Retroperitoneal fibrolipoma: A tumor bigger than its bite

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Abstract

Only five cases have been reported in literature

We present a rare case of retroperitoneal fibrolipoma that presented as a large intraabdominal mass, review essential investigations and treatment given.

Introduction

Lipomas are the most common tumors of adulthood and are morphologically classified as conventional lipoma, fibrolipoma, angiolipoma, spin- dle cell lipoma, myelolipoma and pleomorphic lipoma (1,2). Retroperitoneal benign lipomas, however, are extremely rare, and represent about 2.9% of all primary retroperitoneal tumors (3). Most retroperitoneal lipomas have usually undergone myxomotous change and malignant (liposarcoma) (4). The well differentiated variant is relatively indolent, the myxoid type is intermediate in its malignant behavior while the round cell and pleomorphic variants are aggressive frequently metastasize(5). We present a case showing that it is difficult and sometimes impossible to have a preoperative diagnosis when a large retroperitoneal mass is involved.

Case Presentation

A 41 year old male casual laborer, presented with history of gradual abdominal swelling over a period of eight months. The swelling started as a small mass in the right lower quadrant but gradually grew to involve all quad- rants. It was initially painless until 6 months earlier when the patient started experiencing a dull continuous ache that was severe enough to give him insomnia but was re- lieved partially by analgesics. It was associated with back pain, anorexia and vomiting that was non-bilious and non-projectile. Patient was passing stool normally

There were no urinary symptoms, fever or chills.

He had initially sought care in homeopathic clinics before coming to our hospital, Coast provincial general

hospital.

On examination we found a middle aged man who was wasted and had bilateral pitting edema up to the knees. The rest of the general examination was unremarkable. On abdominal exam he had a grossly distended abdomen that was symmetrical, with veins visible on the abdominal wall. There was a tender palpable mass on light palpation, not adherent to the overlying skin, firm in consistency and had large irregular nodules. Both testes were present in the scrotum. Other systems were essentially normal.

A full heamogram showed normal white blood cell count 6.0 x10° cells/L with a relative osinophilia at 19.8% and basophilia of 12.9%. His erythrocyte sedimentation rate was also increased at 67mm/hr. Liver function tests were normal with alkaline phosphatase 12.98 (39-117 u/L). His renal function tests were normal.

An ultrasound done showed a large intraabdominal mass while a CT scan done showed a well circumscribed intraperitoneal abdominopelvic mass measuring 28 by 24 by 31 cm. The mass displaced all peritoneal contents upwards but did not displace retroperitoneal organs (Figure 1). It compressed the inferior vena cava and the ureters causing a bilateral hydroneprosis. The origin of the mass was not established.

The mass on CT Scan had fat, calcific and soft tissue foci bathed in a thick proteinacious fluid consistent with a teratoma.

The liver and spleen showed no signs of metastasis. The differentials before surgery included teratoma, solid organ tumor, abdominal wall sarcoma, omental tumor or a hydatid cyst.

At surgery, incisions into the pseudo-capsule which included the peritoneum and omentum were made. Many fascial planes were found to exist and at random one was explored bluntly and sharply but found to be adherent as we progressed posteriorly. This was abandoned and another fascial plane anterior to it was explored. Extirpation of the mass was possible through this dissecting plane by separating it from the adherent organs. However, the lower sections of the capsule were adherent to the retroperitoneal structures, including aorta and inferior venacava, so part of the capsule was left behind after consultation with the vascular surgeon. Intraoperative blood loss was 1.5 liters and blood resuscitation was required.

The mass weighed 11.5 kg (Fig 2a) and macroscopically had a well defined margin.

Postoperatively the patient did well, fed on the second day and discharged on the tenth. Gross pathologic exam revealed a gelatinous lobulated vellow mass that microscopically consisted of proteinecous material in a matrix of fibrous tissue with scattered cells. Histologically the mass was reported as a myxoma. A second opinion was sought and the histology report showed an admixture of fat and fibrous tissue consistent with a retroperitoneal fibrolipoma.

Discussion

Retroperitoneal masses are typically divided into benign and malignant causes. Benign tumors account for 15% of all masses. Of the malignant causes, slightly above half are soft tissue sarcomas. [Fig 2(b)]

Retroperitoneal lipomas occur more in women than men. They can reach astounding size and cause little or no symptoms(7). This was the case with our patient who went on with his daily activities unimpeded with an 11 kg mass in his abdomen.

Symptoms attributable to pressure effect include bilateral pitting edema and compression of the ureters with hydronephrosis as demonstrated on the ultrasound. This is usually not the case in small masses.

A physical examination should include testicular examination in men to evaluate the possibility of a primary testicular tumor with metastasis to the retroperitoneum.

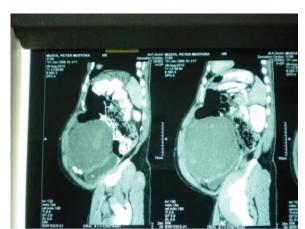


Figure 1 CT scan view of the mass showing a well circumscribed abdomina mass displacing all the abdominal organs



Fig 2a Gross specimen



Figure 2 b Cut specimen showing a yellow homogenous mass

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Our patient had a normal testicular examination.

The other differentials for a retroperitoneal mass include lymphoma, primary retroperitoneal germ cell tumor and sarcoma. Blood test should therefore include germ cell tumor markers including beta-human chorionic gonadotropin (β -hCG) and alpha fetoprotein none of which were available at our institution. Alkaline phosphatase levels in our patient were not elevated as they would be in germ cell tumors (8).

Diagnosis can be difficult but a first step is imaging with CT scan which serves an additional role of staging the tumor. The radiological diagnosis of tera-toma served to add confusion as to whether or not the tumor was a primary retroperitoneal germ cell tumor. A biopsy does not change the management since some teratomas can be treated effectively by surgery alone. The seemingly benign nature of the tumor was confirmed at surgery where a fair line of cleavage was found and the tumor without much difficulty except for the posterior part of the tumor that was adherent to the vessels. Most malignant tumors of the retroperitoneum are without a capsule and a few cases of sarcomas have com-plete or incomplete capsules. However, a majority have a pseudo-capsule due to centripetal expansile growth resulting in a relatively well defined zone of compressed normal tissue.

The first histology report showed myxoma-like cells which pointed towards a liposarcoma of intermediate malignant potential. The second showed a well differentiated fibrolipoma with no malignant potential. Without cytogenetic studies it is difficult to correlate malignant potential with plain histopathological appearance as diagnostic liposarcoma cells may be difficult to identify, even after extensive sampling especially with a large homogenous looking mass as was our case (9). If a malignant tumor was found then a course of chemotherapy would be required (11). It should be noted that in patients with malignant tumor incomplete resection provides symptom palliation and survival prolongation (10).Retroperitoneal lipomas/liposarcomas suffer from a high rate of recurrence and therefore close follow up is required. Our patient did not suffer recurrence up to 4 months post-surgery. In conclusion, retroperitoneal lipomas/liposarcomas can present as large abdominal masses with little or no symptoms. Radiologic investigations may not give you a definitive diagnosis but serve to define the mar-gins of the mass. The nature of surgery is rarely altered by biopsy and definitive

treatment remains complete resection of macroscopic disease. This is usually not possible due to the intimate relationship with unresectable structures (inferior venacava and aorta). Marginal resection however has been shown to provide survival prolongation and symptom palliation.

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