



# Renal Cell Carcinoma, Clear Cell Type with Rhabdoid Differentiation

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## ABSTRACT

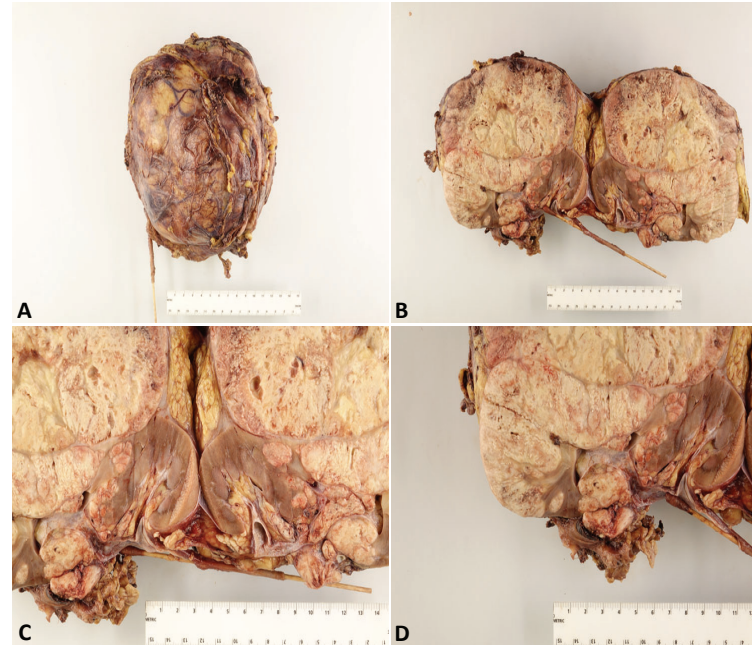
Renal cell carcinoma (RCC) with rhabdoid differentiation is a rare, but aggressive, clinicopathologic phenotype of RCC that is often associated with a higher risk for metastasis and a very poor prognosis. Presented is a case of RCC with rhabdoid differentiation in a 66-year-old female patient. The specimen contained a 17.0 cm, multifocal mass with an extensive amount of necrosis located mainly in the lower pole with extension to the middle aspect and upper pole. The mass was found to involve the renal pelvis, the sinus fat, branches of the renal vein and the perirenal adipose tissue. Also identified was a tumor thrombus in the renal vein which accompanies the diagnosis of RCC in approximately 10% of cases. The tumor was staged as a T3a and the patient was diagnosed with RCC, clear cell type with rhabdoid features and extensive necrosis. The large amount of necrosis prompted the Pathologists' Assistant to increase the sampling size in order to collect all viable tissue that could contain important diagnostic features which, in this case, was the presence of rhabdoid differentiation. Although such features does not constitute a specific morphogenetic subtype of RCC, it could signify early patterns of dedifferentiation which is associated with a poor clinical outcome.

## CLINICAL HISTORY

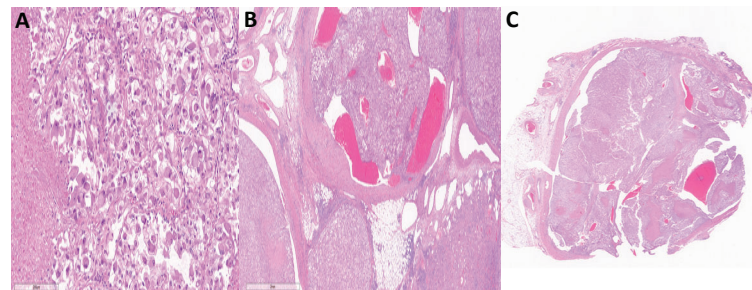
The following case is that of a 66-year-old female patient who presented with abdominal pain, nausea, early satiety and a 55 lb weight loss in late December of 2019. A CT revealed a 14.0 cm left renal mass and the patient underwent a radical nephrectomy in January 2020.

## GROSS DESCRIPTION

The specimen is received in formalin labeled with patients name, initials, medical record number and "1. Left renal mass + renal vein." It consists of a 1561.2 g, 18.5 x 15.7 x 13.0 cm left radical nephrectomy specimen with an attached 5.6 x 1.5 x 1.4 cm adrenal gland. The ureter is 10.5 cm in length by 0.5 cm in diameter, patent and unremarkable. The renal artery is 2.2 cm in length by 0.4 cm in diameter and the renal vein is 1.0 cm in length by 1.2 cm in diameter. Both the renal artery and vein appear to be involved by the mass. The outer surface of the specimen is inked black and bisected at the hilum to reveal a 17.0 x 11.5 x 10.5 cm tan-yellow, focally hemorrhagic, friable, ill-defined mass located predominantly in the lower pole with extension to the middle aspect and upper pole. The mass is multifocal and contains approximately 90% necrosis. It appears to involve the renal sinus and renal pelvis. The remaining uninvolved renal parenchyma is tan-brown with a distinct corticomedullary junction. The adrenal gland is uninvolved. Also identified are 3 tan, firm lymph nodes ranging from 0.8 x 0.6 x 0.4 cm to 1.5 x 0.6 x 0.4 cm.



**Figure 1:** Left radical nephrectomy specimen. **A**, anterior aspect of overall specimen. **B**, something. **C**, multifocality and involvement of sinus fat. **D**, renal vein tumor thrombus.



**Figure 2:** Renal cell carcinoma, clear cell type. **A**, rhabdoid cells. **B**, renal sinus fat invasion. **C**, renal vein tumor thrombus adherent to the vein wall

## FINAL DIAGNOSIS

The patient was diagnosed with Renal cell carcinoma, clear cell type with rhabdoid features and extensive necrosis. The tumor was found to invade the renal pelvis, renal vein branches and sinus fat. Although a renal vein thrombus was present, the renal vein margin was negative for tumor. The renal artery and ureter margins were also negative for tumor and there was no lymphovascular invasion present. Lastly, all three lymph nodes and the adrenal gland was benign. Based on the size of the tumor and the structures involved, the tumor was staged as a T3a, N0, MX.

## DISCUSSION

The most significant prognostic factors for RCC are extent of invasion into surrounding tissues and tumor size. However, the key to diagnosing this patient relied heavily on obtaining an adequate number of sections of the tumor itself to identify the presence of rhabdoid cells. This feature is significant because RCC with rhabdoid differentiation is a highly aggressive neoplasm associated with a poor clinical outcome including a higher risk of recurrence and a higher mortality rate. The presence of rhabdoid features is important also because it has been shown to express p53 more frequently and diffusely than conventional RCC. Typically, p53 is overexpressed with a high mutation rate in sarcomatoid components. Sarcomatoid foci, which are composed of high-grade tumor spindle cells, have been well classified as aggressive components encountered in all subtypes of RCC. It is suggested that rhabdoid features may be related to sarcomatoid transformation as a beginning pathway of tumor dedifferentiation which contributes largely to its aggressive nature.

The presence of rhabdoid differentiation in RCC should not be overlooked. Its presence should be reported even if the rhabdoid area is limited. It is the Pathologists' Assistant that plays a pivotal role in accurately assessing the specimen and collecting the necessary sections needed to not only stage the tumor but to identify unique characteristics such as the rhabdoid features discovered in this case.

## REFERENCES

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