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Diagnosis: Kidney, Right, right kidney (Nephrectomy):
Renal Epithelioid Angiomyolipoma with Atypical Features. The tumor is organ confined and does not have any metastases.

Note: This tumor consists of ~30% epithelioid cells with marked cytologic atypia and extensive necrosis. The mitotic index is $< 1/10$ HPF, however atypical mitoses are identified. The clinical behavior of these tumors is variable, but they may be malignant and metastases can occur. Given the extensive necrosis, atypical mitoses and high degree of cytologic atypia in this case, close clinical follow-up is recommended. In the setting of an angiomyolipoma, somatic sequencing may be useful to document these alterations for future targeted therapy in the case of recurrence. Given the patient's young age, clinical and genetic screening for tuberous sclerosis complex may be warranted, though most of these tumors arise sporadically and the background genetics are remarkable in this case.

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