

# Multilocular Cystic RCC- A Rare Variant of RCC

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## Abstract

We present a case-report of Multilocular cystic renal cell carcinoma (MCRCC) incidentally found in a fifty year old male who presented for renal calculi of contralateral side. Based on characteristic gross findings MCRCC is considered to be a distinct subtype of renal cell carcinoma (RCC). A notable difference between conventional RCC and MCRCC is the absence of nodal /metastatic spread at the time of presentation.

## Introduction

MCRCC is a recently described, uncommon variant of RCC of low grade with a reported incidence of 2.3-2.9% of renal neoplasms and 3.5 - 6% of RCC.<sup>1,2,6</sup> In 1957, Robinson described the first case of the "so" called MCRCC containing clear epithelial cells<sup>3</sup>. In 1998, Eble et al suggested following diagnostic criteria for MCRCC 1) an expansile mass surrounded by a fibrous capsule. 2) interior of tumour entirely composed of cysts and septae with no expansile nodule or solid component confined to less than 10% of entire tumour and 3) septa containing aggregates of clear epithelial cells. These criteria distinguish MCRCC from conventional RCC with extensive cystic change.<sup>1,3,4</sup>

## Case Report

A fifty year old male had frequent loose stools since 10-12 days and pain in loin off and on, since last 2 months. His symptoms were not relieved with local treatment hence, ultrasound of abdomen and pelvis was done which revealed right renal calculus and an echogenic mass 2-3 cm in diameter at midpole of left kidney. There was no history of haematuria or any systemic illness in the past. CT scan was suggestive of Angiomyolipoma for which nephrectomy was done.

## Pathological findings

**Gross:** We received left nephrectomy specimen

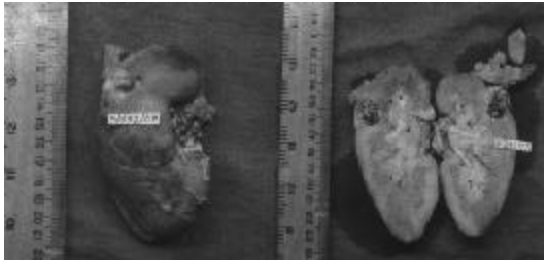
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measuring 10 x 6 x 3 cm along with perinephric fat. Capsule could be stripped off easily. At midpole a nodule was seen. On cut section, there was a well circumscribed, spheroidal yellow to brown nodule measuring 2.6 x 2 x 2 cm in the cortical area with sieve-like locules filled with haemorrhagic, gelatinous fluid (Fig. 1).

**Micro:** Sections from kidney revealed an encapsulated, multicystic tumour with cysts lined by single to multiple layers of epithelial cells having uniform nuclei and eosinophilic to clear cytoplasm. Few nuclei showed irregular nuclear contours with prominent nucleoli - Furhman grade I to II (Fig. 2). Few pseudopapillary areas were seen. The cysts were separated by septae showing clusters of clear cells. The cysts contained proteinaceous fluid and macrophages. No extracapsular extension was noted. The diagnosis of MCRCC - Grade I-II (Furhman grading system), Stage Ia (AJCC/TNM classification) was rendered.

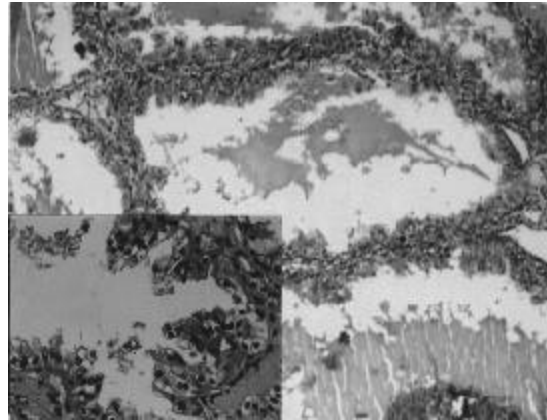
## Discussion

According to the literature, the tumour is incidentally found in 67% of cases and is reported to occur in age range of 33-68 years with a mean of 46 years and a male preponderance, ratio being 3:1.<sup>3</sup> Our case presented at the age of 50 and was detected incidentally. Cystic change may be seen in many renal lesions hence, it is essential to differentiate MCRCC from simple cysts, multilocular cystic nephroma, RCC with cystic degeneration, papillary cystic RCC and clear cell carcinoma.<sup>1</sup> Radiology, cytology and



*Fig. 1 :Gross photograph to show renal cortical tumour at midpole. Cut section shows a well encapsulated yellow to brown mass measuring 2.6 x 2 x 2cm with sieve-like locules filled by haemorrhagic/gelatinous fluid.*

frozen sections are unable to differentiate these tumours.<sup>1</sup> Ultrasonography is a useful screening test but computed tomography and magnetic resonance imaging is the study of choice which differentiates MCRCC.<sup>5</sup> On ultrasonography, tumours are hyperechoic with contrast enhancement limited to solid portions. However, small tumours appear solid and give contrast enhancement due to slight neovascularity.<sup>7</sup> On imaging study MCRCC show multiple cysts with regular, thin septae and no expansile nodule. In contrast cystic RCC show cysts with thick, irregular septa and an expansile nodule 5 mm or more in diameter.<sup>6</sup> MCRCC have characteristic gross and microscopic appearance with a mean tumour size of 3.4 cm.<sup>2</sup> In the presenting case, the size of tumour was 2.5 cm and gross and microscopic criteria for diagnosis were fulfilled. Cystic nephromas may simulate MCRCC but close attention to epithelial lining and fibrous cyst wall leads to correct diagnosis. Cystic nephromas are lined by a single layer of flattened or hobnail epithelium with occasional clear cell and stroma is fibrous, cellular and ovarian type. Septa contains small tubules lined by bland epithelial cells reminiscent of renal tubules.<sup>4</sup> The distinguishing features separating MCRCC from other tumours having similar



*Fig. 2 :Microphotograph shows renal tumour composed of multiple cysts lined by single to multiple layers of epithelial cells with clear cytoplasm and uniform nuclei. (H and E x 10). Inset shows cells having nuclei of Fuhrman grade I-II.(H and E x 40.)*

morphology are summarized in Table 1. The tumour is reported to have no recurrence or metastasis.<sup>1</sup> Since nuclear grade is low (Fuhrman grade I-II), prognosis is good.<sup>2,3</sup> Due to its low malignant potential, nephron-sparing surgery gives good results.<sup>1,5,6</sup>

### **Conclusion**

It is important to differentiate this tumour as it has a low malignant potential thereby carries extremely good prognosis and is amenable to nephron-sparing surgery. Pre-operative recognition of MCRCC is possible using computed magnetic resonance imaging criteria thereby proposing a conservative surgery.

### **References**

1. Tariq Murad, William Komaiko, Ryoichi Oyasu. Multilocular cystic RCC. *Am J Clin Pathol* 1991; 95 : 633-37.
2. Sternberg M. Adult Renal tumours. In: *Diagnostic Surgical Pathology* vol. II, III<sup>rd</sup> ed, Stephen Silverberg Lippincott, William and Wilkins, 17942.
3. Jong Chul Kim, Kie Hwan Kim, Jon Woo Lee. CT and US findings of multilocular cystic RCC. *Korean Journal of Radiology* 2000; 1(2) : 104-9.

**Table 1**

	MCRCC	RCC with cytic change	Papillary cystic RCC	Clear cell earcinoma
Gross	Multicystic with focal solid areas	Solid with focal degenerative cysts	Multifocal, variegated app	Solid, golden yellow, sarcomatoid-areas
Capsule	Well defined	Poorely defined	Pseudocapsule	III defined
Cysts	Numerous with thin septae	Few, as minor component	Few	Cysts in 12%
Content of cysts	Clear or haemorrhagic, macrophages +	Turbid, necrotic material	Necrotic material	Clear Fluid
Septation	Frequent, fibrohyalinised	Rare	Rare	Delicate, fibrovascular
Cellular organisation	Single layer of clear cells or macrophages lining septae, focal areas of clearcells in septal wall	Solid, sheets with tubules	Discrete papillary fronds, fibrovascular core with foamy macrophages	Solid sheets, acinar Papillary.
Cell types	Clear	Clear or granular	Amphophilic, basophilic	Clear, spindle
Nuclear grade	I - II	I-III	I-II	I-IV

- Eble JN, Bonsib SM. Extensively cystic renal neoplasms. *Semin Diag Pathol* 1998; 15(1) : 2-20.
- Nassir A, Jollemin J, Gupta R. Multilocular cystic RCC: a series of 12 cases and review of literature. *Urology* 2002; 60(3) : 421-27.
- Aubert S, Zini L, Delomez, *et al.* Cystic renal carcinoma in adults. *J Urol* 2005; 174 (6) : 2115-19.
- Yamashita Y, Miyazaki T, Ishii A, *et al.* Multilocular cystic RCC presenting as a solid mass: radiologic evaluation. *Abdominal imaging. Springerlink Jour* 1995; 20(2) : 64-168.

### Apology

In an article "Neuroendoscopy" by Vishwanathan Iyer et al published in Bombay Hospital Journal's Special Issue on Laparoscopic Surgery Vol. 2005; 47 (4) : 372-4, the artwork on page 373 is inadvertently published, though it is a copyright of Barrow Neurological Institute 2002. We sincerely regret the error and apologies for the same.