Retroperitoneum: A Rare Location Of Extragonadal Germ Cell Tumour

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Citation

Abstract
Extragonadal germ cell tumours in the retroperitoneal locations are rarely encountered. The imaging findings are distinctive. The present report is a case of a 30-year-old female in whom a retroperitoneal teratoma was found to have classical appearance on imaging. The tumour was resected in toto and histopathological examination confirmed the diagnosis of mature cystic teratoma.

INTRODUCTION
Germ cell tumours are congenital tumors containing derivatives of all the three germ layers (1). They are frequently seen in gonads (1). Their occurrence in extragonadal sites is not unknown (1,2,3). The involvement of extragonadal sites in decreasing order of frequency are mediastinum, sacrococcygeal region, retroperitoneum and pineal gland (1,2,3). Extragonadal primary teratomas are usually encountered in infants and children (1,2,3). Their occurrence in adults is a rare event (2,4-6). The present report describes such a case of primary retroperitoneal teratoma encountered in an adult patient, which was successfully managed by surgical resection.

CASE REPORT
A 30-year-old female presented with dull aching pain in the right flank for the last three years. There were no compressive symptoms. General physical examination and systemic examination was unremarkable. The hematological, coagulation and biochemical parameters including alpha-fetoprotein (AFP) levels were normal.

Plain abdominal radiograph showed a calcification in the right lumbar region (fig 1).

Ultrasonogram of the abdomen showed a 7x6x6cm mass in
the right retroperitoneal region pushing the kidney laterally. Contrast enhanced computed tomography (CECT) of the abdomen revealed a variable-density mass with solid, cystic, and fatty components and evidence of peripheral calcification in the retroperitoneal location. The mass was pushing the kidney laterally, the inferior vena cava anteriorly and was seen to cross the midline (fig 2).

Figure 2
Figure 2a & 2b: CECT axial sections at the level of the kidneys showing a large heterogenous mass in the retroperitoneum with fat (long arrow), soft tissue (short arrow) and calcifications (arrow head)

At laprotomy, an infrarenal retroperitoneal mass of 10x15cm was found below the right renal vein, medial to the right ureter and lateral to the inferior vena cava (IVC). About 6cm of the tumor were extending behind the IVC crossing the midline and lying anterior to the aorta (fig 3).

Figure 3
Figure 3: Intraoperative photograph showing a large tumour (long arrow) occupying the retroperitoneum and displacing the renal vein (short arrow)

The bilateral ovaries were normal. The tumour was removed in toto. Cut section revealed cheesy material and evidence of calcification. The tumour was fleshy and solid at places (fig 4a & 4b). The post-operative course was uneventful.
Figure 5
Figure 4a: Resected specimen showing a dumbbell shaped tumour with variable consistency.

Figure 6
Figure 4b: Cut section of the specimen showing pultaceous material (long arrow) and evidence of focal calcifications (arrow head). The tumour was firm and solid (small arrow) at places.

The histopathology report revealed mature cystic teratoma (fig 5a & 5b).
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DISCUSSION

Germ cell tumours contain the derivatives of all the three germ layers (1,2,3,4). The migratory capacity of germ cells may account for the anatomic variety seen with these tumors and this explains the occurrence of teratoma in the gonads and the midline structures (1,2,3,4). Extragonadal teratomas are thought to arise from primordial germ cells or early embryonic cells (5). These arise from totipotential cells (1,2,3,4). Solid and cystic varieties have been identified macroscopically. The former tends to be malignant while the latter is usually benign (5).

Primary retroperitoneal teratomas account for 1-11% of retroperitoneal neoplasms and are mostly seen in neonates and young adults (1,3). A bimodal age distribution pattern is noted with the disease occurring in the first 6 months of life and subsequently in early adulthood. About half of these are diagnosed in the first year of life (2,4). In adults these tumours have been found to present in their third or fourth decade of life. Those presenting early are more likely to be benign (10).

Benign teratomas are usually asymptomatic and often diagnosed on routine investigation. As the tumor mass increases, obstructive symptoms can develop. Common presenting symptoms include back or abdominal pain, genitourinary symptoms, gastrointestinal symptoms, as well as lower extremity or genital edema secondary to lymphatic obstruction. These tumours have been reported to mimic adrenal tumours also (1). Physical examination may detect a midline or paramedian abdominal mass with limited mobility (9). Teratomas can infect secondarily leading to abscess formation (11). Traumatic rupture with chemical peritonitis has been reported (12). Malignant degeneration, though extremely rare, has been reported (13).

Abdominal radiograph can demonstrate calcification in 61.5% of cases (1,2,3,4) as in the present case. Ultrasonogram demonstrates such a tumour to have a complex echopattern with solid and cystic components. Computed tomogram provides better delineation of fat and calcification (6). MRI allows for improved soft-tissue resolution and is useful in assessing the local extent of the disease. It can differentiate between the benign and malignant variety (14).

Retroperitoneal teratomas can express a variety of tumour markers like CEA, CA19-9, and alpha-fetoprotein (1,2,3,4).

Once the teratoma is detected, surgical resection is mandated. Complete excision in toto is curative for benign lesions. The use of laparoscopic technique in the benign variety has been reported (14). However, the malignant lesions are managed on the lines of their gonadal
counterparts (\(n\)). Incomplete resections may result in recurrence (\(n\)). Prognosis of benign retroperitoneal teratoma is good after complete resection and recurrences have not been reported (\(n\)).

Concluding, the primary extragonadal germ cell tumours in the retroperitoneal location are rarely seen in adults. They are usually asymptomatic or can produce compressive symptoms. The imaging features are suggestive. Complete excision is warranted to prevent recurrences.

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