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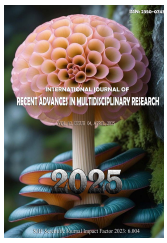

RESEARCH ARTICLE

FETIFORM TERATOMA: A RARE CAUSE OF RETROPERITONEAL MASS: A CASE REPORT

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ABSTRACT

Fetiform teratoma is a rare highly developed mature teratoma with organoid differentiation although it is not as developed as fetus in fetus which is the only differential diagnosis of this entity. It should be distinguished from the fetus in fetu by absence of the axial skeleton. *However, there is controversy in the distinction between Fetus in Fetu and fetiform teratoma and that both entities could represent two aspects of the same pathology at different stages of maturation. A day 6 female child with abdominal massunderwent ultrasonography, CT-scan and surgery.On imaging, Large well defined solid cystic lesion with soft tissue and macrocalcification in retroperitoneum in right side displacing right kidney to left side suggestive of Retroperitoneal Teratoma.. The entire sac with its content were completely excised. Tumor markers wasAFP >2000ng/ml, BHCG =15IU/L. The patient underwent surgery and The mass was excised in toto and observed having three distinct fetal heads with hair and clumped trunks.Very few cases of fetiform teratoma have been reported in English literature, however Highly developed teratoma is a diagnostic dilemma as it resembles fetus in fetu. The absence of a spinal axis differentiates this entity from the fetus in fetu.*

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INTRODUCTION

Fetiform teratoma is a rare highly developed mature teratoma with organoid differentiation although it is not as developed as fetus in fetus which is the only differential diagnosis of this entity. Teratomas are neoplasms made up of tissues that do not belong to the anatomical site where they are found. They show varying degrees of differentiation, from simple somatic elements to highly structured formations with axial and metameric organization. Its most frequent location is in the sacrococcygeal regional though they can also be located in ovaries, retro peritoneum, head, neck and mediastinum. Fetus In Fetu (FIF) is a rare congenital malformation, distinguished from Fetiform Teratomas (FT)by the presence of a vertebral axis in an encapsulated, pedunculated mass, of the fetiform type, with disorganized structures around this axis. It is more frequently located in the retroperitoneal area. However, there is controversy in the distinction between Fetus in Fetu and fetiform teratoma and that both entities could represent two aspects of the same pathology at different stages of maturation.Since FIF is a benign malformed tissue, chances of malignant transformation and postoperative recurrence are very low, and simple surgical resection of mass is sufficient in most cases, with no further follow-up required. On the other hand, teratomas being a true neoplasm, have chances of malignant transformation and require a complete evaluation before surgery, including radiological imaging and serum tumor markers evaluation such as serum Alfa feto protein (AFP) and beta-human chorionic gonadotropin (HCG). Management of teratomas may also include chemotherapy before or after surgery, depending upon the stage of the tumor and other clinical factors[1].

CASE REPORT

A day six female child with abdominal distension since birth as noticed by her mother and was gradually increasing in size. There was no history of any gastrointestinal or genitourinary symptoms. No history of birth asphyxia and was normal vaginal delivery, there was no history of maternal illness or exposure to radiation or drug intake during pregnancy. There was no previous history of twin pregnancy in the mother.On examination, Abdomen was protuberant. Umbilicus was inverted & shifted to left side. Right side flank was full, There was a lump occupying mainly right lumber region, and partly umbilical region right side and right hypochondrium around

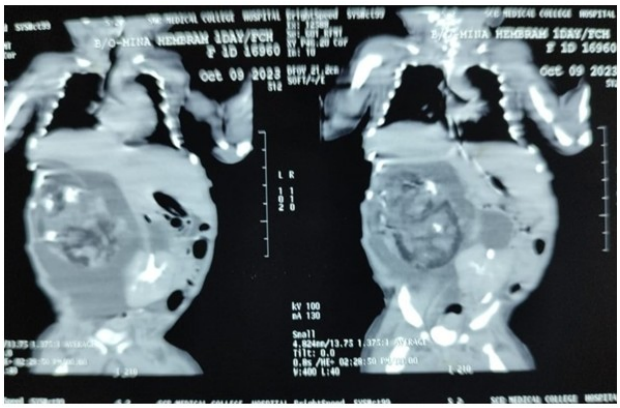

Figure 1. CECT of Abdomen and Pelvis

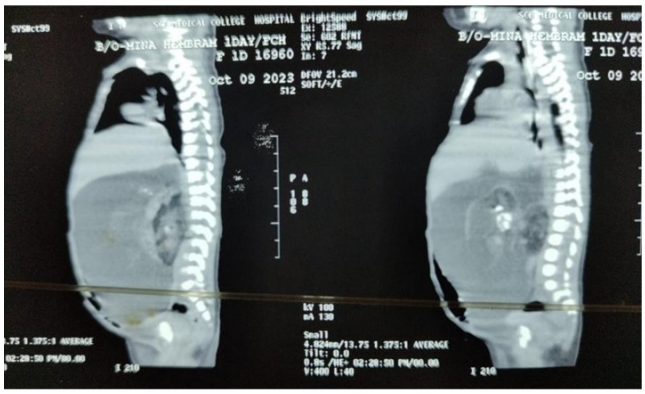

Figure 2. CECT of Abdomen and Pelvis

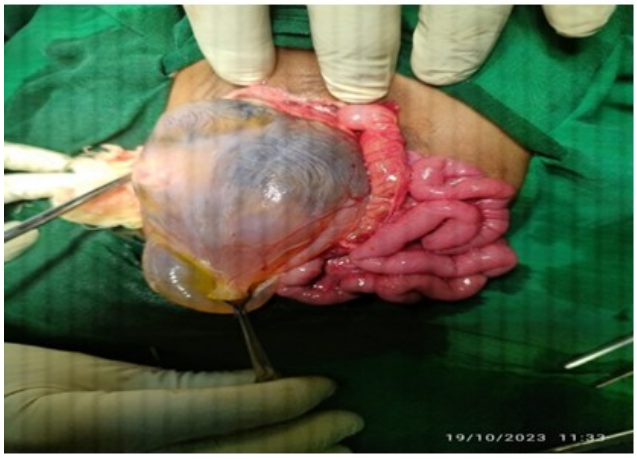

Figure 3. Encapsulated retro peritoneal cystic mass

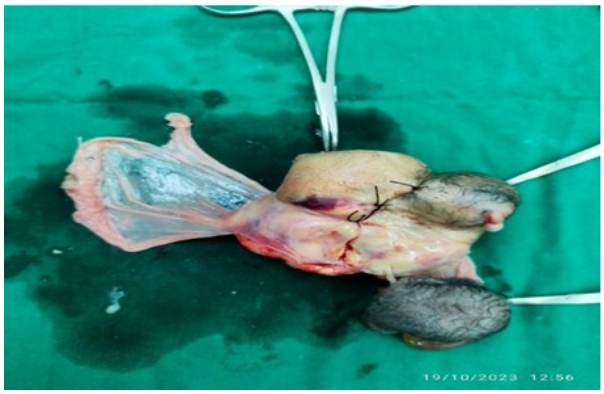

Figure 4. Showing three distinct fetal heads with hair and clumped trunks

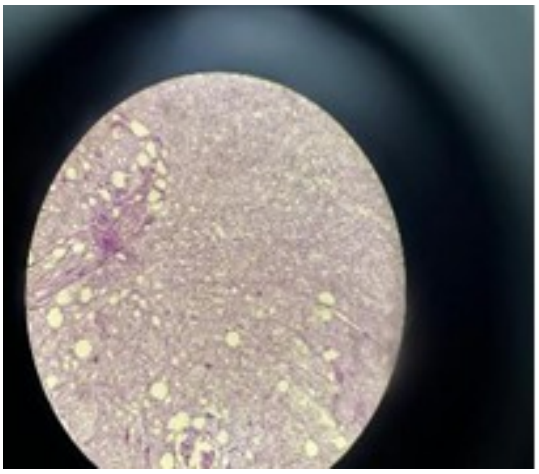

Fig 5. Glial tissue

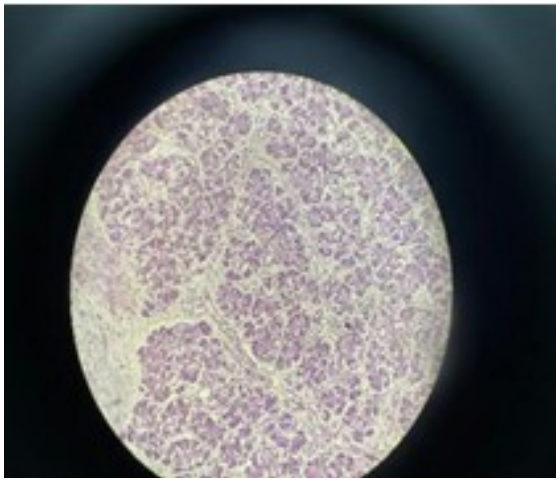

Fig. 6. Glandular tissue

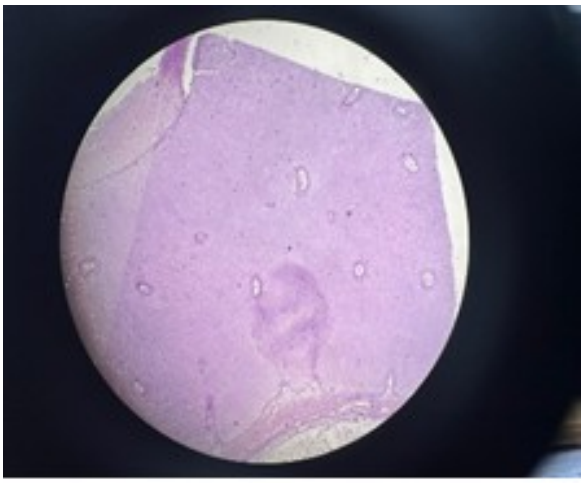

Fig. 7. Cartilage

7*7 cm, movable more in vertical axis than horizontal axis. On USG a large (9.4 *6.1 cm) well defined heterogeneously hyper echoic solid cystic lesion with fat fluid level macrocalcification noted in retro peritoneum displacing bowel loops and right kidney to left side ? Retroperitonal Teratoma. On CECT abdomen and pelvis; -9*8*10cm,large well defined solid cystic lesion with soft tissue and macrocalcification in

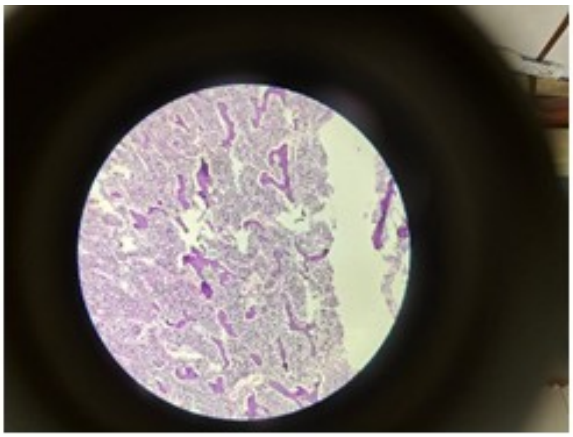

Fig. 8. Bony trabeculae

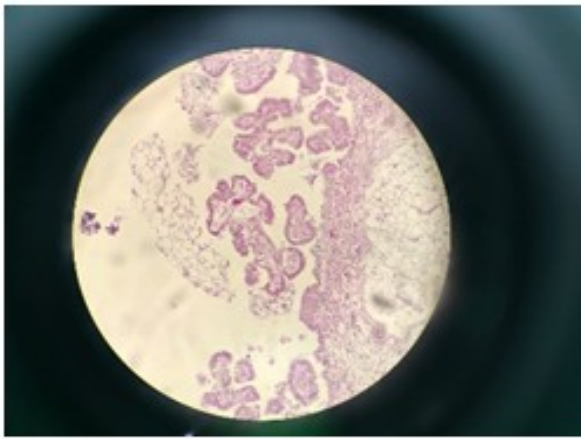

Fig. 9. Skin appendages

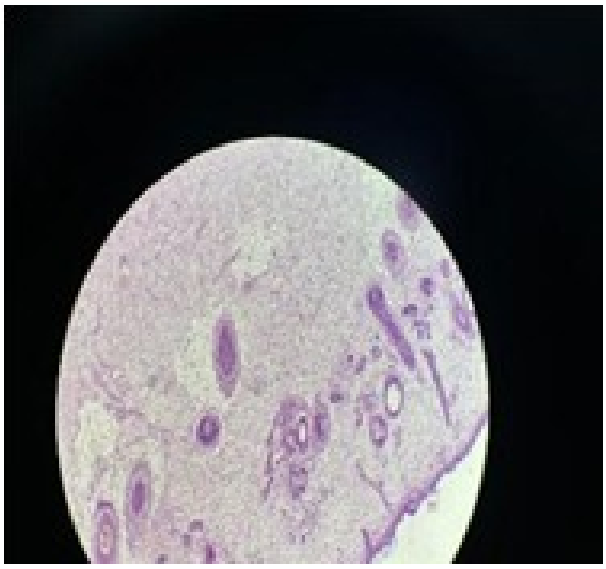

Fig. 10 Choroid plexus

retroperitoneum in right side displacing right kidney to left side suggestive of Retroperitoneal Teratoma. (Figures 1 and 2), serum tumor markers evaluation such as serum Alfa feto protein (AFP) and beta-human chorionic gonadotropin (HCG) was done and *AFP >2000ng/ml, BHCG =151U/L.* The patient underwent a laparotomy which revealed an encapsulated mass measuring 9*8*10cm in retro peritoneum on right side displacing Right kidney to left side, Part of transverse colon is adhered to the cystic mass which was separated by adhesiolysis. The encapsulated cystic mass was excised in toto (figure 3) and observed having three distinct fetal heads with hair and clumped trunks (Figure 4). On Gross pathology skin lined globular mass with cross section shows variegated and cartilaginous areas and onHistopathology examination ;- Elements from all germ layers like glial tissue,Glandulartissue,cartilage,bony trabeculae with marrow elements, skin with dermal appendages was noted suggestive of teratoma(Figure 5-10).

DISCUSSION

Fetiform teratoma (homunculus) is a term that has been given to a rare form of teratoma that resembles a malformed fetus. There are very few reported cases of this entity in the English-language literature. This tumor must be distinguished from both fetus in fetu (a parasitic monozygotic twin usually found inside the body of a newborn or infant) and an ectopic pregnancy[2]. Majority are composed of disorganized, neoplastic, mature tissues of one or more of the embryonic germ layers: ectoderm, mesoderm, and endoderm. Rarely, these tumors develop a high degree of differentiation and organization, resembling a malformed fetus (fetiform structure). It has been proposed that fetiform teratoma and fetus in fetu can be differentiated based on zygosity[3]. Mature cystic teratomas arise from a single germ cell that has completed the first meiotic division. Unspecialised germ cells differentiate into different types of specialized cells, resulting in a tumour that resembles a fetus. Further support for this theory is that the anatomic distribution of this neoplasm is along the primordial germ line[4]. Fetus in fetu has classically been distinguished from teratoma in that the diagnosis of the former requires the presence of a highly developed and segmented axial skeleton[5]. Interestingly, fetus in fetu almost always presents with acardia and anencephaly. In contrast, fetiform teratoma usually does not have complex, well-developed organs[6].

CONCLUSION

To summarize we have described a fetiform teratoma, a rare form of a mature cystic teratoma that is not highly developed and organized, resembling a fetus like structure. The degree of organization and differentiation can vary, blurring its distinction from either fetus in fetu or ectopic pregnancy. Its diagnosis requires both a clinical history and complete evaluation before surgery, including radiological imaging and serum tumor markers evaluation as chances of malignant transformation . Management of teratomas may also include chemotherapy before or after surgery, depending upon the stage of the tumor and other clinical factors,pathological examination. Cytogenetic/molecular studies may be helpful in distinguishing difficult cases[7].

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