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International Journal of Surgery Case Reports

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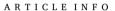


Case report

Retroperitoneal lipoma and bilateral renal cell carcinoma in a rare co-existence

Badi Rawashdeh^{a,*}, Elias Ayoub^b, Alp Demirag^b

- a Division of Transplant Surgery, Medical College of Wisconsin, 9200 W Wisconsin Ave. CFAC 2nd Floor, Milwaukee, WI 53226, United States of America
- ^b Division of Transplant Surgery, University of Virginia, 1215 Lee Street. Charlottesville, VA 22903, United States of America



Keywords: Retroperitoneal lipoma Bilateral renal cell carcinoma End stage kidney disease

ABSTRACT

Introduction: Giant retroperitoneal lipomas are rarely observed clinically, and a retroperitoneal lipoma accompanied by renal cell carcinoma is even more unusual. We present a case of a large retroperitoneal lipoma with bilateral renal cell carcinoma that was definitively diagnosed after resection.

Case presentation: A huge retroperitoneal mass was incidentally discovered in a 58-year-old male with end stage renal disease being evaluated for a kidney transplant. Imaging studies revealed a mixed solid and fat-containing mass displacing the left kidney. Repeat imaging discovered concurrent unilateral renal cell carcinoma and interval enlargement of the mass. Histopathology showed benign adipose tissue, bland spindle cells, and mixed inflammatory infiltrate that was negative for MDM2 amplification. Resection of the mass and bilateral nephrectomy was performed. Final histopathological examination was consistent with bilateral renal cell carcinoma and a large benign retroperitoneal lipoma.

Discussion: The presented case provides a prime example of the diagnostic challenges encountered with retroperitoneal tumors, The final diagnosis of lipoma in this case was only made after review of the resected specimen in its entirety. Retroperitoneal lipomas can present differently based on tumor size and involvement of adjacent organs. The concomitant occurrence of a renal cell carcinoma and retroperitoneal lipoma is extremely rare, and this is the first report published in the literature.

Conclusion: We document a rare case of retroperitoneal lipoma with concurrent bilateral renal cell carcinoma and illustrate frequently encountered challenges during the evaluation of retroperitoneal masses.

1. Introduction

The retroperitoneal space lies between the parietal peritoneum and endoabdominal fascia in the posterior abdominal wall. Superiorly, the space communicates with the posterior mediastinum through the lumbocostal triangle, and inferiorly, with the retrorectal space. It contains the kidneys and the suprarenal glands, the aorta, and inferior vena cava, the third and fourth sections of the duodenum, the pancreas, the rising and the ascending and descending portions of the colon, the ureters, and the rectum [1].

Primary retroperitoneal tumors are rare entities that display significant histological variety. They may originate from lymphoid tissue, muscle, urogenital tract, or other retroperitoneal organs [2–5]. This region's tumor can present challenges for diagnosis and management as they typically present late with nonspecific symptoms [4]. Distinguishing between malignant and benign tumors is an important consideration

and, if done promptly and accurately, allows for better intraoperative management and postoperative follow-up. Most retroperitoneal tumors are malignant, with liposarcoma being the predominant subtype representing 45 % of cases. Other common malignant neoplasms include leiomyosarcoma, lymphoma, and epithelial tumors. Benign tumors of the retroperitoneum are less likely and include schwannomas, neurofibromas, renal angiomyolipomas, and lipomas [6,7].

Lipomas are benign tumors of mature adipocytes and are commonly located in the subdermal tissue of the trunk and extremities but are rarely found in the retroperitoneum, with only 19 cases of retroperitoneal lipomas in adults described in the literature [8]. Differentiating benign retroperitoneal lipomas from liposarcomas is particularly challenging in the pre-operative period. While imaging and biopsies can provide valuable diagnostic information, current radiologic and histopathologic techniques are unable to consistently rule out malignancy [7,9]. Given this risk, the typical approach in equivocal cases is surgical

^{*} Corresponding author at: Medical College of Wisconsin, 9200 W Wisconsin Ave, CFAC 2nd Floor, Milwaukee, WI 53226, United States of America. E-mail address: Brawashdeh@mcw.edu (B. Rawashdeh).

excision if feasible, followed by postoperative pathological assessment of the tumor to make the diagnosis and guide future management [10]. Renal cell carcinoma (RCC) is a common cancer of the urogenital system and accounts for 3 % of all malignant tumors [11]. In this report, we present a case of large retroperitoneal lipoma associated with RCC in a patient on a kidney transplant waiting list, which is to date the first identified case. We describe the radiological and histopathological workup and surgical management. This report has been reported in line with the SCARE criteria [12].

2. Case description

A 57-year-old Caucasian man with end-stage renal disease (ESRD) presented to our transplant center to be evaluated for kidney transplantation and was found to have a giant retroperitoneal mass. His past medical history was significant for hypertension, melanoma, multiple side-branch intraductal papillary mucinous neoplasms of the pancreas, and focal segmental glomerulosclerosis (FSGS), for which he received a living-donor kidney transplant at the age of eleven. The allograft failed, and he has been on hemodialysis since that time. The patient reported decreased appetite associated with an unintentional 17 kg weight loss over the last two years, but was otherwise asymptomatic. During his transplant work up, a large retroperitoneal mass adjacent to the left kidney had been incidentally discovered on abdominal computed tomography (CT) (Fig. 1A). The abdominal CT also revealed a 3.4×3.3 cm exophytic complicated lesion with solid and cystic components arising from the upper pole of the right kidney and a small exophytic lesion arising from the lower pole of the left kidney. The retroperitoneal mass was also visualized on MRI (Fig. 1B); Whole body PET CT did not show any evidence of metastasis. CT-guided core biopsy of the retroperitoneal mass and right kidney lesion were performed. The right kidney lesion demonstrated histology consistent with RCC. Histology of the retroperitoneal mass revealed adipose cells with no overt cytologic atypia suggestive of a benign lipoma. Immunostaining was positive for smooth muscle actin (SMA) and negative for human melanoma black (HMB45) and Melan A. Further testing with fluorescence in situ

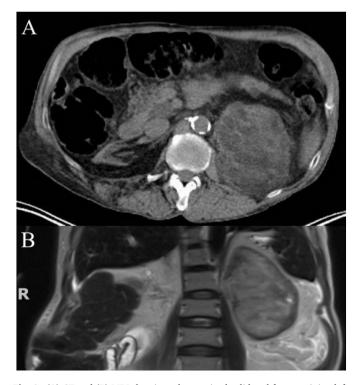


Fig. 1. (A) CT and (B) MRI showing a large mixed solid and fat-containing left retroperitoneal mass.

hybridization (FISH) for *MDM2* gene amplification was negative, providing no support for a diagnosis of well-differentiated liposarcoma.

On physical examination, the patient's abdomen was distended and non-tender to palpation. The patient underwent the resection of the tumor along with bilateral nephrectomies. The left kidney was also removed due to a suspicious radiological lesion which was not successfully biopsied preoperatively. The patient recovered adequately before being discharged on the seventh post-operative day. The patient visited our clinic 5 months after the procedure with no symptoms or signs of recurrence. He received a follow-up chest CT that did not show lung lesions.

Histopathological assessment revealed acquired cystic disease associated RCC and multiple papillary adenomas in both the left and right kidney. On inspection, the retroperitoneal mass measured $17.6 \times 14.6 \times 6.4$ cm. On histopathology, the specimen consisted predominantly of benign adipose tissue with scattered bland spindle cells and mixed inflammatory infiltrate consistent with a benign lipoma (Fig. 2).

3. Discussion

The presented case provides a prime example of the diagnostic challenges encountered with retroperitoneal tumors. The differential diagnosis for fatty tumors of the retroperitoneum includes liposarcoma, angiomyolipoma, and lipoma [9]. The latter is an exceedingly rare diagnosis, with only nineteen cases presented in the literature [8]. Well-differentiated liposarcomas (WDLPS), the lowest-grade subtype of liposarcomas, are particularly difficult to distinguish from benign lipomas [10,13]. The differential can be narrowed in the preoperative period with imaging and histopathology.

Imaging features suggestive of well-differentiated liposarcoma include thickened septa, foci of nodular enhancement, and invasion of adjacent organs [13,14]. However, a significant number of benign lipomas will contain similar features on imaging. Gaskin et. Al.; evaluated the ability of MRI evaluations of fatty masses to distinguish WDLPS from benign variants such as lipomas relibly. They found that 63 % of lesions considered suspicious for WDLPS on MRI were actually benign variants, illustrating the limits of radiological evaluation [15].

For these reasons, histopathological assessment including immunohistochemistry is often required to characterize retroperitoneal tumors. Fine-needle aspiration or core-needle biopsy are commonly performed, with core-needle biopsy having the additional benefit of providing information regarding tissue architecture [6]. Amplification of the CDK4 and MDM2 genes are specific markers of liposarcomas and can be demonstrated with immunohistochemical staining as well as FISH analysis, which was done for our patient. In contrast, angiomylipomas characteristically demonstrate positive immunostaining for HMB45 protein and smooth muscle actin [9]. Despite their utility, a significant portion of these tumors lack these distinguishing features, so they cannot be used to rule out these entities. Given this overlap between features of benign lipomas and angiomylipomas and the much higher incidence of liposarcomas, some recommend surgical excision of retroperitoneal masses of this nature regardless of preliminary biopsy findings [6,10]. The final diagnosis of lipoma in this case was only made after review of the resected specimen in its entirety.

Retroperitoneal lipomas can present differently based on tumor size and involvement of adjacent organs. Common symptoms include abdominal distension and nonspecific pain in the abdomen, back, or flank [5,8,16,17]. They can grow to astonishingly large sizes, with a recent case report describing a $55\times40\times10$ cm tumor weighing 8.95 kg [8]. Compressive symptoms are sometimes present and range from constipation, urinary retention, hematuria, and sciatica and can cause significant morbidity [3,17,18]. Our patient had an uncommon presentation, as the mass was found incidentally on imaging in a patient not complaining of distension, pain, or compressive symptoms. Weight loss has not been described in previously published cases, and it is likely that our patient's weight loss is explained by his bilateral RCC and other

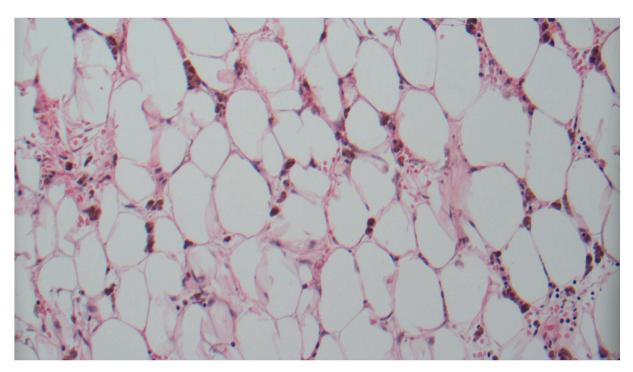


Fig. 2. Hematoxylin and eosin stained section of the retroperitoneal mass showing benign adipose tissue with scattered bland spindle cells and mixed inflammatory infiltrate (magnification ×500).

comorbidities. Imaging and biopsies are often inconclusive in the preoperative period, but MDM2 amplification is consistently negative [5,10,13,16]. In addition, cytogenetic analyses have identified a t(3;12) translocation resulting in HGMA2 rearrangements that further establish the diagnosis of lipoma, but these analyses are performed infrequently. In most cases, complete resection is the mainstay of treatment, and recurrence is possible but uncommon [4].

ESRD is a global health burden, with an overall prevalence of 823 patients per million population [11]. In addition, ESRD is associated with a higher risk of cardiovascular disease and cancer. Worldwide, RCC constitutes 2-3 % of all malignancies and is the third most common urological cancer. It has previously been shown that around 2-7 % of patients with ESRD develop RCC and this risk with time. RCC in ESRD patients presents predominantly as small, low-stage and low-grade tumors [11]. Due to this increased risk, many providers have opted to perform regular screening for patients with ESRD on dialysis, although guidelines describing the optimal imaging modality (ultrasound vs. CT vs. MRI), screening intervals, and target population have yet to be established [19,20]. There is evidence of a survival benefit from screening, especially in younger patients on dialysis, that supports this approach [21]. For patients who are older or in poor medical condition, however, shared decision-making that carefully weighs the risks and benefits of RCC screening is the preferred approach. In our report, this patient had a very long history of ESRD and at the time of detection of the lipoma was found to have also developed small bilateral renal tumors consistent with low-stage RCC. The concomitant occurrence of RCC and retroperitoneal lipoma is extremely rare and this is the first report published in the literature.

4. Conclusions

Lipomas of the retroperitoneum are very rare entity that can mimic other more common retroperitoneal tumors but typically carry a better prognosis. Current diagnostic modalities are limited in their ability to reliably differentiate lipomas from AML and WDLPS, but lipomas should be considered when histopathology does not identify classic markers of other tumors. In summary, abdominal signs, imaging with ultrasound,

CT, and MRI, and histopathological evaluation should be used in combination to make the diagnosis of lipoma and RCC. Complete resection is the preferred treatment.

Funding

N/A.

Ethical approval

N/A.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Registration of research studies

- 1. Name of the registry:
- 2. Unique identifying number or registration ID:
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Guarantor

Badi Rawashdeh, MD

Provenance and peer review

Not commissioned, externally peer-reviewed.

CRediT authorship contribution statement

Badi Rawashdeh, MD: Study design

Alp Demirag, MD: Manuscript writing Elias Ayoub, MD: Data collection

Declaration of competing interest

N/A.

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