

Fetiform Teratoma Presenting as Retroperitoneal Mass in Three-day-old Newborn: A Case Report

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ABSTRACT

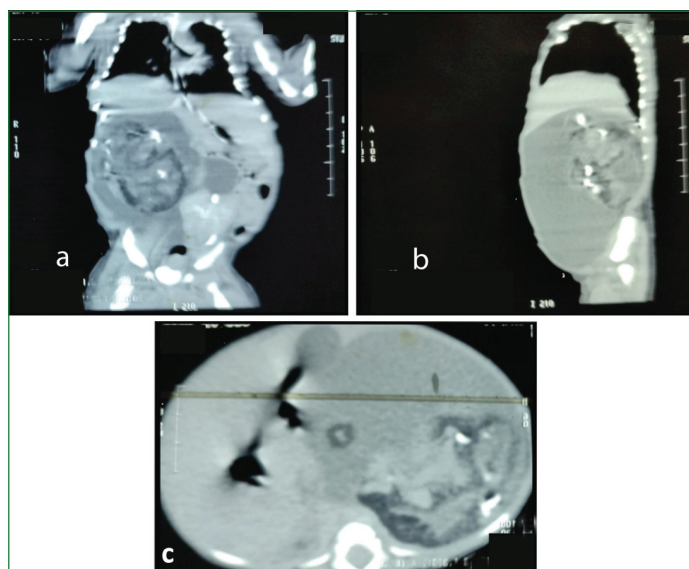
Fetiform Teratoma (FT) is an extremely differentiated neoplasm formed due to unsuccessful migration of primordial germ cells during embryological development. Foetus in Fetu (FIF) is a very rare entity in which a non viable foetus gets reabsorbed within a normally developing foetus, thus mimicking reabsorbed twin. Herein, the authors present a case of three-day-old female who presented with retroperitoneal mass and difficulty passing urine. Ultrasonography (USG) revealed a large, well-defined, heterogeneously hypoechoic solid cystic lesion with internal vascularity noted in retroperitoneum. Histopathology confirmed teratoma due to presence of ectodermal, mesodermal and endodermal components. The authors share the difficulties faced in differentiating between both the conditions. It is of utmost importance to identify FIF and FT antenatally and to diagnose the conditions, as early as, possible to provide adequate management on time, as they are potential mimickers of each other.

Keywords: Foetus in fetu, Hypoechoic, Mature cystic teratoma, Prenatal diagnosis

CASE REPORT

A three-day-old female child was brought by her parents to the Paediatric Surgery Outpatient Department (OPD) with the chief complaints of abdominal mass noticed by the parents one day back. Child also experienced inability to pass urine and stool since one day. Clinical examination revealed a firm, ill-defined mass in the central abdomen, which was felt more towards the right iliac fossa. The USG of abdomen revealed a large, well-defined, heterogeneously hypoechoic solid-cystic lesion with internal fat fluid level, calcification casting shadow and internal vascularity noted in retroperitoneum. The mass was seen displacing bowel loops and right kidney to the left-side, with the presence of gross ascites and debris seen. Contrast-enhanced Computed Tomography (CECT) of the abdomen and pelvis showed a large, well-defined mass in the retroperitoneum, measuring 10×9×8 cm, consisting of numerous macrocalcifications and fat components displacing the bowel loops anterolaterally, abutting the liver surface superiorly and inferiorly abutting right lateral wall of urinary bladder, hence suggestive of retroperitoneal teratoma [Table/Fig-1a-c]. With the provisional diagnosis of retroperitoneal teratoma, the baby was operated on, and tissue was sent for histopathological examination. Grossly, two skin-lined globular structures were received altogether, measuring 11×8.5×3.5 cm, whose cut section revealed variegated areas with predominant cartilage.

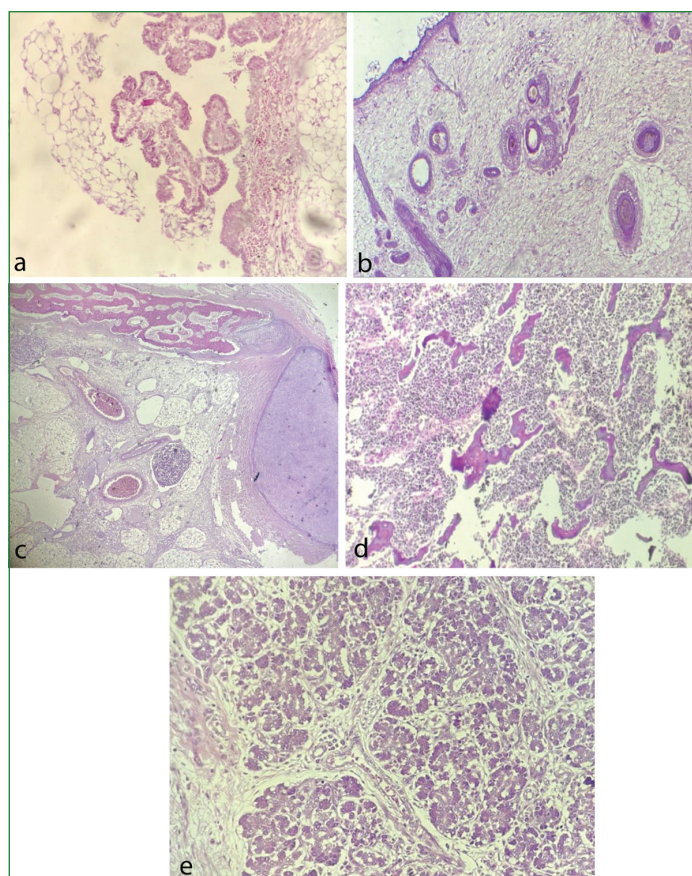
The smaller skin-lined globular mass measuring 4×3.5×1.5 cm showed variegated cartilaginous areas [Table/Fig-2]. Microscopy showed chorionic villi indicating foetal component intermixed with adipose tissue [Table/Fig-3a], lined by stratified squamous epithelium with dermal appendages (hair follicle, sebaceous glands and apocrine glands) and glial tissue [Table/Fig-3b,c]. Other focuses showed presence of fibrofatty tissue, bony trabeculae, mature hyaline cartilage, muscle tissues, salivary gland with acini and intervening marrow elements [Table/Fig-3d,e]. Hence, a final diagnosis of fetiform teratoma was made. Following the surgical removal of the mass, the child was doing well; however, the child was lost to follow-up two months after starting chemotherapy regimen, which included Bleomycin, Etoposide and Cisplatin (BEP).



[Table/Fig-1a-c]: CT abdomen and pelvis shows large well-defined mass in the retroperitoneum, measuring 10×9×8 cm, consisting of numerous macrocalcifications and fat components displacing the bowel loops anterolaterally.



[Table/Fig-2]: Gross image showing two skin lined globular structures altogether measuring 11×8.5×3.5 cm, which revealed variegated areas with predominant cartilage. The smaller skin lined globular mass measuring 4×3.5×1.5 cm showed variegated cartilaginous areas.



[Table/Fig-3]: a) Microsection showing chorionic villi indicating foetal component intermixed with adipose tissue (H&E, 400x); b) Microsection showing stratified squamous epithelial lining with dermal appendages (H&E, 100x); c) Microsection showing bony trabeculae, intervening fibrofatty tissue and glial tissue (to the left) (H&E, 40x); d) Microsection showing bony trabeculae and marrow elements in between vascularised stroma (H&E, 100x); e) There is presence of salivary glands with intervening acinar structures (H&E, 100x).

DISCUSSION

The FT is an extremely differentiated neoplasm formed due to unsuccessful migration of primordial germ cells during embryological development. FIF is a very rare entity in which a non viable foetus is reabsorbed within a normally developing foetus, thus mimicking reabsorbed twin [1]. Hence, it is of utmost importance to identify FIF and FT antenatally and to diagnose these conditions, as early as, possible to provide adequate management on time. Herein, the authors present the case of three-day-old female who presented with retroperitoneal mass and the difficulties faced in differentiating between both the cases.

Teratomas has been derived from the Greek word 'teras', meaning monster, and 'oma', means tumour or swelling. Teratomas contain differentiated tissues derived from germ cells, such as skin from ectoderm, muscle and cartilage from mesoderm, and endodermal derivatives like lining of the respiratory or gastrointestinal tract. Teratomas contain cell derived from two or more germ layers. A mature teratoma shows well-differentiated tissue structures, while an immature teratoma which also shows the presence of neural tissues [2-4]. So far, only 100 cases have been described in literature, according to Alsulaimani AA and El-Zeky AME, reported in 2018 [1].

The four variants of teratoma histologically are mature cystic teratoma, immature, monodermal and teratoma with malignant transformation. However, recently there has been a fifth addition, referred to as FT, grossly visible mature skeleton in the absence of visceral organ and other skeletal tissues, representing a highly organised teratoma. In contrast, FT show a grossly visible, well-developed axial skeleton, suggesting the occurrence of gastrulation in a developing embryo during the third week of development [5-7].

The FIF has been described since 17th century; however, it was Willis RA, who described it as a separate entity consisting of organs, limbs, and vertebral column [8]. Spencer R described the presence of one or more of following criterias for the diagnosis of FIF: normal skin lining, enclosure within a well-defined sac due to persistence of vitellointestinal duct, grossly visible anatomic body parts, connection to the host by few large blood vessels, and location usually near to the attachment of the conjoined twins or to the neural tube or the gastrointestinal system. However, teratoma shows a wide attachment area with abundant smaller blood vessels and less differentiated tissues [9].

The FIF has been seen in 1 in 500,000 births. It is almost always seen in retroperitoneal location, in contrast to FT which can be found anywhere in the body in the midline [8]. FIF presents early in neonates in comparison to FT which presents in later life aged 9-65 years from ovaries [9]. FIF is benign condition, whereas teratomas, being malignant, possess a higher risk of recurrence [10].

Two theories have been proposed in the literature: the parasitic twin theory, which suggests that a parasitic foetus formed inside the body of its host twin, supplying a common blood supply to both; but parasitic twin is usually malformed and usually dies before birth due to lack of blood supply. The second theory is of highly differentiated form of teratoma. An aberration in the process of diamniotic, monozygotic twinning may result in unequal division of the totipotent inner cell mass of the developing blastocyst. As a result, a smaller cell mass may develop within a maturing sister embryo [11].

Antenatal USG is indeed essential for diagnosing conditions in the foetus due to its safety and non invasive nature. Postnatally, Magnetic Resonance Imaging (MRI) can provide a more detailed view, especially for assessing the extent of mass and their involvement with surrounding structures, including the intrapelvic region. When it comes to treatment, surgical resection of the mass is typically the primary approach, especially if there is a risk of malignancy or complications, followed by chemotherapy regimen. Regular follow-up with tumour markers like Alpha-Fetoprotein (AFP) and Beta-Human Chorionic Gonadotropin (β -HCG) is required in teratoma [11]. Additionally, evaluating for potential metastases or secondary growths is crucial in formulating a comprehensive treatment plan. Combining these diagnostic and therapeutic strategies helps optimise outcomes for the patient.

Similarly, Puranik RU et al., found a male baby born at 40 weeks of gestation with a hypoechoic mass in subhepatic region, confirmed to be foetus-in-fetu [11]. The present case, being retroperitoneal, pointed towards FIF, but absence of vertebral column and presence of well-organised dermal appendages (hair follicle, sebaceous glands and apocrine glands), glial tissue, fibrofatty tissue, bony trabeculae, mature hyaline cartilage, muscle tissues, salivary gland with acini, and intervening marrow elements favoured towards a diagnosis of FT.

CONCLUSION(S)

The present case showed clinical heterogeneity due to the presentation of FT in a three-day-old female, with retroperitoneal location would suggest FIF. In a foetus, the differential diagnosis of a retroperitoneal mass includes two close differentials: teratoma showing parts of foetus like mature and immature components whereas FIF may show vertebral column, hence a multimodal approach is required antenatally to provide adequate management on time.

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