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. In 1957, Robinson described the first case of the “so” called MCRCC containing clear epithelial cells. 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.

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 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. 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. Radiology, cytology and
frozen sections are unable to differentiate these tumours. Ultrasonography is a useful screening test but computed tomography and magnetic resonance imaging is the study of choice which differentiates MCRCC. 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. 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. MCRCC have characteristic gross and microscopic appearance with a mean tumour size of 3.4 cm. 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. The distinguishing features separating MCRCC from other tumours having similar morphology are summarized in Table 1. The tumour is reported to have no recurrence or metastasis. Since nuclear grade is low (Furhman grade I-II), prognosis is good. Due to its low malignant potential, nephron-sparing surgery gives good results.

**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**

### Table 1

<table>
<thead>
<tr>
<th>Gross</th>
<th>MCRCC</th>
<th>RCC with cystic change</th>
<th>Papillary cystic RCC</th>
<th>Clear cell carcinoma</th>
</tr>
</thead>
<tbody>
<tr>
<td>Capsule</td>
<td>Well defined</td>
<td>Poorly defined</td>
<td>Pseudocapsule</td>
<td>III defined</td>
</tr>
<tr>
<td>Cysts</td>
<td>Numerous with thin septae</td>
<td>Few, as minor component</td>
<td>Few</td>
<td>Cysts in 12%</td>
</tr>
<tr>
<td>Content of cysts</td>
<td>Clear or haemorrhagic, macrophages +</td>
<td>Turbid, necrotic material</td>
<td>Necrotic material</td>
<td>Clear Fluid</td>
</tr>
<tr>
<td>Septation</td>
<td>Frequent, fibrohyalinised</td>
<td>Rare</td>
<td>Rare</td>
<td>Delicate, fibrovascular</td>
</tr>
<tr>
<td>Cellular organisation</td>
<td>Single layer of clear cells or macrophages lining septae, focal areas of clearcells in septal wall</td>
<td>Solid, sheets with tubules</td>
<td>Discrete papillary fronds, fibrovascular core with foamy macrophages</td>
<td>Solid sheets, acinar Papillary.</td>
</tr>
<tr>
<td>Cell types</td>
<td>Clear</td>
<td>Clear or granular</td>
<td>Amphophilic, basophilic</td>
<td>Clear, spindle</td>
</tr>
<tr>
<td>Nuclear grade</td>
<td>I - II</td>
<td>I-III</td>
<td>I-II</td>
<td>I-IV</td>
</tr>
</tbody>
</table>


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### 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.