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Clinicopathologic Characteristics and Outcomes of MEN1-Associated Bronchopulmonary Neuroendocrine Neoplasms Carcinoid Tumors: The Mayo Clinic Experience

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BACKGROUND

Bronchopulmonary neuroendocrine tumors (bNETs) are uncommon manifestations of multiple endocrine neoplasia type 1 (MEN1), that contribute to morbidity and mortality. Contemporary data describing the presentation, management, and outcomes of MEN1-associated bNETs remain limited.

METHODS

Patients with MEN1 evaluated across three Mayo Clinic sites between 1/1/2005 and 12/1/2025 (n=945) were screened for thoracic, thymic, or mediastinal carcinoid diagnoses. Manual electronic medical record review confirmed concurrent MEN1 and bNET. Demographic, clinicopathologic, imaging, treatment, recurrence/progression, and survival data were abstracted.

RESULTS

Seventeen patients with MEN1-associated bNETs were identified. Median age at diagnosis was 41.1 years; 6 (35.3%) were male. Histology included bronchial carcinoids in 15 patients (88.2%), large cell neuroendocrine carcinoma in 1 (5.9%), and diffuse idiopathic pulmonary neuroendocrine cell hyperplasia in 1 (5.9%). Histologic subtype was available in 9 patients; 5 typical carcinoids (29.4%), 3 atypical carcinoids (17.6%), and 1 mixed tumor (5.9%); 8 tumors (47.1%) were not further subclassified. All tumors with documented hormonal status (n=11) were nonfunctional. Somatostatin receptor PET imaging was performed in 11 patients, demonstrating uniformly avid disease in 6 (55%), heterogenous avidity in 1 (9%), and mild avidity in 4 (36%).

Concurrent MEN1-associated manifestations included primary hyperparathyroidism in 15 patients (88.2%), pituitary tumors in 12 (70.6%), pancreatic NETs in 16 (94.1%), adrenal lesions in 5 (29.4%), and breast tumors in 3 (17.6%). Surgical resection was performed in 9 (52.9%) patients. Among 7 evaluable surgical patients, recurrence occurred in 3 (42.9%), after a median of 23.5 months (range 19.8-27.8).

Management was heterogenous and included somatostatin analog therapy in 7 patients (41.2%), cytotoxic chemotherapy in 3 (17.6%), targeted therapy (everolimus or sunitinib) in 2 (11.8%), radiation therapy in 4 (23.5%), and ablative therapy in 2 (11.8%). Disease progression occurred in 1 non-surgical patient after 6 years of follow-up, and 2 patients (11.8%) died during follow up.

CONCLUSIONS

MEN1-associated bNETs are rare and frequently coexist with other MEN1 manifestations, supporting comprehensive multisystem surveillance. Tumors were predominantly nonfunctional bronchial carcinoids, while treatment approaches were heterogenous, reflecting the lack of standardized management strategies and guidelines. Despite substantial post-surgical recurrence, overall outcomes were favorable, with low rates of disease progression and mortality. Larger, prospective, multi-institutional studies are needed to define optimal screening, surveillance, and management strategies for MEN1-associated bNETs.

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