Multilocular Renal Cysts with Renal Cell Carcinoma: Report of Four Cases

Chia-Hsi Chen, MD; Cheng-Keng Chuang, MD, PhD; Chun-Te Wu, MD; Kwai-Fong Ng[1], MD; Shuen-Kuei Liao[2], PhD

According to Bosniak's classification, renal cysts with moderate calcification, irregular margins, and thickened enhanced septa should raise a suspicion of malignancy. Diagnosis of multilocular renal cysts depends on their histological features. Since 1951, it has generally been considered that a multilocular renal cyst can change from a benign lesion to one which may be combined with malignant change. According to a report from Japan, multilocular renal cysts with renal cell carcinoma are uncommon, and the reported incidence was about 9%. A radical nephrectomy was performed for malignant change. Herein, we present 4 cases of multilocular cyst (Bosniak's class IV) with renal cell carcinoma and review the related literature for their pathogenesis and management. *(Chang Gung Med J 2003;26:772-6)*

Key words: multilocular renal cyst, renal cell carcinoma, Bosniak's classification.

The diagnostic criteria of multilocular renal cysts were documented by Powell et al. to include a well-developed capsule, fibrous stromata, multiple epithelia lining the septa, and no communication with each other.[1] A multilocular renal cyst is usually considered a benign lesion, but malignant changes in the cyst should not be ignored if there is a mass lesion or calcification in the cysts. We report on 4 cases of multilocular renal cysts with renal cell carcinoma which were treated by a nephrectomy.

CASE REPORTS

Case 1

A 63-year-old man was referred to our hospital due to right flank mass without fever, hematuria, or flank pain. He had received extracorporeal shock wave lithotripsy for a right renal stone in a local medical office 3 months before this admission. Renal sonography, an abdominal computed tomographic (CT) scan, retrograde pyelography, and abdominal magnetic resonance imaging (MRI) revealed a right renal cystic lesion with a septum and soft-tissue density (Figs. 1, 2). Under the impression of cystic renal cell carcinoma, a radical nephrectomy was performed.

Case 2

A 47-year-old man had complained of left flank pain for 2 years. Renal sonography and abdominal CT revealed a 7×7×7-cm cystic mass located in the left kidney with a septum and calcification in the cyst. Cystic renal cell carcinoma was suspected, and a radical nephrectomy was performed.

Case 3

A 32-year-old man was incidentally found to have a right renal cystic mass during a physical

From the Division of Urology, Department of Surgery, [1]Department of Pathology, Chang Gung Memorial Hospital, Taipei; [2]Graduate Institute of Clinical Medicine, Chang Gung University, Taoyuan.
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Address for reprints: Dr. Cheng-Keng Chuang, Division of Urology, Department of Surgery, Chang Gung Memorial Hospital. 5, Fushing Street, Gueishan Shiang, Taoyuan, Taiwan 333, R.O.C. Tel.: 886-3-3281200 ext. 2103; Fax: 886-3-3285818; E-mail: chuang89@cgmh.org.tw

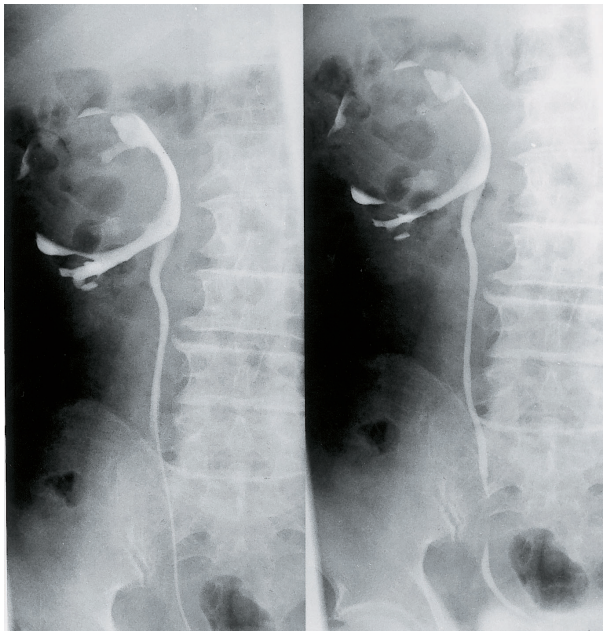


Fig. 1 Retrograde pyelography showing a mass in the right kidney and no communication noted with the collecting system.

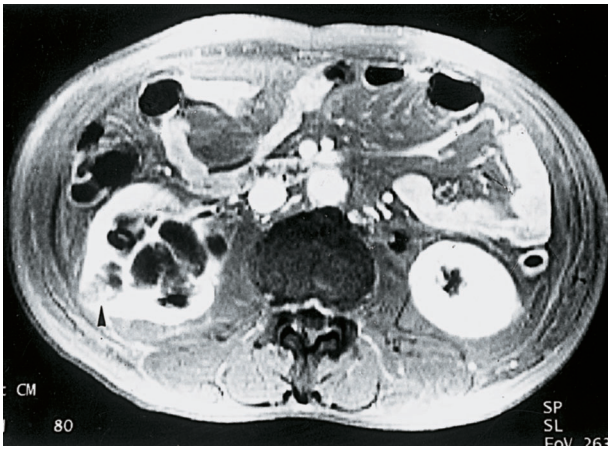
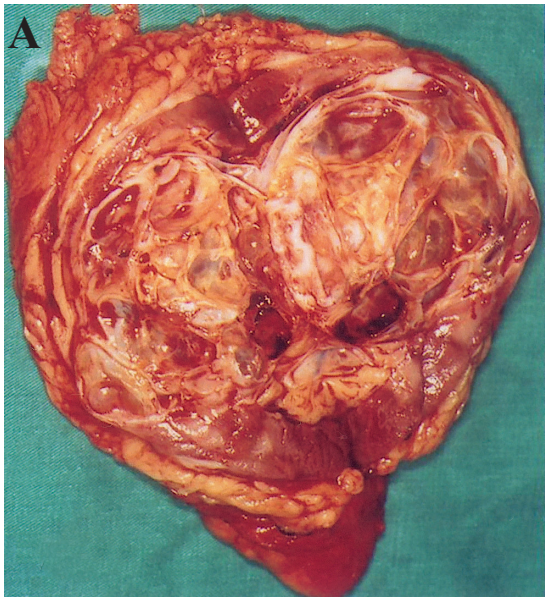


Fig. 2 MRI of the right kidney showing 1 cyst with soft tissue in the septum (arrowhead).



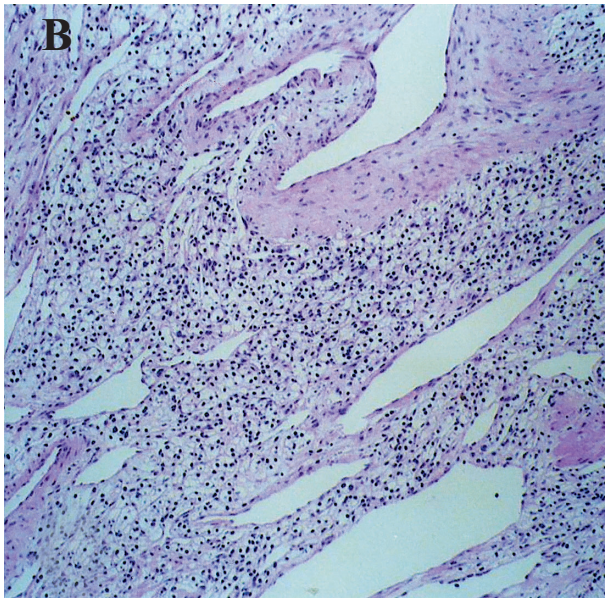


Fig. 3 (A) Gross appearance. The cyst was separated by fibrous septa resulting in a multilocular appearance. (B) A tumor composed of cells of large clear cell carcinoma. These show a number of cystic areas. (H&E stain, ×100)

examination. There were no presenting symptoms. The cystic mass was irregular in contour, and there was a suspicious nodular lesion inside the cyst on abdominal CT. A radical nephrectomy was performed.

Case 4

A 51-year-old man was incidentally noted to have a left renal cystic lesion on abdominal echo during a general health checkup. No hematuria or flank pain was reported by the patient himself. An

abdominal CT was therefore arranged. An enlarged tissue mass between the septum of the cyst was noted. We therefore performed a radical nephrectomy on this patient.

Grossly, the 4 tumors had a cystic appearance with septa, and all contained serous fluid within the cyst. Microscopically, the 4 tumors showed a clear cell carcinoma morphology with Fuhrmann's grade I nuclear atypia arranged in a tubulocystic pattern. All 4 cases showed multilocular cystic features of carcinomas (Fig. 3A, B) and were classified as Bosniak's class IV. These 4 patients were closely followed-up for 9 months to 3 years, and no evidence of disease was found in any of them.

DISCUSSION

Gallo and Penchansky hypothesized the pathogenesis of multilocular renal cysts as follows: (i) those of dysgenetic origin are topographically confined to a single lobe, and (ii) those of neoplastic origin are mainly based on observations of clinical and histologic similarities with Wilms' tumor.[2] Diagnosis is mainly based on histopathological features such as a well-developed capsule, fibrous stromata, multiple epithelial lining septa, and no communication with each other.[1] According to Bosniak,[3] renal cysts can be classified into 4 types: (I) simple cysts; (II) mildly complicated but clearly benign cysts with thin septa and minimal calcification but with no enhancement; (III) more-complicated cysts with moderate calcification, irregular margins, and thick enhanced septa; and (IV) cystic neoplasms. If any one of the following 3 criteria are met--moderate calcification, irregular margins, and a thickened (> 1-mm) septum--one should consider the possibility of malignant changes in the cyst.[2,3] Traditionally the clinical management of multilocular renal cysts included unroofing-marsupialization, a partial nephrectomy, or enucleation for benign lesions and a radical nephrectomy for malignant changes. But there are also some reports of nephron-sparing surgery for the management of multilocular renal cell carcinoma.[4]

Multilocular renal cysts are usually benign, but malignant changes may occur in them. However, the molecular basis of malignant transformation remains unknown. In the previous literature, there is a unifying concept that links Wilms' tumors, cystic partially differentiated nephroblastomas, and cystic nephromas in a spectrum of differentiation. In view of the fact that renal cell carcinoma arises in a variety of longstanding cystic diseases of the kidney, by analogy it is possible that renal cell carcinoma may in the same way occasionally arise in cysts of cystic nephromas, accounting for some cases of multilocular cystic renal cell carcinoma.[5,6] Most of the malignancies in multilocular renal cysts were clear cell carcinomas. The incidence of multilocular renal cysts with clear cell carcinoma in Japan was around 9% (20/223).[7] In a Spanish study, the incidence of multilocular cystic renal cell carcinoma was about 11.2% (23/206).[8] In this study, all 4 cases of multilocular renal cyst also showed clear cell carcinoma histology. The most recent Japanese study of 21 patients with cystic renal cell carcinoma reported that after surgery, 5-year and 10-year disease-specific survival rates for these patients were both 100%. It is of interest to note that consistent with the status of tumor grade for the 21 cystic renal cell carcinomas,[9] the 4 cases presented in this study were all found to be grade I tumors. Taken together, we stress the clinical relevance of multilocular renal cysts with irregular margins or mass lesions within the cyst, both of which should raise suspicions of malignancy; after pathological confirmation, early surgery on these patients is likely to achieve long-term survival.

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多房性腎囊腫合併腎細胞癌：四病例報告

陳家熙 莊正鏗 吳俊德 吳桂芳[1] 廖順奎[2]

依據Bosniak對腎囊腫在X-光下的表現並配合病理組織的發現將其分爲四大類，若在中隔處發現鈣化或有增厚的情形，或是不規則邊緣均有可能合併惡性腫瘤。傳統上多房性腎囊腫之診斷主要依賴其病理組織之特徵。自從1951年多房性腎囊腫被提出至今，對此病理變化之想法已由傳統上單純良性腫瘤轉變成爲具有惡性變化之可能性的腫瘤，根據日本於1996年提出的報告來看，多房性腎囊腫約有9%的機率會合併了腎細胞癌，如果合併惡性病變則必須採取根除性腎切除術。在此吾人提出四個病例爲多房性腎囊腫合併腎細胞癌，皆屬Bosniak分類第四型，此四個病例接受根除性腎臟切除手術。在此吾人回顧醫學文獻討論可能的病理機轉及其臨床處理方式。(長庚醫誌 2003;26:772-6)

關鍵字：多房性腎囊腫，腎細胞癌，Bosniak分類。

長庚紀念醫院 台北院區 泌尿一科，[1]病理科；[2]長庚大學 臨床醫學研究所
受文日期：民國91年10月14日；接受刊載：民國92年3月28日。
索取抽印本處：莊正鏗醫師，長庚紀念醫院 泌尿一科。桃園縣333龜山鄉復興街5號。Tel.: (03)3281200轉2103; Fax: (03)3285818; E-mail: chuang89@cgmh.org.tw