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

M. A. Choti†1; S. Bobbia†; J. R. Strosberg†; A. B. Benson, III; M. Bloomston†; J. C. Yaun; C. Zornosa; E. Bergsland; M. H. Kulke; E. K. Nakakura; M. H. Shah†

1The Sidney Kimmel Comprehensive Cancer Center at Johns Hopkins, Baltimore, MD; National Comprehensive Cancer Network, Fort Washington, PA; M. H. Lurie Comprehensive Cancer Center, Chicago, IL; The Ohio State University Comprehensive Cancer Center - James Cancer Hospital and Solove Research Institute, Columbus, OH; The University of Texas MD Anderson Cancer Center, Houston, TX
2Dana-Farber Cancer Institute, Boston, MA; Northwestern University Feinberg School of Medicine, Chicago, IL; University of Maryland Medical Center, Baltimore, MD; Washington University School of Medicine, St. Louis, MO; The University of Texas MD Anderson Cancer Center, Houston, TX; UC San Diego Comprehensive Cancer Center, San Diego, CA; The University of New Mexico Comprehensive Cancer Center, Albuquerque, NM

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 NET.

• 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 = 165) which included:
    - Pheochromocytoma (n = 45)
    - Adrenal cortical carcinoma (n = 26)
    - Paragangioma (n = 34)
  - The study population consisted of patients presenting with eligible histologies between January 2004 and December 2010 (n=1,497).

RESULTS

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

<table>
<thead>
<tr>
<th>Year</th>
<th>N (%)</th>
<th>Functional Tumor Status</th>
<th>%</th>
<th>%</th>
<th>%</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>All</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Local/Regional</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Distant</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>All</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Carcinoid</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>pNET</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>NET-NOS</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Adrenal</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

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 functioning 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 versus 65% in carcinoid tumors (30%).

• Approximately one quarter of carcinoid patients without metastatic disease had metastatic disease in the liver at diagnosis (Carcinoid: 84%, pNET: 88%)

CONCLUSIONS

• Prevalence of carcinoid syndrome in this NCCN database (30%) was slightlyhigher 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 metastases.

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 Tumors by Primary Site and Stage (N = 788)