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Renal Neuroendocrine Tumors: A Single Center Clinicopathologic and Molecular Analysis

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BACKGROUND

Renal neuroendocrine tumors (NETs) are a particularly rare subset of NETs, with fewer than 100 cases described in the literature. Of these, next generation sequencing (NGS) findings have been described in fewer than 10 cases, all but one of which had grade 1 disease.

METHODS

A single-center, IRB-approved retrospective analysis of patients with grade 2 or 3 well-differentiated renal NETs treated at our institution from 2016-2023 for whom NGS data was available was conducted. Clinical, radiographic, and pathologic data including NGS were abstracted from the electronic medical record.

RESULTS

A total of three patients were found and the median age at diagnosis was 42.7 years (range 24.2-51.7 years). The presenting symptom was back/flank pain in two patients and hematuria in the third. One patient had a horseshoe kidney. All patients had large primary renal tumors measuring >9cm at the time of diagnosis and metastatic disease at or shortly following diagnosis. Clinical, radiologic, and molecular findings are described in the table below.

Table 1: Clinicopathologic and molecular characteristics

	Patient 1	Patient 2	Patient 3
Tumor Grade (Ki-67 index)	2 (8%)	3 (>20%)	3 (22%)
Pathogenic mutation	TP53	None	None
Dotatate PET Imaging uptake	Heterogenous	Homogenously +	Heterogenous
Nephrectomy	Yes	Yes	No
Liver Debulking	No	Yes	Yes
Liver embolization	Yes	Yes	No
Systemic therapies	SSA	SSA, Lu177*, Lenvatinib*	SSA, everolimus, CT
Alive	Yes	Yes	No
Diagnosis to last follow-up (years)	1.7	10.5	2.1

Abbreviations: PET positron emission tomography, SSA somatostatin analogue, Lu177 lutetium 177 dotatate, CT capecitabine/temozolomide.
*confirmed radiographic response

CONCLUSIONS

This retrospective analysis includes to our knowledge the largest cohort of patients with grade 2 or 3 renal NETs for whom NGS analysis is reported to date. In this cohort, known pathogenic mutations were infrequent and no actionable mutations were found. These tumors demonstrated heterogeneous clinical behavior and radiographic characteristics.

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