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A Rare Case of a Giant Retroperitoneal Teratoma In An Adolescent; A Case Report.

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Abstract: *Teratoma originates from multipotential germ cells that recapitulate normal somatic development by developing elements of the three embryonic germ layers. Primary retroperitoneal teratoma develops from totipotential germ cells that have failed to migrate their normal gonadal locations.*
Case summary: Here we present a case of an adolescent girl presenting with a huge abdominal swelling. On clinical examination it was a hard abdominal mass. Radiological investigations revealed that a large retroperitoneal mass extending from epigastrium to hypogastrium filling bilateral flanks, displacing bowel loops laterally, pancreas superiorly and bilateral kidneys inferiorly. The mass was composed of cystic areas with fat and bony components within.
Conclusion: *Laparotomy with excision of the retroperitoneal mass was performed and pathology report confirmed the diagnosis of a mature cystic teratoma with no evidence of malignancy.*

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I. Case Report

A 15 years old girl presents with gross abdominal distention with mass per abdomen.She felt the mass 6 years ago, since then the mass was gradually increasing but she sought medical advice recently only when she felt difficulty in breathing due to the mass. She denied any associated symptoms like fever, loss of appetite, weight loss, nausea, vomiting, pain ,bowel or urinary symptoms. She had no any known underlying disease. On clinical examination a large hard lump was noted extending from epigastric region to hypogastric region filling both the flanks. It was non-tender with lobulated surface. The mass was fixed and did not move with respiration Routine blood tests showed mild anaemia .A frontal abdominal radiograph(Fig.1) shows a large heterogenous abdominal mass with multiple areas of sheet like and foci of calcifications within. Bowel loops were displaced laterally by the mass.

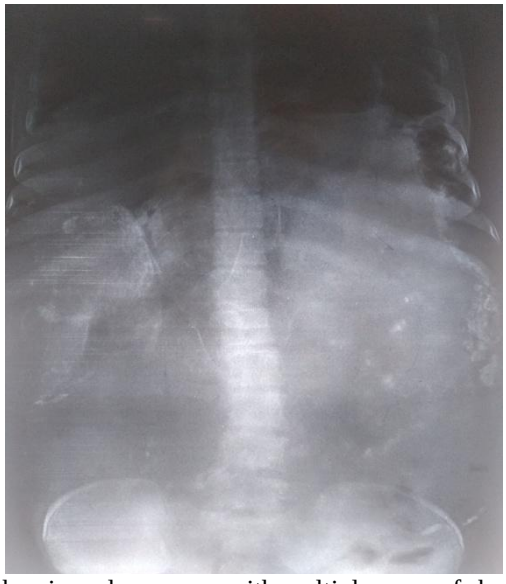
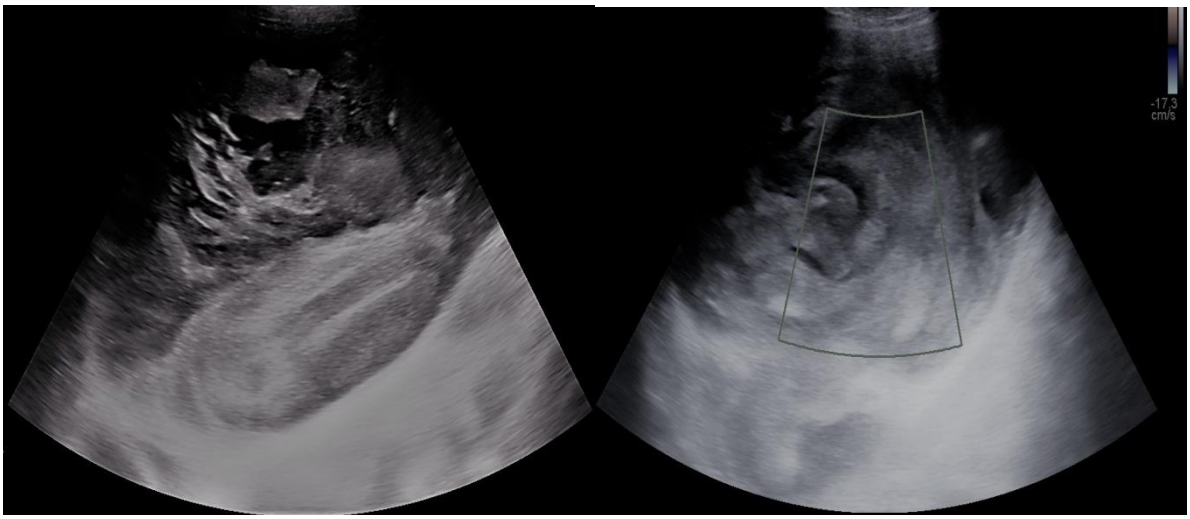


Fig. 1. Plain X ray abdomen showing a large mass with multiple areas of sheet like and foci of calcifications within. Bowel loops are displaced laterally and superiorly.

Ultrasonography demonstrates a large heterogeneous abdominal mass (Fig.2a&b) measuring approximately 29.2cmX18.2cm extending from epigastric region to hypogastric region and extending laterally to bilateral flanks with multiple linear and foci of calcifications within. Few cystic areas are noted within the mass. Pancreas was displaced superiorly, bilateral kidneys displaced inferiorly and bowel loops were displaced laterally. On color Doppler the mass was not showing vascularity.



2a 2b

Figure 2a: image of abdominal ultrasound showing heterogeneous predominately hypoechoic solid-cystic mass in abdominal cavity with multiple linear and foci of calcification within.Fig2b:On color Doppler the mass was not showing vascularity.

CT abdomen revealed a huge retroperitoneal mass (Fig.3a, 3b,3c and3d) with multiple areas of linear and foci of calcifications. The mass composed of good amount of cystic and fat attenuated areas. There was no retroperitoneal lymphadenopathy, ascites, or intestinal mechanical obstruction. The ovaries were well developed and unremarkable .

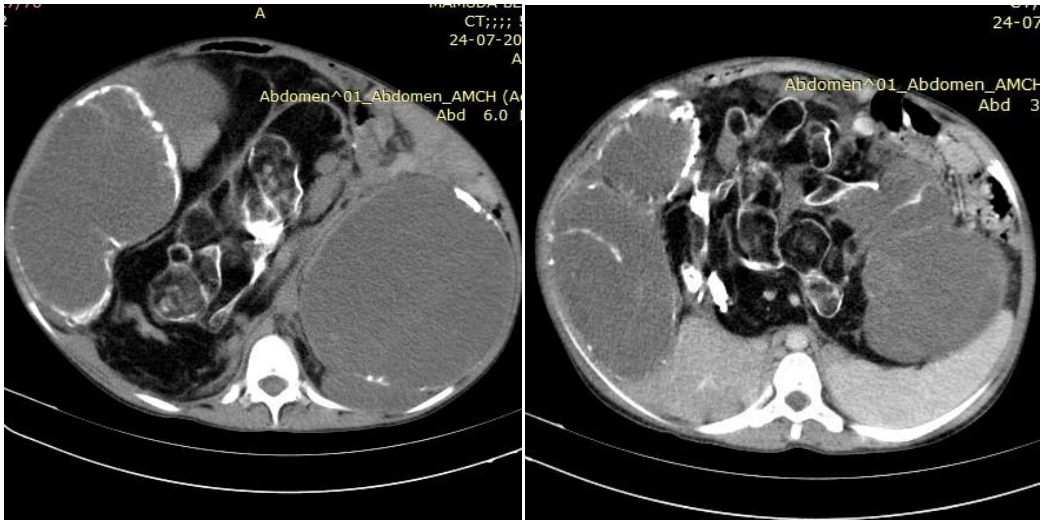


Fig:3a Fig:3b

Fig:3a. Axial noncontrast enhanced computed tomography image of abdomen and pelvis showing a large solid-cystic mass with fatty attenuated and multiple calcific areas within.**Fig:3b:**The mass does not show any significant enhancement on post contrast study.

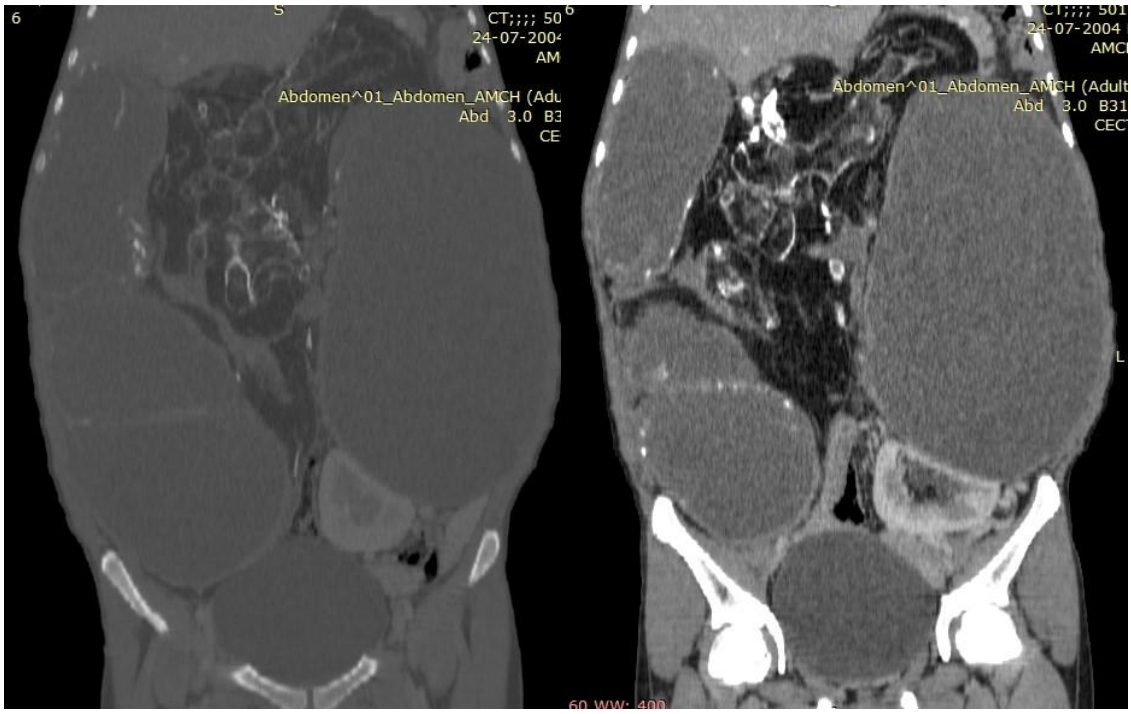
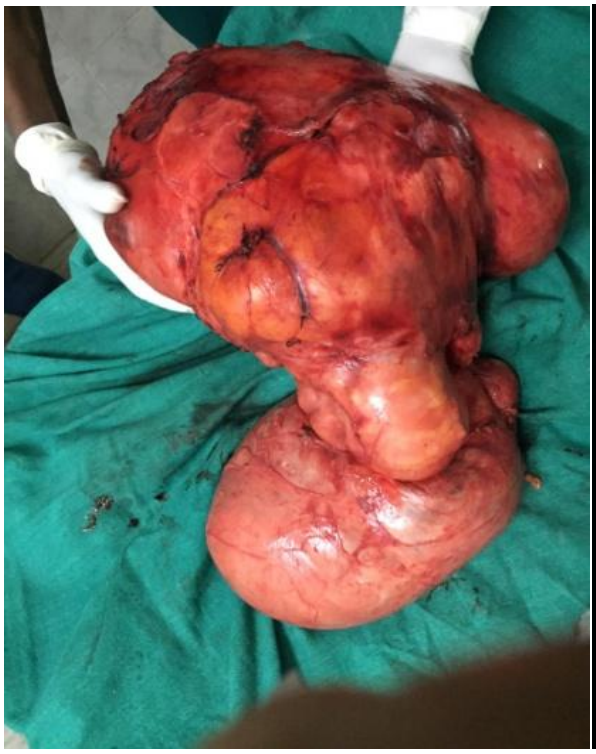


Fig:3c Fig: 3d

Figure 3c. Coronal bone window computed tomography image of abdomen and pelvis showing a large solid-cystic mass in abdominopelvic cavity extending from epigastric region to hypogastrium filling both the flanks with multiple areas of calcifications within.**Fig3d:** Coronal contrast enhanced image does not show any significant enhancement of the mass. Left kidney is noted to be displaced inferiorly and pancreas superiorly (not shown in image) by the mass.

Provisional diagnosis of retroperitoneal teratoma was made and an open exploration was performed with midline incision .There was a large horse-shoe shaped solid cystic mass in retroperitoneal compartment. The entire mass was excised. Post operatively the mass(Fig:4a) measuring 30.1 x 25.8cm is obtained and found to be filled with bony elements, cystic areas and yellowish creamy material. The mass weighted 19.3kgs. Histopathologic examination (Fig:4b) revealed a teratoma with no malignant cell. Stratified squamous epithelium with sebaceous and sweat glands, hair follicles, cartilaginous tissue, bony spicules, bone marrow elements all were demonstrated. The post operative course was uneventful and she had no significant complaint till 3 months of follow up.



Fig;4a

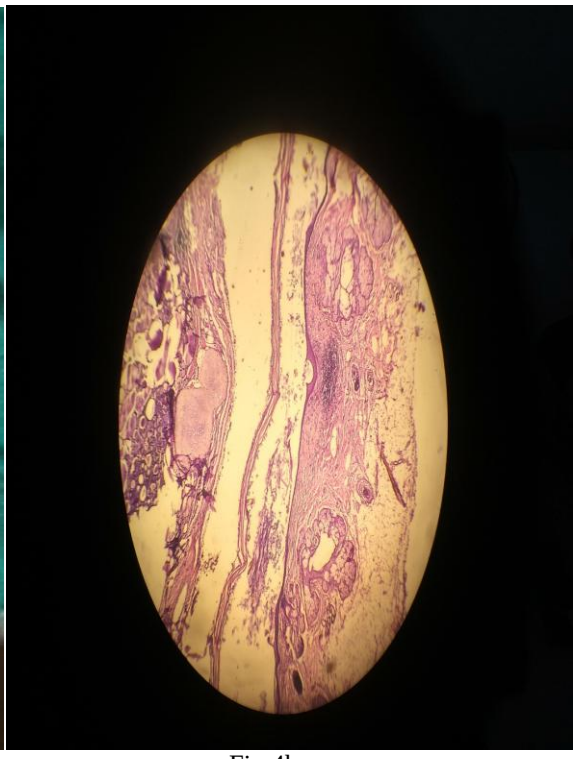


Fig:4b

Fig4a; Post operative gross specimen of the tumor. **Fig:4b** Photmicrograph of dermoid cyst showing squamous epithelium, cartilage and skin adnexal structures without any malignant cell; findings are consistent with benign teratoma

II. Discussion

RT were first described by Morgagni[(1)] in 1769. In 1937, urography was first used to diagnose RT[(2)(3)] .

Teratoma is a germ cell tumour that originates from pluripotent germ cells that have been interrupted in their normal migration to the genital ridges. Less than 10% of teratomas are found in the retroperitoneum.Teratoma accounts for as many as 11% of primary retroperitoneal tumors and is the third most common tumor in the retroperitoneum in children, after neuroblastoma and Wilms tumor[(4)]. There are various theories regarding the origin of primary RT. They were once thought to arise from the earliest segmentation of the fertilized ovum. This theory is also known as the Blastomere Theory[(5)]. The most widely accepted theory is that they are vestiges of the wolffian and müllerian ducts or that they arise from pronephric or mesonephric tubules as it correlates with their midline and paramedian location[(4)].

Extragonadal teratomas can be located in decreasing order of frequency, in the anterior mediastinum, retroperitoneum, presacrum, coccygeal region, intracranium, neck, and abdomen[(6)] .

The location of the teratoma would coincide with the symptoms of compression of the adjacent structures such as vomiting, constipation, lumbar back pain, abdominal distention, and edema. Systemic symptoms may also be present such as fever, chills, night sweats, and weight loss [(4),(7),(8)].RT may present with chemical peritonitis from rupture of the cyst[(9)].

Retroperitoneal cystic teratoma can also get infected and a case presenting as a subhepatic abscess has been reported in a postpartum woman[(10)].

Unusual presentations include ocular myasthenia gravis and hypertension which both resolved after excision of the cystic mass [(11),(12)].

The size of RT in adults is variable and can reach as large as 41 cm. RT have been reported to weigh as much as 31.6 kg and contain 3 L of cystic fluid[(4),(13),(12)].

Teratoma is more common in females, with a bimodal age distribution (<6 months and early adulthood). Teratoma can be benign or malignant, and benign teratoma can be either mature or immature. Mature teratoma (dermoid cyst) contains well differentiated tissues from at least two germ cell layers. Ectodermal layers are seen in all, mesodermal layers in 90% of lesions, and endodermal layers in the majority of lesions. Mature teratomas are predominantly cystic. Calcification (tooth like or well defined) and fat can be seen in 56% and 93% of cases, respectively [(14)]. A fat-fluid (sebum) level and chemical shift between fat and fluid are pathognomonic. A villi form solid component known as a Rokitansky protuberance is seen in 81% of cases.

Malignancy has been reported in 2%–3% of mature teratomas, more commonly in children (26%) than adults (10%), and is associated with wall thickening, irregular margins, and infiltration of adjacent organs.

Compared with mature teratoma, immature teratoma is less common (<1%), contains more than 10% undifferentiated tissue, and is seen in a younger age group (<20 years). The most common location of immature teratoma is near the upper pole of the left kidney.(14) Immature teratoma is predominantly solid, with scattered areas of fat and calcification (coarse and ill defined), but cystic components are found occasionally. Malignant teratoma can have germ cell or non–germ cell malignant tissue. Malignant transformation is less common in the retroperitoneum[(4)]. Malignant tumors are irregular, with invasion of adjacent structures and vascular invasion. A poor prognosis is associated with lesions with germ cells or lesions with rhabdomyosarcoma or neural differentiation. An elevated a-fetoprotein level is found in 50% of malignant teratoma. Complete surgical resection is required for definitive treatment.

III. Conclusion

Retroperitoneal teratoma is a uncommon entity which has distinctive imaging findings. Majority of the lesions are benign. Plain radiographs findings of calcifications, bone, or teeth are pathognomonic. Ultrasonography and CT are useful tools for further evaluation of the extent of the lesion. The lesions are mostly amenable for curative surgical excision. Prognosis is generally good and recurrence can be monitored with tumor marker like AFP

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Informed consent ;Written informed consent was obtained from the parents of the patient for their anonymized information to be published in this article

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