CASE REPORT

Cystic renal cell carcinoma – rare clinical finding Radiographic variations of tumor/cyst appearance and further diagnostic work-up

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Abstract

Objectives: **To review the cases of cystic renal cell carcinoma and multilocular cystic nephroma, point out the radiographic variations and define further diagnostic work-up.**
Materials and methods: **Between 2003 and 2005 5 patients with suspected cystic renal cell carcinoma were treated surgically (1 pt underwent radical nephrectomy, 1 pt laparoscopic cyst decortication, 3 pts ablation), 2 patients with multilocular cystic nephroma underwent ultrasound guided biopsy.**
Results: **Histopathologic examination confirmed cystic renal cell carcinoma (CRCC) T1aN0M0 Fuhrman grade 1 in 3 cases, T1bN0M0 Fuhrman grade 2 in one case. One patient with suspected tumor inside the cyst wall who underwent laparoscopic cyst decortication was excluded (final histology confirmed organized hematoma in the cyst wall). Biopsy in 2 patients with multilocular cystic nephroma did not confirm the presence of malignant cells. The mean tumor size was 4.2 cm (range 3.7 to 5.5) for CRCC and 4.7 cm (range 4 to 4.5 cm) for multilocular cystic nephroma. All 4 cases of CRCC were clear cell type.**
Conclusion: **In conclusion according to the data described and from our study, tumor/cyst co-existence requires further surgical exploration in group 2, 3, 4. Small cystic renal cell carcinomas up to 4 cm in diameter have usually favourable pathology and prognosis, which offers the minimally invasive nephron-sparing treatment options such as excision, ablation or partial nephrectomy *(Fig. 9, Ref. 18)*.**
Key words: cystic renal cell carcinoma, multilocular cystic nephroma, cystic lesions.

According to the data the incidence of CRCC is approximately 4 % to 15 % of renal cell carcinomas (1, 2, 3). Cystic renal cell carcinoma includes any malignant neoplasm of renal tubular epithelium which presents as a fluid-filled mass (1).

CRCC may arise by intrinsic cystic growth ab initio- multiloculated or uniloculated, by cystic necrosis or by development of adenocarcinoma in the wall of a pre-existing simple cyst. There are three basic radiologic patterns of cystic CRCC: unilocular cystic mass, multiloculated cystic mass, and discrete mural nodule in a cystic mass (1).

Gibson's classification of tumor/cyst co-existence describes 4 appearances: 1), cancer and cyst distant from each other, 2) cyst arising in a tumor, 3) tumor arising in the cyst wall and 4) cyst distinct from cancer, but sufficiently for a possible pathologic relation (4).

At ultrasound, cystic renal cell carcinomas may be mistaken for simple cysts, unless the criteria for the diagnosis of a simple cyst are not fulfilled. Solid components, wall thickening or internal echoes should give a suspicion of malignancy. In these cases CT and MRI are neccessary to perform.

The aim of our study was to point out some radiographic variations of tumor/cyst appearance and define further diagnosic work-up.

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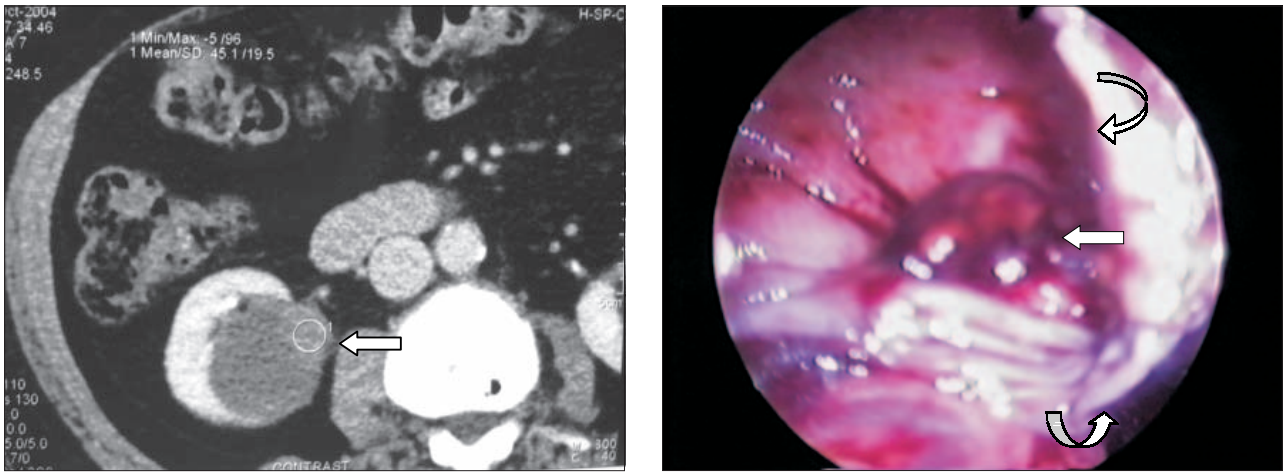

Fig. 1. Solid component located inside the wall of the right renal cyst (Bosniak type III), perioperative finding and final histology – organized hematoma – white arrow, cyst wall- - curved arrows.

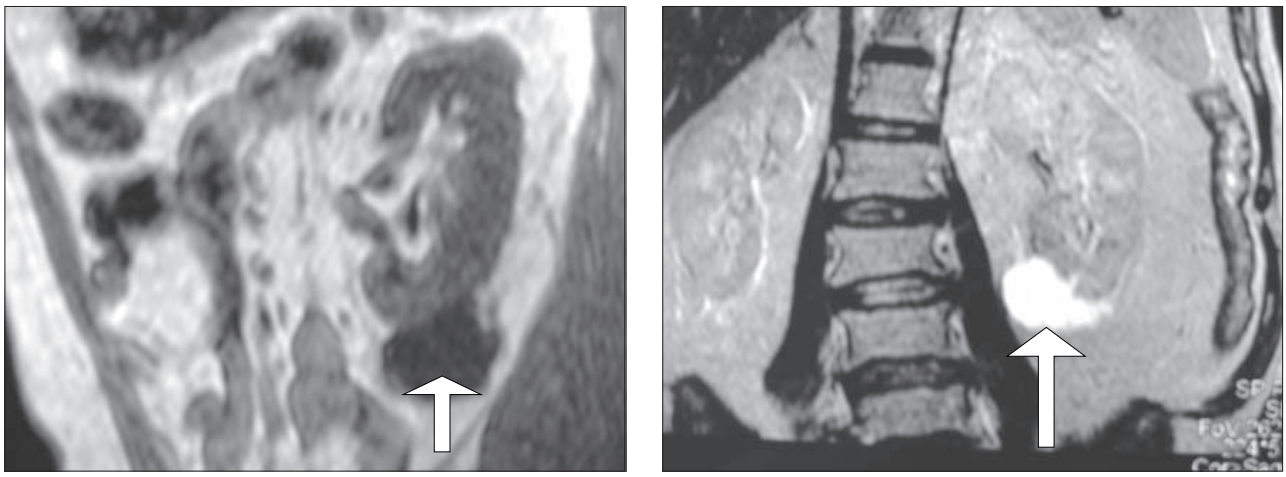

Fig. 2, 3. Multilocular cystic nephroma – lower pole of the left kidney, patient underwent biopsy under ultrasound guidance, histology – negative for cancer, the lesion needs to be followed-up.

Material and methods

Between 2003 and 2005 5 patients with suspicious cystic renal cell carcinoma were treated surgically (1 pt underwent radical nephrectomy, 1 pt laparoscopic cyst decortication, 3 pts ablation), 2 patients with multilocular cystic nephroma underwent ultrasound guided biopsy. All cases were solitary, detected as suspected renal cystic tumor lesions on ultrasound and computed tomography with pathological enhancement features. Tumor stage was assessed according to the EAU guidelines (2001) and tumor nuclear grade with Fuhrman's grading system.

Results

Histopathologic examination confirmed cystic renal cell carcinoma (CRCC) T1aN0M0 Fuhrman grade 1 in 3 cases, T1bN0M0 Fuhrman grade 2 in one case. Biopsy in 2 patients with multilocular cystic nephroma did not confirm the presence of malignant cells. The mean tumor size was 4.2 cm (range 3.7 to 5.5) for CRCC and 4.7 cm (range 4 to 4.5 cm) for multilocular cystic nephroma. Our study group included 7 male patients, mean age 62.3 years (range 59 to 67), one patient with suspected tumor inside the cyst wall who underwent laparoscopic cyst decortication was excluded (final histology confirmed organized hematoma in the cyst wall) (Fig. 1). All 4 cases of CRCC were clear cell type. All lesions were incidentally detected.

Discussion

True CRCC must be distinguished from conventional RCC with extensive necrosis or haemorrhage that could result in a false cystic appearance (5). It tis not enough to find cystic spaces filled with fluid on US, CT or gross examination. True cysts covered by neoplastic cells must be identified histologically and cases

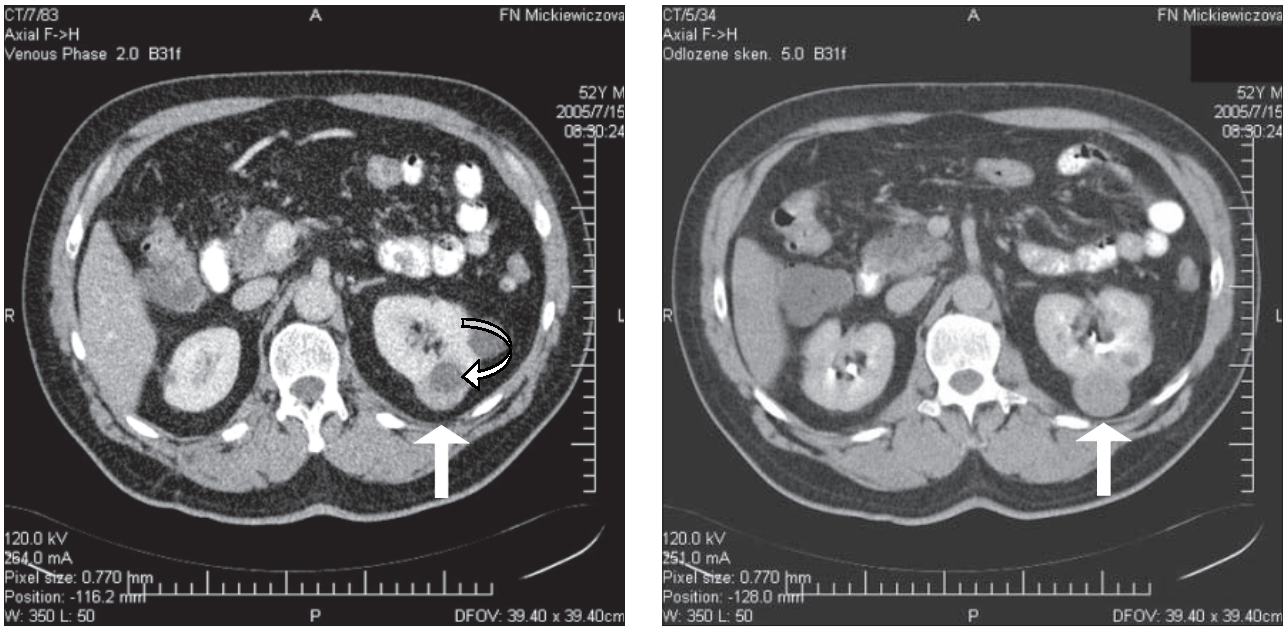

Fig. 4, 5. Tumor of the left kidney (before contrast 35-40HU, after contrast 70 HU – white arrow), simple cyst in the intimate possition with the tumor – curved arrow. Histology – RCC Fuhrman gr. 3. T1bN0M0, there was no infiltration of the cystic wall.

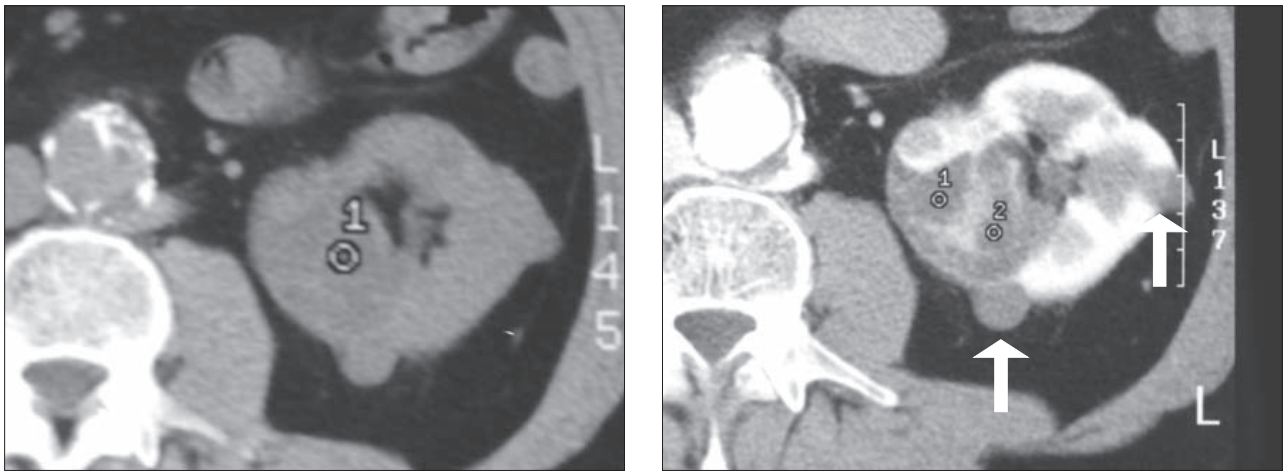

Fig. 6, 7. Cystic tumor of the left kidney (before contrast 20HU, after contrast material infusion 47,29 HU), histology – cystic clear cell renal Ca, Fuhrman nuclear gr. 1, T1bN0M0. Simple cortical cyst – white arrows.

with superimposed haemorrhage or necrosis must be excluded (6, 7). The presence of haemorrhage inside the cystic cavity of CRCC was reported only in few cases (8, 9).

Bielsa et al (3) identified 12 % (25 pts) of CRCC cases from 206 patients with RCC (multilocular in 92 %, unilocular in 4 %). There was no difference in clinical manifestation between CRCC and RCC proportion of cases, however presence of asthenia, anorexia and weight loss were much more unusual in CRCC.

In differential diagnosis of CRCC we have to take into account: 1) multilocular cystic nephroma (Fig. 2, 3), 2) radiographic finding of pseudocystic foci in RCC and (3) the presence of haemorhagic or secondary infected cysts.

Tumor/cyst coexistence have been previously described by several authors (4, 9, 10). Gibson's classification describes 4 appearances: 1), cancer and cyst distant from each other, 2) cyst arising in a tumor, 3) tumor arising in the cyst wall and 4) cyst distinct from cancer, but sufficiently for a possible pathologic relation (4). Group 3 – is highly suspected cystic lesion for malignancy, according to Bosniak classification system it corresponds to Bosniak 3 category (lesion that requires surgical exploration or US/CT guided biopsy (11, 12)) (Fig. 1). Group 4 may correspond to different phases of the same process: the tumor initially induces obstruction of a tubule or an artery, leading to the formation of a cyst [10]. In this situation only a final his-

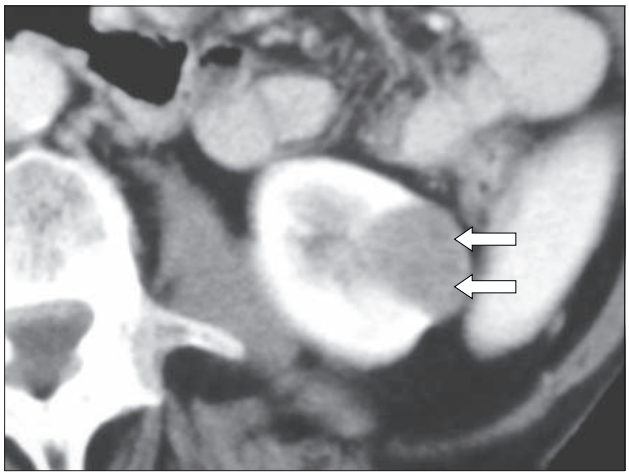

Fig. 8. Solid tumor of the left kidney, with pathological enhancement (>20 HU), susp. cystic foci-white arrows, histology – multiloculated cystic renal cell Ca, Fuhrman grade 1.

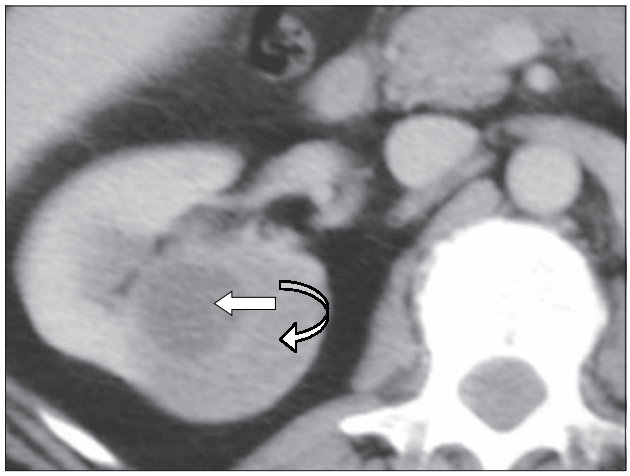

Fig. 9. Hyperdense cyst Bosniak type IIF, after 10 yrs of follow-up no signs of progression, simple cyst – white arrow arising inside the complex cyst – curved arrow.

topathological examination will confirm the presence or absence of malignant cells (Fig. 4, 5).

Masses that appear predominantly cystic should be classified according to the Bosniak system (13). Attenuation coefficients of renal masses should be evaluated on both pre-enhancement and postenhancement CT images.

Collimation should be 10 mm or less, and patients in whom a renal abnormality is specifically suspected, 5 mm scan collimation should be used. If the baseline units exceed 20 HU, thin collimation (3–5 mm) should be used to minimize partial volume artifacts (14).

It is of major importance to measure attenuation coefficients in multiple locations of suspected CRCC lesion, otherwise in case of measurement in only one location of postenhancement CT scan we can possibly downstage the lesion to the less aggressive category (Fig. 6, 7).

CT evaluation of MCRCC lesions is less reliable when the lesion is less than 4 cm in diameter, because it may appear as a solid mass (Fig. 8) (15). Nassir et al (16) observed enhancement in all cases of MCRCC during MRI evaluation (12 pts). However according to this data until today there are no specific preoperative diagnostic tools that allow safely to identify CRCC or MCRCC. That is why histopathological examination is always required to confirm the presence of malignant cells in the cyst wall in most cases (3). CRCC can pose serious diagnostic problems during intraoperative frozen-section analysis for pathologist, it not always so easy and clear to confirm or exclude malignant potential of the specimen. Immunostain with cytokeratin antibodies is required do differentiate the small and bland cells of CRCC from histiocytes (3).

From our and literature data pathological parameters (stage and grade) were favourable. Fuhrman nuclear grade was 1–2 (in most of the cases 1) a pathological stage 1 or 2 (3, 6, 8, 16, 17).

Conclusion

Limitation of our study is small subset of patients. The purpose of our study was to point out the possible variations of tumor/cyst appearance and to define the furtherdiagnostic work-up.

In conclusion according to the data described and from our study tumor/cyst co-existence requires further surgical exploration in group 2, 3, 4. In group 2 urologist might face the situation when simple cyst is arising in hyperdense cyst (>20 HU) category IIF – which might appear as a solid mass on computed tomography (Fig. 9). In this situation follow-up with serial CT examinations is required (18).

The principal diagnostic tool in the evaluation of cystic renal cell carcinoma CRCC is US, CT or MRI. However final histological finding will confirm the presence or absence of neoplastic cells in the cystic wall. The standard care management for small cystic renal neoplasms is partial nephrectomy, excision or ablation. Biopsy of suspicious renal masses or multilocular cystic lesions (that can hide renal cancer) can provide essential information for the treatment decission-making.

Histopathology parameters of CRCC are usually favourable which allows the nephron-sparing surgical approach.

In specific cases when patient is at high risk of morbidity and perioperative mortality, older age or does not want to undergo surgery – watchul waiting should be considered as another option, but it is still experimental. Patient should be well informed of possible progression into the metastatic state apart from the fact that the small cystic renal tumors have low-metastatic potential.

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