A Rare Case Of Retroperitoneal Mature Cystic Teratoma In A Young Male Patient

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Abstract: Retroperitoneal mature cystic teratomas are exceedingly uncommon in young male. They are more common in infants and in young females. We report an unusual case of 27 year old male presented with abdominal distension, abdominal fullness and vomiting. Patient underwent ultrasonography and contrast enhanced computed tomography (CT) scan of abdomen which were suggestive of large mature cystic lesion. Patient was planned for surgery and laprotomy was done. A huge retroperitoneal mass was observed and was totally excised. On histo-pathological examination (HPE) stratified squamous epithelium with keratin flakes, underlying fibromuscular tissue reveal hair shaft & sebaceous glands and mature cystic teratoma was diagnosed. Retroperitoneal mature cystic teratoma in an older male is extremely rare. Primary gonadal teratoma with retroperitoneal metastasis should be excluded first. Evaluation of age and location of tumor are critical for its prognosis. [Kagathara V Natl J Integr Res Med, 2022; 13(3): 56-61, Published on Dated: 10/05/2022]

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Introduction: Mature cystic teratomas, also known as dermoid cysts, are neoplasms composed of tissues from at least two of the three germ layers. They are most commonly located in the gonads and infrequently in extragonadal sites such as the retroperitoneum, intracranial, cervical, mediastinal, and sacrococcygeal. Retroperitoneal teratomas account for 4 % of all primary teratomas.

Metastasis of gonadal malignancies to the retroperitoneum comprises most of the retroperitoneal neoplasms; however, primary retroperitoneal neoplasms comprise only 0.1%–0.3% of all the tumors and 1%–11% of those are teratomas.

In children, the most common retroperitoneal tumors are neuroblastoma and Wilms' tumor. Primary retroperitoneal teratomas (RT) are characterized by absence of attachment to other organs such as kidneys, adrenal glands, and pancreas. Cases are reported usually early in infancy or in young females.

The most common site is the sacrococcygeal area in infants and the left suprarenal region in adults. The aim of this article is to present a rare case of a large retroperitoneal mature cystic teratoma located on the right infrarenal region in a 27-year-old male.

Case Study: A 27 year-old man presented with a mass in abdomen. He first palpated the lesion five years ago. Recently he noticed an increase in the size of the mass and referred to the clinic.

He denied any associated symptoms including fever, loss of appetite, weight loss or pain.

He had no underlying diseases. He had not undergone any surgery.

He denied taking any medications, smoking or alcohol consumption and his family history was unremarkable.

He was normotensive with a blood pressure of 118/84 mmHg, his pulse was 84 per minute, his respiratory rate was 16 per minute and his oral temperature rate was 35.6 °C. His hemoglobin was 13.8 mg/dl and other investigations under normal limits.

His physical examination was unremarkable except for a firm, non-mobile fullness palpated of the abdomen without any tenderness or abdominal guarding.

The extent of the mass could not be established. As it covers whole abdomen. The patient underwent for x-ray abdomen and ultrasonography of abdomen.

Figure 1: X-Ray Abdomen

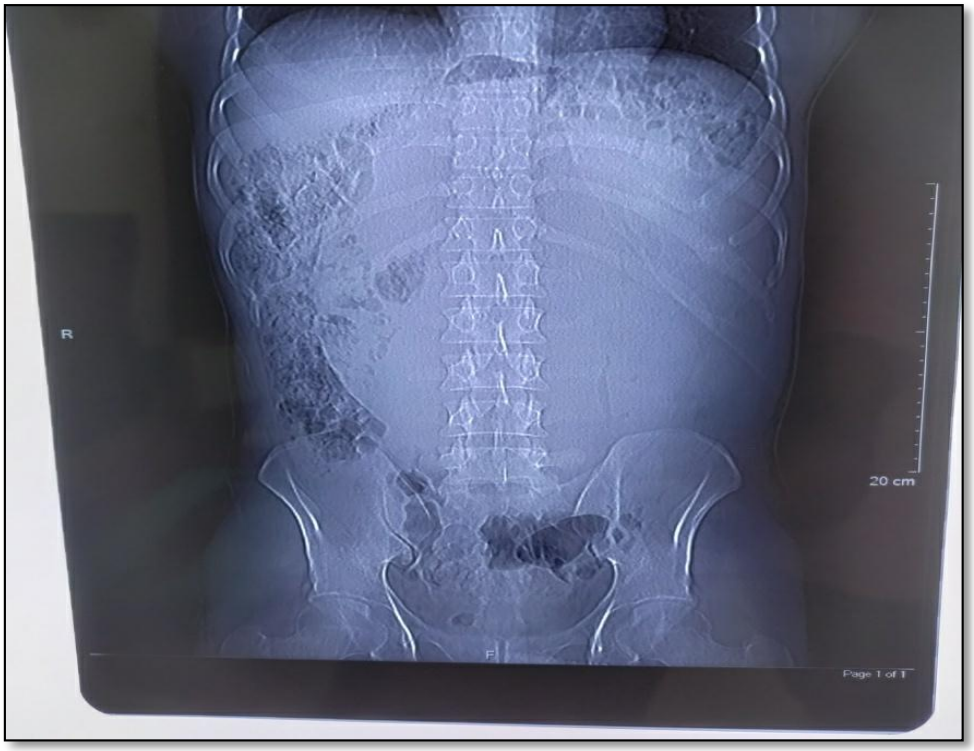

Figure (1) X-ray abdomen showed large soft tissue density mass covering the left half of the abdomen, crossing the mid line and extending to the right lumbar region. The mass displaces the bowel loops superiorly in left hypochondriac region, right laterally and inferiorly in the pelvis.

Figure 2: An ultrasonography

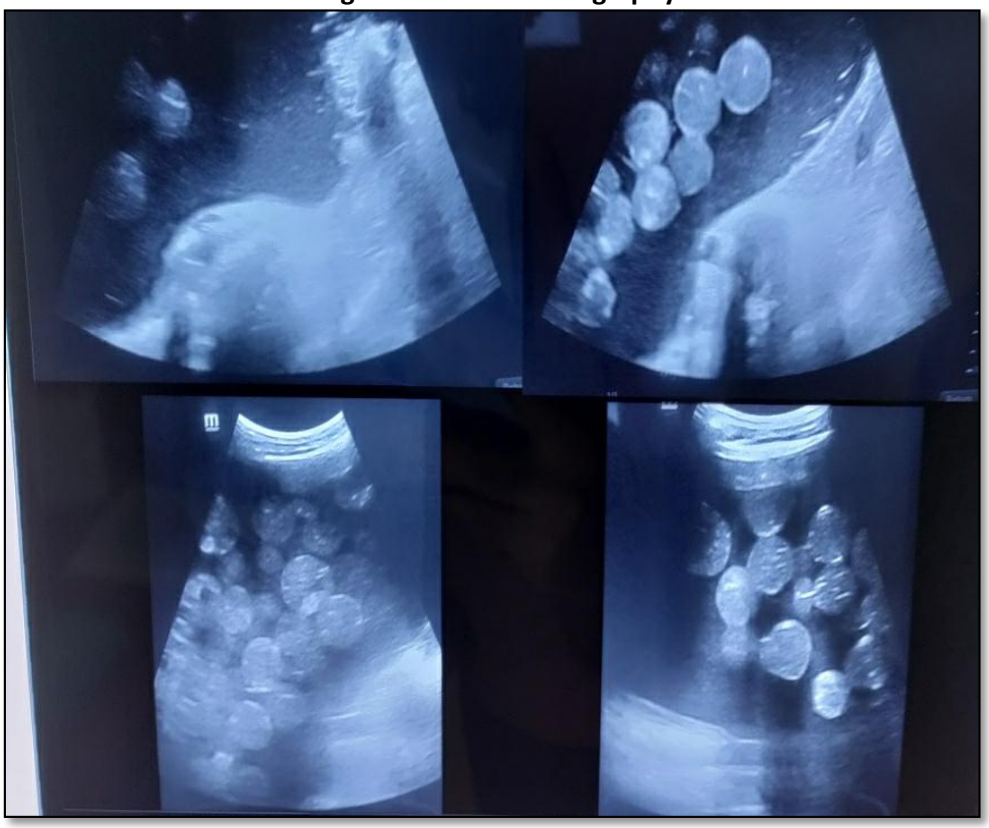

Figure (2) An ultrasonography revealed large well defined intraperitoneal cystic lesion measuring approximately 30 x 22 cm with internal echoes and multiple well defined spheroid isoechoic lesions and multiple linear hyperechoic foci (hair/debris) within it with few hyperechoic foci casting posterior acoustic shadow noted within the lesion, largest measures 28 mm, the lesion extends from the epigastric region to pelvis.

Figure 3: Coronal Section

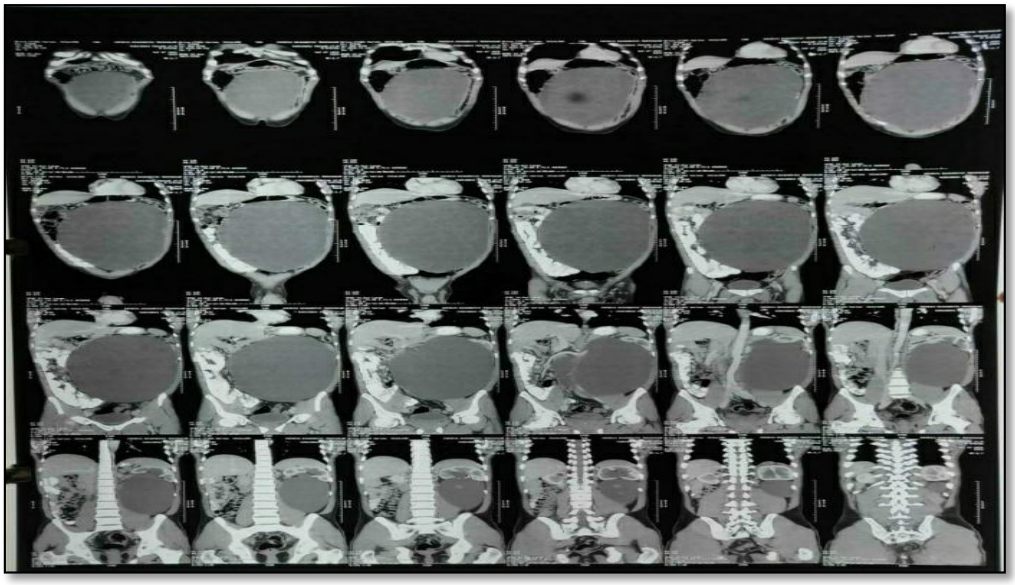

Figure 4: Axial Section

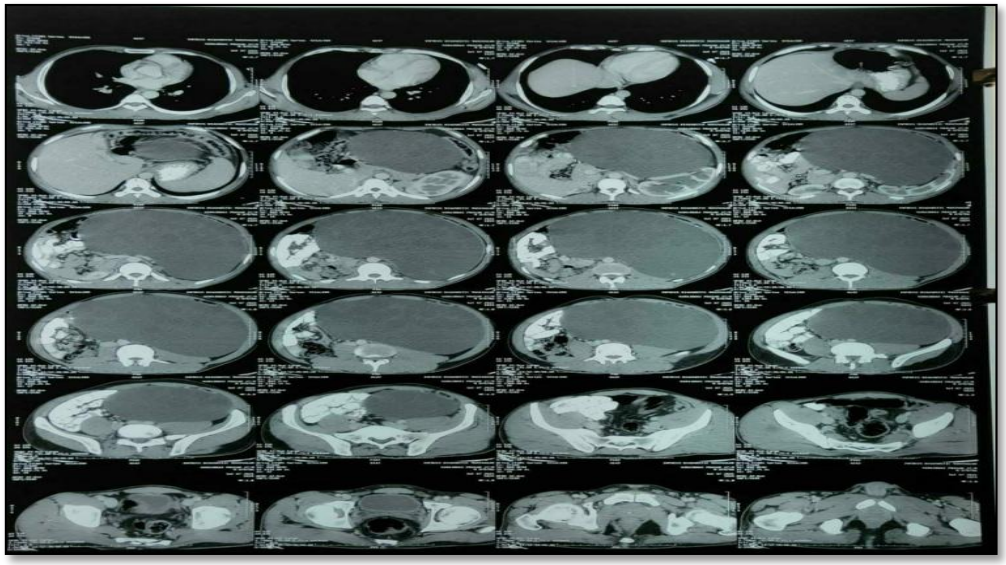

Figure (5) Coronal Section

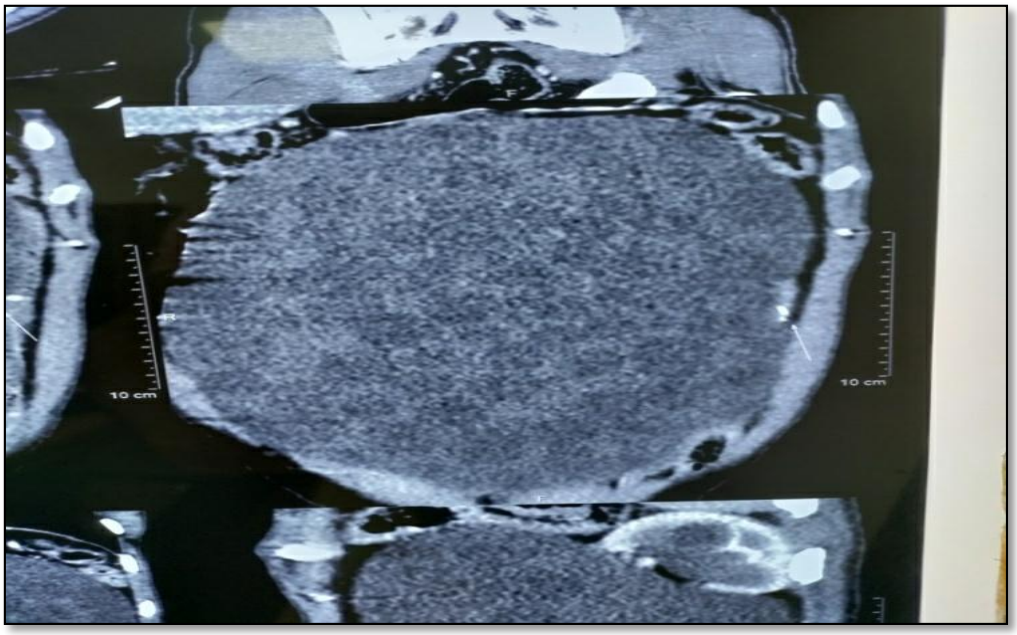

Figure (3), (4) & (5): A Computed Tomography Scan (CT) was performed for the patient that revealed approximately 31 x 23.6 x 7.8 cm (AP x TR x CC) sized intraperitoneal thin walled cystic lesion with calcification within it (arrow in figure 5) involving abdominal cavity more on left side extending from epigastric region to left iliac fossa region and reaching upto the anterior and left lateral abdominal wall.

It causes mass effect in the form of postero-superior displacement & compression over the left kidney with secondary malrotation and resultant mild hydronephrosis. Left ureter coursing along the medial wall of the cyst and is displaced towards the right side.

The lesion causes significant compression over pancreatic parenchyma and proximal part of splenic vein.

It entirely displaces small bowel loops on right side along with displaced & right sided duodenojejunal flexure.

It also effaces left psoas muscle and infra renal abdominal aorta with marginal displacement of aorta towards right side.

Figure 6: The Patient Underwent Laparotomy. A Huge Retroperitoneal Mass Was Observed. Mass Was Totally Excised And Sent For Histo-Pathologic Evaluations

In gross examination reveal large, well defined mass having smooth surface and prominent vascular markings, on cut section thick walled cystic mass lesion having multiple well defined grayish-white ball like structures with raised protuberance and soft consistency was noted.

Histo-pathological assessment reported stratified squamous epithelium with keratin flakes, underlying fibromuscular tissue reveal hair shaft and sebaceous glands, mature cystic teratoma was pathologically diagnosed.

Discussion: The differential diagnosis of a primary retroperitoneal mass could be divided between solid and cystic. Cystic retroperitoneal masses can be either neoplastic or non-neoplastic. Mature teratomas, mucinous cystadenomas, and cystic mesotheliomas encompass the neoplastic cystic masses, while lymphangiomas, müllerian cysts, epidermoid cysts, pancreatic pseudocysts, lymphoceles, urinomas, and hematomas comprise the non-neoplastic cystic masses.

However, primary retroperitoneal masses with calcification include malignant fibrous histiocytoma, paraganglioma, and ganglioneuroma. Immature teratomas usually present as solid masses containing undifferentiated tissues but may also include fat and calcification.

Extragonadal teratomas can be located in decreasing order of frequency, in the anterior mediastinum, retroperitoneum, pre sacrum, coccygeal region, intracranium, neck, and abdomen.

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.

Retroperitoneal teratoma may present with chemical peritonitis from rupture of the cyst.

Teratomas are part of the family of non-seminomatous germ cell tumors. Totipotent cells differentiate into ectoderm, mesoderm, and endoderm components of the teratoma.

They can be classified according to the number of layers present (mono-, bi-, or tridermal), epithelial lining (epidermoid, dermoid, or teratoid), degree of maturity of tissues (mature or immature), presence of malignant tissues, and content of tumor (cystic, solid components, or mixed).

Risk of malignancy of these neoplasms ranges from 6.8% to 36.3% and increases with age, male sex, and presence of immature tissues and solid components. Serum markers such as alpha-fetoprotein, carcinoembryonic antigen(CEA), lactate dehydrogenase, beta-human chorionic gonadotropin, CA 19-9, and CA 125 were detected from retroperitoneal teratoma, but these were more associated with malignancies arising from the teratoma.

Elevated alpha-fetoprotein levels were found in 50% of immature teratomas and 6% in mature teratomas. These serum markers are not specific to teratomas as they can be found in other diseases. These markers are evaluated preoperatively to exclude other germ cell tumors and are followed up post-operatively to detect recurrence of the retroperitoneal teratoma.

Plain abdominal radiographs are part of the initial evaluation which may show calcifications but the findings are nonspecific. Ultrasound may show echogenic spots with acoustic shadows or dermoid plugs (Rokitansky body) but has limited sensitivity which makes CT scan the more appropriate imaging.

CT scan findings indicative of primary retroperitoneal teratoma include well-circumscribed fluid component, hypoattenuating fat and calcifications which were found in our patient. Complete excision of the mass was the primary method of management of these neoplasms with involved significant morbidity and mortality.

Laparoscopic surgery is the preferred approach, especially when the cyst is well circumscribed on imaging. Surgical resection through laparotomy was done in our case due to the massive cyst size and the 3%–6% rate of malignant transformation in older males.

Adjuvant chemotherapy and radiotherapy are necessary when retroperitoneal teratoma harbors malignancy, especially germ cell carcinomas. Malignant teratomas behave like advanced testicular germ cell tumors, thus platinum-based chemotherapy is used.

Teratomas with malignancies are highly resistant to chemotherapy and radiotherapy with an overall response rate of 61%. Since there are fewer reports of retroperitoneal teratoma, there is little data for assessment of prognosis which depends on the type of tissues that the teratoma contains. With complete excision of the benign cyst, prognosis is excellent. However, if left untreated, mortality and morbidity are much higher due to obstruction of nearby viscera or

metastasis from malignancies arising from the retroperitoneal teratoma. Continued long-term postoperative follow up is recommended to detect early recurrence.

Conclusion: Retroperitoneal mature cystic teratoma in an older male is extremely rare. Primary gonadal teratoma with retroperitoneal metastasis should be excluded first. Evaluation of age and location of tumor are critical for its prognosis. Complete excision of tumor is necessary to evaluate whether there are immature and solid elements which need long-term follow up due to the increased risk of malignancy.

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