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Type III Gastric Neuroendocrine Tumor: Case Report of a Rare Gastric Carcinoma

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Background

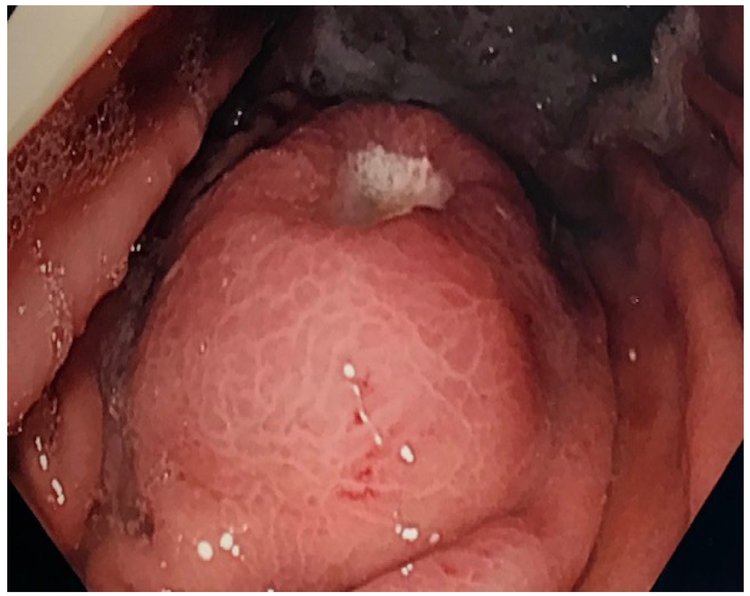
- Gastric neuroendocrine tumors (GNETs) are rare lesions that comprise 0.1-0.6% of all gastric cancers. The frequency of GNETs is 2.5 cases per 100,000 persons per year, with recently increased incidence in the United States due to widespread use of upper endoscopy. GNETs are classified into three groups, based on pathogenesis and histomorphologic characteristics. These types differ in biologic behavior and prognosis, ranging from benign to highly malignant with extremely poor prognosis. Type I is the major subtype of GNETs, associated with chronic atrophic/ autoimmune gastritis with pernicious anemia (PA). Type II GNETs are associated with multiple endocrine neoplasia-1 (MEN-1), and Zollinger-Ellison syndrome (ZES). Type III GNETs represent the second most common type, which behaves aggressively with rates of metastasis higher than 50%, and tumor related death over 25-30%.

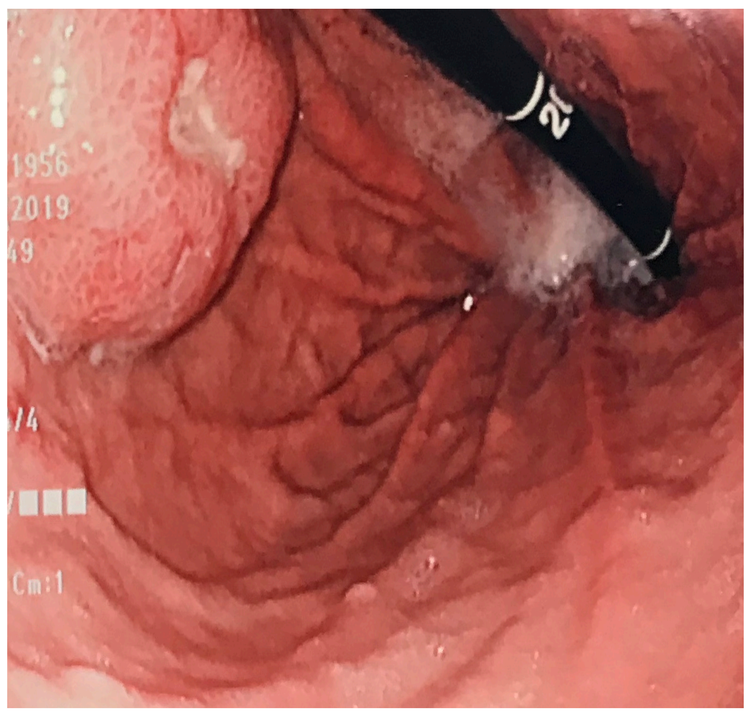
Case Description

- The patient was a 63-year-old Caucasian male who presented for evaluation of recurrent anemia. Upon admission, patient was found to be anemic and required packed red blood cells (PRBC) transfusions. Esophagogastroduodenoscopy (EGD) revealed a 2-3cm ulcerated mass with a clean based ulcer with rolled over edges in the proximal body along the greater curvature of the stomach. Tissue was sent for an expert opinion and the patient was diagnosed with an unusual neuroendocrine carcinoma. Three endocrine marker stains: chromogranin, synaptophysin, and CD56, all tested strongly positive confirming the diagnosis.

Gastric Carcinoids

Carcinoid	% Total	Assoc.	Gastrin	Acid Sec.	Mets
Type I	75	PA / atrophy	Inc.	Low	V. rare
Type II	5 - 10	ZES / MEN-1	Inc.	High	Rare
Type III	15 – 25	None	Normal	Normal	Common





Discussion

- Type III GNETs develop in the absence of hypergastrinemia, and tend to purse an aggressive clinical course compared to Type I and Type II GNETs. Type III GNETs have the greatest potential to metastasize, with five year survival also being worse (75-80%) compared to (90-95%) for Type I GNETs. They are sporadic and usually present as a single large (>2cm) mass with normal gastrinemia. Immunohistochemical analysis is essential in diagnosing GNETs. It allows diagnostic confirmation and permits classifying the lesion according to histological grade. For diagnostic confirmation chromogranin A and synaptophysin are necessary; while for prognostic definition the proliferation index Ki-67 and the number of mitoses per high magnification field are required. Poor prognostic factors are: lesion > 2 cm, deep invasion into the submucosa or beyond, Ki-67 >3%, vascular invasion, low degree of structural differentiation, presence of atypia and/or necrosis. Type III lesions should be managed aggressively with total or subtotal gastrectomy, and if metastases are found systemic cytotoxic chemotherapy should be administered.

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