



Cystic Renal Cell Carcinoma – Sometimes A Diagnostic Dilemma: A Case Report on Rare Multilocular Cystic Renal Cell Carcinoma

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Abstract:

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Cystic renal tumors often create diagnostic dilemma. It might be a cancer in a complex cyst, cystic degeneration of a renal cell carcinoma, Papillary cystic renal cell carcinoma or multilocular cystic renal cell carcinoma (MCRCC). MCRCC is a rare cystic tumor of the kidney. It is recognized as a separate subtype of renal cell carcinoma in WHO classification of adult renal tumors in 2004 with excellent prognosis. A 47 years old male presented with an incidentally detected large right renal cystic lesion in the upper and hilar region. After discussion with the patient about the possibilities, right radical nephrectomy was performed. Histopathology was done by one of the renowned Pathologists of the country and was reported as a rare variety Multilocular Cystic Renal Cell Carcinoma. It is very important to differentiate this type of cystic renal cell carcinoma from cystic degeneration of conventional RCC.

Introduction:

Cystic renal tumors or complex renal cysts frequently misdiagnosed and creates diagnostic dilemma. As Urologists, we would like to know the exact diagnosis from our Pathologist Colleagues to explain the prognosis to our patients. For example, multilocular cystic renal cell carcinoma has an excellent prognosis, reported as rare variety and cured by simple nephrectomy.¹ On the other hand, cystic degeneration of conventional RCC with necrosis and sarcomatoid changes has worst prognosis.

Cystic renal disease is based on Bosniak Classification system.^{2,3} But some renal cell carcinomas like multilocular cystic renal cell carcinoma sometimes looks like simple renal cysts or like Bosniak type 1 or type 2 cysts. Multilocular cystic renal cell carcinoma may be mistaken for cystic degeneration of clear cell carcinoma.⁴ MCRCC comprises only 1-2% of all renal tumors.⁵ There are other types of renal tumors from which MCRCC has

to be differentiated like – cystic nephroma, cystic clear cell carcinoma, clear cell papillary renal cell carcinoma and tubulocystic carcinoma. Immunohistochemistry (IHC) helps to reach exact diagnosis in many cases.

Case Report: A 47 years old male presented with an incidental finding of a mass in right kidney on Ultrasonogram. It was reported on Ultrasonogram as a mixed echogenic cystic lesion of about 7.8x6.9 cm in the upper medial aspect of right kidney. There was no significant family history or signs or symptoms suggestive of Von Hippel Lindau syndrome or renal tumors of any kind. All other biochemical investigations including renal function were normal. There was no significant finding on general or systemic examination. Contrast enhanced CT abdomen and Pelvis (Fig: 1) revealed a fairly large (8.6x6.73 cm) mass in the right renal pelvic region. Density of the mass at the center was -3HU and at the periphery 16 HU. Post contrast scan showed no significant enhancement. The

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mass deformed collecting system and pushed the right renal vein upwards. Renal vein and IVC were free of tumor. There was no lymphadenopathy.

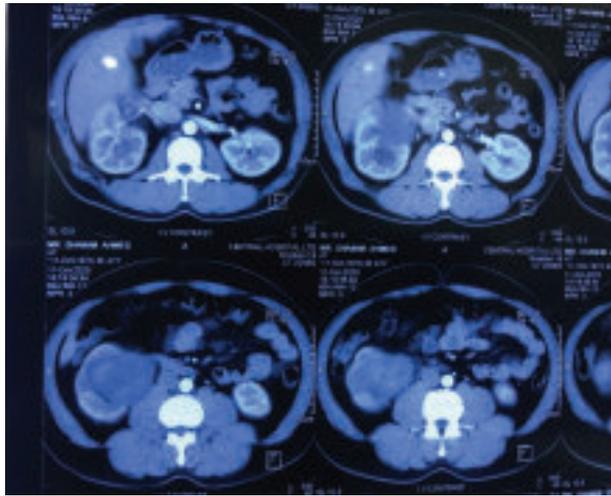


Fig: 1: CT scan of the patient

No mural nodule or calcification noted in the cyst. Considering the size and location of the cystic lesion, I planned for right nephrectomy with arrangement of frozen section. But when I exposed the hilum and observed extension of cystic lesion with chocolate colored cyst fluid, upward stretching of renal vein, I performed right radical nephrectomy without frozen section. No lymphadenopathy, renal vein or IVC thrombus was noted. There was no visceral metastasis or ascites.

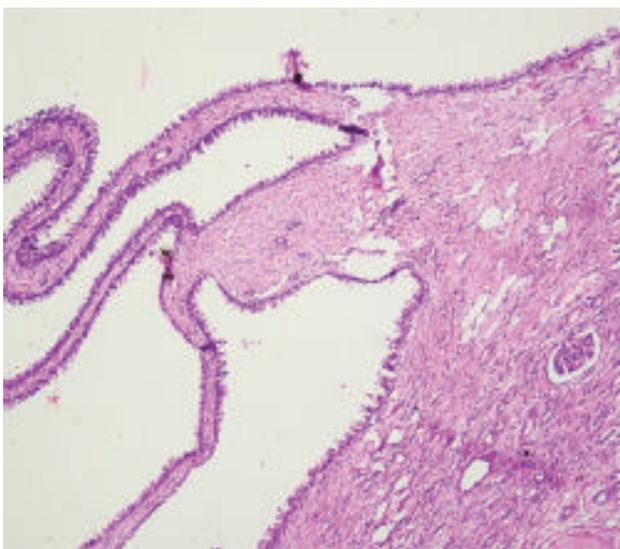


Fig.-3a: Photomicrograph of Morphology

Excised specimen showed thick wall cystic structures containing thick chocolate brown mucinous fluid. No definite solid component to tumor or tumor necrosis seen. Gross (of Histopathology) was reported – cut surface of the kidney showed (Fig: 2) multilocular cysts. The largest one measured 5x4 cm containing dark brown material.



Fig: 2: Cut surface of kidney

Microscopic examination (Fig: 3a & 3b) revealed many cystic spaces lined by clear cells. Some of these cells showed stratification and prominent nucleoli. Neither necrotic area nor expansile nodule was appreciated. Some foci revealed intra cystic papillary structures.

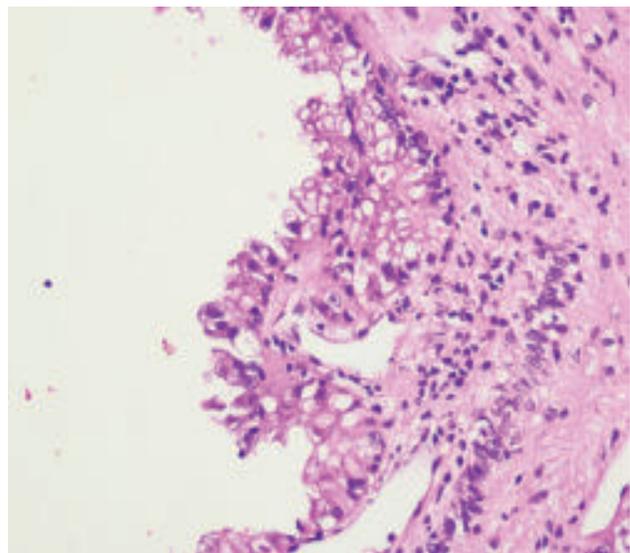


Fig-3b: Photomicrograph of tumor morphology

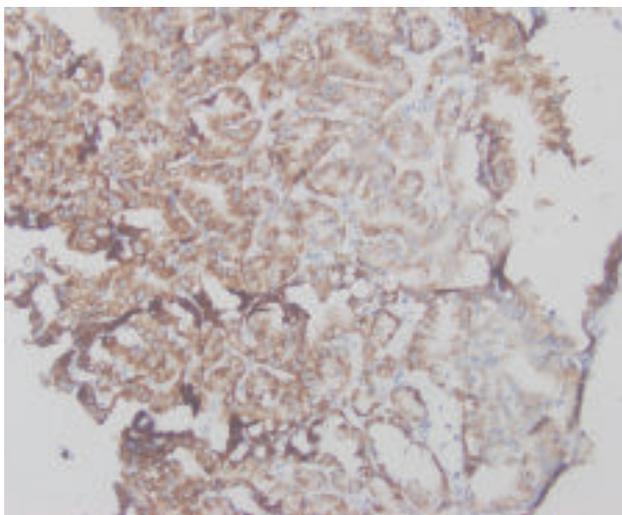


Fig: 4a: AMACR IHC slide

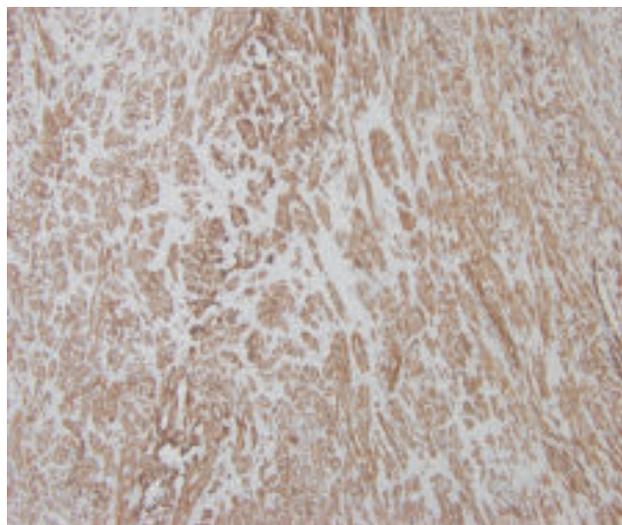


Fig: 4b: Vimentin IHC slide

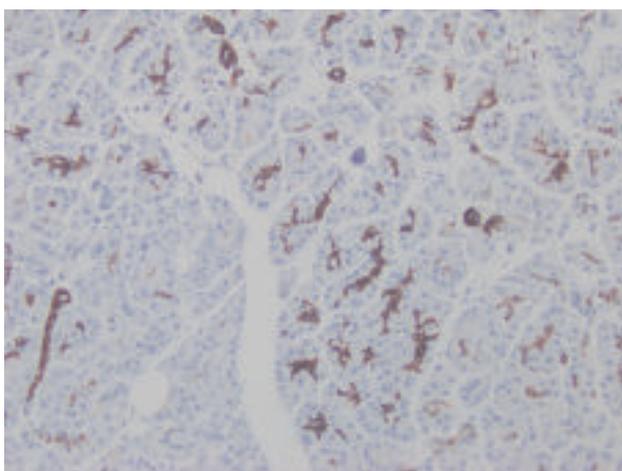


Fig: 4c: CK7 IHC slide

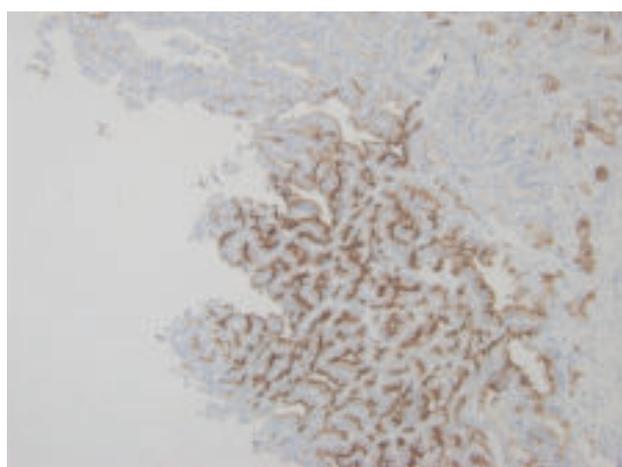


Fig: 4d: CD10 IHC slide

No capsular or lymphovascular invasion seen. Renal pelvis, ureter, renal vessels, perinephric fat were free of tumor. Feature suggestive of histopathological diagnosis of Multilocular Cystic Renal Cell Carcinoma (MCRCC).

Immunohistochemistry (IHC) was requested from a renowned institute of Dhaka and it was reported as – AMACR, Vimentin, CK7, CD 10 (Fig: 4a, 4b, 4c, 4d) all positive for tumor cells, which goes more in favor papillary renal cell carcinoma (PRCC). But when IHC slides were reviewed by two renowned Pathologists from North America, it was reported as CK7 and Vimentin negative. Immunostains were not conclusive in this case and not typically fitted either with MCRCC or PRCC.

So, gross morphology was taken into consideration and reviewed by two North American Anatomical Pathologists and was reported as Multilocular Cystic Renal Cell Carcinoma. MCRCC typically low-grade tumor. In our case, TNM stage was T2aN0M0 and Fuhrman nuclear grading was grade 2.

Discussion:

Cystic degeneration of kidney tumors and renal cysts are very common renal lesions. However, MCRCC is a relatively rare condition.⁶ Sometimes it is difficult to make diagnosis and provide treatment of MCRCC. Because MCRCC and benign renal cystic disease have similar manifestations and imaging characteristics. A retrospective study (by Zhang and his group) analyzed 13 patients with MCRCC and among those, 2 cases

completely looked like benign cysts on preoperative imaging, were diagnosed as MCRCC at frozen section. Those two cases underwent simple nephrectomy later on. Morphology and immunohistochemistry of those 2 cases confirmed MCRCC.⁷ As these tumors are low malignant potential and have excellent prognosis, they recommended simple or partial nephrectomy for tumors of favorable size and location.⁷

Our patient had a hilar cystic lesion and more than 8 cm size with distorted renal vein and calyceal system. We decided to go for radical nephrectomy. Intraoperative frozen section is recommended in all cases of cystic lesions by some authors; because benign looking cysts sometimes ended up in MCRCC on frozen section of excised capsule. During follow up (range 6-60 months), no tumor recurrence or metastasis occurred in their series.⁷

Careful inspection of imaging, many a times, gives clue for diagnosis of this type of cystic lesions. Typical B ultrasonography images are: Cystic echo-free masses with hyperechoic septa, thick capsular walls and several hyperechoic nodules attached to septa.⁷ CT scan can show thick and irregular capsule walls surrounding the cysts with hyperdense septa and nodules. In enhanced images, capsule walls, septa and nodules might show intense early enhancement.¹⁰ There might be coarse and crescent calcification in the capsule wall, septa and nodules. Septa tends to be uneven thickness (often >1 mm in diameter) and nodular thickening can appear at junctions of capsular walls. Hydatid fluid contains debris, floc and blood clots, which appear uneven on CT scan. They usually have unclear border adjacent to renal parenchyma.⁷ However, some MCRCC, have similar characteristics to those of simple cysts.⁷ Preoperative FNAC (Cyst puncture cytology) can assist diagnosis theoretically.⁷ low positive rate limits its use in clinical settings.⁷ Intraoperative frozen section is more accurate for diagnosis.⁷

Nephrectomy (radical or simple or partial) is the most effective treatment for MCRCC.⁷ Corica and his group compared surgical approaches and their results showed- the best approaches were radical and partial nephrectomy.⁹ Nephron sparing surgery is suitable for small polar lesions, patients with contralateral renal insufficiency or for solitary kidney. Five years survival was reported 100% for MCRCC.¹⁰

On imaging, 10-22% RCC appear unilocular or multilocular cystic mass.¹⁰ Four types of RCC with cystic features have been described – intrinsic unilocular cystic growth (Papillary cystic carcinoma), Intrinsic multilocular cystic growth, tumor necrosis results in cyst formation (Pseudocyst) and tumor arising from preexisting simple renal cyst.⁴ The term MCRCC should be used exclusively to identify cystic RCC with a small volume of (25% or less) of neoplastic clear cells in the cyst walls. WHO criteria of diagnosis for MCRCC⁴ are - Circumscribed, noncommunicating, expansile nodule composed entirely of cysts and septa. Cysts should be lined by single layer of low-grade clear cells. No papillary growth, cysts separated by fibrous septa. Septa contains low grade clear cells. These groups must not be expansile nodules and must not show infiltrative growth. Nuclei should be low grade (Fuhrman grade 1 or 2). Presence of any expansile nodules of clear cells in the septa between cysts, indicate an ordinary fully malignant, clear cell carcinoma with cystic changes. Mean age for MCRCC is 51 years and M:F ratio is 3:1.¹

Conclusion

Multilocular Cystic Renal Cell Carcinoma (MCRCC) is a rare cystic special type of renal cell carcinoma.¹⁰ Sometimes it is very difficult to diagnose MCRCC because of its clinical manifestations and imaging characteristics.⁶ It can be mistaken as a simple renal cyst even. As Urologists, we need to know the exact type of RCC to explain the prognosis to the patients. This case was reported because of its rarity, diagnostic dilemma that raised and also to avoid misdiagnosis as conventional clear cell RCC which has worse prognosis. Multilocular Cystic Renal Cell Carcinoma should always be kept in mind in case of cystic renal disease. Immunohistochemistry plays an important role in diagnosis of renal tumors and should be advised for such cases. Per operative frozen section should be considered by the Urologists for proper planning of Surgery. Both morphology and Immunohistochemistry should be advised for exact diagnosis.

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