It's Gross to Me: The role of the PA in diagnosing a renal cell carcinoma, clear cell type with rhabdoid differentiation

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Introduction

Renal cell carcinoma (RCC), clear cell type with rhabdoid differentiation is an uncommon, aggressive tumor most commonly found in adult men over the age of 40. The rhabdoid component of the tumor is thought to be due to dedifferentiation of the parent cells and can present in other types of renal cell carcinomas, though it is most often associated with clear cell type.

These tumors usually present with minimal symptoms and metastasize quickly, making it difficult to identify and treat the tumor. The late presentation, low incidence of this tumor, and aggressive nature have made it a challenge to establish a standard treatment for this tumor type, and studies have shown minimal success in reduction of mortality with various treatment options.

Clinical History

A 68-year-old male presented to the emergency department with severe hematuria, anemia, and hypertensive urgency with an unremarkable history. Abdominal imaging studies showed a large left renal mass, a left renal vein thrombus, and a left lower lung lobe mass and the patient subsequently underwent a left radical nephrectomy.

Methodology

The surgical pathology department received a left radical nephrectomy specimen that was bivalved to reveal a 10.4 x 10.2 x 9.7 cm soft, bulging, partially encapsulated pale tan mass (figure 1) with focal hemorrhage extending from the superior pole, obliterating more than 90% of the renal parenchyma and the calyceal system. The mass extended into the superior pole sinus fat, the renal vein (figure 2), and beyond the renal capsule but did not grossly extend beyond Gerota's fascia.

Histologic assessment of the mass showed characteristic features of RCC, clear cell type with rhabdoid features, including extreme nuclear pleomorphism, discohesive eosinophilic rhabdoid cells, and 60% necrosis. Immunohistochemical results were consistent with the diagnosis, positive for CD20 and vimentin and negative for cytokeratin 7 and desmin.[1,2] Confirmation of invasion into the renal vein, renal sinus fat, perinephric fat, and regional lymph nodes (figure 3 and 4), but no invasion beyond Gerota's fascia staged the tumor as pT3a, pN1.[3]

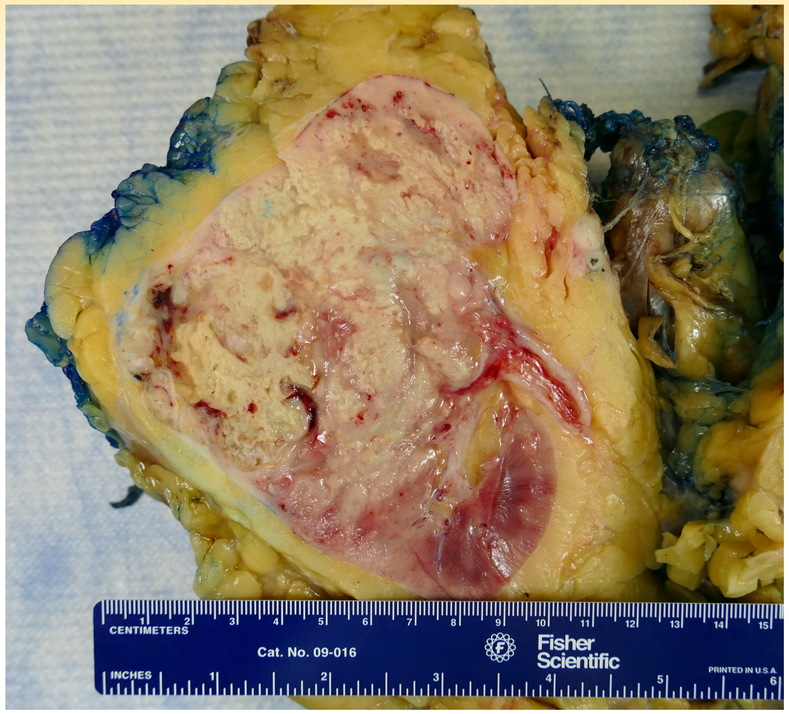

Figure 1. Bivalved specimen showing overall mass

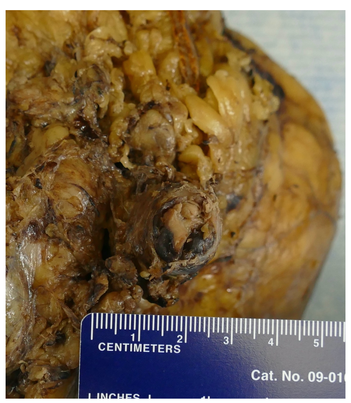

Figure 2. Renal vein occluded by tumor

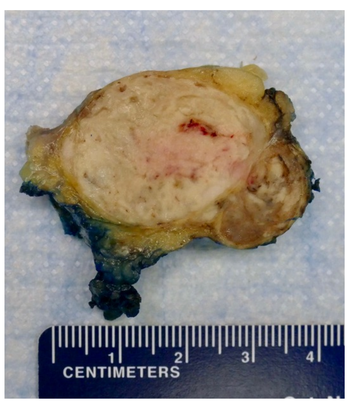

Figure 3. Grossly positive regional lymph node

References

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2: Godken N, Nappi O, Swanson P, et. Al. Renal Cell Carcinoma with Rhabdoid Features. *The American Journal of Surgical Pathology*. October 2000; 24:1329-1338

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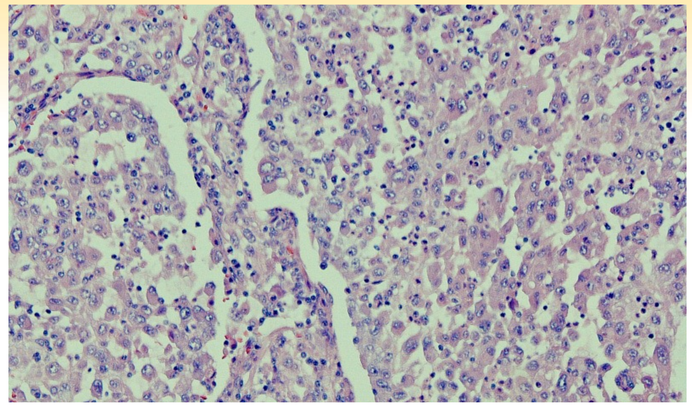

Figure 4. Histologic appearance of fig. 3 lymph node, displaying tumor features with nuclear pleomorphism and eosinophilic rhabdoid cells

Discussion

The pathologists' assistant (PA) plays a vital role in the accurate diagnosis of renal neoplasms, which is particularly emphasized in the case presented. Staging markers established by the College of American Pathologists allow for clear guidance in the development of grossing guidelines put forth by the American Association of Pathologists' Assistants (AAPA).[3] Critical components of these guidelines include demonstration of tumor with respect to the renal vein, sinus fat, and perinephric fat, in addition to tumoral invasion of lymph nodes, the adrenal gland, or beyond Gerota's fascia. Many of these aspects are shown in figures 1-3 and were crucial in the diagnosis of this patient, highlighting the importance of the PA in their knowledge and competency of the given AAPA grossing guidelines.

Conclusion

RCC, clear cell type with rhabdoid features is a rare subtype of renal neoplasm that is extremely aggressive and presents with minimal symptoms. These tumors carry an increased risk of metastasis as compared to other renal cell carcinoma types and patients frequently have metastases at the time of presentation. The PA plays a critical role in the diagnosis of these neoplasms due to the prognostic importance of numerous gross features. Because of the aggressive nature, late presentation, and lack of research due to the low incidence, treatment options are limited for this tumor and have low success rates.