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Localized Cystic Disease of the Kidney: Progression in a Case Followed-Up for 14 Years

Localized cystic disease (LCD) is the accumulation of multiple simple cysts, partially involving the kidney which are usually detected incidentally. Imaging findings of LCD are important to differentiate it from other renal cystic diseases and to avoid unnecessary surgery. Although it is known as a non-progressive condition, we present here the imaging findings of a case with LCD showing progression in long-term period.

Keywords: Impotence, Computerized Tomography, Cavernosography

Introduction

Localized cystic disease (LCD) is an uncommon presentation of renal cystic disease which has been confused with other renal cystic masses. It has also been called by different names such as segmental cystic disease or unilateral cystic disease.¹⁻³ LCD is a benign condition that may be followed under the guidance of imaging. Although it is known as a nonprogressive condition, we present here the imaging findings of a case with LCD showing progression in long-term period.

Case Presentation

An asymptomatic 29-year-old man was referred for abdominal sonography to evaluate hepatosteatosis. During sonographic examination, multiple various-sized cysts that did not communicate with each other were found incidentally in the upper half of the right kidney (Fig. 1A). These non-encapsulated cysts were randomly scattered throughout the cortex and medulla, some of which contained thin septa. The parenchymal septa separated the upper portion of the right kidney from the lower portion.

The patient had no family history of autosomal dominant polycystic kidney disease (ADPKD), or urinary complaints or symptoms. Serum creatinine and blood urea nitrogen (BUN) levels were 0.7 mg/dL and 9.0 mg/dL, respectively. The liver function tests were normal (alanine aminotransferase, 13 U/L; aspartate aminotransferase, 20 U/L; alkaline phosphatase, 112 U/L; and lactate dehydrogenase, 327 U/L). At the time of initial diagnosis, normal parenchyma between the cysts and intervening septae could be seen on sonography. No involvement was observed in the lower half of the same and the complete contralateral kidney. There was no other organ involvement.

Sonographic findings and the final diagnosis of "localized cystic disease of the kidney" were confirmed by pre- and post-contrast enhanced computed tomography (CECT) and renal angiography. Digital subtraction angiography (DSA) of the kidneys showed vascular displacement in the upper part of the right kidney. A few vessels surrounded the upper pole and normal vascular distribution was

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seen in the uninvolved portions of the kidney (Fig. 1B). Axial CECT scan demonstrated multiple cysts and normally enhancing renal tissue (Fig. 1C). The family of the patient was invited to search for the presence of ADPKD. His family members had no cysts on abdominal US.

Fine needle aspiration biopsy (FNAB) revealed only fibrous elements, serous fluid and benign epithelial cells. Since a benign process was strongly considered by urologists, partial nephrectomy or surgical biopsy was not performed. Diagnosis was made in a weekly interdisciplinary meeting on the basis of FNAB and imaging findings.

After a 7-year follow-up of the patient with annual sonographic examinations, the cysts were stable. Additional CT scan also confirmed radiological

stability. The uninvolved lower portion of the right kidney and left kidney remained normal. In the 10th year of follow-up, a progressive increase in the number and size of cysts was noticed on serial ultrasound examinations. The cysts distorted the adjacent renal parenchyma. Almost all the upper half of the kidney was involved with various-sized multiple cysts without any solid component or renal parenchyma among them. The upper half of the right kidney was markedly enlarged and there was no sign of renal failure. Fourteen years after the diagnosis, the longitudinal length of the right kidney reached 266 mm. Parenchymal septae and the lower part of the kidney were still intact (Fig. 1D).

The patient still remained clinically stable in the 14th year of the follow-up.

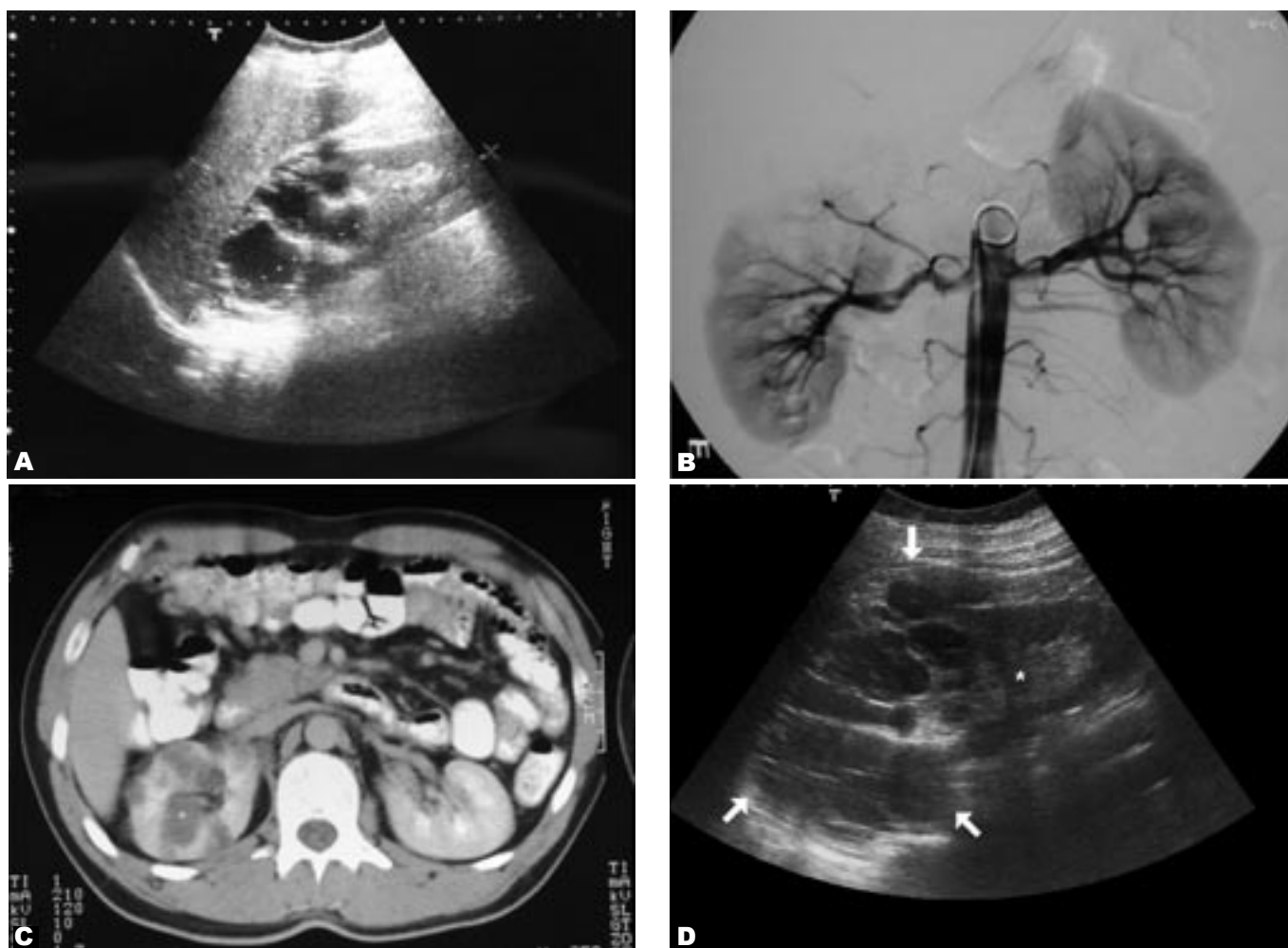


Fig. 1. A 29-year-old man with a localized cystic disease.

A. Sonography shows a cluster of cysts separated by parenchyma in the upper part of the right kidney.

B. DSA shows vascular displacement in the upper part of the right kidney. The left kidney and uninvolved portions of the right kidney have normal vascular distribution.

C. Contrast-enhanced abdominal CT demonstrates multiple simple cysts of various sizes separated by a normally enhancing renal tissue.

D. After 14 years, a progressive increase in the number and size of the cysts on sonography is noticed. The cysts occupy almost all the upper half of the kidney without any solid component among them. The right kidney is markedly enlarged. The lower part and parenchymal septa (*) is still intact.

Discussion

LCD is the accumulation of multiple simple cysts in one portion of the kidney. This entity is non-familial and is usually detected incidentally.¹⁻⁴ CT scan and sonography are the main imaging methods to characterize LCD. Magnetic resonance imaging findings are parallel to those of corresponding CT scan. DSA is not necessary for diagnosis, but it helps us differentiate LCD from renal cystic neoplasm. In this case, the CECT finding was presence of multiple simple cysts of various sizes separated by normally enhancing parenchyma. Sonography revealed multiple scattered simple cysts in one portion of the right kidney and delineated internal structure of the cysts better than CT scan.

LCD is a rare non-progressive clinical presentation. Slywotzky and Bosniak stated that LCD should not be labeled as a hereditary, progressive renal cystic disease like ADPKD. Unilateral and partial involvement, absence of family history, no progression to chronic renal failure, and no associated cysts in any other intra-abdominal organs help us to distinguish LCD from ADPKD.¹⁻³ However, both of them have morphologic similarities. The pathological findings of LCD are not different from those of ADPKD. Regarding our patient, we diagnosed LCD on the basis of CECT, DSA and sonographic findings. After 10 years of stability, we realized that the disease showed a progression in the affected segment similar to ADPKD. The cysts increased in number and distorted the renal parenchyma. Almost all the upper half of the kidney was involved with various-sized multiple cysts and consequently, the kidney was markedly enlarged. Some authors proposed that there may be a somatic mutation within the ADPKD gene only in the affected segment of the kidney.³⁻⁵ Although Slywotzky and Bosniak¹ reported that four out of nine followed-up patients showed progression in the size of the cysts,

they did not report any renal enlargement in these patients. The total follow-up time in these four cases, which is necessary to decide whether the condition is progressive or not, was not clear in their article.

Benign cystic lesions of the kidney can probably be managed with regular follow-ups. In this way, unnecessary surgery may be avoided in many patients. Sonography is an appropriate technique to follow-up such lesions, because it shows stability or progression. There has been no consensus on the total follow-up time for LCD yet. Some authors suggest that a 5-year follow-up may be adequate.^{4,6} However, a longer follow-up period, as we preferred in our patient, may be necessary, especially in young patients with LCD-like lesions, to show real progression in terms of enlargement and parenchymal loss. Studies in large series having long-term follow-ups are necessary to confirm this approach.

In conclusion, LCD may show an ADPKD-like progression in the affected segment, and long-term follow-ups with sonography provide proper information about the progression. Radiologists should gain familiarity with imaging characteristics of LCD in order to prevent unnecessary surgery.

Conflict of interest statement: All authors state that they have no conflict of interest.

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