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Primary Neuroendocrine Neoplasms of the Kidney, A Distinct Entity but Classifiable-Like the Gastroenteropancreatic Neuroendocrine Neoplasms

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BACKGROUND: Primary neuroendocrine neoplasms of the kidney are a distinct and rare entity, but classifiable-like the gastroenteropancreatic neuroendocrine neoplasms. Due to rarity of these tumors, not much is known about histopathology and behaviors of these tumors. We attempted to review pathology of primary kidney neuroendocrine tumor patients at our institution.

METHODS: Retrospective chart review identified 8 primary kidney neuroendocrine tumors from Siteman Cancer Registry database from 1/1/2000 until 1/1/2018. Pathology review was done for all the patients to confirm their diagnosis and other pathological features.

RESULTS: In our cohort, we identified eight cases of neuroendocrine neoplasms of the kidney. Three of the cases were poorly differentiated neuroendocrine carcinoma. All cases of well-differentiated neuroendocrine tumor (either grade 1 or grade 2) were identified in females (age range 44 – 60). All the tumors characteristically extended to the perirenal fat. These tumors showed diffuse positivity for synaptophysin, variable positivity for chromogranin, and did not stain for markers specific for renal differentiation (PAX-8). The growth pattern in well differentiated neuroendocrine tumors is predominantly trabeculated, but a diffuse plasmacytoid growth is also noted, which is unusual in gastroenteropancreatic neuroendocrine neoplasms. Two patients had Primary tumor measuring 9 cm and 14 cm respectively also presented with positive lymph nodes and lymphovascular invasion developed metastatic disease in 2 years.



CONCLUSION: Primary kidney neuroendocrine tumors are very rare. These tumors also do not stain for markers-like renal cell carcinoma (negative for WT-1, AMACR). The growth pattern in well differentiated neuroendocrine tumors is predominantly trabeculated, but a diffuse plasmacytoid growth is also noted, which is unusual in gastroenteropancreatic neuroendocrine neoplasms.