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Cystic Renal Cell Carcinoma vs Cystic Degeneration in Renal Cell Carcinoma: Major Disparity in Prognosis in a Seemingly Same Entity

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Received: Jul 22, 2022

Accepted: Aug 12, 2022

Published Online: Aug 15, 2022

Journal: Case Reports of Oncology: Open Access

Publisher: MedDocs Publishers LLC

Online edition: http://meddocsonline.org/

Introduction

Cystic degeneration is common in Renal Cell Carcinoma (RCC) in about 15% of the cases [1]. This has to be differentiated from the cystic variant of RCC. This is difficult due to the expanding subtypes being described as the number of cases seen and being studied are increasing. A seemingly cystic degeneration in RCC can easily be a multilocular variant of cystic RCC with solid component of less than 25%. The prognosis of the patient varies according to the diagnosis and thus the aggressiveness of the treatment needs to be tailored. Here we present such a diagnostic dilemma.

Case report

A 35 years old gentleman presented to our outpatient department with history of abdominal mass for the past 7months which was gradually progressive in size. He had no history of bowel or bladder disturbances. On palpation, there was a huge abdominal mass of about 30x20cms covering all quadrants on the left and central part of abdomen. The mass was extending to the left flank. Contrast enhanced CT of the abdomen showed a large retroperitoneal predominantly cystic mass lesion measuring 29 x 19.5 x 19cms with eccentrically enhancing nodular solid components. The left kidney was not separately visualised. The case was discussed in multidisciplinary tumor board and planned for surgery in view of the huge size and later chemotherapy based on the final histopathology. We performed an exploratory laparotomy and left radical nephrectomy.

Cite this article: Sankar KV, Sahoo PK, Agrawala SK, Panda SK, Sadangi S, et al. Cystic Renal Cell Carcinoma vs Cystic degeneration in Renal Cell Carcinoma: Major Disparity in Prognosis in a Seemingly Same Entity. Case Rep Oncol Open Access. 2022; 1(1): 1002.

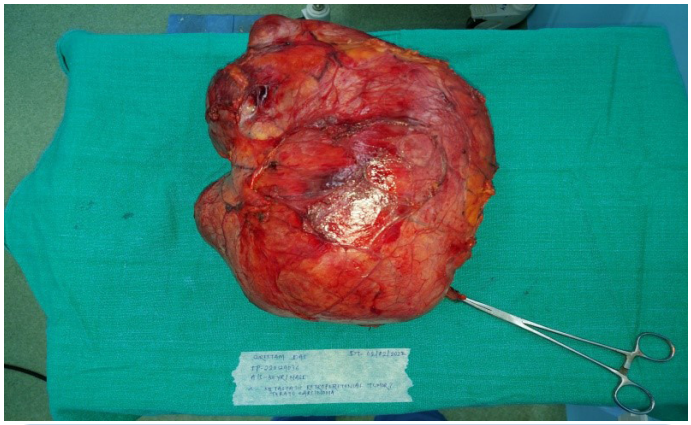

Figure 1: The excised left nephrectomy specimen.

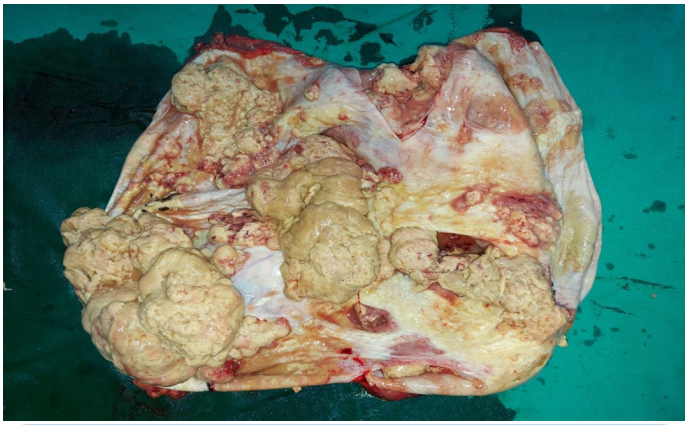

Figure 2: Cut open specimen (after drainage of fluid).

The specimen weighed approximately 7kgs and 4litres of light brownish cystic fluid was drained after giving multiple nicks. On cutting open the specimen, multiloculated cystic renal mass with peripheral solid components was noted with no gross renal tissue being identified. The cut open specimen is shown in Figure 2.

Final Histopathology was suggestive of Renal cell carcinoma, unclassified, ISUP grade 4. Due to the scarcity of renal tissue in the specimen, immunohistochemistry (IHC) has been used to arrive at the diagnosis. The IHC images are shown in Figure 3.

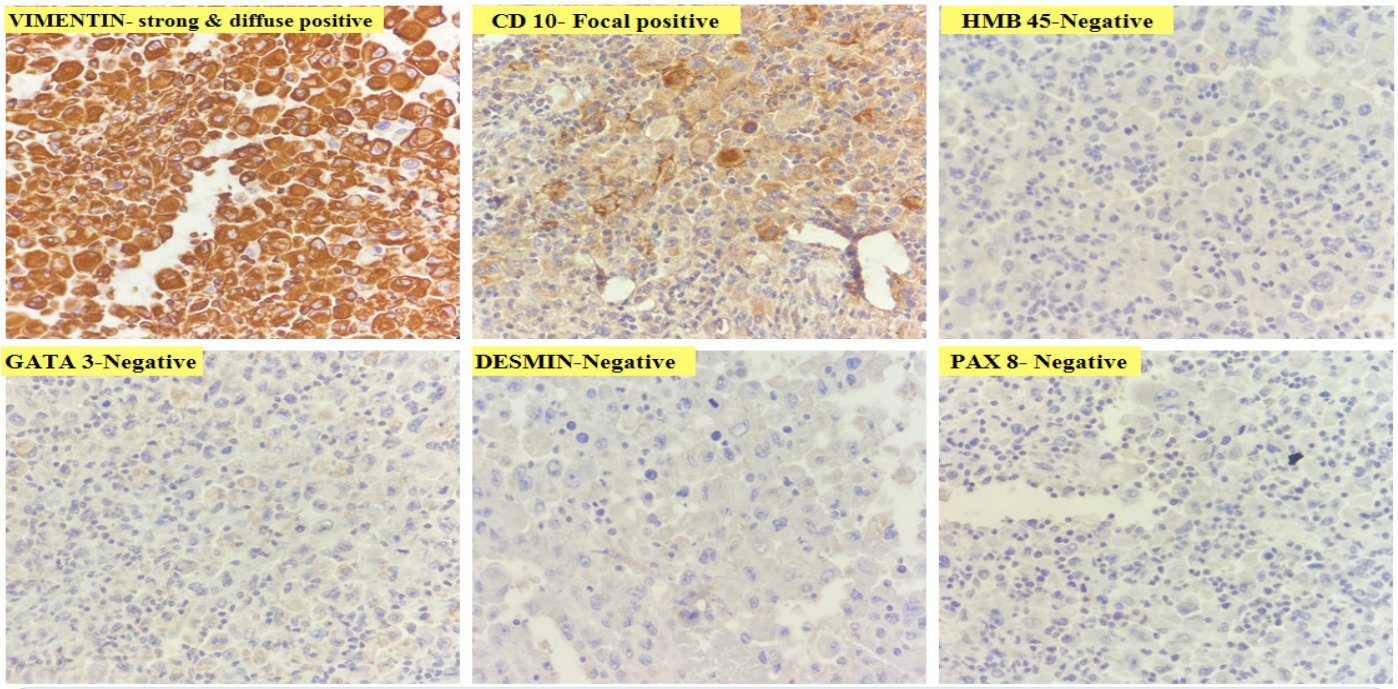

Figure 3: IHC images.

Post surgery, recovery of the patient was uneventful and discharged in stable condition. The patient rapidly developed an Inferior Venacava (IVC) thrombus which extended to the bifurcation of iliacs and caused swelling of bilateral lower limbs. He was started on therapeutic dose of low molecular weight heparin and chemotherapy and planned for IVC filter placement. Later he lost to follow up.

Discussion

Renal cell tumors with cystic changes were divided into 4 groups by Hartman et al [2]. Recent series have focused on the outcomes of unilocular and multilocular tumors based on the definition that the cystic component of these tumors constituted at least 75% of the tumor without evidence of necrosis [3-5]. It is important to distinguish tumors with cystic necrosis from unilocular or multilocular cystic RCCs. Tumors with cystic necrosis carry a significantly worse prognosis [4,6]. Furthermore, necrosis has recently been identified as a predictor of poor outcome in patients with kidney cancer [7,8]. Fuhrman grading system is the universally used grading system for renal tumors but recently it has been recommended that for clear cell RCC and papillary RCC, grading should focus upon nucleolar prominence [9,10]. This gave way to the International Society of Urological Pathology (ISUP) grading system [11]. The tumors were graded 1 to 3 according to the degree of nuclear prominence—ie, grade 1: absent or inconspicuous nucleoli at -400 magnification; grade 2: nucleoli conspicuous at -400 magnification but inconspicuous or invisible at -100 magnification; and grade 3: Nucleoli conspicuous at -100 magnification. Tumors showing extreme nuclear pleomorphism with or without multinucleate tumor giant cells were assigned grade 4, although tumors with a sarcomatoid or rhabdoid component were also assigned grade 4.

Conclusion

The presence of cystic component in an RCC is a prognostic dilemma. Cystic RCC has very good prognosis as they tend to present with small size, lower T stage and low grade whereas cystic degeneration in RCC has poor prognosis especially when associated with tumor necrosis. Better characterisation of differences in radiological features and gross features might help in prompt diagnosis and aggressive treatment.

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