Primary retroperitoneal transitional cell carcinoma presenting as a dumb-bell tumour

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ABSTRACT

We report a retroperitoneal transitional cell carcinoma arising from the primitive urogenital remnants of a 56-year-old married Indian woman. She presented with a huge cystic mass in the hypogastrium and right iliac fossa, which extended into the right thigh as a massive dumb-bell tumour. On exploration, it was found not to be arising from any known retroperitoneal structure. The mass was excised, and the histopathology confirmed transitional cell carcinoma with positive margins. Though she received postoperative chemotherapy with cyclophosphamide, adriamycin and cisplatin, she developed extensive local recurrence and hepatic secondaries, and succumbed to the disease after ten months of follow-up. We highlight the rarity of the disease, its atypical presentation as a cystic dumb-bell lump, its diagnostic challenges and aggressive behaviour, and review the literature on primary retroperitoneal transitional cell carcinomas.

Fig. 1 Clinical photograph of the patient shows a massive lower abdominal lump with another in the right thigh.

INTRODUCTION

Primary retroperitoneal tumours arise from the tissues of the potential retroperitoneal space. These tumours develop independently, without any apparent connection to any organ or major vessel. The embryonic remnants of the primitive urogenital ridge form variable components of the retroperitoneal space, and they can give rise to both benign and malignant tumours in this region. The tumours arising from these remnants are usually benign and cystic, and are known as nephrogenic cysts.[1] Malignant tumours are rare and are called urogenital ridge tumours. We report a rare case of malignant urogenital ridge tumour, which presented as a dumb-bell shaped cystic swelling in the abdomen and right thigh, and discuss the challenges to the surgeon.

CASE REPORT

A 56-year-old Indian woman presented with complaints of a rapidly increasing lump in the lower abdomen, which was associated with pain, weight loss and anorexia for the past six months. She also complained of an inability to completely flex the right hip for the last one month. General physical examination was unremarkable, except for moderate pallor. The abdominal examination showed a large cystic lump of 25 cm × 20 cm in the right lower abdomen extending to the hypogastric region. Another 10 cm × 10 cm cystic lump was present in the right upper thigh (Fig. 1). Both these lumps were apparently communicating with each other as a dumb-bell cystic mass, as evidenced by cross fluctuation. The femoral pulse was feeble and was felt superficial to the lump. There was no gross neurovascular deficit in the right lower limb. Movements of the right hip joint were present but restricted, and both flexion and extension were painful. Except for moderate anaemia (haemoglobin 7.2 g/dL), her haematological and

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Fig. 2 Contrast-enhanced reformatted coronal CT image shows the cystic nature of the lump with a dumb-bell appearance. Note that the abdominal lump is continuous, with the extension in the thigh.

Fig. 3 Preoperative intravenous urogram shows the displacement of the right ureter to the left.

biochemical tests were within normal limits. Contrast-enhanced computed tomography (CT) showed a huge thick-walled retroperitoneal cyst in the right iliac region extending into the pelvis and right thigh (Fig. 2). An intravenous urogram showed a displacement of the right ureter to the left (Fig. 3). There was no evidence of infiltration into surrounding structures or metastasis on CT. Based on these findings, a provisional diagnosis of a retroperitoneal dermoid cyst or lymphatic cyst was made. Although the rapid progression was a cause of concern and indicative of malignancy, it was not confirmed on CT.

On exploratory laparotomy, the metastatic evaluation was negative. The tumour was not found to be arising from, or infiltrating into any known retroperitoneal structure or organ. The right ureter was displaced to the left, across the midline by the tumour, along the lower part of its course without any tumour infiltration. The lump could be excised through a combined approach via the abdomen and thigh. Histopathology confirmed it as a transitional cell carcinoma of high grade, with microscopic positive resection margins (Fig. 4). A final diagnosis of a primary retroperitoneal urogenital ridge tumour with transitional cell carcinoma was made. The patient was started on multidrug adjuvant chemotherapy with cyclophosphamide, adriamycin and cisplatin. After ten months of follow-up, she developed multiple secondaries in the liver and extensive local recurrence in the retroperitoneum, and finally died from hepatic and renal failure.

DISCUSSION

Adult primary retroperitoneal tumours are rare, but a diverse group of neoplasms constitute only 0.2%–

0.5% of all malignant tumours.[1,2] They are either mesodermal (50%–60%), lymphatic/reticuloendothelial (30%–40%) or neurogenic (10%–20%) in origin,[3] and may arise from any structure in the retroperitoneum, such as the fat, fascia, muscle, vascular tissue, nerve tissue, lymph vessels, lymph nodes, and also from remnants of the urogenital ridge. The two most common malignant primary tumours of the retroperitoneum are lymphosarcoma and liposarcoma.[4] In the early part of this century, Handfield-Jones observed that a majority of the retroperitoneal cysts had their origin from the remnants of the urogenital apparatus.[5] A similar origin of these tumours was reported in a series of 17 cases by Hansmann and Budd.[6] In a review of 101 cases of primary retroperitoneal tumours, Braasch and Mon found only three cases of urogenital ridge tumours. Two of them died within six months of the diagnosis and the third case, treated by radiation, died within three years. A complete excision was not possible in any of these cases.[7] In a report by Gupta et al, a urogenital ridge tumour with transitional cell carcinoma was found to be inoperable in a 50-year-old woman, who finally died within two years of the diagnosis and treatment.[8] The present case is the fifth of its kind, and the patient died within one year of surgery and adjuvant treatment. These cases indicate that urogenital ridge tumours are highly malignant and have a poor life expectancy. Surgery is the main treatment modality, as response to adjuvant therapy is meagre.

In the present case, the tumour was a primary retroperitoneal one, as evidenced by the absence of origin from any defined retroperitoneal organ or structure. Other differential diagnoses would have been a retroperitoneal dermoid cyst, retroperitoneum malignant lymphatic
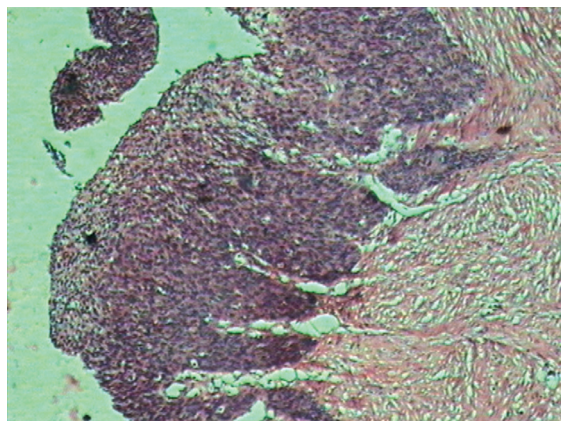
Fig. 4 Photomicrograph of the excised tumour shows features consistent with transitional cell carcinoma (Haematoxylin & eosin, × 50).

cyst, cloacal cyst with transitional cell carcinoma,[9] or mucinous cystadenocarcinoma.[10] Though CT did not demonstrate any evidence of malignancy or metastasis, the rapid progression of the tumour, its huge size and aggressiveness were suspicious signs of malignancy. The dumb-bell shape of the tumour was unique and has not been reported before. Primary retroperitoneal tumours tend to remain localised in the retroperitoneum, and an extension into the thigh is extremely unlikely. This occurred in our case, probably due to the rupture of the Gerota's fascia and progression through the psoas fascia into the thigh. An ultrasonography or CT-guided fine needle aspiration cytology (FNAC), though an invaluable preoperative investigation in solid tumours, should be done with caution in case of a cystic malignancy. Tumour perforation and dissemination can occur, and hence, FNAC should be done from the solid components of a complex tumour, which was not so in our case, or the needle pass should be made through healthy uninvolved tissue, which was difficult in the retroperitoneum. Proliferation markers and tumour markers may help in the diagnosis, but some of these are nonspecific, and one needs to know for sure which marker would be useful. These procedures are costly and often unaffordable for most patients. Probably a positron emission tomography-CT fusion scan would have been of help in diagnosing its malignant nature. The positive microscopical resection margins can be explained by the fact that in large retroperitoneal tumours, there is often microscopic breach of the capsule without infiltration. In such cases, an apparently R0 resection may be histologically R1, taking into consideration the lack of adequate margins in the retroperitoneum. Furthermore, in a large cystic malignancy, the chance of rupture during extirpation, which leads to tumour dissemination, remains high,. When one is unsure of the malignant

potential, an intraoperative frozen section biopsy can be of help, but in large tumours, it may be difficult to determine the area of suspicion for biopsy unless any macroscopic breach of margins has occurred.

Retroperitoneal tumours, due to their deepest location in the abdomen and unrestricted growth in the loose retroperitoneal tissue, usually attain a large size before producing any symptom. The symptoms are mainly due to compression, obstruction or invasion of the adjacent organs. Urogenital ridge tumours are said to occur with much greater frequency in females because the major portion of the ridge may remain in an unused vestigial state.[8] In the male, however, the mesonephros (wolffian body) is completely utilised in forming the gonads, the epididymis and vas deferens. Surgery remains the mainstay of treatment, as the adjuvant therapies are not very promising.[11] A complete excision of these tumours is not easy and may not be possible in most cases, due to the proximity of the vital structures and adjacent vessels. In difficult situations, partial excision and vascular resection with reconstruction may have to be done as disease-free survival and overall survival depend mainly on the adequacy of resection. The most important reason for incomplete resection is vascular invasion. Incomplete resection is justified in patients with widespread metastasis in the abdominal cavity or whose complete resection is not possible due to extensive vascular or organ invasion. The sole purpose is then to reduce the tumour burden and relieve pressure symptoms. Infiltration into adjacent structures is common, and the inability to achieve negative margins leads to early local recurrence, poor postoperative outcome and decreased long-term survival. Only about 25% of the malignant retroperitoneal tumours can be completely excised with an operative mortality of 10%–25%.[8] The progression of the disease produces local compressive effects and may result in intestinal or urinary tract obstruction. The terminal events involve widespread metastasis, haemorrhage from invasion of major blood vessels and renal/hepatic failure.

In conclusion, primary retroperitoneal tumours are rare, particularly those arising from the urogenital remnants. Most of them are benign cysts, but malignant ones are often aggressive, and being in the retroperitoneum, they tend to grow to huge dimensions before diagnosis. Local infiltration and metastasis are telltale signs of malignancy on CT. However, caution should be exercised during surgery in an apparently benign cystic tumour, lest it turns out to be malignant on histopathology, and hence utmost effort must be made for complete excision. Most of these are advanced at

diagnosis and complete removal is often not possible. Surgery is the mainstay of treatment as these tumours respond only partially to adjuvant therapy. The prognosis is poor, as most patients die within a few months or years.

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