

Delayed Diagnoses of Retroperitoneal Cystic Adenocarcinoma Mimicking Renal Cyst

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INTRODUCTION

Primary retroperitoneal cystadenocarcinomas are extremely rare.⁽¹⁾ In the English literature, only 28 cases of primary retroperitoneal cystadenocarcinoma have been published.⁽²⁻⁸⁾ Because of the rarity of these tumors, it is hard to determine their exact origin and define an optimal treatment regimen. When this type of lesion is adjacent to the kidney, it can mimic a renal cyst. Preoperative diagnosis is difficult because the condition can be mistaken for renal cystic disease.⁽⁹⁾ With the increasing availability and use of abdominal imaging tools, cystic renal disease has become a very common finding and the detection of complex cystic masses in the kidney has increased dramatically.⁽¹⁰⁾ However, there is no agreed upon follow-up schedule for complex cystic lesions of the kidneys.

We report our experience with a patient with delayed diagnosis of primary retroperitoneal cystadenocarcinoma involving hepatic metastasis that mimicked a renal cyst. It is our hope that physicians will be made more aware of this type of tumor and include it in their differential diagnoses.

CASE REPORT

A 67-year-old woman was admitted to our hospital with left flank discomfort and a palpable mass. Her medical history included an extracorporeal shock wave lithotripsy (SWL) procedure for a urinary stone in the left ureterovesical junction 14 years earlier. At that time, she had a 12 × 9 cm left renal cyst and cytology following cystic aspiration was negative. After 7 years, another left renal cyst, 7.4 cm in size, was detected; cytology following cystic aspiration in that instance was also negative. She failed to appear at a follow-up appointment. When she presented to our hospital, physical examination was unremarkable. She was afebrile and normotensive, her pulse was 96 beats/min, and her respiratory rate was 20 breaths/min. Routine laboratory findings including urinalysis results were normal. A chest X-ray and electrocardiogram showed no remarkable findings. Abdominal computed tomography (CT) scan demonstrated a retroperitoneal multi-septated cystic lesion with a solid enhancing mass in the left lower pole of the kidney, which met the criteria for a Bosniak category IIF cyst (**Figure 1A**). Magnetic resonance imaging (MRI), performed to evaluate the cause of the patient's symptoms, showed a 7 × 6.5 × 5.1 cm well-defined lobulated hemorrhagic cystic mass abutting the lower pole of the left kidney, which was in agreement with the diagnosis of a Bosniak category IIF cyst.

The patient visited our outpatient clinic for a follow-up every six months, and there was no significant change in the cystic lesion except for an increase in size (the cyst grew from about 7 cm to 8.8 cm over 2 years), as determined by ultrasound imaging. Given that the cyst significantly increased in size and flank discomfort persisted, surgery was considered.

An abdominal CT scan revealed a 8.4 × 7.3 × 5.5 cm septated cystic mass with solid enhancing portions in the left retroperitoneum abutting the lower pole of the left kidney and the descending colon, suggesting a malignant cystic tumor, several variable-sized peripheral enhancing mass lesions in the liver, suggesting metastases, and several small (1 cm or smaller) lymph nodes in the para-aortic space (**Figures 1B and 1C**).

The patient underwent ultrasound-guided 18G needle biopsy of the left hepatic lobe and the retroperitoneal cystic

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Figure 1. Computed tomography (CT) scan. **A)** A Bosniak category IIF cystic lesion in the lower pole of the left kidney. **B)** Multiple heterogeneous masses in the liver. **C)** A cystic mass in the retroperitoneum, which is invading the psoas muscle.

lesion. Both pathological exams were consistent with adenocarcinoma, so it was clear that metastasis had occurred (**Figure 2**). For lesions not amenable to surgery, chemotherapy is the treatment of choice.

DISCUSSION

Cystic renal disease is very common and the rate of detection of complex cystic lesions in the kidney has increased dramatically over the last few decades because of the increased availability of cross-sectional imaging tools.⁽¹⁰⁾ Cysts may be complicated by hemorrhage or infection and develop calcification, septation, wall thickening, or high attenuation, which are all features that are shared by cystic tumors. To characterize these lesions and provide an approach for their management, the Bosniak classification system based on CT features is used by urologists and radiologists in clinical practice. Category I lesions require no follow-up or other intervention. Category II lesions are considered benign

and also do not require follow-up. Where there is some ambiguity, however, about the extent or character of the septation, calcification, or other features, follow-up with repeat imaging studies at 3 months, 6 months, and 1 year is recommended. Category III lesions can be either malignant or benign tumors. Because it is difficult to accurately distinguish between different kinds of multilocular renal masses, such lesions must be surgically removed. If clinical and imaging features suggest multilocular cystic nephroma, kidney-sparing surgery can be performed. Category IV lesions have more obvious malignant characteristics and are regarded as unequivocally malignant, requiring surgical management.⁽¹¹⁾ In the case of our patient, the lesion was determined to be a category IIF, which requires annual follow-up studies. However, in the 7 years after the patient saw a specialist for the first time, the lesion was evaluated only once. After 14 years, the lesion had evolved into an adenocarcinoma with liver metastases.

Retroperitoneal adenocarcinomas are extremely rare.⁽¹⁾ Because of the rarity of these tumors, it is hard to determine their exact origin and decide on an optimal treatment regimen. Preoperative diagnosis is difficult because such lesions are easily mistaken for renal cystic disease.⁽⁹⁾ The ability to differentiate between benign and malignant tumors represents a major challenge. The clinical course may appear to be indolent; however, these tumors can become aggressive. Overall, the prognosis and pathophysiology of such tumors are uncertain because of their rarity and the lack of long-term follow-up studies. The longest reported follow-up study was done over the course of 6 years.⁽¹²⁾ Of the 26 female patients in that study, 4 died of metastatic disease, which involved the lungs, liver, adrenal gland, peritoneum, and regional lymph nodes. Adjuvant chemotherapy was attempted in 6 patients with limited success. Three patients died within 4–18 months after surgery, 1 experienced recurrence after 21 months, and 2 others were

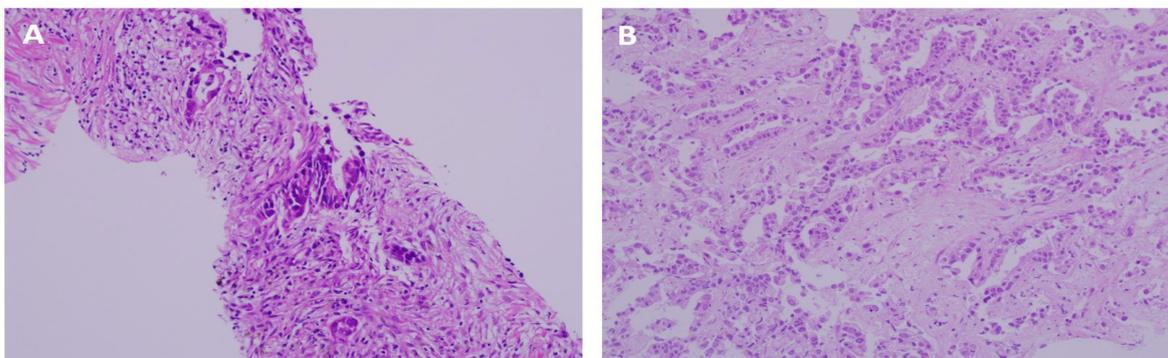


Figure 2. **A)** Histological evaluation of the retroperitoneal mass, demonstrating adenocarcinoma (Hematoxylin and eosin staining, $\times 10$). **B)** Histological evaluation of the hepatic mass, demonstrating adenocarcinoma (Hematoxylin and eosin staining, $\times 10$).

free from recurrence at 18 and 33 months.⁽⁴⁾

Mucinous cystadenocarcinomas can also occur in the liver, pancreas, and ovaries. Our patient had a renal cystic mass and several hepatic masses. Preoperative evaluation could not exclude benign renal cystic disease as the origin of the cystic retroperitoneal mass. Renal lymphangiomas can also present as slow-growing flank or abdominal masses that take an indolent clinical course; in those cases, imaging reveals large perirenal and peripelvic cysts. Parasitic cysts or renal hydatidosis are uncommon conditions but should also be considered.^(7,14) Diagnosis can often only be made through surgical excision.⁽¹³⁾ Needle biopsy may be of use in these situations because a biopsy may reveal adenocarcinoma, which implies that a tumor, such as a primary retroperitoneal adenocarcinoma, is likely to exist. However, needle biopsy is not a good tool for identifying malignancy in cystic tumors.

Physicians should consider primary retroperitoneal adenocarcinoma in the differential diagnosis of large retroperitoneal cystic lesions, because such lesions are often mistaken for renal cysts. The tumors usually present with mass effects, as in our patient, take an indolent course, and can become very large before they cause clinical problems. Preoperative diagnosis is difficult, and in most cases, a laparoscopic procedure is the best method for diagnosis and treatment. Aspiration and cytology of the cystic lesion was not effective in this case. However, needle tract seeding is a rare event; the incidence is estimated to be less than 1 in 10,000 biopsies. Only eight cases of needle tract seeding during a renal mass biopsy have been described in the medical literature.⁽¹⁵⁾

CONCLUSIONS

Even though a cyst may not progress rapidly, it can display aggressive clinical behavior, and the appropriate treatment regimen has yet to be defined. To date, extirpative surgery appears to be the treatment of choice, and the role of adjuvant radiation or chemotherapy has yet to be determined.

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CONFLICT OF INTEREST

None declared.

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