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ACTH-secreting Pancreatic Neuroendocrine Neoplasms: A Case-Series

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BACKGROUND

Pancreatic neuroendocrine neoplasms rarely secrete ACTH resulting in ectopic Cushing's syndrome. Data are limited to case reports and very small series.

METHODS

Cases of ectopic Cushing's syndrome were identified from a database of pancreatic NEN seen at the Moffitt Cancer Center between 1/2008 and 4/2022. Tumor characteristics, clinical signs and symptoms, therapies and outcomes were evaluated.

RESULTS

13 patients with ACTH-producing pancreatic NENs were seen, ranging in age from 16 to 65 years at time of NEN diagnosis (median 42). 12 of 13 patients had metastatic disease at presentation. Four patients also had ZE syndrome. All tumors were well-differentiated at diagnosis although 2 were described as transformed to poorly differentiated after re-biopsy. Bilateral adrenalectomy was performed in 5 patients for control of Cushing's syndrome. Tumor responses to systemic therapy were very poor. Median overall survival was 56 months from time of initial cancer diagnosis but only 18 months from diagnosis of Cushing's syndrome.

CONCLUSIONS

Ectopic Cushing's syndrome is a morbid condition when occurring in pancreatic NENs and is generally associated with aggressive metastatic disease. Bilateral adrenalectomy can be considered for syndrome control.

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