Prevalence of Functional Tumors in Neuroendocrine Carcinoma: An analysis from the National Comprehensive Cancer Network Neuroendocrine Tumors Database

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INTRODUCTION

- Neuroendocrine tumors (NETs) are increasing in incidence and prevalence. Identification and treatment of specific clinical NET syndromes are established, yet there is uncertainty regarding the prevalence of NET with hormone-related symptoms versus nonfunctional tumors.
- The primary aim of this analysis was to describe demographic and clinical characteristics of NET patients by functional status at diagnosis.

METHODS

- The National Comprehensive Cancer Network (NCCN) created a comprehensive database to characterize patients treated for NETs.
- This database consists of patients presenting to 7 NCCN institutions with a confirmed NET diagnosis between 2004 and 2010.
- Institutional IRB approval was obtained at all participating institutions:
  - The Sidney Kimmel Comprehensive Cancer Center at Johns Hopkins
  - Dana-Farber Cancer Institute
  - The University of Texas MD Anderson Cancer Center
  - UCSF Helen Diller Family Comprehensive Cancer Center
  - The Ohio State University Comprehensive Cancer Center - James Cancer Center and Solove Research Institute
  - Robert H. Lurie Comprehensive Cancer Center of Northwestern University
  - H. Lee Moffitt Cancer Center and Research Institute
- This analysis was limited to the following histology sub-types:
  - Carcinoid (n = 788)
  - Pancreatic NET (n = 426)
  - Neuroendocrine carcinoma-NOS(n = 178)
  - Adrenal (n=105) which included:
    - Pheochromocytoma (n = 45)
    - Adrenal cortical carcinoma (n = 26)
    - Paraganglioma (n = 34)
- The study population consisted of patients presenting with eligible histologies between January 2004 and December 2010 (n=1,497).
- Functioning NET refers to an association between a clinical syndrome and elevation in blood or urine hormone level among patients with a confirmed NET.

RESULTS

Table 1: Demographic and Clinical Characteristics (N=1497)

	Functional Tumor Status: Yes N	Yes Row %	No N	No Row %	Unknown N	Unknown Row %	All N	All Col %	X^2	p
Age at Diagnosis (yrs)									16.8	<.01
18 to 44	96	29	208	62	31	9	335	22		
45 to 54	112	27	284	68	19	5	415	28		
55 to 64	126	32	236	60	32	8	394	26		
65+	79	22	245	69	29	8	353	24		
Median = 55 (Range: 18-88)										
Gender									0.8	0.66
Male	187	27	458	66	48	7	693	46		
Female	226	28	515	64	63	8	804	54		
Race									7.7	0.11
White	348	28	791	64	99	8	1238	83		
African American	20	20	76	75	5	5	101	7		
Other	45	29	106	67	7	4	158	10		
Tumor Type									20.7	<.01
Carcinoid	237	30	484	61	67	9	788	53		
pNET	97	23	302	71	27	6	426	29		
NET-NOS	44	25	128	72	6	3	178	12		
Adrenal	35	33	59	56	11	10	105	7		
Metastatic Disease at Initial Diagnosis									81.9	<.01
Local/Regional	108	16	476	75	54	9	638	43		
Distant	294	36	480	58	51	6	825	55		
Not staged	11	32	17	50	6	18	33	2		
Metastatic Site (distant stage patients)									11.4	<.01
Liver	248	38	358	55	43	6	649	43		
Extra-hepatic	46	26	122	69	8	5	176	12		
Vital status									11.4	<.01
Alive	223	25	593	66	78	9	894	60		
Deceased	190	32	380	63	33	6	603	40		
Total	413	28	973	65	111	7	1497	100		

Figure 1: Prevalence of Functional Tumors by Tumor Type and Stage (N = 1497)

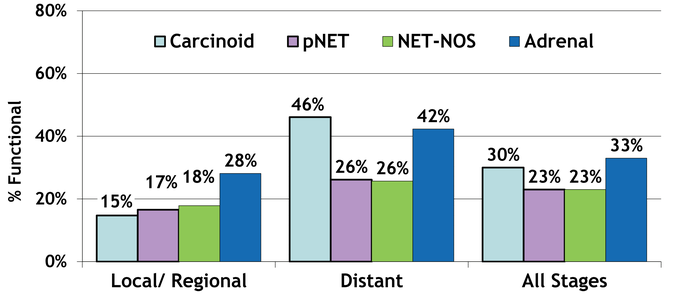


Figure 2: Functional Tumors by Histology (N = 413)

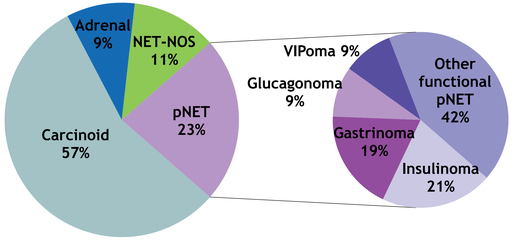
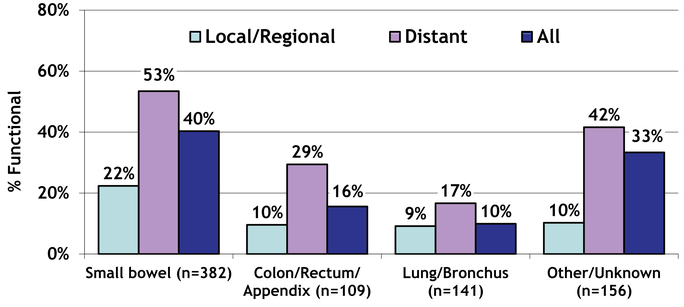


Figure 3: Prevalence of Functional Carcinoid Tumors by Primary Site and Stage (N = 788)



SUMMARY OF FINDINGS

- Functional tumors accounted for 28% of the cohort (413/1,497).
- Prevalence of hormonal syndrome was higher for carcinoid tumors (30%), compared to pNET (23%).
- The most common symptoms prompting the NET diagnosis among patients with functional tumors included change in bowel habits, weight loss, abdominal cramping, and nausea.
- Among carcinoid patients with elevated 5-HIAA (n=200), 58% had functional tumors.
- The majority of functional pNET had an unspecified sub-type, with insulinoma (21%) and gastrinoma (19%) also prevalent.
- The majority of carcinoid(75%), pNET (71%), and NET-NOS (88%) patients with functioning tumors had distant disease.
- Most patients (84%) with functional tumors and distant disease had metastatic disease in the liver at diagnosis (CARC: 84%, pNET: 88%, NET-NOS: 89%).
- The majority of patients with functional adrenal tumors had local/regional disease (n=51%). Among distant stage patients with functional adrenal tumors, 6 of 11 had liver metastases.

CONCLUSIONS

- Prevalence of carcinoid syndrome in this NCCN database (**30%**) was slightly higher than the 10% previously reported in the literature.
- The prevalence of hormonal syndrome among pNET pts (23%) was lower than previously reported.
- Approximately one quarter of carcinoid patients without metastatic disease had carcinoid syndrome, warranting further analysis as carcinoid syndrome most often occurs in the presence of liver metastasis.

NANETS 2012 Abstract: Scan QR code to view poster online.

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