Case report: a left upper quadrant complex cystic mass

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1. Introduction

We describe an unusual case of a patient presenting with left-sided abdominal pain. The differential diagnosis for a large left upper quadrant complex cystic mass would include lesions of the pancreas, spleen, adrenal gland, kidney, and mesentery. On resection, the lesion was found to be a cystic nephroma. These are rare, almost always benign tumors of the kidney that can grow to massive size. They are usually removed due to symptoms and/or lack of clarity as to diagnosis. The clinical features and pathology of these tumors are discussed.

2. Presentation of case

A 61-year-old woman presented with left upper quadrant and flank pain that had been present for 3 months. She also complained of increasing abdominal girth during this time. She denied nausea, vomiting, change in bowel habits, blood per rectum, melena, hematuria, weight loss, or fever. She had no major medical comorbidities and was on no medications. She did not drink alcohol and was not a smoker. There was no family history of malignancy. On physical exam, she was afebrile, and her vital signs were normal. Abdominal exam revealed a palpable, mildly tender mass in the left upper quadrant. Stool hemoccult was negative. Her serum electrolytes and BUN/creatinine were normal. White blood cell count, hemoglobin, and platelet count were unremarkable and serum levels of CA 19-9, CEA, and CA-125 were all within normal limits. CT scan with oral and IV contrast revealed a large complex, multi-cystic lesion in the left upper quadrant (Fig. 1). Some mild left hydrenephrosis was noted. Primary differential diagnosis based on imaging included retroperitoneal lymphangiomatosis vs. pancreatic cystic neoplasms. The patient was taken for surgery. Ureteral stenting was followed by laparotomy. A large mass was found attached to the left kidney (Fig. 2). There was no involvement of the pancreas or spleen. Left nephrectomy was performed (Fig. 3). The patient recovered uneventfully and was discharged on the 4th postoperative day. At one-year follow-up, no recurrence was noted, and her renal function was normal.

3. Discussion

The differential diagnosis for a large left upper quadrant complex cystic mass would include lesions of the pancreas, spleen, adrenal gland, kidney, and mesentery. Of these, pancreatic cystic neoplasms would likely be the initial clinical diagnosis in a middle-aged woman presenting with left-sided abdominal pain. Although the size of the lesion was impressive, our patient had no weight loss or cachexia to suggest obvious malignancy. All tumor markers were negative. CT scan showed a multi-septated complex cystic tumor with no evidence of metastatic disease. Based on this study, the primary pre-operative radiologic diagnosis was lymphangiomatosis. A cystic lesion of the pancreatic tail was thought less likely. We elected to proceed with surgery since resection would be primary treatment for either of these potential diagnoses. Preoperative biopsy was not obtained due to concerns about possible cyst rupture as well as our belief that the results would not change management of this large and symptomatic lesion. At operation, a 25 cm renal tumor was found and resected en-bloc with the left kidney (Fig. 3). Pathology revealed cystic nephroma (Fig. 4).
Cystic nephromas are rare, almost uniformly benign tumors whose age incidence can span from infancy into the elderly. Diagnosis is most frequently made in the sixth decade, and there is increasing female predominance as age increases.3–5 There is some controversy as to whether the disease is the same in children vs. adults, with some authors arguing that in infancy, these lesions may actually be a benign form of nephroblastoma (Wilm’s tumor).4 Genetic predisposition has been reported but appears to be rare.6,7 The tumor may present as an incidental finding on imaging studies done for other reasons, or it may present with pain and/or mass effect, as in this case.8 Symptoms appear to correlate with increasing size of the lesion.3 Hematuria is not uncommon although urinary tract infection (presumably due to ureteral obstruction) is unusual.3,9,10 Radiographically, cystic nephroma is difficult to distinguish from other cystic diseases of the kidney, both benign and malignant, such as cystic renal cell cancer.11,12 When massive, such as in our case, it may not be possible to even clearly define a renal etiology. On CT imaging, these tumors appear as thinly septated, multilocular mass.
lesions. There is minimal to no contrast enhancement, and contrast excretion into the cysts is never seen. The borders are irregular and peripheral calcifications may be noted. May be extension into the renal pelvis as seen here (Fig. 1). Cyst fluid has Hounsfield measurements similar to that of water.\textsuperscript{11,13} On magnetic resonance imaging, the cysts may be hyperintense on T2 images and have varying signal intensity on T1 images. The septa may enhance with gadolinium administration.\textsuperscript{13,14} Despite these characteristics, definitive radiologic diagnosis is impaired by the rarity of the lesion as well as its similarities to other cystic lesions.

On gross inspection, the resected specimen forms a distinct mass lesion and consists of multiple non-communicating cysts separated by a thin stroma. In terms of histopathology, the diagnostic criteria for cystic nephroma require that they be unilateral, solitary, and multiloculated. There must be no communication between the individual locules or with the renal pelvis. Finally, the cysts should have a well-differentiated epithelial lining and contain no renal elements.\textsuperscript{4,15} The lining is usually characterized by flat or cuboidal epithelium with no features of malignancy, such as atypia or mitotic figures. The fibrous stroma contains cellular elements similar to ovarian tissue; hence these were once thought to be Mullerian developmental defects.\textsuperscript{5,16} Treatment is almost always surgical, either due to symptoms or a question as to diagnosis. Pre-operative biopsy is controversial with some authors arguing that if the diagnosis can be confirmed, observation may be an option,\textsuperscript{12} as only a single case of malignant transformation has been reported.\textsuperscript{17} This may be a consideration, however, we as well as others,\textsuperscript{5,10,13,14,18} would favor resection in the presence of symptoms, increasing size, or uncertain diagnosis. Most cases have historically been treated with nephrectomy. Recently, nephron-sparing techniques have been suggested for this benign lesion, especially when small.\textsuperscript{10} Larger tumors such as our patient’s continue to be treated with nephrectomy, especially when involving the collecting system.

4. Conclusion

Cystic nephromas are benign tumors of the kidney. When very large, they may mimic other cystic lesions of the abdomen and thus should be on the surgical oncologist’s radar. They should be removed when symptomatic, increasing in size, or there is uncertainty as to diagnosis.

Conflict of interest statement

None.

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Consent

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

Author’s contributions and competing interests

LC drafted the manuscript and performed the literature review. NM provided the micrographs and performed the pathologic analysis. SK drafted and completed the manuscript, aided in the literature review, and provided the gross and intra-operative photographs. All three authors have no competing interests.

References