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Case report

Per Magna Cystic Renal Cell Carcinoma

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Abstract

Renal cell carcinoma (RCC) rarely presents with a cystic appearance. Its cystic form accounts for 3-14% of all RCC. We present a rare case of an enormous 30 cm in diameter cystic renal cell carcinoma (cRCC) that was successfully removed with a nephron-sparing surgery in our institution. The pathology report revealed a 1330 g cyst, size 22x15x9 cm containing two solid tumor formations 4x3 cm and 3x2 cm located on its inner wall and diagnosed as Clear Cell Papillary Renal Cell Carcinoma (CCP-RCC) a subtype of RCC recently established by WHO in 2016. It was our goal in this case report to emphasize that occasionally RCC could present atypically by displaying unusual imaging and clinical appearances even to bizarrely large size and weight.

Keywords: per magna cystic renal tumors, RCC, clear cell papillary renal cell carcinoma, nephron-sparing surgery

Introduction

Renal cell carcinoma (RCC) represents 3-5% of all cancers being the 6th most frequently diagnosed cancer in men and the 10th in women [1].

RCC rarely presents with cystic appearance and this form accounts for only 3-14% of all RCC [2-4]. Cystic RCC commonly present at a lower stage and grade, with almost no loco regional and distant metastases at the time of presentation (localised RCC pT1-pT2, N0/x, M0/x, Fuhrman nuclear grade I-II) with a favorable prognosis after operative treatment [3]. Most patients are asymptotic and are detected incidentally. It was our aim to present an extremely rare case of a bizarrely 30 cm in diameter big and 14 L heavy cystic renal cell carcinoma and emphasize the wide range of possibilities it can present in the human body.

Material and methods

We report a case of a 58 year old male with a per magna cystic tumor of the right kidney. He presented mostly asymptomatic, with only a largely distended abdomen and a mild abdominal discomfort gradually developing in the past year. The patient had no history of any associated pain, hematuria, loss of appetite or weakness. On palpation, a non-tender tensed mass was felt on the entire anterior and lateral abdominal wall. He was aware of having a cyst in his right kidney for a minimum of 5 years incidentally discovered on a routine US examination. Due to his negligence over the time the cyst grew to bizarre size. US and CT scan revealed a massive unilocular cystic lesion in size 30x29x28 cm with extensive dislocation of the surrounding organs and no signs of loco regional lymph node or contact visceral infiltration nor distant metastases. Contrast phase CT confirmed compressed but not obstructed right kidney and ureter with a free contrast passage. A nodule enhancement within the inner cystic wall was reported by the radiologist placing it in a category IV of the Bosniak classification of renal cystic masses with a clear malignant prediction. Preoperative serum creatinine and GFR level were within the normal range. A chest X-ray was performed and revealed no abnormalities. Urinalysis was also normal.

Due to the large proportions of the cyst and the inability to access laparoscopically an open nephron sparing surgery (NSS) was performed through a right subcostal incision, seen on Figure 1. The cyst was punctured and 14 L of chocolate fluid was carefully aspirated. It was then by sharp and blunt dissection removed in-toto and the kidney residue was properly preserved. No signs of visceral metastases were found intraoperatively. Operative material (cyst and aspiration fluid) was sent to pathology for histopathology and cytology examination. The patient had a good postoperative recovery and was discharged on the fifth postoperative day.

Results

The pathology report of the specimen revealed a 1330 g cyst, size 22x15x9 cm containing two solid tumor formations 4x3cm and 3x2 cm located on the inner cyst wall. It was diagnosed as Clear Cell Papillary Renal Cell Carcinoma (CCP-RCC) a subtype of RCC recently established by WHO in 2016. In the report no malignant infiltration outside the cyst was confirmed.

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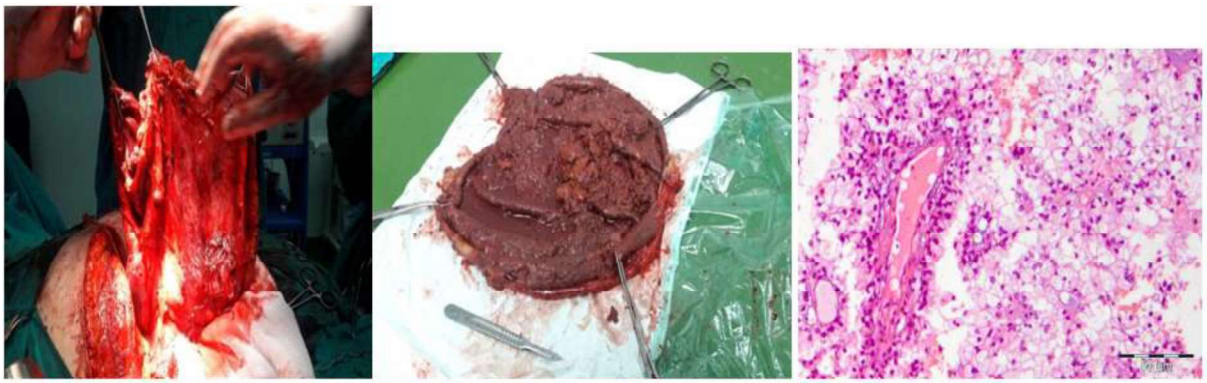

Fig. 1. Nephron sparing surgery of cystic tumor of the right kidney (H.Ex100 Papilla and clear cells)

Malignant cells were not present in micro lymphovascular area. (pTNM=pT2b pNx pMx Fuhrman II). Cytopathology report proved no cell presence in the cyst aspirate (Classification group I).

Discussion

About 50% of individuals over 50 years have cystic renal disease. However, CRCC is relatively rare [5]**.** Accurate diagnosis and treatment are sometimes difficult because clinical manifestations and imaging characteristics of CRCC can be similar to those of benign renal cystic disease. The Bosniak system of cyst classification is a widely followed system for imaging classification of renal cystic masses. It classifies renal cysts into five categories based on CT imaging appearance to predict malignant risk and advocates treatment for each category. Category I and II are treated as benign ad don't warrant treatment or follow up. Category IIF cyst are rarely malignant and request follow up up to 5 years. Category III cysts are over 50% malignant and request surgery or active surveillance. Bosniak category IV are considered clearly malignant with surgery as method of treatment. Alternatively, contrast enhanced US and MRI are additional method used in diagnosing cystic RCC [2,6].

Cystic RCC comprises a wide category of renal cancers, including multilocular cystic RCCs, unilocular cystic RCCs, RCCs with extensive cystic necrosis, and unilocular cysts with mural tumor nodules [3,4]. Cystic RCCs represent a mix of various histological subtypes similar to the solid RCC, most commonly clear cell RCC, papillary RCC and chromophobe RCC and other less common subtypes. They all commonly present at a lower stage and grade, with almost no loco regional and distant metastases at the time of presentation (localised RCC pT1-pT2, N0/x, M0/x, Fuhrman nuclear grade I-II) and come with a favorable prognosis after operative treatment. [7].

In our case a pathology report confirmed a cystic type of Clear Cell Papillary Renal Cell Carcinoma (CCP-RCC). It is a recently recognized histological subtype of RCC and encompasses only 1-5% of all RCC. That makes it the fourth most common histological subtype of RCC after clear cell RCC (ccRCC), papillary RCC (pRCCr) and chromophobe RCC (chRCC). [8,9]. Microscopicaly it is characterized by clear cell cytology in papillary architecture with a low-grade nuclei (Fuhrman grade 1 or 2). Macroscopicaly it presents mainly as encapsulated by variably thick fibrous capsule limited to the renal parenchyma (low stage localized predominantly T1). The low stage and low grade parameters of CCP-RCC and the cystic RCCs in general, provide an excellent prognosis of these tumors following surgical treatment [10,11].

Based on current available oncological and quality of life outcomes, localised RCC stage T1is best managed by partial nephrectomy (PN) (nephron-sparing surgery-NSS) rather than radical nephrectomy (RN). Partial nephrectomy can be performed, either with an open, laparoscopic or robot-assisted approach, based on surgeon's expertise and skills. Patients with localized T2 tumors not treatable by partial nephrectomy should be offered radical nephrectomy performed laparoscopically. [2,12].

Koga *et al*. [13] reported an excellent 5-year survival rate of 80-100% for cystic RCC vs 55% for conventional solid RCCs as well as Onishi *et al*. [14] with an overall 5-year survival rate of 88,6% vs 50-60% for conventional RCCs. No evidence of recurrent or metastatic disease was reported in these groups.

Conclusion

It was our goal to emphasize that occasionally RCC could present atypically by displaying unusual imaging and clinical appearances even to bizarrely large size and weight. In this particular case we encountered and removed a cystic renal tumor grown to enormous proportions of 30cm in diameter containing 14L of fluid. Due to the short postoperative terms, no imaging follow up was conducted at the moment of presenting this report.

Conflict of interest statement. None declared.

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