Original Article

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Magnetic Resonance Imaging Findings of Multilocular Cystic Renal Cell Carcinoma and Clinical-pathologic Comparison

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Abstract

Objective: Multilocular cystic renal cell carcinoma (RCC) is a unique type of renal carcinoma characterized by multi-loculated cystic masses. The aim of this study was to retrospectively evaluate the magnetic resonance imaging (MRI) findings of multilocular cystic RCC.

Materials and Methods: All patients were examined by MRI. Two radiologists retrospectively evaluated MRI features, and compared radiological findings and Bosniak category with histopathological findings.

Results: The patient population comprised seven men and three women with a mean age of 52.9 years (range:37-61 years). The margins of the multi-loculated cystic masses were well defined in all patients, and there was no sign of infiltration of the adjacent tissue and metastatic lymphadenopathy.

Conclusion: Multilocular cystic RCC exhibits non-malignant behavior and frequently has a long survival. The size of the lesion at diagnosis is variable, but there is no evidence of infiltration and metastasis in patients at diagnosis.

Keywords: Multilocular cystic renal cell carcinoma, MRI, renal cell carcinoma

Introduction

Multilocular cystic renal cell carcinoma (RCC) is a distinctive subgroup of RCC that constitutes approximately 2.3-3.1% of all RCCs (1,2). This group of tumors was listed as a rare variant of RCC by the World Health Organization (WHO) in 2016. Multilocular cystic RCC is a low-grade tumor and prognosis is considered to be favorable compared to conventional clear cell RCC (3), which is the most common RCC subtype. Patients with cystic multilocular RCC who undergo resection have excellent prognosis. The multilocular cystic RCC is considered benign by some pathologists (4) due to the lack of progression or metastasis (5,6).

The imaging finding of multilocular cystic RCC is septated, multilocular, solitary renal cyst with or without solid portions similar to other cystic renal masses. The differential diagnosis of multilocular cystic RCC includes cystic renal cell carcinomas, cystic nephroma, and complicated renal cysts (7).

The aim of this study was to evaluate the magnetic resonance imaging (MRI) characteristics of multilocular cystic RCC, and to present typical and atypical imaging aspects of multilocular cystic RCC.

Materials and Methods

Patients

This study was based on the patients with renal tumors who were investigated by MRI between January 2005 and May 2016, and was performed retrospectively in our institution (the protocol number of non-interventional investigation ethical committee approval was 3476 and decision number was

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2017/25-30). The requirement for informed consent was waived due to the retrospective nature of the study. Of 698 cases with a diagnosis of renal tumor, 10 patients were diagnosed as having multilocular cystic RCC. All patients with multilocular cystic RCC were enrolled in this study. The patients were between the ages of 37-61 and female/male ratio was 3/7. All patients were evaluated with abdominal MRI.

Imaging Methods

All MRI studies were obtained on a 1.5-T MR scanner (Gyroscan Intera, release 8.1; Philips Medical Systems, Best, the Netherlands) using a 4-channel phased-array coil, including routine and post-contrast (0.1 mmol/kg body weight Gadolinium-chelates at 2-2.5 mL/s) acquisitions. MRI was comprised of axial and coronal T2-weighted fast spin-echo images (TR, 519 ms; TE, 120 ms; section thickness, 5 mm; gap, 1 mm; ETL, 77; matrix size, 256x256; field of view, 40.5 cm), axial dual-echo T1-weighted in-phase and opposed-phase gradient-echo images (TR, 154; TE, 2.3-4.6; section thickness, 5 mm; gap, 1 mm; matrix, 256x256; field of view, 40.5 cm), axial spectral fat-saturated T2-weighted fast spin-echo images (TR, 2145 ms; TE, 70 ms; section thickness, 7 mm; gap, 1 mm; ETL, 24; matrix size, 256 x 256; field of view, 40.5 cm), and axial three dimensional frequency-selective fat-saturated T1-weighted gradient-echo images (TR, 316 ms; TE, 5 ms; section thickness, 5 mm; matrix, 512x512; field of view, 40.5 cm). Dynamic contrast-enhanced axial T1-weighted images were performed in the corticomedullary and nephrographic phases after administration of a bolus of 0.1 mmol per kilogram of body weight gadolinium chelates.

Image Interpretation

Two radiologists (M.S., C.A.) evaluated MRI in consensus. To characterize the renal cysts, Bosniak radiological classification was used (8,9,10). Tumor location and size, Bosniak category, tumor shape, contour, regional lymph node metastasis, and presence of tumoral invasion to perirenal fat, sinus, adrenal or renal veins were investigated.

Results

Clinical and Pathological Results

The frequency of multilocular cystic RCC in our study group was 1.4%. The mean age of the patients with multilocular cystic RCC was 52.9 years (range: 37-61 years). Three patients were female and seven were male. There was no evidence of additional systemic malignancy, renal stone disease or congenital renal abnormality in our study group. The tumor sizes ranged from 3.5 cm to 10 cm (mean=6.1 cm).

According to assessment of macroscopic specimens, all patients had cystic renal tumors. These tumors were located in the renal parenchyma and there was no evidence of extension into the renal sinus or local invasion to adjacent tissue or organs in all patients. The diagnosis of multilocular cystic RCC was confirmed histopathologically in all patients. Nine patients were Fuhrman grade 1 and one patient was Fuhrman grade 2. Four patients (40%) had stage T1a, four (40%) had stage T1b, and two (20%) had stage T2. None of the patients had Fuhrman grade 3 or 4, stage T3 or T4, renal vein tumor thrombus or distant metastasis at the time of diagnosis. All cases were N0 and M0. All patients were followed up clinically and radiologically for 36-96 months (mean=77.1 months). During the follow-up, no local recurrence, regional lymph node metastasis, or distant metastasis was observed.

MRI Features

All patients underwent MRI. Because of the cystic part, multilocular cystic RCCs had low signal intensity on T1-weighted images (T1WI) and high signal intensity on T2-weighted images (T2WI). The lesions had well-defined outer margins with thick capsules. After the administration of paramagnetic contrast agent, variable enhancement was observed on the septations and wall of cysts of Bosniak category 3 and 4 cysts. On MRI, the cystic renal lesions were found in five patients on right side and in five patients on the left side. These lesions were located at the upper pole of the kidney in seven patients, at the lower pole in two patients and at the interpolar region in one patient.

All of the renal tumors were in cystic and none of the renal tumors that were diagnosed as multilocular cystic RCC had a complete solid structure. The Bosniak categories of the patients were II in one patient, IIF in one patient, III in six patients and IV in two patients (Figures 1,2,3 and 4). There was no evidence of multifocal or bilateral multilocular cystic RCC. One patient had a chromophobe RCC with Bosniak category II cyst in the same kidney.

In MRI, there were no changes in signal intensity due to hemorrhage or calcification. In our study, a synchronous solid or

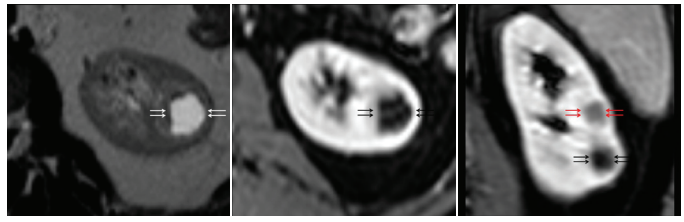

Figure 1. a-c. Bosniak category II cyst; the lesion was detected incidentally during intraoperative ultrasonography for neighboring solid renal tumor (red arrows) and was histopathologically confirmed as multilocular cystic renal cell carcinoma. Axial (a) T2-weighted image shows few small septations in the renal cyst at the lower pole of the left kidney (black arrows). The corticomedullary phase of the dynamic study (b) and late phase post-contrast coronal T1 image (c) show no enhancement of the few septa compared to the renal cortex in the mass (white and black arrows)

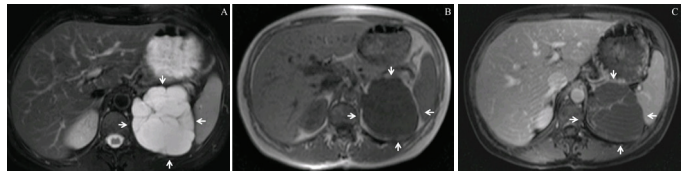

Figure 2. a-c. Bosniak category IIF cyst; after a follow-up period of 1 year, the lesion increased in size and was histopathologically confirmed as multilocular cystic renal cell carcinoma. The axial (a) T2-weighted image shows a giant septated cystic mass at the upper pole of the left kidney. The axial (b) T1-weighted image shows well-demarcated cystic renal mass. The corticomedullary phase of the dynamic study (c) shows mild enhancement in a few septa compared to the renal cortex in the mass

cystic renal neoplasm was found in both kidneys in one patient. The demographic characteristics of patients were comparable in each group with Bosniak category II, IIF, III, and IV lesions. Table 1 provides the MRI, pathological and demographic features of all patients.

Significance of the Study

This study revealed that multilocular cystic renal cell carcinomas are characterized by multi-loculated cystic masses. The Bosniak categories of multilocular cystic renal cell carcinomas were observed II, IIF, III and IV. The differential diagnosis of multilocular cystic renal cell carcinoma includes cystic renal cell carcinomas, cystic nephroma, and complicated renal cysts.

Discussion

Multilocular cystic RCC is a rare subgroup observed predominantly in men with an incidence varying between 1-1.5% of all renal malignancies and generally found in the fifth-sixth decades (11,12,13). In our study, the incidence was similar to previous studies. The clinical symptoms of the patients with multilocular cystic RCC are non-specific and patients are usually asymptomatic (13,14). In practice, multilocular cystic RCC is a coincidentally detected tumor. If totally resected, multilocular cystic RCC has an excellent prognosis. Local recurrence and distant metastasis have not been reported in the English literature (11,14,15).

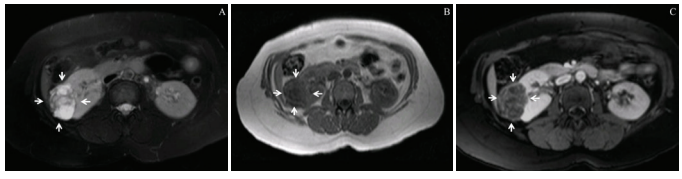

Figure 3. a-c. Bosniak category III cyst; histopathologically confirmed as multilocular cystic renal cell carcinoma. The axial (a) T2-weighted image shows a septated cystic mass with heterogeneous appearance and hypointense rim in the right kidney (arrows). The axial (b) T1-weighted image shows well-demarcated hypointense renal mass (arrows). The corticomedullary phase of the dynamic study (c) shows enhancement in the septa and cyst wall compared to the renal cortex in the mass (arrows)

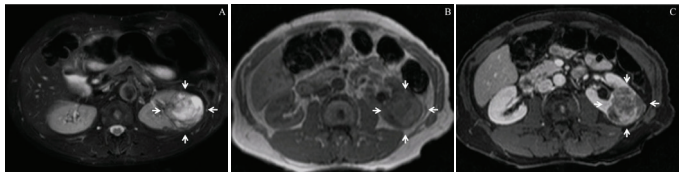

Figure 4. a-c. Bosniak category IV cyst; histopathologically confirmed as multilocular cystic renal cell carcinoma. The axial (a) T2-weighted image shows a heterogeneous cystic mass with solid portion in the left kidney (arrows). On the axial (b) T1-weighted image, the mass has a well-demarcated and hypointense appearance (arrows). The corticomedullary phase of the dynamic study (c) shows prominent enhancement of the septa and solid portion of the mass (asterisk) compared to the renal cortex (arrows)

The multilocular cystic RCC was defined as a rare entity with excellent prognosis according to WHO 2004 criteria. According to WHO, the diagnostic criteria for multilocular cystic RCC are a multilocular cystic appearance, a limited solid component in the small areas with no expansile nodules and no tumor necrosis, and microscopically low Fuhrman grade (16). Histopathologically, these tumors are well demarcated and separated from the kidney by a thick capsule, that may contain fluid, clear cell lining septations, vascularized or non-vascularized fibrosis (1). They consist of multiple fibrous septa composed of malignant epithelial cells with clear cytoplasm (10). In immunohistochemical staining, multilocular cystic RCC is usually positive for vimentin, EMA and CD10 (1).

In MRI, multilocular cystic RCC is defined as septated, variable sized multilocular cystic tumors with fibrous capsule, and they cannot be differentiated from complicated non-malignant renal cysts and other cystic RCC types (1,15). Conventional MRI sequences provide the data of tumor location and internal structure of the multilocular components. The MRI signal of multilocular cystic RCCs depends on the content of the cyst fluid. The fluid portion of multilocular cystic RCC usually appears as hypointense on T1WI and hyperintense on T2WI. Fibrous septations are usually isointense on T1WI and markedly hypointense on T2WI. However, overall or partial portion of the tumor may be observed as hyperintense on T1WI and variable hypointense on T2WI due to intra-tumoral hemorrhage. The signal alteration due to intra-tumoral hemorrhage may contain fluid-fluid level. In our series, the presence of the limited hypointensity on T2WI and the hyperintensity on T1WI was detected in 40% of the patients.

After contrast agent administration in MRI, the enhancement pattern of the tumor is variable, depending on the presence of solid portion and distribution of the cellular component and the fibrous tissue. On contrast enhanced sequences, asymmetric septal, irregular cystic wall or solid part enhancement may be observed. One patient (10%) had Bosniak type II cyst and no

Table 1. Imaging, pathologic and demographic features of the study group

Case	Age	Gender	Side	Location	Bosniak category	Tumor size (cm)	Fuhrman grade	Tumor stage
1	52	M	Left	Upper pole	IV	5.2	2	T1b
2	61	M	Left	Upper pole	IV	5.5	1	T1b
3	60	M	Right	Upper pole	III	5.3	1	T1b
4	59	M	Right	Upper pole	III	3.5	1	T1a
5	50	F	Left	Upper pole	IIF	8	1	T2
6	52	F	Right	Interpolar	III	5.2	1	T1b
7	37	M	Left	Lower pole	III	10	1	T2
8	43	M	Right	Upper pole	III	3	1	T1a
9	54	M	Left	Lower pole	II	2	1	T1a
10	61	F	Right	Upper pole	III	5	1	T1a

M: Male, F: Female

contrast enhancement was observed in the septations. In one patient (10%) with Bosniak type IIF cyst, the septations of the tumor showed tiny enhancement in our study. The remaining eight patients (80%) have asymmetric septal and irregular cystic wall enhancement in MRI compatible with Bosniak type III and IV cysts. Furthermore, enhancement of the solid portion was observed in two patients (20%) with Bosniak type IV cyst.

For the first time, the renal cysts were classified by Bosniak in 1986 (7). Bosniak revised his classification in 1997 and 2012 (8,9). According to the Bosniak classification, renal cysts are divided into 4 groups. Bosniak I, II and IIF cysts contain benign features. Bosniak III cysts are complicated cystic lesions with septal enhancement and thickening. Bosniak IV cysts are clearly malignant lesions and contain solid portion. The distribution of the Bosniak category in multilocular cystic RCC ranged from type IIF cyst to type IV cystic tumor. Similar to previous studies, multilocular cystic RCCs tend to be Bosniak type III cyst (60) and to have a multilocular appearance in our study.

Patients usually present with non-specific symptoms similar to other types RCC, such as low back pain and hematuria (13). In our study, tumor size, gender distribution, clinical symptoms, tumor lateralization, Fuhrman grade and Tumour, Node, Metastasis stage were found to be similar with the literature (11,14,17). Complete resection of the renal cyst was performed in all patients. In six patients, cystic tumors were resected by nephron sparing surgery and radical nephrectomy was performed in the remaining four patients. Nephron-sparing surgery may be a preferable treatment method in patients with multilocular cystic RCC, especially in Bosniak type 3 cysts (18). During the follow-up period, all patients were uneventful in our study.

Study Limitations

Our study has some limitations. First, this is a retrospective study performed in renal carcinoma patients who were met during the diagnostic procedures or somehow discussed in multidisciplinary meetings; hence the selection bias is inevitable. Second, the study reflects the results of a single institute; larger series may be achieved in multi-institutional studies. However, our study group consisted of patients with images in 12-year PACS archive and with close clinical and radiological follow-up.

Conclusion

Multilocular cystic RCC is a rare malignant renal tumor and should be kept in mind in the differential diagnosis of complicated cystic renal masses. In our study, multilocular cystic RCCs appeared as complicated cystic lesions and were identified as Bosniak II-IV cysts in MRI.

Ethics

Ethics Committee Approval: Because of the study was designed as a retrospective study, ethics committee approval was not obtained.

Informed Consent: Because of the study was designed as a retrospective study, informed consent was not taken from the patients.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Concept: C.A., Design: C.A., Data Collection or Processing: O.B., Ö.D., G.A., B.T., K.Y., Analysis or Interpretation: C.A., M.S., Literature Search: O.B., Ö.D., G.A., B.T., K.Y., Writing: C.A.

Conflict of Interest: No conflict of interest was declared by the authors.

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