Renal Neuroendocrine Tumors: Definitive Surgery is the Cornerstone of Longevity

Andrew Nguyen1; James De Andrade1; Christopher Larocca1; Camille Stewart1; Phillip Ituarte1; Jonathan Kessler1; Gagandeep Singh1; Sue Chang1; Daneng Li1

1City of Hope

BACKGROUND: While pancreatic, lung, and gastrointestinal primary neuroendocrine tumors are uncommon, renal neuroendocrine tumors are many times rarer with less than 100 cases reported in the literature. The natural history of these tumors has not been well characterized. We performed the first population-based study that describes patient and tumor characteristics as well as survival patterns of patients diagnosed with renal neuroendocrine tumors.

METHODS: A retrospective study from the SEER database including patients from 1973 to 2014 with a histologic diagnosis of a primary renal neuroendocrine tumor was performed. Patients were stratified by stage and operative versus non-operative management. Kaplan-Meier and multivariate Cox proportional hazards analyses were performed.

RESULTS: 114 patients were identified with primary renal neuroendocrine tumors. For all patients, 5-year overall survival was 62% and disease-specific survival was 67%. Median age at diagnosis was 56, where patients who were managed operatively were younger (operative, median age 53 versus non-operative, median age 65; p=0.0019). Those who underwent operative management had a 5-year overall survival of 78% and a disease-specific survival of 82%, whereas those with non-operative management had a 5-year overall survival of 28% and disease-specific survival of 34%. In addition to being younger, patients who underwent operative management had earlier stage disease (63/80, 78.8% patients with local or regional disease). In the 34
patients who did not have an operation, 26 patients (76.5%) had distant disease at presentation. On univariate analysis, operative management reduced risk of all-cause mortality (HR 0.19, 95% CI 0.11-0.34, p<0.001), yet in the multivariate model, this risk reduction was no longer significant. Younger age, lower tumor grade, local disease was associated with reduced risk of mortality in both univariate and multivariate analyses.

**CONCLUSION:** Renal neuroendocrine tumors are rare. Surgery remains the cornerstone for longevity. Increased survival was observed in patients of younger age, earlier stage and lower grade tumors.