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Clinical Characteristics and Outcomes of MEN1-Associated Thymic Neuroendocrine Tumors Across Mayo Clinic Sites

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BACKGROUND

Thymic neuroendocrine tumors (tNETs) are an uncommon but historically lethal manifestation of multiple endocrine neoplasia type 1 (MEN1), accounting for a disproportionately large share of MEN1-related mortality. Evidence guiding surveillance and management strategies remains limited to small retrospective series. We evaluated the clinical characteristics, management, and outcomes of MEN1-associated tNETs across Mayo Clinic sites.

METHODS

We retrospectively reviewed 945 patients with MEN1 evaluated at three Mayo Clinic sites between 1/1/2005 and 12/1/2025. Patients were further filtered for a thoracic, thymic, or mediastinal carcinoid diagnosis, followed by manual electronic medical record review confirming concurrent MEN1 and tNET diagnosis. Demographic, clinicopathologic, imaging, treatment, recurrence/progression, and survival data were abstracted.

RESULTS

Four patients with MEN1-associated tNETs were identified. Median age at diagnosis was 36.4 years, and 3 patients (75%) were male. Histology demonstrated well-differentiated atypical carcinoid tumors in 2 patients (50%) and well-differentiated NET subtype not specified in 1 patient (25%). One diagnosis was based on characteristic imaging findings. One tumor (25%) was functional, manifesting as ectopic Cushing syndrome. Somatostatin receptor PET imaging revealed avid lesions in all 4 patients.

Concurrent MEN1-associated manifestations were common, including primary hyperparathyroidism in 3 patients (75%), pituitary tumors in 3 patients (75%), pancreatic NETs in 3 (75%), adrenal lesions in 3 (75%). None had breast tumors. Surgical resection was performed in all 4 patients. Among the 3 patients with available follow up data,

recurrence occurred in 2 (66%), with a median time to recurrence of 63.3 months (range 40.8-85.8 months).

Management was heterogenous and multimodal, including somatostatin analog therapy in 2 patients (50%), cytotoxic chemotherapy in 3 patients (75%), targeted therapy (everolimus or sunitinib) in 2 patients (50%), and radiation therapy in 3 patients (75%). One patient (25%) died during follow up due to complications of his tNET.

CONCLUSIONS

MEN1-associated tNETs are rare and present alongside a high burden of concurrent MEN1 manifestations, reinforcing the need for systematic multisystem surveillance. Despite surgical resection being performed in all patients in this cohort, recurrence rates were high (66%) and multimodal therapy was frequently required. These findings reinforce the high morbidity associated with MEN1-related tNETs and highlight the need for prospective, multi-institutional studies to optimize surveillance, surgical timing, and adjuvant treatment strategies in this high-risk patient population.

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