Case report

Surgical excision of a Giant Mature Retroperitoneal Teratoma Encasing the Superior Mesenteric Artery: A Case Report and review of the literature

Abstract

Introduction:
Teratomas are uncommon tumors derived from all three embryonic germ layers. Primary retroperitoneal teratomas (PRT) constitute 1-11% of retroperitoneal tumors and are most often seen in neonates and young adults. This case report presents a rare instance of a massive mature retroperitoneal teratoma in a 16-year-old female, which uniquely encased the superior mesenteric artery (SMA).

Case Presentation:
A 16-year-old girl presented with back pain and an abdominal lump. Clinical examination and computed tomography (CT) revealed a 16.4x9.3x10.9 cm retroperitoneal mass containing fat, cystic elements, and calcifications. The mass displaced surrounding organs and encased the proximal SMA but did not invade other structures such as the inferior vena cava (IVC) or renal arteries. Tumor excision was performed via laparotomy. During surgery, the tumor was meticulously dissected from the SMA, IVC, and surrounding vessels. The surgery was uneventful, with minimal blood loss and no complications. The patient had an uneventful postoperative recovery and was discharged on the fifth postoperative day. Histopathological analysis confirmed the diagnosis of a mature cystic teratoma, with no evidence of malignancy.

Discussion:
PRT is a rare extragonadal tumor often found in the midline structures Although mostly benign, malignant transformation is possible in 1-2% of cases, more so in adults. Radiological imaging, particularly CT and MRI, plays a crucial role in diagnosing and planning the surgical approach. Surgical excision is the gold standard for treatment. The uniqueness of this case lies in the tumor's large size and its encasement of the SMA, which has not been previously reported.

Conclusion:
Primary retroperitoneal teratomas are rare, and this case represents a unique presentation of a giant mature teratoma encasing the SMA. Preoperative imaging and meticulous surgical dissection are essential for successful management. This case highlights the importance of careful surgical planning in complex cases involving vital vascular structures.

Introduction:

Teratomas are uncommon neoplasms comprised of mixed dermal elements that develop from the three embryonic germ layers[1]. Primary retroperitoneal teratomas(PRT) make up 1 to 11% of retroperitoneal tumors and are most frequently observed in neonates and young adults[1]. RPTs are commonly located near the upper pole of the kidney, with higher preponderance on the left side[2]. In this article, we report an unusual case of a massive mature retroperitoneal teratoma in a 16-year-old female encasing the superior mesenteric artery with emphasis on the evaluation and management strategies.

Case report:

A 16 years aged girl presented with complaints of back pain since 6 months which was intermittent, non radiating and not related with postural changes. She felt an abdominal lump since 2 months. She didn't have weight loss and her appetite was normal. Clinical examination revealed an ill defined firm, non tendermass palpable in right upper quadrant extending to epigastric and right lumbar region which doesn't move with respiration and dull on percussion. She was evaluated with CECT abdomen (Figure 1) which revealed 16.4x9.3x10.9cm large well defined lobulated retroperitoneal mass with predominantly fat containing with cystic components and, tooth like structures. The mass is displacing pancreas and D2/D3 anteriorly; encasing proximal 5cm of superior mesenteric artery(SMA). It also was closely associated withaorta and bilateral proximal renal arteries (arc of 180*-270*); exerting mass effect on infrahepaticinferior vena cava (IVC); but there is no IVC thrombus. Boththe ovaries were normal. There was no ascites/ lymph node enlargement. LDH was 168 U/L (normal lab reference value : 125-220U/L for age >12 years). CA-125 was 8.8U/ml (normal lab reference value :<35U/ml).

With diagnosis of suspected retroperitoneal tumor- possibly teratoma, laparotomy and excision of the tumor was planned. Intraoperatively, there was a large lobulated retroperitoneal tumor wedged between aorta,SMAand celiac trunk(Figure 2 &3) extending on both sides of aorta with major component on right side extending from inferior surface of liver till the level of caecum. Structures like pancreas, duodenum, portal vein were pushed anteriorly by tumor. Both kidney with renal vessels were displaced inferiorly. Tumor was compressing infrahepatic IVC. Hepatic flexure mobilized down. Duodenal kocherization done; IVC and renal vessels were dissected off tumor. Inferior mesentic vein also dissected off tumor. Then SMA which was passing between the lobes of tumor was carefully dissected from tumor and looped.The infra SMA twig of tumor was divided Tumor dissected off retroperitoneal attachements and excised enbloc(Figure 4). Intraoperative period was uneventful with estimated blood loss of around 120ml and operative duration of around 4.30 hours. She was started on oral sips on 1st postoperative day (POD) then gradually progressed to solid diet on POD-5 and discharged on POD-5.

Histopathological examination of tumor revealed encapsulated nodular mass of size 18x14.5x8cm on macroscopy. On cut surface, predominantly yellow homogenous with cyst measuring 5.5x4.5x2.5cm. Cyst is filled with hair shaft, greyish white pultaceous material and focal areas which are hard to cut. On microscopic examination, cyst cavity showed acellular keratin flakes with skin appendages, benign adnexal glands supported by fibromuscular stromal tissue. Fragments of cyst lined with squamous epithelium, apocrine ducts and glial tissue along with odontogenic epithelial clusters (Figure 5). No

evidence of immature neuroepithelial components or atypical cells. All these said features are consistent with retoperitoneal mature cystic teratoma.

Discussion:

Teratomas are rare tumors made up of diverse tissue types originating from all three germ cell layers. Theyusually occur within the ovaries of young women and testes of young men. Extragonadal teratomas are commonly found in midline structures such as the anterior mediastinum, retroperitoneum, sacrococcygeal region, and pineal gland in the descending order of frequency[1]. Primary retroperitoneal teratomas account for 1–11% of retroperitoneal neoplasms and are most commonly found in neonates and young adults. The age of onset shows two distinct peaks: one occurring around 6 months after birth and another in early adulthood. Incidence of PRT is more common in females than males.

Several hypotheses about pluripotent cells have been proposed to relate the origin of teratoma, with the germ cell origin being the most widely accepted. The embryonic (Blastomeric) cell theory proposes that teratomas originate from pluripotent embryonic cells that have escaped the guidance of the primary organizer during early embryonic development[1]. Extra-embryonic cell origin hypothesis states that teratomas arise from ectopic visceral yolk sac which contain primordial germ cells in early embryonic development[3].

Teratomas are generally classified into three main categories: mature (cystic or solid, benign), immature (malignant), and monodermal (highly specialized). Each type can appear alone or in combination with others. A mature teratoma is characterized by well-differentiated, adult-type tissue, whereas an immature teratoma contains elements with incomplete somatic differentiation, resembling embryonic or fetal tissues[5].

Retroperitoneal teratomas often occur near midline, positioned commonly in the left upper quadrant. Benign teratomas are typically asymptomatic and are often found incidentally[3]. With increase in size of the tumor, a range of symptoms related to visceral obstruction may emerge which include abdominal or back pain, urinary issues such as hydronephrosis, gastrointestinal problems like nausea, vomiting and constipation as well as edema in the lower extremities or scrotum due to lymphatic flow obstruction. On abdominal examination, a midline or para-median abdominal mass would be palpable with restricted mobility. Retroperitoneal malignant teratomas can cause nausea, weight loss, fever, dyspnea, and chest discomfort.

There is a risk of malignant transformation in 1% of PRT. This risk is higher in adults than in children (26% vs. 10)[6]. Malignant teratomas (0.2%-2%) have the potential to metastasize to the lung and lymph nodes[7]. The main pathological indicators of malignancy include the presence of germ cell components and immature or undifferentiated tissue[3]. In contrast to children, adults with retroperitoneal teratomas might endure a chronic malignant process. Squamous cell carcinoma is the most common type of malignant transformation observed in teratoma. Malignant transformation of retroperitoneal teratomas occurs less frequently than in mediastinal teratomas.CECT abdomen is the ideal tool for diagnosis of retroperitoneal teratoma[3].Mature teratomas can be identified by the presence of fluid attenuation or signal intensity, fat-fluid levels, and calcifications[8].MRI provides superior image resolution for soft tissue compared to CT. It is useful for assessing whether the tumor has encased or invaded blood vessels, as well as for identifying various structures within the mass[3].

Surgical resection is the primary treatment approachforprimary retroperitoneal teratoma[3]. It is essential for both diagnosing and treating the condition, as the final diagnosis relies on the histological analysis of the specimens.Trans-abdominal approach allows for a broader view and

better control of feeding vessels. Usually, median and transverse abdominal incisions are used. Whereas thoracoabdominal incision is reserved for very large tumors located in the upper abdomen, posterior to liver. If feasible, laparoscopic resection may also be considered for small benign looking teratoma based on CT findings.

This case is unique in terms of its location and size. PRT are unilateral and excised without organ resection. Encasement of SMA for initial 5 cms and extending to left side and enlarging on left side has never been reported so far. Although, it is closely associated with multiple major vessels of abdomen, careful dissection enables us to preserve vital structures as described in this case report.

Conclusion:

Primary retroperitoneal teratoma is a rare clinical condition. Preoperative imaging is essential for accurate diagnosis and surgical planning. Dumb bell shaped giant mature cystic teratoma with wedge of tissue between celiac and superior mesenteric artery was excised in this case, which makes this case, first of its kinds reported.

Abbreviations:

PRT- Primary Retroperitoneal Teratoma

SMA- Superior Mesentric Artery

CECT – contrast enhanced computed tomography

IVC- Inferior Vena Cava

POD- PostOperative Day

COMPETING INTERESTS DISCLAIMER:

Authors have declared that they have no known competing financial interests OR non-financial interests OR personal relationships that could have appeared to influence the work reported in this paper.

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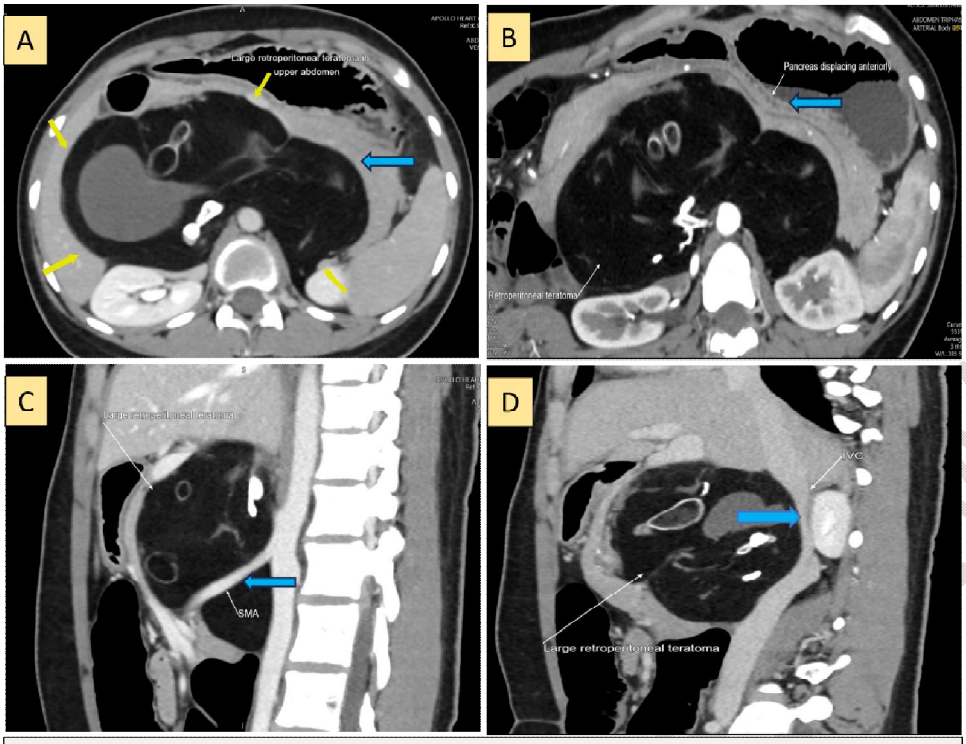

Figure 1: CECT abdomen images. A & B- axial views showing retroperitoneal tumor 16.4x9.3x10.9cm displacing pancreas anteriorly
C- SMA coursing through tumor
D- Tumor compressing infrahepatic IVC

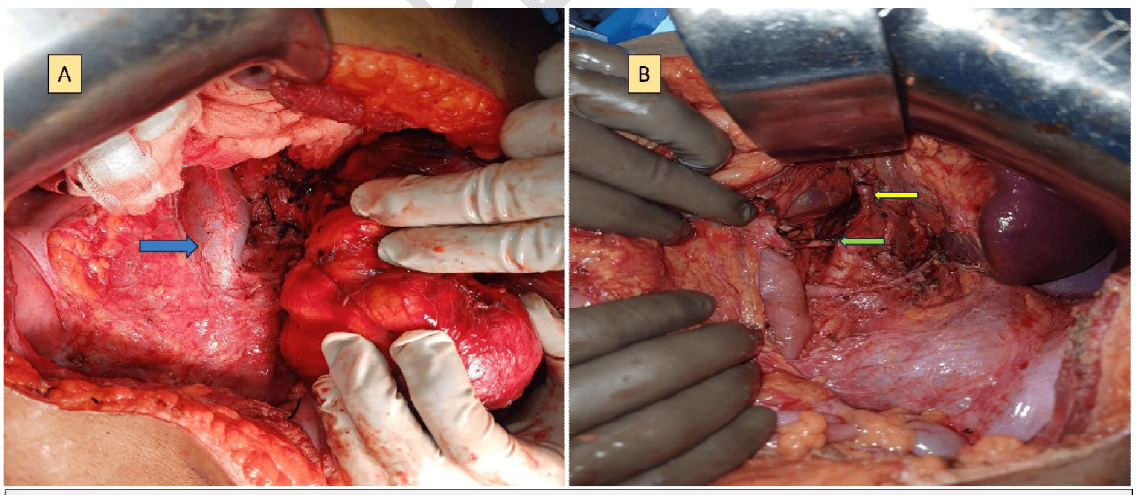

Figure 2: A - tumor been dissected from retroperitoneum and lifted to left, exposing IVC behind (blue arrow).
B - origin of celiac trunk (yellow arrow) and SMA (green arrow) from abdominal aorta, SMA coursing between the lobes of tumor.

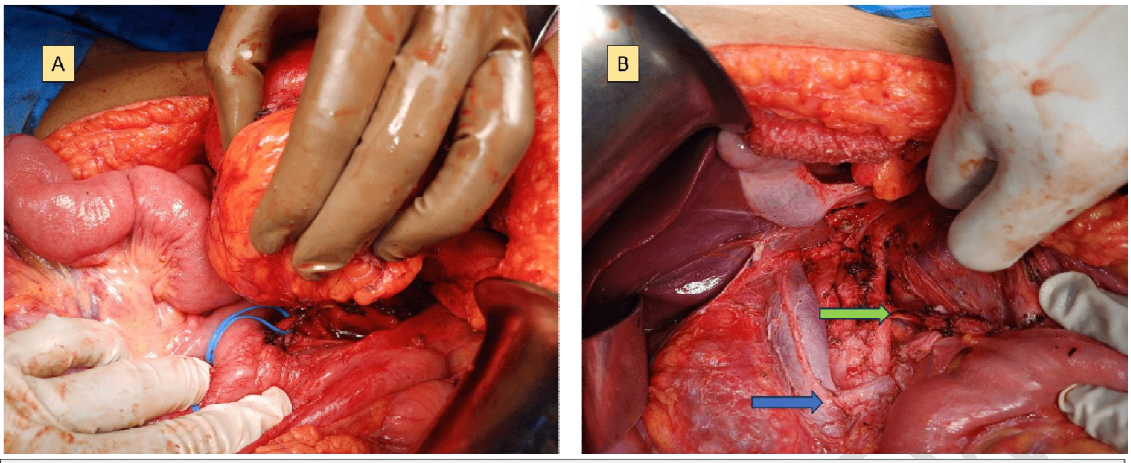

Figure 3: A – SMA coursing through tumor and looped with vessel loop.
B – IVC with left renal vein draining into it (blue arrow) and SMA origin from abdominal aorta (green arrow).

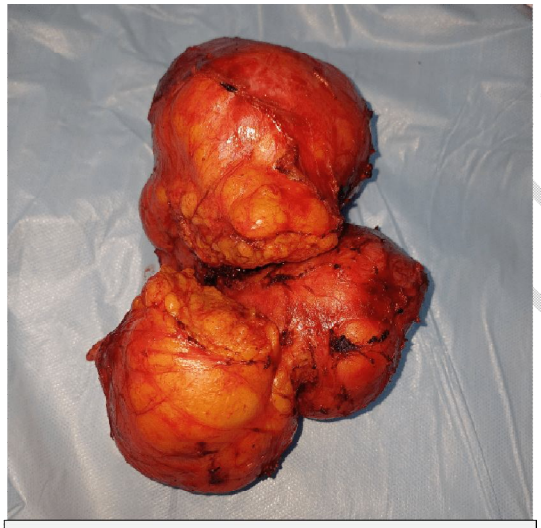

Figure 4: specimen image

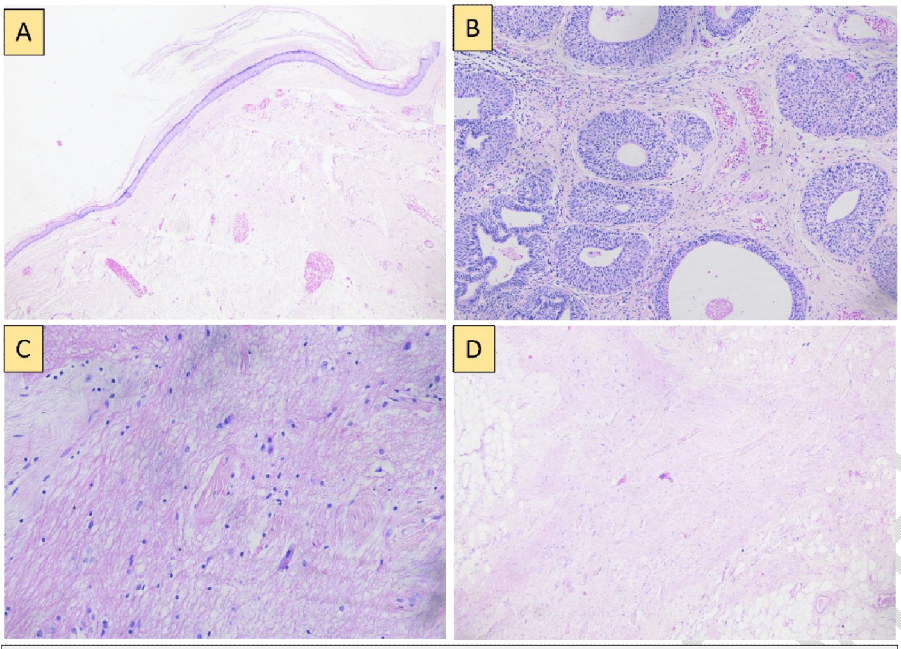

Figure 5: HPE images. A- keratinized skin layer, B- glandular architecture, C- Glial tissues, D- Glial tissues with adpiocytes