

CASE REPORT

Giant Retroperitoneal Teratoma in an Adult Presenting with Abdominal Mass: A Case Report and Literature Review

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ABSTRACT

A teratoma is a true neoplasm that comprises tissues that are either distinct from the initial site of origin or histologically heterogeneous and represent several embryonic germ layers. Here we present a case report of a 24-years-old female patient who presented with chief complaints of swelling in the right upper abdomen for 4 months. Per abdomen, examination revealed a lump of around 20 cm in size in the right hypochondriac region extending to the Lumbar and umbilical region. Contrast-enhanced computed tomography abdomen revealed a large, well-encapsulated, heterogeneous mass lesion of approximate size 30.3x18.4x2 in the right retroperitoneal/suprarenal region. The lesions show fat components with few incomplete septations and few specks of calcification. No evidence of solid component or hemorrhage within the lesion. The patient was taken up for exploratory laparotomy and a tumor was found in the retroperitoneum and was excised with due care. Histopathological examination features were suggestive of Dermoid Cyst. The post-operative stay was uneventful.

Keywords: Abdominal mass, Mass lesion, Retroperitoneal Teratoma, Tumour

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INTRODUCTION

Teratomas are rare neoplasms made up of mixed dermal components generated from the three germ cell layers. They might be either mature benign without malignant cells or immature with malignant cells.¹ In adults, retroperitoneal teratomas are uncommon, making for just 4% of all primary teratomas.² Although extragonadal locations, including cerebral, cervical, mediastinal, retroperitoneal, and sacrococcygeal, have also been recorded, the testes and ovaries are where these tumors are most frequently detected.³ We report a case of giant retroperitoneal teratoma in a 24-years-old lady who presented with an abdominal mass.

Case Report:

A 24-years-old lady presented to our outpatient department with complaints of a painless abdominal mass for the past 4 months, which was progressive and associated with mild pain over the mass with no other urinary or abdominal complaints. She had neither fever nor loss of appetite and weight. Clinical examination revealed a huge lump of size 20x18 cm involving the right hypochondrium, right lumbar, right iliac fossa, and

umbilical region and the mass was crossing the midline. The mass was firm in consistency, immobile and dull on percussion. Her Routine blood investigations were within normal limits. The patient underwent abdominal Computerized Tomography (CT), which revealed a large, well-encapsulated, heterogeneous mass lesion of approximate size 30.3x18.4x25 (AP x Transverse x CC) in the right retroperitoneal/suprarenal region. The lesion was also extending across the midline to the left side. The lesions show fat component (avg, CT attenuation value -2711U) with few incomplete septations and few specks of calcification. No evidence of solid component or hemorrhage within the lesion. No invasion of fat planes with adjacent organs and vessels is seen. Superiorly, the lesion was abutting and compressing the gallbladder and right lobe of the liver without frank infiltration. Inferiorly, the Right kidney was pushed anterior to the aorta across the midline inferiorly at the level of the umbilicus. Medially, it was abutting and displacing the duodenum, inferior vena cava and pancreas. Anteriorly, fat planes with ascending colon were well maintained. The CT scan featured was suggestive of retroperitoneal teratoma. She underwent laparotomy with excision of the retroperitoneal

dermoid cyst through a right subcostal incision with a left subcostal extension which revealed the following findings: a firm to cystic retroperitoneal mass of size 34x18x30 cm occupying most of the right hemiabdomen.

Superiorly adhered to the inferior surface of the liver and falciform ligament, it medially displaced IVC from its position and adhered to IVC from the inferior portion of hepatic veins to renal veins. An outpouching of size 6x6 cm was present in between IVC and the aorta. The right kidney was displaced inferiorly; laterally, it has adhered to the parietal wall. The weight of the dermoid was 7 kg and on gross examination, the cut surface was grey-brown to grey-white filled with brown-coloured fluid, pultaceous material, hair, and a few fatty areas suggestive of dermoid tumor of retroperitoneum. The post-operative course was uneventful.

DISCUSSION:

Teratomas develop from pluripotent embryonal germ cells and are non-seminomatous germ cell tumors. During embryogenesis, germ cells grow and often descend along a midline path towards the pelvis, where they give rise to ovarian cells or the scrotum, which gives rise to testicular cells. Germ cells may be deposited in extragonadal locations and are susceptible to neoplastic conversion if they fail to move via the urogenital ridge. As a result, in decreasing order of frequency, teratomas can develop in the ovaries, testicles, anterior mediastinum, retroperitoneum, and skull.⁴ Primary retroperitoneal tumor incidence ranges from 1% to 11%.⁵ According to several studies, PRT is most common in newborns and young people, making up 4% of all retroperitoneal neoplasms.⁶ They are seen in females twice as commonly as males.⁷ The incidence of retroperitoneal mature cystic teratomas peaks twice in the first 6 months of life and early adulthood. The Primary retroperitoneal tumor's genesis is the subject of several theories. They were originally believed to develop from the first segmentation of the fertilized ovum. It is often referred to as the Blastomere Theory. Another hypothesis is that incomplete conjoined twins are the source of extragonadal teratomas. The blastula doesn't divide evenly to allow for the separation of a twin; instead, an imperfect division takes place, and just a fraction of the cells are split, resulting in the teratoma. The most commonly accepted idea, which explains their midline and paramedian locations, is that they are remnants of the wolffian and müllerian ducts or originate from pronephric or mesonephric tubules.⁸ To further categorize teratomas, factors such as the number of layers present (monodermal, bidermal, and tridermal), the epithelial lining (epidermoid, dermoid, and teratoid), the degree of differentiation (mature and immature), the content (solid, cystic, or mixed), and the presence of malignancy (present or not) are taken into consideration.⁹ The retroperitoneum, close to the upper pole of the left kidney, is the typical presentation site. Normally asymptomatic, they might still exhibit abdominal pain, distension, nausea, vomiting, or compressive symptoms.¹⁰ The symptoms of compression of the nearby structures, such as vomiting, constipation, lumbar back discomfort, abdominal distention, and edema, would match with the

site of the teratoma. There may also be systemic symptoms such as fever, chills, night sweats, and weight loss. Chemical peritonitis caused by the rupture of the cyst may accompany retroperitoneal teratoma. It has been documented that a postpartum lady had a case of retroperitoneal cystic teratoma that manifested as a subhepatic abscess. Ocular myasthenia gravis and hypertension are two unusual manifestations resolved when the cystic tumor was removed.^{11,12} Although it might be challenging to distinguish between primary retroperitoneal teratoma and gonadal metastasis, the two require distinct treatments and have different results. Before a teratoma is classified as a Primary Retroperitoneal Teratoma, the gonadal primary must be ruled out by clinical examination and imaging. Teratomas don't have any distinct tumor markers. However, certain immature teratomas may have high AFP levels in the serum. Retroperitoneal teratomas can have high levels of AFP, CEA, and CA-19-9 in the blood, among other tumor markers. These indicators can track a patient's response to therapy and if their tumor recurs. A CECT or magnetic resonance imaging scan is frequently used to diagnose PRT. Calcification is a distinctive feature of 74% of benign and 25% of malignant teratomas. Imaging aids in identifying PRT and its relationships to surrounding structures, which is helpful for both diagnosing the condition and organizing the operation. It is possible to do a biopsy under CT or USG guidance, and CECT is the preferred test for differentiating between various retroperitoneal masses. Surgical excision in its entirety is the treatment's cornerstone. Malignant teratomas frequently recur despite full surgical excision. The reported five-year survival rate for benign teratomas is 100%, but for malignant tumors, it is around 67%.^{13,14} It is important to consider differential diagnoses for several conditions, including growing teratoma syndrome (the development of a mature teratoma after chemotherapy for a non-seminomatous germ cell tumor), soft tissue sarcoma, neurogenic mass, tuberculosis, Kaposi sarcoma, Castleman disease, and secondary from genitalia. Although imaging is frequently used to make the diagnosis, surgery and pathology are the cornerstones for the definitive diagnosis. Retroperitoneal teratomas harbor cancer, notably germ cell carcinomas, and require adjuvant chemotherapy and radiation. Platinum-based chemotherapy is employed because malignant teratomas exhibit characteristics similar to advanced testicular germ cell cancers. Malignant teratomas have a 61% overall response rate but are very resistant to radiation and chemotherapy. There are not as many reports of RT, thus there isn't as much information to determine the prognosis, which relies on the kinds of tissues the teratoma comprises. The prognosis is favorable with thorough resection of the benign cyst. However, death and morbidity are significantly greater if untreated because of blockage of surrounding viscera or metastases from cancers resulting from the RT. The prolonged post-operative follow-up is advised to find the early occurrence.

CONCLUSION

Retroperitoneal teratomas are a rare entity. These should be differentiated from gonadal teratoma with retroperitoneal

metastasis. Radiological evaluation for location and extent is a must for management. Complete resection is the mainstay of treatment. Follow-up is necessary with CT and tumor markers when malignancy is confirmed.

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