

## Cystic Renal Cell Carcinoma Arising From Multilocular Cystic Nephroma of the Same Kidney

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### ABSTRACT

Multilocular cystic nephroma is an uncommon benign entity grouped among the cystic non-genetic diseases. It is characterized by variable-sized, non-communicating cysts separated by irregular thin walled septa. Though multilocular cystic nephroma is usually considered a benign lesion, malignant changes in the cysts should not be overlooked.

**Key words:** kidney; cyst; renal cell carcinoma

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### INTRODUCTION

The kidneys are prone to a variety of cystic disorders that includes developmental, acquired, and neoplastic lesions. However, the synchronous occurrence of two different renal tumors in the same kidney is a rare event. Here we report a case of cystic renal cell carcinoma arising in a multilocular cystic nephroma in a 65-year-old woman.

### CASE REPORT

A 65-year-old white woman presented with a 10-month history of weight loss and malaise. There was no history of urinary or bowel symptoms. A mass was palpated in the right loin. Urine analysis was normal and urine cytology was negative for malignancy. Full blood count, liver function tests and renal function tests were all normal. An ultrasound scan showed a large complex mass arising from the lower

pole of the right kidney. CT scanning displayed a huge multilocular heterogeneous mass in the lower pole of the right kidney (Figure-1). The differential diagnosis of a malignant versus a benign lesion was not possible with imaging, and a right radical nephrectomy was performed.

On pathology, a large multi-loculated cyst with occasional more solid areas replaced the kidney. The lesion had an intact fibrous capsule with no involvement of the peri-renal tissues. Microscopically, the cysts were thin walled and epithelial lined, with varying amounts of intervening cellular stroma containing numerous small tubular cysts and focal smooth muscle (Figure-2). The appearances were those of a multilocular cystic nephroma (Figure-3). However, in some of the variegated and thickened areas noted macroscopically, the typical clear cells of cystic renal cell carcinoma were found (Figure-4). The renal cell carcinoma did not breach the capsule or invade adjacent renal tissues.

**COMMENTS**

The etiology of multilocular cystic nephroma (MCN) has been the subject of controversy for a long period of time. However, many currently believe that MCN is a benign neoplasm and lies at the benign end of a continuum that includes the cystic partially differentiated nephroblastoma (CPDN) variants of Wilms' tumor (1,2).

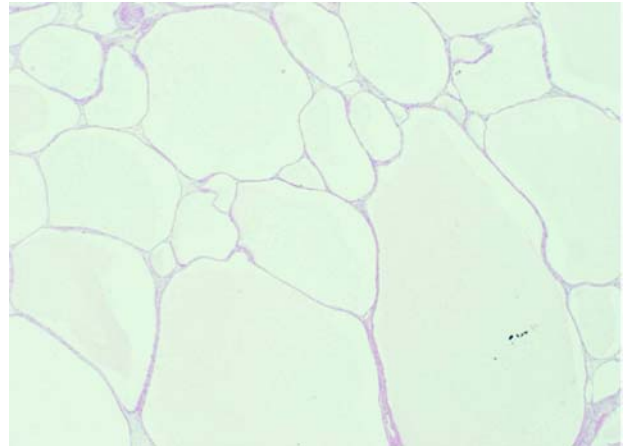
There is a bimodal age distribution. Under the age of 5 years, MCN occurs most frequently in males, whereas the adult group has a female predominance between the ages of 40 and 60. The most common presenting symptoms are a painless abdominal mass, abdominal or flank pain, and occasionally hematuria (2).

Several authors have reported on the difficulty of differentiating between MCN and multilocular cystic renal cell carcinoma (MLCC). However, definitive diagnosis is always histological.

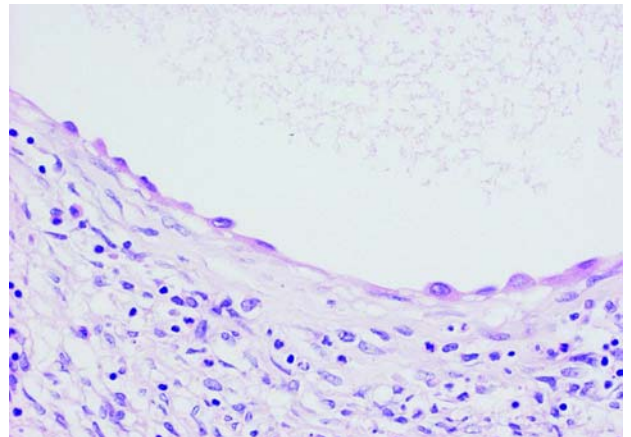
Renal cell carcinoma may occasionally arise in a variety of longstanding cystic diseases of the kidney, including the cysts of multilocular cystic nephroma. However, most of the reported malignancies in multilocular renal cysts have been clear cell carcinomas.



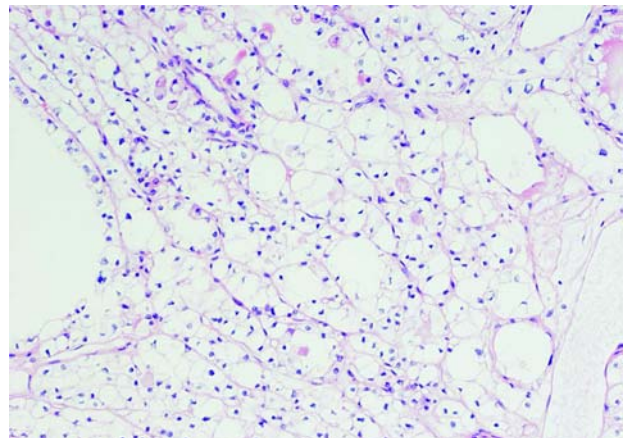
**Figure 1** – Contrast-enhanced CT scan of the abdomen showing the large right-sided multiseptated renal mass.



**Figure 2** – Photomicrograph showing the multiple thin walled cysts, with varying amount of intervening stroma (HE, X20).



**Figure 3** – Photomicrograph showing a cyst lining by the flattened epithelial cells with hobnail appearance (HE, X200).



**Figure 4** – Photomicrograph showing the neoplastic clear cells lining the wall of a cyst (HE, X250).

The prognosis for CRCC is excellent. A recent study of 21 patients from Japan reported that after surgery, 5-year and 10-year disease free specific survival for these patients were both 100% (3). Our patient remains free of recurrence with 4 years follow-up.

In summary, a multilocular renal cyst is usually considered a benign lesion, but malignant changes in the cyst can occur, indicating that thorough sampling should be undertaken for histology. However, the prognosis is excellent and most patients are likely to achieve long-term survival.

### CONFLICT OF INTEREST

None declared.

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### EDITORIAL COMMENT

Epithelial hyperplasia evolving to neoplasia is common to several forms of human renal cystic disease, both congenital and acquired. Most frequently occurs in von Hippel-Lindau disease, a dominantly inherited syndrome of cerebellar and retinal hemangioblastomas, pancreatic, epididimal and renal cysts and tumors. Tumors also occur in acquired renal cystic disease in end-stage kidneys especially those of patients on long-term hemodialysis, in the dominantly inherited disorder tuberous sclerosis complex and, less frequently, in the adult polycystic renal disease. The arising of cystic renal cell carcinoma from multilocular cystic nephroma is a very controversial condition. The present case is worth reporting because the multilocular cystic nephroma is well

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documented. Thin septa of either fibrous tissue or resembling ovarian stroma are present and covered by flattened or low cuboidal epithelium with small amounts of cytoplasm occasionally with hobnail appearance. However, this cystic lesion may be coincidental to the renal cell carcinoma. Many other cases should be reported in order to consider cystic multilocular nephroma a predisposing lesion to kidney neoplasias.

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