



# Renal Neuroendocrine Tumors: A Single-Center Experience

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## Abstract

### Background:

- Renal NETs are exceedingly rare neuroendocrine neoplasms
- Very little is known about their clinical behavior

### Objective:

- To review all cases of renal NETs at a large referral center and to describe the mode of presentation, response to therapy and outcomes
- To estimate annual incidence using a population based database

### Methods:

- All patients diagnosed with renal NETs at Mayo Clinic during the period 2000 to 2015 were identified
- Poorly differentiated neuroendocrine carcinomas and small cell carcinomas were excluded
- Data on patient characteristics and outcomes of therapy were extracted and analyzed
- Cases from the SEER Registry (2000 – 2013) were identified and incidence and survival calculated

### Results:

- 14 patients with renal NETs were identified
- Median age at diagnosis was 48.5 years (range: 21 – 82).
- Nine patients were females (64%)
- Pain was present in 8 patients (57%)
- Two patients had flushing and hematuria
- Four patients had a horseshoe kidney
- WHO grade was G1: 36%, G2: 43% and G3: 7%
- T stage was T1: 29%, T2: 29% and T3: 36%
- Nine patients (64%) had a recurrence with a median time to recurrence of 15.5 months
- Responses to systemic therapy were uncommon
- Median overall survival from diagnosis was 99.9 months

### Conclusion:

- Renal NETs are rare
- The most common presenting symptoms was pain
- There may be an association with horseshoe kidneys
- Recurrences are common
- Renal NETs are resistant to systemic therapy
- Despite treatment resistance, the overall survival is long

## Methods

- All patients with renal NETs diagnosed at Mayo Clinic were identified using the Mayo Clinic Cancer Registry
- Baseline characteristics as well as presenting features, behavioral factors, diagnostic methods, therapy details and outcomes were recorded
- Time to recurrence defined as the time from resection until tumor recurrence was estimated using the Kaplan-Meier method
- Cases were identified in the SEER 2000 – 2013 registry
- Incidence was calculated using SEER data and survival calculated with the Kaplan-Meier method

## Results

### Patient Characteristics

- 14 patients were identified
- Median age at diagnosis: 48.5 years (range: 21 – 82).
- Nine patients were females (64%)
- The most common presenting feature was pain (57%) followed by hematuria and flushing (14%)
- Only 3 patients (21%) were asymptomatic
- Two patients had flushing (a functional tumor)
- The mean duration of symptoms prior to diagnosis was 6 months (range: 1 – 24 months)

### Diagnostic Methods

- 4 patients (29%) had a biopsy prior to resection and in all but one patient, the diagnosis was confirmed with resection of the tumor
- Somatostatin receptor scintigraphy (OctreoScan) was done in only 2 patients and showed uptake in both
- Only one patient had chromogranin A measured and it was normal

Presenting features at diagnosis	No. of Patients (%)
Bone metastases	7 (15%)
Horseshoe kidney	4 (29%)
Pain	8 (57%)
Hematuria	2 (14%)
Flushing	2 (14%)

## Results: Grade and Stage

### Tumor Characteristics

Tumor Grade (WHO 2010)	No. of Patients (%)
Grade 1	5 (36%)
Grade 2	6 (43%)
Grade 3	1 (7%)
Grade not reported	2 (14%)

### Tumor Stage at Diagnosis

Tumor Stage (AJCC)	No. of Patients (%)
T1	4 (31%)
T2	4 (31%)
T3	5 (38%)
T4	0
Node Positive	9 (64%)
Distant metastases	4 (28%)
Mean primary tumor size	9 cm

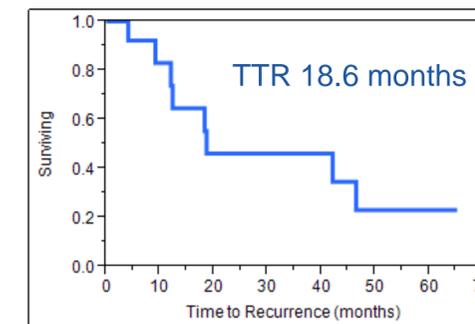
## Surgical Therapy

### Therapy and Patient Outcomes

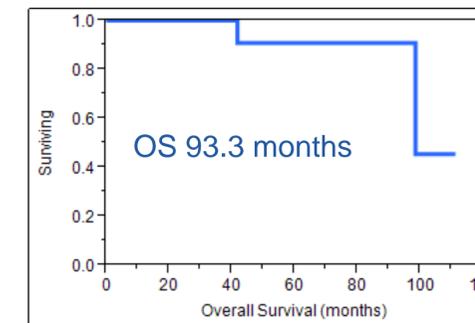
- 13 (93%) patients underwent surgery
  - 12 nephrectomies
  - 1 partial nephrectomy
  - R0 resection obtained in 10 (77%)
- Ablation of metastatic lesions and/or nodes was performed in 9 patients (64%)
- Of the resected patients, nine (69%) experienced recurrence
  - The median time to recurrence in these patients was 18.6 months (range 4 -65 months)

## Results: Survival

### Time to Recurrence (TTR)



### Tumor Characteristics and Outcome

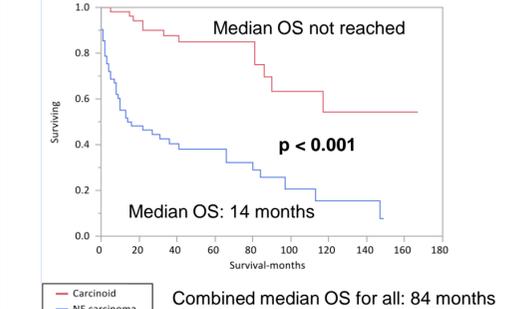


## Results: SEER Data

### Renal NETs are rare

- 117 cases from 2000 – 2013
- Annual incidence: 1.3 cases/million
- Median age at diagnosis: 57 years
- 26% with distant metastases at diagnosis

### Survival Plot



## Conclusions

- Renal NETs are rare tumors
- Association with horseshoe kidneys
- Lymph node metastases are common at diagnosis but distant metastases are seen in less than a third
- Most patients were able to undergo a complete resection but metastatic recurrences are common
- Most recurrences occur within 2 years
- Bone metastases are common
- There is no standard therapy for metastatic disease
- Radiographic responses are uncommon, especially beyond first-line therapy
- Despite limited options for systemic therapy and lack of responses, the overall survival is long

## References

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