Case Report

Multilocular cystic renal cell carcinoma: A rare case report

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ABSTRACT

Multilocular cystic renal cell carcinoma (MCRCC) is an uncommon low grade RCC with unique morphologic features. MCRCC also known as multilocular clear cell RCC is a rare cystic tumor of the kidney with an excellent outcome. MCRCC has been recognized as a separate subtype of RCC in the 2004 World Health Organization (WHO) classification of adult renal tumors. MCRCC is diagnosed on the basis of strict histological criteria according to 2004 WHO classification. 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 43-year-old female patient who presented with pain in left lumbar region and hematuria. Left-sided radical nephrectomy was performed, and on histopathologic examination it was diagnosed as MCRCC with Fuhrman nuclear grade 1. Immunohistochemistry with epithelial membrane antigen and vimentin confirmed the diagnosis.

Key words: Clear cell, kidney, multilocular cystic renal cell carcinoma

INTRODUCTION

Multilocular cystic renal cell carcinoma (MCRCC) is a distinct subtype of clear-cell RCC with a favorable biological behavior. These tumors are a rare entity, comprising approximately 1–2% of all renal tumors. It occurs in about 3.1–6% of the conventional RCC.[1] The 2004 World Health Organization (WHO) classification of kidney tumors recognizes MCRCC as a rare variant of clear cell carcinoma with a good prognosis. MCRCC is also known as multilocular clear cell RCC and multicystic clear cell carcinoma. It is important to differentiate this entity from the clear cell RCC since it is associated with a good prognosis. There are other entities from which MCRCC has to be differentiated such as cystic nephroma, cystic clear cell carcinoma, clear cell papillary RCC and the tubulocystic carcinoma.[2] We report this rare entity in a 43-year-old female patient.

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CASE REPORT

A 43-year-old female presented with on and off pain in left lumbar region and hematuria since 5 months. Complete hemogram and other investigations were within normal limits. Patient had a history of renal stones. Ultrasonography revealed a 58 cm × 55 cm × 54 cm mass arising in the anterior part of the lower pole of cortex of the left kidney. Contrast enhanced computed tomography showed a heterogeneously enhancing, well-defined, encapsulated multiseptated mass lesion with central necrosis and calcification in the lower pole of the left kidney. Left nephrectomy was performed, and the specimen was sent for histopathological examination. Grossly, the nephrectomy specimen including peri-nephric fat measured 15 cm × 10 cm × 5 cm. Cut-surface of the kidney showed a well circumscribed, well encapsulated growth measuring 7 cm × 7 cm situated in the cortex of the lower pole of the kidney. Cut-surface of the growth was multiloculated, cystic with thin septae in between the cysts [Figure 1]. The cysts were filled with brown colored gelatinous material and clots. The ureter was grossly free from any growth. Separately received left adrenal gland measured 3.5 cm × 2 cm × 1 cm. Cut-surface of adrenal was grossly unremarkable. Microscopic examination of the growth revealed a complex cystic tumor comprising of variably sized noncommunicating cysts separated by septae

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containing tumor cells showing uniform, hyperchromatic nucleus, inconspicuous nucleoli (Furhman nuclear grade 1), and abundant clear cytoplasm with distinct cell borders [Figure 2]. Mitotic figures were sparse. Lumen of the cysts contained eosinophilic secretions/hemorrhage. Septae between the cysts also contains cords and clusters of similar tumor cells. Immunohistochemistry showed positivity for vimentin, epithelial membrane antigen (EMA) and CD10 [Figure 3]. The ureteric resection margin, peri-nephric fat and renal vessels were free from tumor cell invasion. Adjacent renal parenchyma showed various degenerative changes in the form of focal glomerular hyalinization and sclerosis, tubular casts, interstitial fibrosis, and chronic inflammatory cell infiltrate. The adrenal was within normal histological limits. The histomorphological and immonohistochemical features were compatible with MCRCC, Fuhrman nuclear grade 1. The patient is well without any recurrence on follow-up.

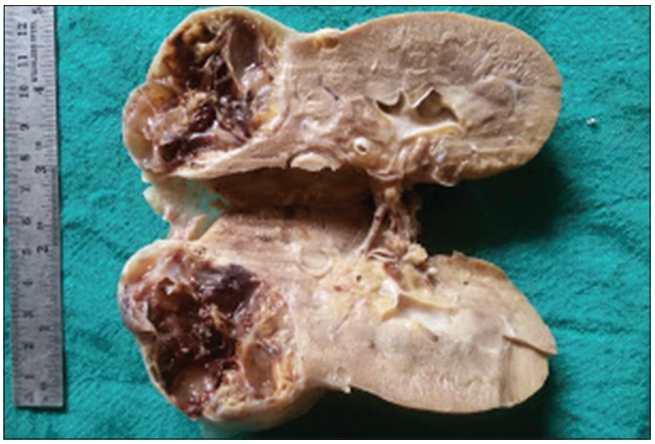

Figure 1: Gross examination of kidney revealed multiloculated cyst in lower pole

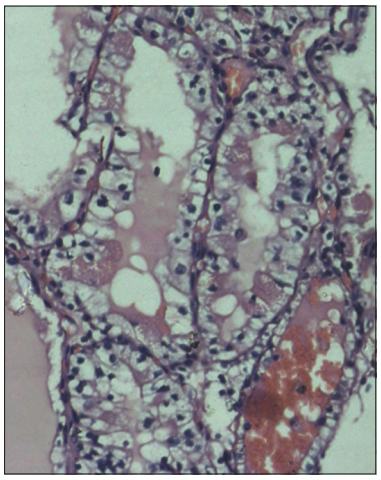

Figure 2: Histological examination showed cyst wall lined by multiple layers of clear cells with well-defined cytoplasmic borders and small nuclei (H and E, ×400)

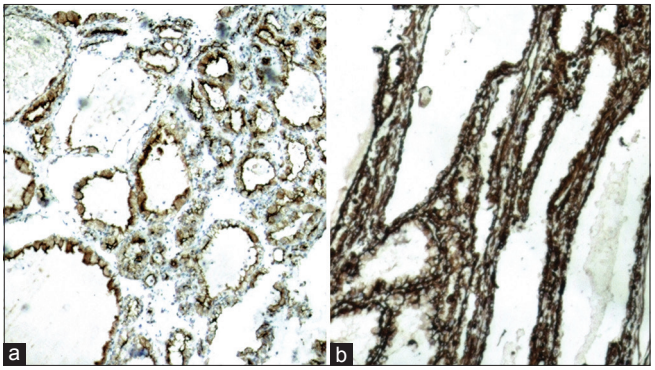

Figure 3: Immunohistochemistry showed strong positivity for CD 10 (a) (IHC, ×200) and vimentin (b) (IHC, ×200)

DISCUSSION

Multilocular cystic renal cell carcinoma is almost always discovered incidentally even before the onset of symptoms. The age of presentation ranges between 20 and 76 years with a mean age of 51 years. The tumor has a male predominance with male to female ratio of 3:1. MCRCC is considered a distinct subtype of clear cell RCC in the 2004 WHO classification based on the characteristic gross features with a multilocular cystic appearance and a variegated, yellowish solid component limited to small areas with no expansive nodules or necrosis. Histologic examination reveals cysts lined by occasionally flattened cuboidal clear cells and septa that contain aggregates of epithelial cells with clear cytoplasm; tumors usually are Fuhrman grade 1.[1] The differential diagnosis of this entity consists of other cystic lesions of the kidney primarily cystic nephroma, extensively cystic clear cell RCC, clear cell papillary RCC, and the tubulocystic carcinoma. Although cystic nephroma may have at least some clear cells lining the septa, the lining clear cells tend to be focally rather than diffusely distributed, and there are no clusters of clear cells in the walls.[3] The ovarian-like stroma in cystic nephroma, if present, distinguishes it from MCRCC, which does not exhibit this type of stroma. Extensively cystic clear cell RCC is distinguished from MCRCC by the fact that solid areas may be evident grossly, or expansile nodules of clear cells are observed microscopically. Clear cell papillary RCC is usually cystic and the cyst walls are lined by clear cells; however, much of the tumor exhibits papillary architecture, a feature not found in MCRCC. Tubulocystic carcinoma is another multicystic tumor that has been increasingly recognized in the past 15 years. It differs from multilocular cystic RCC in that the cystic spaces, rather than being lined by clear cells, are lined by flat cuboidal and sometimes hobnail-type cells with eosinophilic cytoplasm and variable nuclear atypia, typically with nucleolar prominence in the range of Fuhrman grade 2 or 3, another feature incompatible with a diagnosis of MCRCC[2] which is a low grade tumor. In addition, the septal structures of tubulocystic

carcinoma do not harbor clusters of clear cells.[4] On immunohistochemistry, MCRCC is usually positive for CD10, vimentin, and EMA as seen in our case thereby confirming our diagnosis.[1]

Multilocular cystic renal cell carcinoma is a low grade tumor with better prognosis and survival rate as seen in a recent large series of 45 cases of MCRCC by Suzigan *et al*., where they found 82% of cases in T1 stage and low nuclear grade was present in 62% of their cases. As seen in our case where it was graded as Furhams grade 1. In their series, the 5 years survival rate was 100%.[5] Based on the fact that this is a tumor with low nuclear grade and is usually confined to the kidney when diagnosed, the survival rate is significantly better than conventional RCC.

The treatment options available are either simple nephrectomy or nephron sparing surgery. Nephron sparing surgery is performed in patients with unilateral kidney or where the contralateral kidney is at risk of failure.[6] In our case, as the other kidney was absolutely normal, radical nephrectomy was done for treatment.

CONCLUSION

Multilocular cystic renal cell carcinoma is an uncommon renal neoplasm, which developed in our case in a patient with long-term history of renal stones. Hence, it must be kept in mind because it has an excellent prognosis when compared to conventional RCC.

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