CASE REPORT

Multilocular Cystic Renal Cell Carcinoma in an HIV Patient: A Diagnostic Dilemma

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Abstract
A radical nephrectomy specimen was referred for pathological examination. The history, course and the pathological findings proved very interesting. The specimen belonged to a forty four year old HIV positive man who was admitted with pain and tenderness in the right hypochondriac region. Sonography demonstrated fatty liver and incidental multilocular enhancing cystic lesion in the left kidney. A left radical nephrectomy was performed and pathological examination of the specimen showed a multilocular cystic renal cell carcinoma (MCRCC). The clinical presentation, management and outcome of this new entity are discussed.

Key words: Kidney, Cyst, Multilocular cystic renal cell carcinoma (MCRCC).

Introduction

Multilocular cystic renal cell carcinoma (MCRCC) is relatively a new entity. It was included in 2004 World Health Organization's classification of renal tumors as "a rare histologic variant of renal cell carcinoma with very good prognosis" (1). However, the term itself has been indeed introduced as early as 1982 (2). These tumors represent approximately 3% of all renal cell carcinoma (3). Less than 100 cases have been reported in the world literature. They differ from conventional renal cell carcinoma in their clinical presentation and gross appearance. We report a rare presentation of incidental case of MCRCC in an HIV patient and discuss the importance of distinguishing it from other other forms of renal cystic disease.

Case background

History and examination

A 44 year-old man was admitted to Tripoli Medical Center with a history of pain in the right upper quadrant of the abdomen. There were no other symptoms except passing dark urine once and specifically no history of weight loss.

He had a history of surgery on haemorrhoids. He is a known case of HIV, HBV and HCV. He is a diabetic patient on premixed insulin (30/70) 36 IU in the morning and 18 IU in the evening. The patient was on prophylactic treatment for tuberculosis as he had had primary tuberculosis at the age of 7 years. He was not pale and not jaundiced. No lymphadenopathy and no ascites were recorded. He was conscious, oriented, not dyspnoeic and his temperature was 37^0 C. Abdominal examination revealed tenderness in the right upper abdomen with palpable soft liver one finger width below costal margin. Examination of the cardiovascular, respiratory and central nervous systems was unremarkable.

Preoperative investigations

All his haematological investigations, blood urea and serum creatinine were normal. His liver function tests were slightly deranged with marginally elevated serum GPT 52 IU/L (normal: 5-41 IU/L) and serum GOT 48 IU/L (Normal: 5-38 IU/L). Urine sediment showed many red blood cells but no significant pus cells. Records of abdominal sonography demonstrated fatty liver and incidental multilocular enhancing cystic lesion in left kidney. Pre and post contrast MRI confirmed hepatomegaly with fatty change. An enlarged left kidney with multilocular enhancing cystic lesion in lower and middle parts was seen enclosed within renal capsule with distortion and replacing of calyces. The mass measured about 6.5 x 5.5 x 4.5 cm. with radiological impression of possibly stage 1 renal cell carcinoma.

Surgeon's notes: Left kidney showed normal renal capsule, perirenal fat and vessels with a cystic mass occupying middle and lower part. The left renal artery was identified and ligated. Radical nepherectomy of left kidney was done.

Pathology Report

On gross examination, the kidney measured 16x11x4.5 cm. The upper pole had a normal appearance, but the middle and lower poles were cystic and fluctuant with circumscribed borders. The mass lesion measured 6x4 cm. Membranous septa subdivided the cyst into smaller spaces focally filled with brown haemorrhagic material (Figure 1). Microscopically, sections from the renal tumor revealed multilocular cystic lesions with fibrous sepatae (Figure 2), the cysts are essentially lined by single layer of epithelial cells with clear cytoplasm (Figure 3), and few areas with multilayering. The cells show small nuclei with mild anisonucleosis, dense chromatin, some with conspicuous nucleoli. The adjacent renal tissue was normal (Figure 4). No evidence of invasion to renal capsule, preinepheric fat and renal artery. Sections of the ureter and adrenal

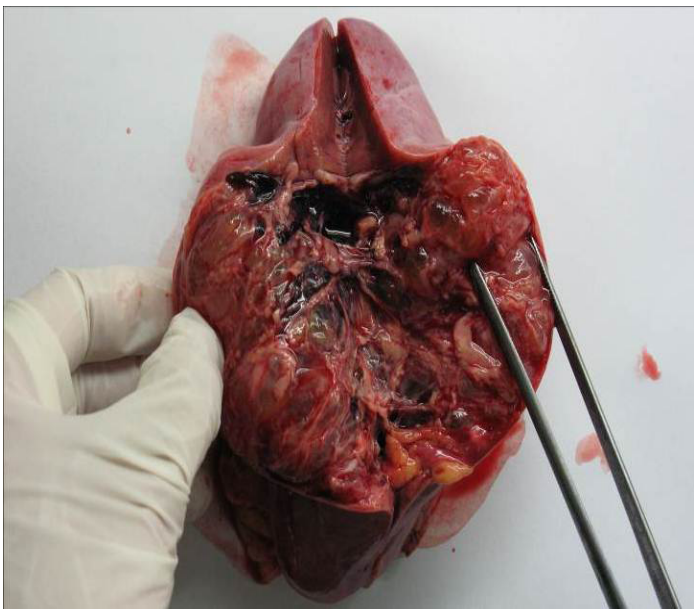

Figure 1. Cross section of the left kidney show multilocular cystic lesion. As described in the text, a cystic mass occupied the middle and lower part.

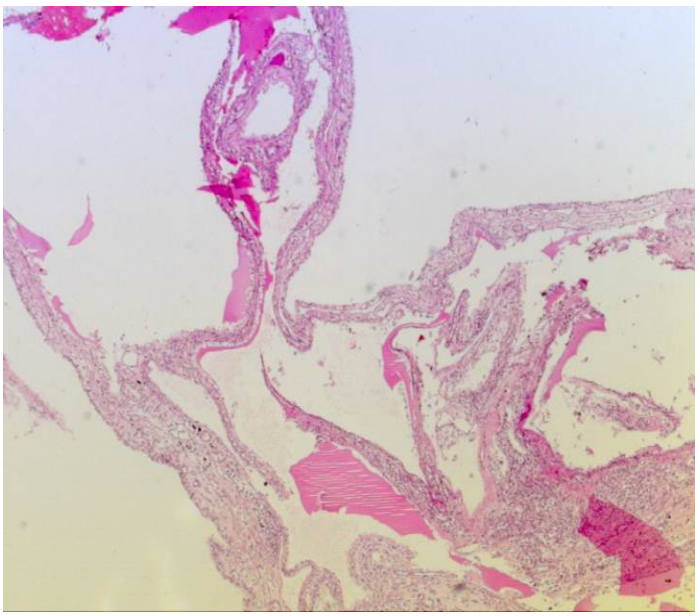

Figure 2. Photomicrograph reveal multilocular cystic lesions (x5 power) (H & E stain).

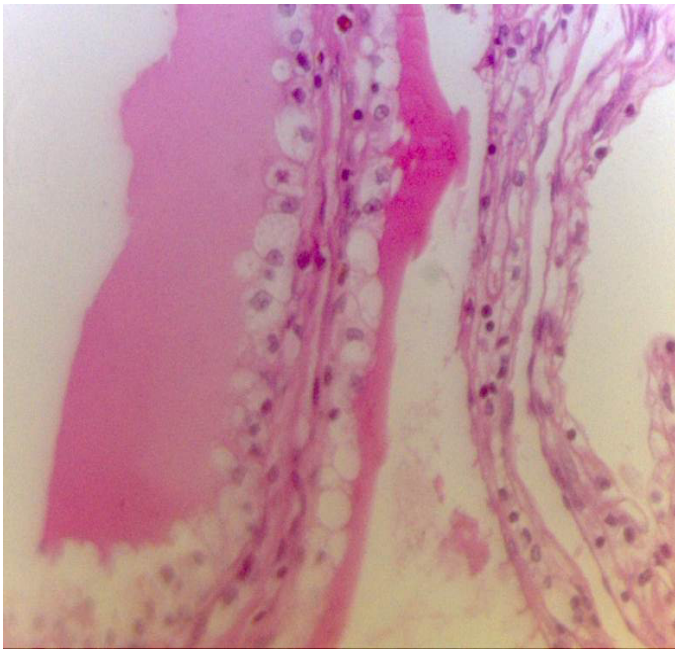

Figure 3. Thin walls of cysts covered by single layer of epithelial cells with small dark nuclei and clear cytoplasm (x40 power) (H & E stain).

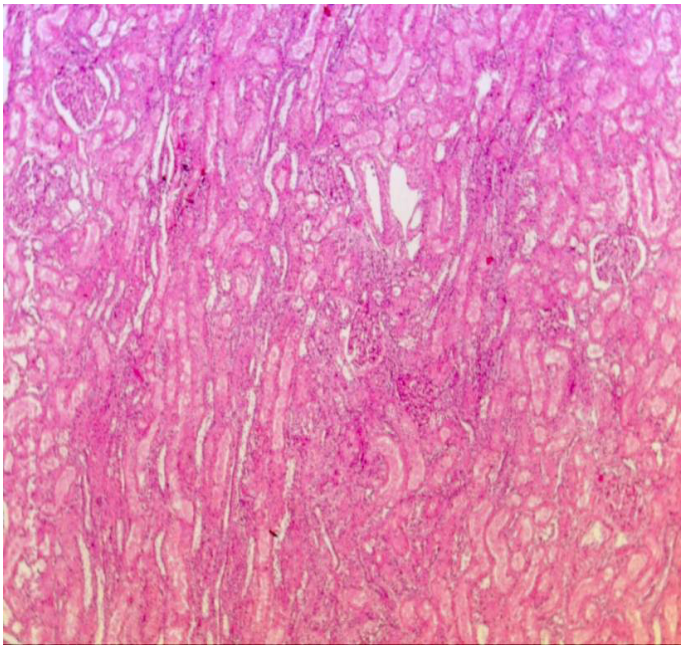

Figure 4. The adjacent normal renal parenchyma (x40 power) (H & E stain).

glands were free of tumor. The features are consistent with Fuhrman multilocular cystic renal cell carcinoma Grade 2 (TNM staging would have been T1a N0 Mx).

Outcome

The post-operative period was uneventful and the patient went home one week after the operation. The patient was referred to the oncology department for regular follow-up every 3 months with no further treatment. No sign of local recurrence were reported up to a year after the operation.

Discussion

Multilocular cystic renal cell carcinoma is an uncommon renal neoplasm with low malignant potential and its histogenesis is unclear (1,2). It is considered as a subtype of clear cell carcinoma because of resemblance of neoplastic cells to classical clear cells, VHL gene mutations and chromosome 3p deletion status similar to clear cell RCC (1). MCRCC is characterized by variable-sized, non-communicating cysts separated by irregular thin walled septa with no expansive tumor nodules. MCRCC is slightly more commonly seen in men and tumor less than 4 cm is more likely to have Fuhrman grade 1 (4). Our patient was also a male but the Fuhrman grade 2 as maximum tumor dimension of 6 cms. No local spread or metastasis were recorded. Our patient fulfilled the other classical features of MCRCC tumors. They are usually seen incidentally in 67% cases and symptomatic cases present usually as loin pain with mass and only rarely present with hematuria (3). Relationship of HIV and hepatitis virus with MCRCC is not well established. MCRCC tumors are noted for their excellent clinical outcome with 5 year survival of 100% (4). This may possibly be attributable to their innate features of circumscribed tumor, smaller size, low Fuhrman nuclear grade and lack of metastasis (5). Nassir et al observed no recurrence on 42 months of follow up, even with partial nephrectomy cases in 33% of their 12 cases (3). The differential diagnosis of unilateral focal cystic renal mass in an adult should include multilocular cystic nephroma, malignant change in a multilocular cystic nephroma, MCRCC, conventional RCC with cystic degeneration, mixed epithelial and stromal tumor of kidney (6-8). In addition, non-neoplastic lesions like hydatid cysts and renal tuberculosis with cystic degeneration should be considered to0 (6-8). MCRCC is characterized essentially by non-communicating cysts with thin fibrous septae lined by clear cells, without any expansive nodules, as exemplified by the present case. On the other hand, RCC with cystic degeneration shows classic variegated pattern consisting of yellowish solid part with haemorrhage, necrosis and cystic change without lining epithelium. In 4-15% of renal cell carcinoma patients, cystic changes that appear on the

imaging studies are mainly due to degenerative process within the neoplasm (9). MCRCC neoplastic cells are likely to react with distal nephron markers like mucin 1 (MUC1) unlike those of RCC with cystic degeneration, which react with proximal nephron markers like lymphotoxin α (LTA) reflecting their different histogenesis (10). In cystic nephroma, the cysts are lined by flat epithelium without clear cells. Though they show focal solid area with classic clear cell carcinomatous change (7). Careful imaging studies to know the size and type of unilateral cystic renal disease are warranted to enable the biopsy of representative areas to demonstrate the exact pathology. This may facilitate planning for proper conservative nephron sparing surgery or radical nephrectomy accordingly. Though the preoperative diagnostic dilemma persists in a few cases, surgery remains the treatment of choice in the management of multilocular cystic tumors of the kidney. Some workers suggested to rename MCRCC as "multilocular cystic renal cell neoplasm of low malignant potential" to reflect its excellent clinical outcome.

In conclusion, despite the relatively benign nature of MCRCC, urologists should be constantly aware of such entity because non-specific clinical findings and poor contribution of medical imaging examinations to make the right preoperative diagnosis.

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