Primary Benign Cystic Retroperitoneal Teratoma in an Asymptomatic Adult

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Abstract—Primary retroperitoneal teratoma is a rare entity in adults with a distinctive imaging appearance. We describe an unusual case of a 19-year-old male patient referred to our hospital due to a large mass in his abdomen. Radiological work-up revealed a large retroperitoneal mass with no other masses or abnormalities. Exploratory laparotomy with tumor resection was performed. Histopathological diagnosis confirmed a benign cystic teratoma. The patient is doing well on follow up 5 years later.

Index Terms—Laparotomy, Retroperitoneum, Teratoma

I. INTRODUCTION
Teratomas are uncommon neoplasms that contain derivatives of all three germ layers (ectoderm, mesoderm, and endoderm). Historically, teratomas were attributed to demons, sexual misconduct, and abnormal fertilization. As with teratogenic, the name derives from the Greek word teras, meaning ‘monster’. Teratomas belong to a class of tumors known as non-seminomatous germ cell tumor and are typically located in the gonadal region. However, extragonadal sites such as the sacrococcygeal region, mediastinum, neck, diaphragm and retroperitoneum have also been reported. Retroperitoneal teratomas often occur in infancy and childhood but are rare in adults. In this article, we describe an unusual case of a retroperitoneal teratoma in a 19-year-old asymptomatic male patient who subsequently underwent successful surgical resection.

II. CASE REPORT
A 19-year-old male presented with upper abdominal distension for 3 months. He had no weight loss, bowel or urinary complaints. On examination, a large mass was palpable in the epigastric and left hypochondrium moving with respiration and dull to percussion. A clinical diagnosis of pseudopancreatic cyst was made and patient was sent for various investigations. Routine blood tests and urinalysis were all within normal limits. However, ultrasound demonstrated a large, complex, densely mixed echogenic mass, suggestive of a fatty nature to the mass with sheet like calcifications (Image 1) and no ascites.

Contrast computed tomography (CT) of the abdomen revealed a large retroperitoneal mass anterior to the head of pancreas, initially suspicious for a retroperitoneal sarcoma. However, the presence of fat and bone inside the mass was highly indicative of a benign cystic teratoma. CT-guided fine needle aspiration cytology (FNAC) of the mass was consistent with the diagnosis of teratoma. Chest X-ray prior to surgery, revealed no lung metastases or lymphadenopathy.

Exploratory laparotomy revealed a large cystic retroperitoneal mass (Image 2) situated anterior to the pancreas and posterior to the stomach. The tumor extended from the left hemidiaphragm down to the umbilicus beyond the midline. Macroscopically, the encapsulated mass measured 14.86 cm x 11.03 cm (Image 3). Microscopically, it appeared to be a cystic tumor with extensive necrosis and amorphous debris. There were few mature squamous cells along with few anucleate squamous cells. Background showed an infiltrate of polymorphs, lymphocytes and plasma cells. Within the mass itself, skin...
Adnexa, bone, fat and nerve were seen, consistent with a trilineage teratoma. Histopathology confirmed the diagnosis of a primary cystic teratoma, benign in nature, with no malignant cells present. Postoperative course has been uneventful.

III. DISCUSSION

Overall, primary retroperitoneal teratomas constitute about 1-11% [1-3] of all primary retroperitoneal tumors. Approximately half of all teratomas found in children are discovered in first decade of life with 43-45% of retroperitoneal teratomas diagnosed within the first year of life. Less than 10-20% of retroperitoneal teratomas present in patients after 30 years of age [4].

Teratomas arise from germ cells that fail to mature normally in the gonadal locations. These totipotent cells can differentiate into tissue components representing derivatives of mesoderm, ectoderm and endoderm. The distribution of teratomas listed in order of decreasing frequency is: ovaries, testes, anterior mediastinum, retroperitoneal space, pre sacral and coccygeal areas, pineal and other intracranial sites, neck and abdominal viscera other than gonads. The developmental migratory properties of germ cells would explain teratomas in these extragonadal sites, which generally occur along midline structures [1, 5].

Retroperitoneal teratomas are usually asymptomatic except when compression of the surrounding structures occurs. Patients with compressive symptoms may present with back pain, genitourinary symptoms, gastrointestinal symptoms (abdominal distension, pain, nausea and vomiting), as well as lower extremity or genital edema secondary to lymphatic obstruction [1, 6].

The differential diagnosis of retroperitoneal teratomas include ovarian tumors, renal cysts, adrenal tumors, retroperitoneal fibromas, retroperitoneal sarcomas (usually liposarcomas), hemangiomas, xanthogranulomas, enlarged lymph nodes and perirenal abscesses [7, 8]. Testicular ultrasound is necessary to rule out testicular germ cell tumor in male patients. This is a necessary step since 50% of men with retroperitoneal tumors also have testicular carcinoma in situ, a precursor for testicular germ cell tumors [9].

Between 1932 and 1987, 32 adult cases were reported out of which 15 occurred in female patients and 17 occurred in males. Retroperitoneal teratomas in these patients were most often located near the upper pole of the kidney, mostly on the left side [5, 6].

Malignant degeneration was higher in adults than in children, with incidences of 25.8% and 6.8%, respectively. Malignant teratomas may cause rise in serum alpha-fetoprotein (AFP) [6].

Calcifications can be demonstrated in 61.5% of teratoma cases on a plain anteroposterior and lateral abdominal films and are useful for the pre-operative diagnosis [1]. Such calcifications may be within the tumor or on the rim of the cyst wall. Even though 74% of benign teratomas contain calcification, they also occur in 12.5% of malignant teratomas [10].

Several imaging modalities elucidate different characteristics of a teratoma. For example, ultrasound can identify the cystic, solid or complex components of the tumor [11]. The cystic portion may be further differentiated into sebum, non-fat fluid and structures resembling fetal parts. However, ultrasound has its limitations as Davidson et al found that ultrasound poorly identified fat and calcifications, which are suggestive of teratoma.

CT has several advantages over ultrasound. First, it gives more specific information on the fat, proteinaceous fluid and calcification components of the teratoma through Hounsfield units, which allows quantitative comparison of substances of different radiodensities. The presence of fatty portions of the tumor in the horizontal interface with dependent fluid, which probably represents sebum, is virtually pathognomonic of a teratoma [11, 12]. However, a fat fluid level has also been described in case of a well differentiated liposarcoma of the retroperitoneum [13]. Second, CT appears superior to
ultrasound at defining extent in to surrounding organs and for evaluating the cyst wall [11].

Magnetic resonance imaging (MRI) and angiography offer other benefits. MRI is superior to both ultrasound and CT in defining the anatomical relationship of the teratoma with adjacent organs and local tumor spread [14, 15]. MRI can also distinguish fluid, fat, calcium and soft tissue elements, as well as predict resectability and evaluate recurrence [16]. Angiography is beneficial for detecting the presence of hypervascularity, arterial encasement and organ invasion, features often suggesting malignancy [17].

In our case, the initial differential diagnosis included retroperitoneal sarcoma. Even with modern preoperative imaging studies, retroperitoneal sarcomas can be confused with retroperitoneal teratomas [18]. Pre- or post-operative radiation therapy can be challenging in retroperitoneal sarcomas because of radiosensitive adjacent normal structures and the need for relatively high doses, especially in unresectable cases or when positive margins are left behind. In such situations, fast neutron therapy [19] or more recently, intensity-modulated radiotherapy, intraoperative electron beam radiotherapy or proton therapy has been considered [20]. While retroperitoneal sarcomas are often treated with a combination of preoperative radiation therapy and surgery [21, 22], retroperitoneal teratomas, if benign, usually do not warrant radiotherapy.

IV. CONCLUSION

In summary, teratomas can macroscopically be divided into 2 categories: cystic or solid. Cystic teratomas are mostly benign, containing sebaceous materials and mature tissue types. On the other hand, solid teratomas are often malignant and composed of immature embryonic tissues in addition to adipose, cartilaginous, fibrous and bony components. The prognosis is excellent for benign retroperitoneal teratoma if complete resection can be accomplished.

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REFERENCES