Case Report

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Multilocular Cystic Renal Neoplasm of Low Malignant Potential: A Case Report and Literature Review

낮은악성가능성을 가진 다방성낭성신장생성물: 증례 보고와 문헌 고찰

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Multilocular cystic renal neoplasm of low malignant potential (MCRNLMP) is a rare subtype of renal cell carcinoma that has a favorable outcome. Most cases of MCRNLMP usually present as distinct multilocular cystic lesions; however, they may appear as small complicated cysts with hemorrhagic components. Herein, we present a case of MCRNLMP and provide a review of the literature.

Index terms Kidney; Neoplasm; Cyst

INTRODUCTION

Multilocular cystic renal neoplasm of low malignant potential (MCRNLMP) is a rare subtype of renal cell carcinoma (RCC) with favorable outcome (1). This entity was previously known as multilocular cystic RCC or multicystic clear cell carcinoma. The 2012 International Society of Urological Pathology (ISUP) consensus conference on renal neoplasia adopted the new terminology of MCRNLMP owing to its similar pathological features to renal clear cell carcinoma, but different natural history (2). The World Health Organization (WHO) classification of 2016 subsequently accepted this change in terminology and the diagnostic criteria for MCRNLMP is defined as a neoplasm of kidney composed entirely of cysts with small groups of non-expansile clear cells in the septa (3). Several studies have been conducted to assess radiologic findings and most reports described the imaging features of these tumors as multilocular cystic lesions

without enhancing solid nodule (4, 5). Even if variable imaging appearance can be occur in MCRNLMP, a case of small complicated cyst is not common (4). Furthermore there is a lack of literature detailing imaging features. Here, we describe a case of MCRNLMP presenting small complicated cyst and review of literature.

CASE REPORT

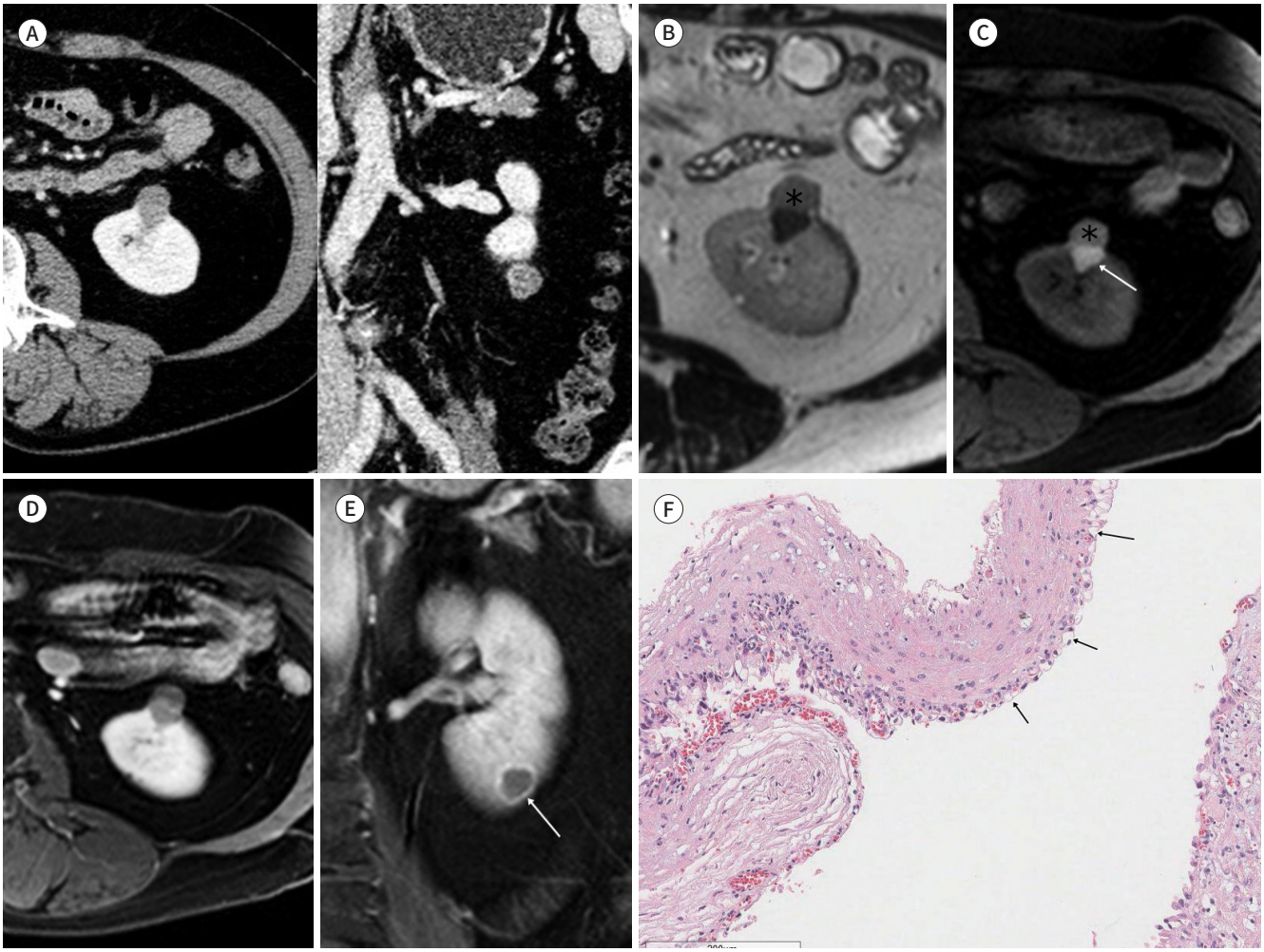
A 62-year-old female underwent right partial nephrectomy 2 years earlier. Postoperative pathological diagnosis revealed stage I RCC, clear cell type. Also, she underwent endoscopic submucosal resection due to early gastric cancer 7 years ago. Preoperative Contrast-enhanced abdominal CT scan using a single portal venous phase was obtained in the initial assessment of RCC 2 years ago. It revealed a well-defined, slightly high density mass, about 70 Hounsfield unit (HU), size 1.7 cm × 1.4 cm, in the lower pole of kidney. The radiological findings diagnosed the lesion as benign such as hemorrhagic renal cyst. During routine follow-up assessment, an enhanced CT scan showed a slightly increase in the size of the slightly high attenuating mass, 60 HU, approximate measuring 2.1 cm × 1.7 cm. The mass demonstrated partly exophytic growth and equivocal peripheral enhancing septa (Fig. 1A). Moreover, there was no fat-density lesion or enhancing solid component in the mass. On MRI, the lesion correlating with CT image appeared as a well-defined tumor with dependent hemorrhagic component. Relative to the normal renal cortex, the nondependent portion showed intermediate signal intensity on axial T2-weighted image and T1-weighted image (Fig. 1B, C). On dynamic contrast-enhanced scan, the mass showed a few minimally thickened enhancing septa, about 3 mm in thickness in the peripheral portion (Fig. 1D, E). There was no metastasis or lymph node enlargement. Our initial differential diagnosis included renal tumor with hemorrhage such as RCC, complicated renal cyst with hemorrhage. The patient underwent partial nephrectomy due to history of RCC in contralateral kidney. Invasion of the surrounding tissue was not observed. Grossly, the mass was a 2.3 cm sized, well-demarcated multilocular cystic mass filled with serous and hemorrhagic fluid. Microscopic examination revealed that the mass was composed by variable sized cysts without expansile growth. The cysts were lined by single layer of tumor cells with abundant clear or granular cytoplasm with low grade nuclei (WHO/ISUP grade 2). The final diagnosis was confirmed to be MCRNLMP (Fig. 1F). The patient has been regulatory followed up for 1 year without recurrence or metastasis.

Written informed consent was obtained from the patient for the use of data for research purposes.

DISCUSSION

MCRNLMP is rare type of RCC and accounts for less than 1% of all kidney cancer (1). The true incidence of MCRNLMP is not known due to rarity of these tumor and the variance of pathologic criteria used to diagnose this subtype (6). There tumors develop due to extensive cystic regression or growth within renal tubules, causing obstruction ultimately forming cysts. Histopatholgically, the tumor is composed entirely of numerous cysts with the septa

Fig. 1. Imaging and pathologic features of multilocular cystic neoplasm of low malignant potential in a 62-year-old female.
A. Axial (left) and coronal (right) contrast-enhanced images show a well-defined slightly high-density mass of approximately 60 Hounsfield unit located in the left renal cortex with partly exophytic growth.
B, C. Axial T2-weighted MR image (B), pre-contrast T1-weighted image (C) display fluid-fluid levels in the mass lesion and dependent high signal intensity on the T1-weighted image (arrow), suggesting hemorrhage. The non-dependent portion of the mass shows intermediate signal intensity (asterisks) on T2-weighted (B) and T1-weighted MR image (C).
D, E. Axial (D) and coronal (E) contrast-enhanced T1-weighted MR images show mild enhancement of the peripheral, minimally thickened wall and septa of the mass (arrow).
F. Histopathology confirms the lesions as multilocular cystic renal neoplasm of low malignant potential, which is lined by a single layer of tumor cells (arrows) with abundant clear or granular cytoplasm with low-grade nuclei (hematoxylin and eosin stain, × 100).



containing individual or groups of clear cells without expansile growth or mural nodules. Based on the 2016 WHO's definition, it is important to differentiate MCRNLMP from conventional clear RCC with cystic change because it clearly does not share the same prognosis (3). The latter has expansile growth of clear tumor cells and maybe have necrosis and hemorrhage. Expansile solid nodule, necrosis, vascular invasion were not identified in this case and there were no histological features of clear cell proliferation.

Several studies have been evaluated imaging features of MCRNLMP (4, 5, 7). Kim et al. (4) in their study on 10 patients, found that they appeared as a well-defined multilocular cystic mass in all cases. The tumors were filled with serous and/or complicated fluid and there was

no expansile solid nodules found in the thin septa. In 3 cases, hyperdense areas in some loculues on CT scan correlated with hemorrhage or gelatinous fluid on pathologic examination, like in this case. Aubert et al. (7) described 9 cases of these tumors and correlation with imaging features were recorded in 8 patients; 6 multilocular cystic masses as Bosniak category III, one unilocular with small mural nodule as category IV, one solid tumor. Among them, two cases was misinterpreted due to smaller tumor size (less than 3 cm); multiple septa simulated solid component and a thick cystic wall was confused with expansile nodule. They mentioned enhancing nodule of 5 mm or greater and the Bosniak classification were significant to distinguish MCRNLMP from clear cell RCC. Hindman et al. (5) reported 23 cases of MCRNLMP and they were seen as complicated cystic lesions. Seven of pathologic confirmed MCRNLMP were considered as Bosniak category IIF, 13 as category III and 3 as category IV and the average tumor size is about 2.9 cm. They insisted that the percentage of vascularized fibrosis reliably correspond to the solid internal enhancement seen in category IV lesions on CT scan. These studies showed good correlation between imaging findings and pathologic features, however it was not correlated for small tumors, less than 3 cm.

Unlike foregoing studies, intermediate signal intensity on T2-weighted image of our case is difficult to differentiate MCRNLMP initially. It revealed well-defined mass with minimally thickened enhancing septa and no expanding nodule like other studies, so it can be categorized as Bosniak classification IIF.

It would be clinically significant, but any imaging feature cannot differentiate complicated renal cyst, partially cystic RCC and MCRNLMP (7). Hindman et al. (5) reported that there was overlap between category IIF, III and IV lesions with respect to clear cell percentage in the lining of septa in their study.

The majority of MCRNLMP cases reported in the literature are low stage tumors, and therefore extension of this type of kidney tumors into perinephric fat is extremely rare (8). Generally, it has indolent nature with negligible risk of recurrence or metastasis, because of fewer malignant cells in the tumor and the low nuclear grade of the tumor cells (1). Li et al. (9) reported case series of 76 patients concluded that these tumors were predominantly of a low nuclear grade irrespective of tumor size and staging and suggested longer follow-up interval to minimize unnecessary investigations. Weiss et al. (10) insisted than when MCRNLMP is suspected preoperatively and confirmed intraoperatively, nephron sparing surgery should be recommended due to good prognosis.

In summary, due to variable and non-specific imaging features, diagnosis of MCRNLMP is a challenge to clinicians and radiologists. When a well-defined complicated mass that contains hemorrhagic component and septal enhancement, it can be considered as a differential diagnosis as it can be managed as less invasive surgery.

Author Contributions

Conceptualization, L.J.; investigation, L.J., S.J.Y.; methodology, L.J., K.D.W.; data curation, L.J., S.J.Y.; formal analysis, all authors; supervision, Y.S.K.; writing—original draft, L.J.; and writing—review & editing, all authors.

Conflicts of Interest

Seong Kuk Yoon has been an Section Editor of Journal of the Korean Society of Radiology since 2014; however, he was not involved in the peer reviewer selection, evaluation, or decision process of this ar-

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낮은악성가능성을 가진 다방성낭성신장생성물: 증례 보고와 문헌 고찰

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낮은악성가능성을 가진 다방성낭성신장생성물은 신종양 중 비교적 드문 타입으로 비교적 좋은 예후를 보인다. 이 종양은 주로 다방성의 신낭종으로 관찰되며 출혈을 동반한 복합신낭종으로 보이는 경우는 흔치 않다. 본 연구에서는 낮은악성가능성을 가진 다방성낭성신장생성물 증례를 보고하고 논문을 고찰하고자 한다.

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