Neuroendocrine Tumors in First Three Decades of Life

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Background: Neuroendocrine tumors (NET) are rare tumors with limited data on young NET patients less than 30 years. We performed this analysis to investigate the demographics, clinicopathological characteristics and management of the young and adolescent well-differentiated NET patients.

Methods: We conducted a retrospective review of the medical records of patients diagnosed with NETs at age less than 30 years at Vanderbilt University Hospital, between 1990 and 2015.

Results: A total of 77 patients (pts) with NET were identified. Mean age at presentation was 19 years (range, 10 to 29 y). Male to female ratio is 0.6:1. The majority were incidentally diagnosed (n=70; 91%), non –functional tumors (n= 66; 86%) and only 5% (n=4) had carcinoid syndrome at presentation. Stage at diagnosis was: stage 1 -60%; stage 2 – 4%, stage 3 – 12%; stage 4 - 23%; unknown – 1.3%. Of the patients with metastatic disease, 89% (n =16) had synchronous metastases and 11% were metachronous (n = 2). The most common primary site of origin was midgut (n=45; 58.4%) followed by foregut (n=25; 32.5%) and hindgut (n= 5; 6.5%). Midgut tumors arising from appendix was the most common primary site of origin (n = 38, 49%) The grade of the tumor was available in 44 patients of whom 73% were WHO grade 1. Ki-67 was reported in 35 patients and the majority had Ki-67 <3% (n=22, 65%). Mitotic rate was reported in 29 patients with low mitotic rate of less than 2 per 10 hpf in 20 (69%). Most pts had complete surgical resection with negative margins (n=59; 77%). Only 10% (n=8) were treated with chemotherapy and 19.5% (n=15) were treated with somatostatin analogues. Only 3 patients (4%) were noted to have MEN1 syndrome, but an additional 2 (2.6%) of pts had family history of NET. There were no reported second malignancies. Median follow up was 24 months (range 0.6 – 193 mo). Most pts were alive either without active disease (75%) or with active disease (20%). The disease specific mortality was 5% (n=4).

Conclusions: In this single-institution retrospective review, the survival of young pts < 30 years of age was excellent, probably largely due to the fact that most were incidental findings and early stage with about half of all patients having NET of the appendix.