Multilocular Cystic Renal Cell Carcinoma: A report of two cases with a rare feature

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Citation

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Abstract

Multilocular cystic renal cell carcinoma (MCRCC) is a relatively are cystic tumor of the kidney. Because of its distinct characteristics in prognosis and natural history, MCRCC was recognized as a separate subtype of renal cell carcinoma in the 2004 WHO classification of adult kidney tumors.[1]We report two cases of MCRCC, one from antemortem surgical specimen and the other from the post mortem (autopsy) which was an incidental finding of smallest size which is a rare occurrence and probably the first reported case.

INTRODUCTION

Multilocular cystic RCC is a distinct subtype of clear-cell RCC and appears to have a favorable biology. These tumors are a rare entity, comprising approximately 1 to 2% of all renal tumors. [2] We report two cases of MCRCC, one from antemortem surgical specimen and the other from the post mortem (autopsy) which was an incidental finding of smallest size which is a rare occurrence. To the best our knowledge, this is probably the first reported case.

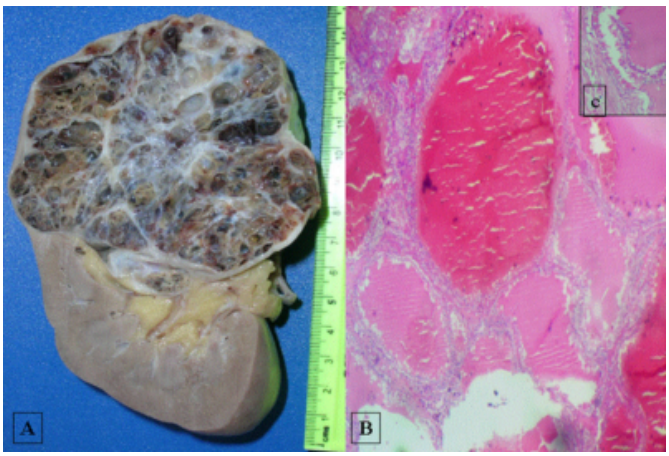
CASE HISTORY

Case 1: A 55-year-old male presented with left flank pain and hematuria since 2 months. Ultrasound revealed a mass in upper pole of the left kidney. A left nephrectomy was carried out. Grossly, the specimen measured 18 × 10 × 10 cms which included the tumor of 9 cms diameter which was well circumscribed and encapsulated. The renal capsule was grossly free. The cut section was multiloculated, cystic. The cysts ranged from 2 mm to 2 cm. The content of the cyst included clots and gelatinous material. Microscopically, the section showed a multicystic tumor. The cysts were separated by fibrocollagenous connective tissue lined by clear cells. The clear cells had small hyperchromatic nuclei with mild anisonucleosis and inconspicuous nucleoli. Renal capsule, vessels, and perinephric fat were free. The histomorphology was compatible with multilocular cystic renal cell carcinoma, Fuhrman nuclear grade I. After 2 years follow up, the patient is well with no recurrences.

Figure 1A: Gross of kidney mass showing cystic tumor with multiloculations and no solid foci seen.

Figure 1B: Microscopy of the cytic mass showing cysts lined by clear cells. Inset(c) shows high power of the cyst lined by clear cells.

Figure 1

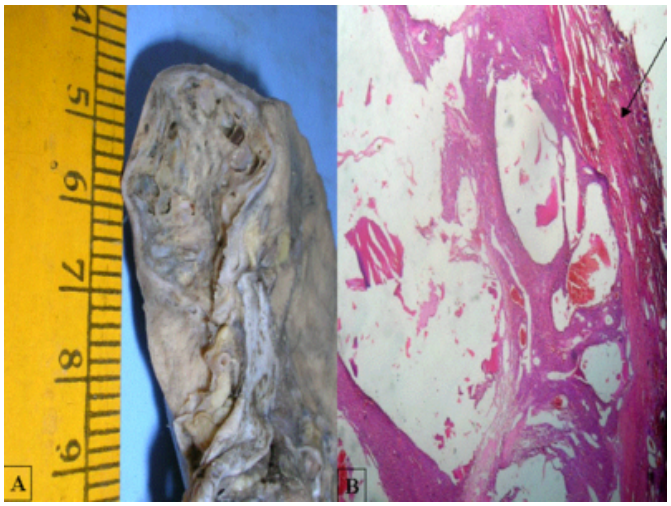


Case 2: A 40 year old male admitted with perforative peritonitis. Emergency exploratory laporotomy was done and patient subsequently expired 2 days later. A complete autopsy was done. The gross and microsopic findings of all the organs were studied and septicemia was given as final cause of death. During the gross evaluation of all the organs, we found a mass in the kidney of size 2.2cm x1.5cms. The cut section was multiloculated and cystic, however contents of cyst was clear fluid. There was no blood or clot seen. The histomorphology was compatible with multilocular cystic renal cell carcinoma.

Figure 2 A: Gross of the kidney mass which is 2.2cms(scale) and multicystic appearance.

Figure 2 B: Microscopy shows compressed renal parenchyma (arrow) and cysts separated by the fibrocollagenous tissue.

Figure 2



DISCUSSION

Multilocular cystic RCC is a distinct subtype of clear-cell RCC and appears to have a favorable biology. These tumors are a rare entity, comprising approximately 1 to 2% of all renal tumors.[2] Renal cell carcinoma (RCC) accounts for 80% to 85% of primary malignant neoplasms of the kidney.[3] It usually presents as a solid mass; however, in 10% to 22% of cases, RCC appears as a unilocular or multilocular cystic mass on imaging studies.[3]Four mechanisms have been described to account for renal cell carcinomas with cystic features: intrinsic unilocular cystic growth (papillary cystic adenocarcinoma), intrinsic multilocular cystic growth, tumor necrosis resulting in cyst formation (pseudocyst), and tumor arising in a preexisting simple renal cyst.[3]

The 2004 WHO classification of kidney tumors recognizes multilocular cystic renal cell carcinoma (MCRCC) as a rare variant of clear-cell RCC with a good prognosis.[1]While cysts are common in clear cell renal cell carcinoma, only rarely is the tumor entirely composed of cyst. The term MCRCC should be used exclusively to identify cystic RCC with a small volume (25% or less) of neoplastic clear cells in the cyst walls.[4]

The differential diagnosis of a multilocular cystic renal mass includes numerous cystic diseases of the kidney, most of which are easily excluded based on CT findings.[5] However, multilocular cystic nephroma, formed by multiple separate cysts (which are also known as multilocular cyst) is a rare benign tumor of the kidney, frequently cannot be distinguished from multilocular cystic RCC by imaging. Microscopic examination of surgically resected tissue is often necessary to distinguish between these two neoplasms. Microscopically, multilocular cystic nephroma demonstrates multilocular cysts lined by a single layer of flattened, low cuboidal epithelium without nuclear atypia. The cytoplasm is usually eosinophilic and the fibrous septae within cyst wall is cellular resembling ovarian stroma and may contain mature tubules. In this case, clear cells must not be found in the walls and the intercystic stroma. The cystic nephroma does not have any relation to the multilocular clear cell carcinoma (despite certain similarity with it) [1]Multilocular cystic renal cell carcinoma is distinguished from the other subtypes of RCC based on the high incidence of stage I disease at initial presentation (83% to 88%), infrequent metastases, and an extremely high cure rate following surgical resection of the tumor.[1,6] To rename this tumor as multilocular cystic renal cell neoplasm of low malignant potential might help urologists approach the patients conservatively.

The second case was a smallest MCRCC, incidental renal mass discovered during autopsy. Autopsy results have shown that approximately 50 percent of persons older than 50 years have one or more renal cysts.[7] Other studies indicate that almost one third of persons in this age group have at least one renal cyst that is identifiable on a CT scan.[7] Most of these lesions are benign simple cysts that require no further evaluation, intervention or urologic consultation.

The renal cortical adenomas are detected at autopsy with an incidence of 1-2%. The size of the lesion cannot be used as criterion to distinguish between benign and malignant., because,it is well known that some of the small clear-cell tumors have metastasis capacity and therefore currently the existence of an adenoma of clear cells is not accepted, instead it is considered that all the clear-cell tumors are carcinomas, with greater or lesser aggressiveness irrespective of size provided there is clear cells present. Among the papillary neoplasms the WHO 2004[1]renal cell tumors classification are considered as papillary adenomas tumors with a maximum diameter of 5 mm and may represent a continuum biological process to papillary renal cell carcinoma. Our second case, though was a cystic mass of small size, the cysts were lined by the clear cells, which was similar to the first case. The literature on MCRCC

reports the average diameter ranging from 3.5 cms – 13 cms[1] and there were no reports on incidentally detected MCRCC. Hence our second case was a rare occurrence detected at autopsy and is probably the first case of incidental MCRCC of smallest size.

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