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A QUANDARIUS CASE OF KIDNEY TUMOR-A MORPHOLOGICAL ILLUSION

Pathology

Sayantan Paul* 3rd Year PGT, MD Pathology, Burdwan Medical College. *Corresponding Author
Moulima Banerjee Demonstrator, Department of Pathology, Burdwan Medical College.
Rupam Karmakar Professor & Head, Department of Pathology, Burdwan Medical College.

ABSTRACT

Introduction: Acquired cystic kidney disease associated renal cell carcinoma is a rare clinical entity with unique and specific histological features, commonly but not always associated with dialysis patients. **Case Report:** A 33-year-old female patient presented to the general surgery outdoor with complaints of lower abdominal pain, nausea and burning sensation in lower abdomen for last two months. CECT whole abdomen was performed, and it revealed a homogenously enhancing mass lesion over upper pole of left kidney with some foci of calcifications. Left sided radical nephrectomy was performed subsequently. Gross examination of the specimen showed a greyish white tumor located in the upper pole of left kidney. Histopathological examination revealed a high-grade malignant tumor showing large eosinophilic cells with prominent nucleoli, some areas of reticular and clear cell differentiation along with large numbers of intratumoral calcium oxalate crystals. It also showed a sieve-like architecture characterized by inter and intracellular lumina separated by thin strands of cytoplasm. Perinephric fat, fascia of gerota, and pelvicalyceal systems were not involved. A diagnosis of "Acquired Cystic Disease Associated Renal Cell Carcinoma" was provided. **Conclusion:** The outcome data for acquired cystic kidney disease associated renal cell carcinoma remains extremely limited. While most follow an indolent course, a few of them have shown themselves to behave aggressively and metastasize. Because of unique histological features and different clinical outcomes, it is important to recognize this tumor as a distinct renal cell tumor entity.

KEYWORDS

Acquired cystic kidney disease associated renal cell carcinoma, sieve like architecture, calcium oxalate crystals.

INTRODUCTION:

Our patient, a 33-year-old female presented with complaints of lower abdominal pain followed by accidental trauma. She was also suffering from burning sensation and pain in the lower abdomen for last 2 years. After initial clinical examination, several differential diagnoses were put forward including chronic urinary tract infection, kidney or ureter stone, a ruptured cyst and gynaecological problems like endometriosis. Rare possibilities like malignancy of the urinary or reproductive system were also kept in mind. To determine the cause, patient immediately underwent a CT whole abdomen, which revealed a homogenously enhancing mass in the upper pole of left kidney along with a cystic lesion in segment VII of liver. With these findings in mind, renal cell carcinoma was suspected as a principal differential. As the kidney cyst was accompanied with a hepatic cyst, autosomal dominant polycystic kidney disease was also considered a possibility. A left sided radical nephrectomy was performed. Microscopic examination revealed a high-grade renal cell carcinoma with predominantly cribriform architecture with intratumoral calcium oxalate deposits. Several different morphological diagnoses for subtyping were put forward including clear cell renal cell carcinoma (RCC), translocation associated RCC and acquired cystic kidney disease associated RCC (ACKD RCC). After putting together all the puzzle pieces, acquired cystic kidney disease associated RCC was signed out as the final morphological diagnosis.

Case Report:

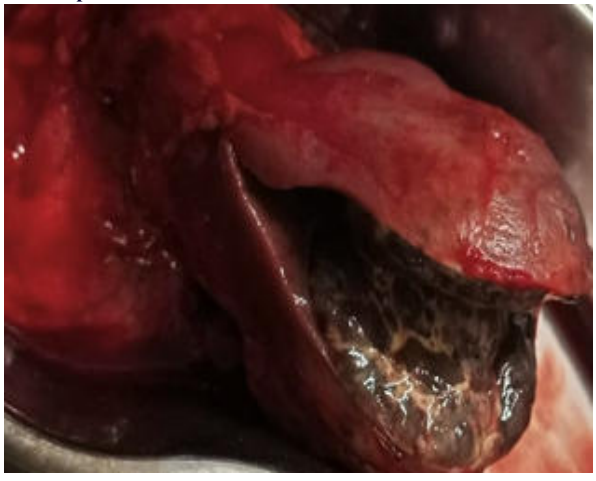

Fig 1: A greyish multiloculated cystic tumour

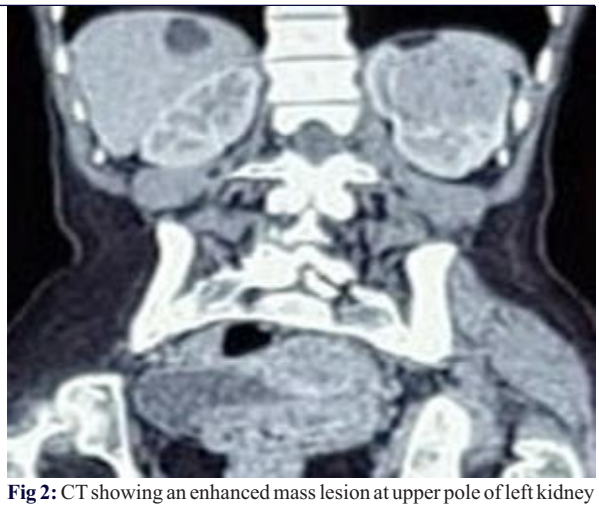

Fig 2: CT showing an enhanced mass lesion at upper pole of left kidney

A 33-year-old female patient presented to the general surgery outdoor with complaints of lower abdominal pain followed by accidental trauma. She also had history of burning sensation and pain in the lower abdomen for last 2 years. The patient was not on dialysis and did not have any history of end stage kidney disease or renal transplant. CT whole abdomen revealed a homogenously enhancing mass lesion (approximately 7 cm * 5 cm size) in the upper pole of left kidney. It also showed a cystic lesion measuring 2.1*1.9 cm in segment VII of liver. The renal lesion had extended to the renal sinus without any hydronephrotic changes. A left sided radical nephrectomy was performed.

Gross examination of the specimen showed left kidney with perinephric fat and gerota's fascia measuring14.0*8.0*3.0 cm. On cut section, it revealed a greyish multiloculated cystic undermeasuring 6.5*5.0*4.0 cm located in the upper pole of left kidney (Fig 1). Pelvicalyceal systems and renal sinus were free of tumor. The renal vascular cut margin was 5.5 cm away and ureter cut margin was 6.5 cm away from the tumor.

Microscopic examination revealed a tumor with sieve like architecture with multiple intra and intercellular holes (Fig 3), along with clear cell morphology in some areas. Multiple large scattered eosinophilic rhabdoid cells (Fig 5) were present. The most unique morphological feature was some intratumoral crystals identified as calcium oxalate

crystals (Fig 4). Many differential diagnoses were put forward including clear cell RCC, translocation associated RCC, ACKD RCC and then putting together all the puzzle pieces the diagnosis of Acquired Cystic Disease Associated Renal Cell Carcinoma (ACKD RCC) was made.

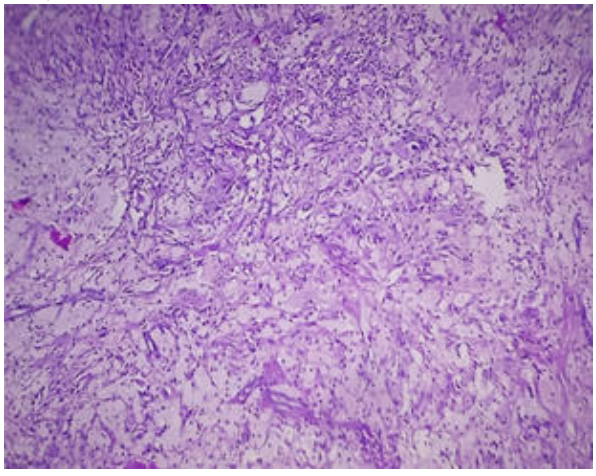

Fig 3: H&E, 100x magnification, showing characteristic sieve-like architecture with inter and intracellular spaces

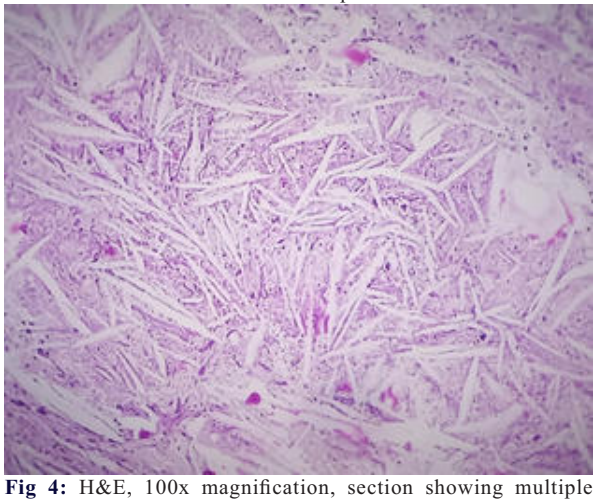

Fig 4: H&E, 100x magnification, section showing multiple intratumoral calcium oxalate crystals.

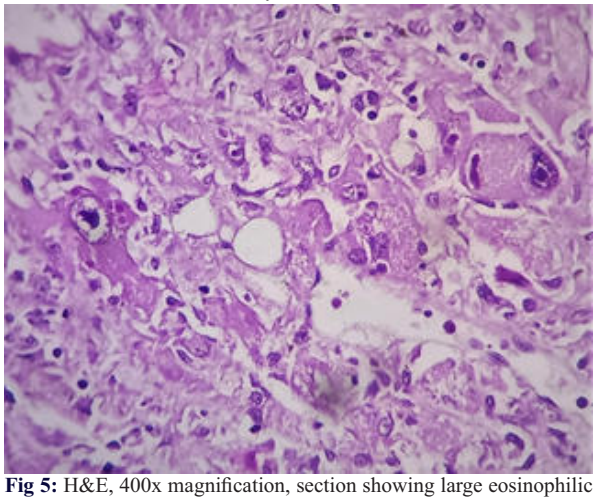

Fig 5: H&E, 400x magnification, section showing large eosinophilic rhabdoid cels.

DISCUSSION:

In a patient without polycystic kidney disease, ACKD is characterized as having at least three cysts per kidney or cysts that make up more than 25% of the renal parenchyma. Renal cell cancers occur in about 3–7% of ACKD patients. ACD-RCC is the most common RCC subtype, even though several other subtypes have been identified in the context of ACKD. Only individuals with ACKD develop ACD-RCCs, which make for 36% of all ESRD epithelial neoplasms and 46% of those in ESRD with ACKD(Chrabańska et al., 2021)

The differential diagnoses of this case are varied, but the most important ones are clear cell renal cell carcinoma, papillary renal cell carcinoma, chromophobe renal cell carcinoma and translocation associated RCC. ACKD RCC may have areas of both clear cell differentiation and papillary architecture. Granular eosinophilic cells and solid architecture are also observed in ACKD RCC. But the characteristic sieve-like architecture and intratumoral calcium oxalate crystals can help to give the diagnosis in favour of ACKD RCC(Abdelwahed & Tretiakova, 2025).

Both ACKD RCC and clear cell RCC can show areas of clear cell differentiation but finding other areas with typical sieve like morphology and intratumoral calcium oxalate crystals can help in establishing diagnosis of the former entity(Abdelwahed & Tretiakova, 2025).

Similarly areas of papillary architecture and granular eosinophilic cells with solid architecture may be present in this entity, resembling papillary and chromophobe RCC respectively, However, these areas will always be accompanied by other areas showing classical ACKD RCC morphology(Abdelwahed & Tretiakova, 2025).

Translocation associated RCC (TFE3 rearranged and TFEB altered) can also mimic this entity as both are high grade RCC with varying morphological similarity. However, the former one classically presents with a triad of papillary architecture, large pale or clear cells with prominent nucleoli and psammoma bodies. As these features were not present in our case, this possibility was also ruled out(Rizkalla & Tretiakova, 2025).

The predisposing factors for this type of renal tumor generally but not always are patients with history of end stage kidney disease, long term haemodialysis and renal transplant history(Kojima et al., 2020).

Our case represented a very unusual presentation where the patient was a middle-aged female with no history of end stage kidney disease, haemodialysis and renal transplant. Thus, this entity should be kept in mind when a cystic renal tumour shows sieve like architecture and intratumoral calcium oxalate crystals under microscopy.

While ACKD RCC generally follows an indolent course, several have shown to metastasize and behave aggressively. Presence of rhabdoid and sarcomatoid morphology typically are associated with an aggressive course(Kuroda et al., 2011). Long dialysis sessions, tumor size, pathological stage, grade 4 tumor, and lymphovascular invasion were also indicators of adverse outcomes(Kondo et al., 2018).

ACKD RCC is characteristically known to show positivity for AMACR, PAX 8 and CD 10 while being negative for CD 57, CD 68 and PAX 2(El-Zaatari & Truong, 2022). However, these are not specific markers and show overlap with other common renal cell neoplasms.

We could diagnose our case based solely on morphology as it presented with classical features such as sieve-like architecture and intratumoral calcium oxalate crystals. In difficult cases without classic morphology, immunohistochemistry and molecular study can prove to be valuable diagnostic tool.

CONCLUSION:

There is still a dearth of outcome data for renal cell carcinoma linked to acquired cystic kidney disease. A small number of them have been proven to act aggressively and spread, while the majority have a generally slow course. It's critical to identify this tumor as a distinct renal cell tumor entity due to its distinctive histologic characteristics and potentially varied therapeutic consequences.

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