow with hematopoietic elements (h), mature lamellar bone (i), cartilage and salivary glandular tissue (j), sweat glands (k), large arteries and vein (l), glial tissue (m), skeletal muscle and fat (n) and hair (o)

it, while the third limb-like structure showed only fat content. The entire grid of the specimen revealed many tissues such as skin with adnexal structures, nerves, lymph nodes, respiratory epithelium, bone marrow with hematopoietic elements, mature lamellar bone, cartilage, salivary glandular tissue, sweat glands, large arteries and veins, glial tissue, skeletal muscle and fat [Figure 4c-o]. However, no definite structures resembling gonadal tissue, thyroid tissue, lung parenchyma, gastrointestinal tract tissue or immature neuroepithelium were identified. Based on gross and microscopic features, the possibility of a FIF or FT was rendered.

In a follow-up of nearly 3 years, there has been no clinical or radiological evidence of recurrence.

Discussion

FIF is a rare condition where a parasitic twin foetus is found inside the other. It has a prevalence of 1 in 500,000 pregnancies, with only about 200 cases reported since it was first described by Meckel in the late 18th century.[3,4] FIF occurs in an anomalous monozygotic, diamniotic and monochorionic pregnancy, where an unequal division of totipotent cells of a blastocyst results in the inclusion of these cells into the more mature embryo. However, this diagnosis is closely mimicked by a rarer aberration where a teratoma becomes highly differentiated to take a foetus-like appearance, known as FT.[5]

Differentiation of a FIF from FT is imperative as the latter has malignant potential in contrast to FIF which is mostly benign. Only one case of malignant transformation in FIF is reported in the literature.[6] Some of the differences between a FIF and FT clinicopathologically are sex preponderance, axial skeleton, presence of specialised tissues and others [Table 1]. The presence of the vertebral column is quintessential to the diagnosis of FIF, unlike that of the FT.[7] It was, however, absent in our case. Histopathology report of the previous surgery evidenced that the contents of the cyst, including a

lot of hair and some bones, had been removed in the index surgery. This put us in a dilemma if the vertebral column was essentially absent as in an FT, or if that was lost to the previous evacuation of the contents of the cyst in the index surgery, of what was probably a FIF. This situation was unique to our case as no other instance of a retained or a recurrent FIF or FT has ever been reported in the literature. A review of available literature revealed that in an adult male, the possibility of FT is extremely rare, with only two cases described to the best of our knowledge [Table 2]. However, with the atypicality of our situation, it is quite impossible to come to a satisfactory conclusion. Some recent articles suggest that both entities could be a part of the spectrum of the same condition at different stages of maturation.[14]

The diagnosis of a FIF can be easy and even be diagnosed antenatally by ultrasonography when the axial skeleton is present. After birth, computed tomography (CT) angiography is an excellent tool for diagnosis. It delineates surrounding vascular relations in addition to the identification of bony components in mass. The pre-operative CECT scan and the previous operative note in our case were the first concrete clues to anticipate difficulty due to morbid adhesions to the ureter and the vessels. The CECT helped us plan our plane of dissection intraoperatively so that the iliac vessels and ureter could be carefully dissected and preserved.

Residual FT or FIF has never been reported earlier in English literature to the best of our knowledge. In either situation, complete removal of the mass is recommended to avoid recurrence. Partial removal of the tumour, as done during the indexed surgery is to be condemned, as subsequent completion surgeries would be more difficult owing to the prior interference of the planes of dissection and ensuing inflammation and fibrosis. Moreover, a definitive diagnosis can remain elusive if relevant tissues are lost to debridement. Follow-up with Agence France-Presse level is recommended to diagnose the recurrence of an FT.

Table 1: Difference between foetus-in-fetu and fetiform teratoma

Feature	FIF	FT
Sex	Male preponderance	Female preponderance
Age	Newborn and infancy	Adolescents and young adults
Presentation	Abdominal mass	Abdominal mass
Axial skeleton	Highly developed and segmental	Not well-developed
Organ development	Well-developed	Absence of well-developed organs
Specialised organs	Yes	No
Skeletal muscles	Usually present	Rare
Zygosity testing	Homozygous	Heterozygous

FIF: Foetus-in-fetu, FT: Fetiform teratoma

Conclusion

FIF is a benign condition, whereas FT has malignant potential. Conclusive pre-operative diagnosis may not always be possible, and distinguishing them intraoperatively is difficult; hence complete removal, in either case, should be the rule for both surgical and diagnostic ease. Extensive pre-operative evaluation, including a CECT scan, should be mandatorily done to anticipate intra-operative difficulty and plan the dissection accordingly.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published, and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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Table 2: Clinicopathological diagnosis of foetus-in-fetu/fetiform teratoma in adult males: A review of the literature

Case	Age (years)	Site	Presentation	Duration	Vertebral column	Limbs	Size	Genetic analysis	Pre-operative diagnosis
Shrivastava *et al*.[8]	27	Retroperitoneum	Pain and mass progressive	3 years	Present	NR	20 cm×18 cm cystic	NR	Teratoma
Awasthi *et al*.[9]	30	Retroperitoneum	Pain, constipation and mass	Childhood	Present	4	26 cm×15 cm×12 cm	NR	FIF
Massad *et al*.[10]	27	Retroperitoneum	Dysphagia and abdominal swelling	Childhood	Present	4	14 cm	46 xy Absence of inversion 12p	
Lee *et al*.[11]	39	Intraperitoneum	Abdominal discomfort	Incidental	Unclear	2	NR	NR	FIF
Sharma *et al*.[12]	36	Retroperitoneum	Abdominal discomfort	NA	Present	4	27 cm	NR	FIF
Daga *et al*.[13]	20	Retroperitoneum	Abdominal distension	Childhood	A. Present B. B-present	A-2 B-2	NR	NR	FIF (2)
Present (2018)	14	Retroperitoneum		Childhood	Unclear	3	15 cm×10 cm	NR	Teratoma

FIF: Foetus-in-fetu, NR: Not reported

Conflicts of interest

Pankaj Kumar, Tushar S Mishra, Sindhu Sankaran, Mukund N Sable, Swagatika Samal, declare that they have no conflicts of interest.

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