Case Report

Multilocular cystic renal cell carcinoma a diagnostic dilemma: A case report in a 30-year-old woman

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Abstract

Multilocular cystic renal cell carcinoma (MCRCC), also known as multilocular clear cell renal cell carcinoma (RCC), is a rare cystic tumor of the kidney with an excellent outcome. It occurs in about 3.1-6% of the conventional RCC. It is usually included in the group of tumors of undetermined malignant potential with low nuclear grade. We present a case of MCRCC in a 30-year-old female patient presenting incidentally as an apparently benign-looking multicystic space occupying lesion in the upper pole of right kidney. Right-sided simple nephrectomy was performed, and on histopathologic examination it was found to be MCRCC, stage 1 with Fuhrman nuclear grade 1. Immunohistochemistry with epithelial membrane antigen and vimentin confirmed the diagnosis.

Key Words: Multilocular cystic renal cell carcinoma, nephrectomy, cystic tumor

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INTRODUCTION

Multilocular cystic renal cell carcinoma (MCRCC), also known as multilocular clear cell renal cell carcinoma (RCC), is a rare cystic tumor of the kidney with an excellent outcome.[1] Diagnostic criteria for MCRCC were defined by the 2004 World Health Organization (WHO) classification of kidney tumors based on previous reports and the suggestions of Eble and Bonsib as they may be mistaken for cystic degeneration of conventional RCC.[2] We present a case of MCRCC diagnosed incidentally in a 30-year-old female patient. MCRCC is usually positive for CD10, vimentin, and epithelial membrane antigen (EMA).

CASE REPORT

A 30-year-old female patient was referred for the management of an incidentally diagnosed right-sided cystic renal lesion.

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Postcontrast computed tomography (CT) scan showed a well-defined multiseptate cystic space occupying lesion in the upper pole of right kidney without any obvious lymphadenopathy in the hilum or the aorto-caval area. No mural nodule or calcification was noted in the cyst and there was no hydronephrotic change either [Figure 1]. There was no family history of constellation of signs or symptoms suggestive of von Hippel Lindau syndrome or renal tumors of any kind. Biochemical assessment of renal function was normal. Complete blood count and other preoperative investigations were within normal range. There were no significant findings on general and systemic examinations. Since cystic renal RCC could not be excluded a right-sided nephrectomy was performed, and the gross examination of the kidney revealed a multiloculated cyst in the upper pole of 3.5 × 3 × 3 cm dimension containing mucinous material. Neither necrotic area nor expansile nodule was appreciated [Figure 2].

Histopathological examination revealed cyst wall lined by single or multiple layers of clear cells with well-defined cytoplasmic borders and small nuclei. There were areas with alveolar arrangement of clear cells surrounded by fibrovascular septae. The tumor did not invade the renal capsule and hilar structures [Figure 3]. Immunohistochemistry with vimentin

and EMA showed strong membranous positivity [Figure 4]. TNM staging and Fuhrman nuclear grading were stage T1a and grade 1, respectively.

DISCUSSION

Among all the renal neoplasms MCRCC occurs in about 3.1-6% of clear cell RCC.[3] They have been found in the mean age of 51 years with the age range of 20-76 years. This particular type has a male preponderance (male to female ratio of 3:1).[4] MCRCC is usually included in the group of tumors of undetermined malignant potential as they are of low nuclear grade and usually confined to the kidney. Main pathologic features of MCRCC are divided into gross and microscopic features. The gross features are encapsulated multilocular cystic appearance, yellowish small solid component, absent expansive nodules, and absent tumor necrosis. The microscopic features include cysts lined by cuboidal clear cells or flattened epithelium with septa containing aggregates of epithelial cells with clear cytoplasm and of low Fuhrman grade.[4] These features differentiate them from the cystic RCC which is nothing but the cystic degeneration of conventional RCC carrying worse prognosis. In our case, there was no solid component or any kind of tumor necrosis and microscopically the septae were lined by multilayered clear cells.

Cystic nephroma and benign multilocular cysts are another important differential diagnosis of this condition, but close attention to the monolayered flattened or hobnail epithelial cell lining and presence of ovarian stroma like cellular stroma with or without tubules in the septae will differentiate them from MCRCC.[3]

MCRCC is usually positive for CD10, vimentin, and EMA, and in our case it was positive for both vimentin and EMA confirming our diagnosis.[1]

In a recent large series by Suzigan *et al.* of the 45 cases of MCRCC, they found 82% of their cases in T1 stage and low

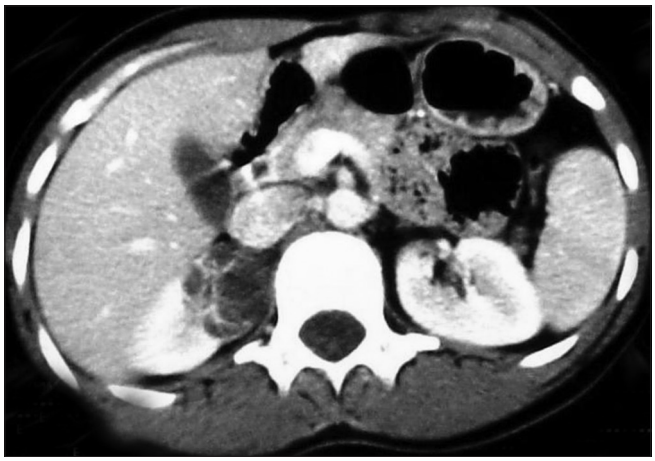

Figure 1: Postcontrast CT scan showed a well-defined multiseptate cystic space occupying lesion in the upper pole of right kidney

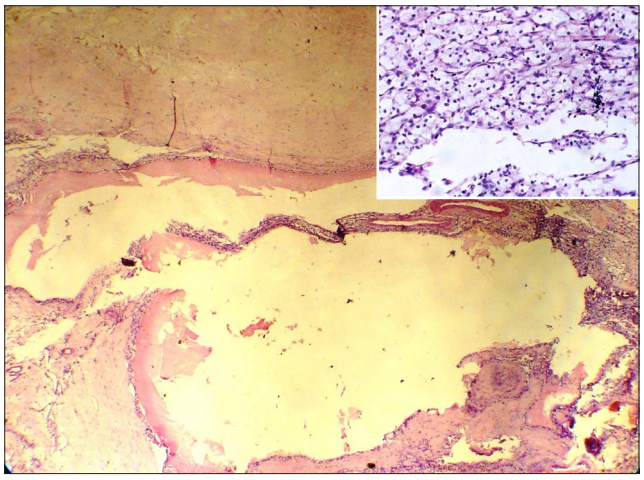

Figure 3: Histopathological examination revealed cyst wall lined by multiple layers of clear cells with well defined cytoplasmic borders and small nuclei. (Hematoxylin and eosin stains; ×40 magnification, *inset* – ×400 magnification)

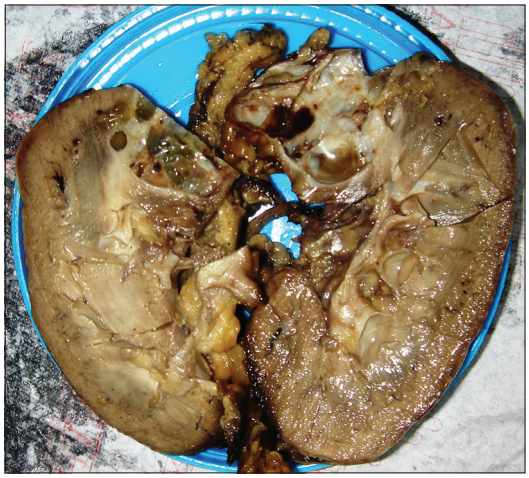

Figure 2: Gross examination of the kidney revealed a multiloculated cyst in the upper pole containing mucinous material

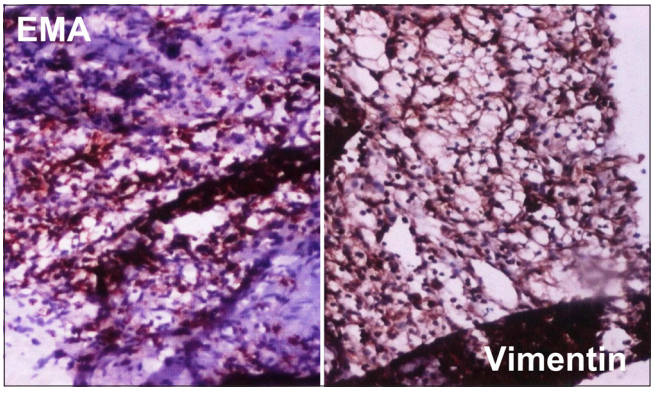

Figure 4: Immunohistochemistry showed strong membranous positivity with vimentin and epithelial membrane antigen

nuclear grade found in 62% cases of them. In their series the 5-year survival rate was 100%.[5]

Based on the fact that this is tumor with low nuclear grade and confined to the kidney, their survival rate is significantly better than conventional RCC. The treatment options are either simple nephrectomy or nephron sparing surgery. In our case as the other kidney was absolutely normal we planned to offer simple nephrectomy for treatment. Nephron sparing surgery is only advisable to those patients with single kidney or those where contralateral kidney tend to be jeopardised in near future.[6,7] We currently favor elective simple nephrectomy as the treatment of choice for MCRCC. Families are provided with balanced information, fully informing them of the risks of surgery, and the potential morbidity associated with it, thereby allowing them to take an active part in the decision-making process.

This is a case report of a patient with cystic renal cell carcinoma that had computed tomographic features of benign multicystic renal disease. Although this entity is a possibility in some patients, thick-walled cystic lesions of the kidney must be considered malignant until proven otherwise and surgery cannot be avoided.

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