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A patient with MEN-1 presents with upper gastrointestinal bleeding: what is the cause and where is the primary tumor?

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Disclosures

No disclosures to report

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Patient History

65 yo male with **multiple endocrine neoplasia type 1 (MEN-1)** with a **one year history of epigastric pain** presents with **anemia and bloody stools**

Medical History
MEN-1
Type 2 Diabetes
Gastroesophageal reflux disease

Surgical History
3½ gland parathyroidectomy
Partial pancreatectomy
Splenectomy

Family History
Two daughters with MEN-1

Social History
Denies tobacco, alcohol, and illicit drugs

Allergies
NKDA

Medications
Amaryl
Lantus
Zofran
Omeprazole 20mg daily
Promethazine
Metoprolol

Physical Exam & Laboratory Data

- Unremarkable
- Pertinent Findings
  - Abdomen: soft, nontender, nondistended. No palpable masses. No hernia. Surgical incision well healed.
  - Digital Rectal Exam: no gross blood
  - Heme occult: positive

142	105	21	202
5.0	27	1.38	

6.5	3.7
0.1	<0.2
22	21
69	

13.6 | 39.4, 1.02

10.7, 6.77, 354, 33.6

Gastrin: 17,290 (<100)

Chromogranin A: 18,710 (<93)

Vasoactive Intestinal Peptide: 50

Pancreatic Polypeptide: 785 (<312)

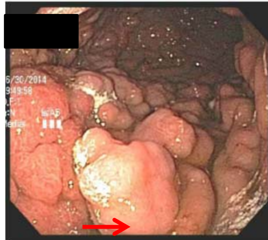
Neuron Specific Enolase: 9.1

Parathyroid Hormone: 146 (15-65)

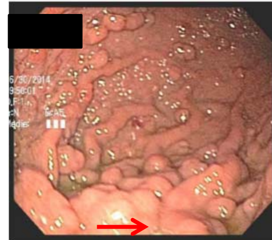
Calcium: 2.39

Esophagogastroduodenoscopy (EGD)

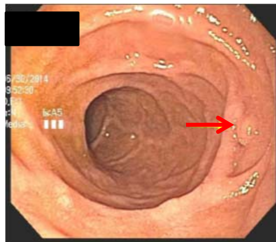
Nodules in Stomach



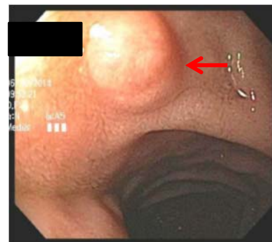
Enlarged Gastric Folds with Nodules



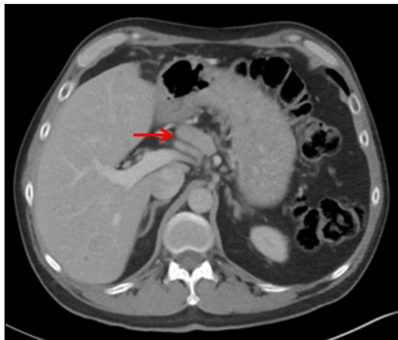
Nodule in duodenum

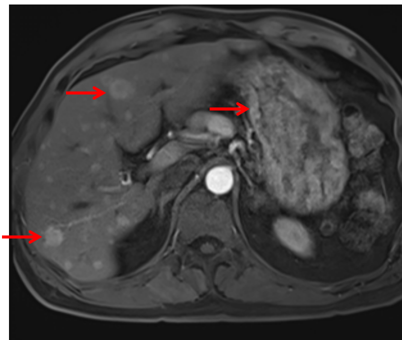


Nodule in duodenum

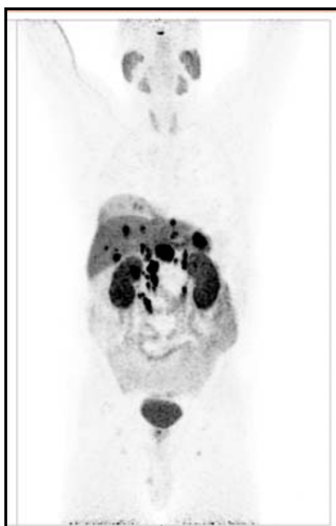


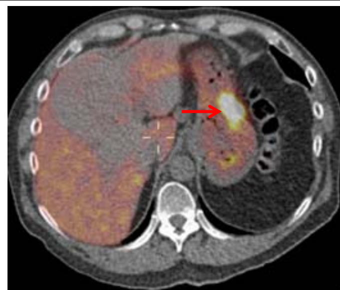
Metastatic Workup

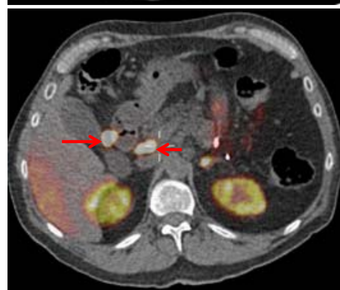




Metastatic Workup
Ga-68 DOTATATE PET/CT







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Management & Patient Course

- After discussing treatment options including surgery and targeted therapy, the patient elected to continue with active surveillance.
- Patient instructed to **increase omeprazole** to 40mg twice daily
- Systemic therapy (somatostatin analogs, targeted therapy and PRRT) discussed
- Four months later
  - dizziness
  - **hematemesis** requiring **hospitalization**
  - **one blood transfusion**
- **EGD: no source of bleeding**
- Patient reported **inconsistency** with **proton pump inhibitor** treatment
- Patient stabilized and returned for workup and further management

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Surgical Exploration

- **exploratory laparotomy**
  - extensive lysis of adhesions
  - lymphadenectomy of aortocaval, retropancreatic and porta hepatic lymph nodes
  - resection of dominant duodenal mass involving the pylorus
  - wedge resection of the greater curvature
  - cholecystectomy
  - biopsy of segment III liver lesion
- **Intraoperative findings included**
  - 5 cm gastric mass
  - numerous small nodules and a dominant 1cm lesion in the duodenum
  - subcentimeter peripancreatic lesions

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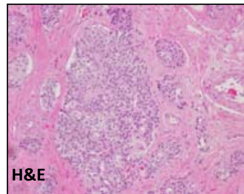
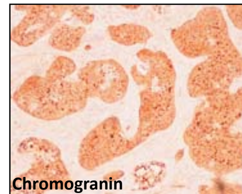
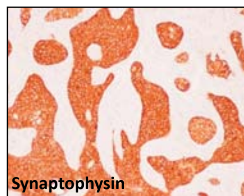
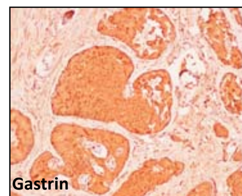
Pathology Findings

Duodenal Mass


H&E


Chromogranin


Synaptophysin


Gastrin

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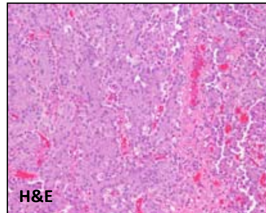
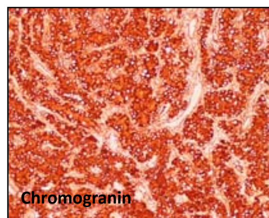
Pathology Findings

Gastric Mass





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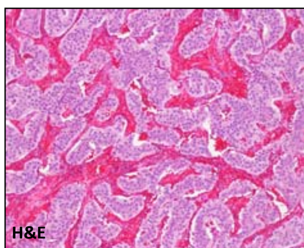
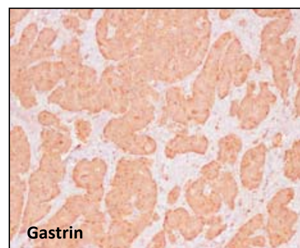
Pathology Findings

Porta Hepatic Lymph Node





Final Pathology: metastatic gastrinoma (pT2N1M1)

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Postoperative Course

- Uncomplicated postoperative recovery
- Patient continued on his proton pump inhibitor
- **Gastrin level decreased to 794 pg/ml**
- **No further UGI bleeding** has occurred
- Treatment options for metastatic neuroendocrine tumor including targeted therapies discussed with patient.
- Due to social reasons patient has chosen **active surveillance**.

Discussion

Key Teaching Points

- The **most common cause of GI bleeding in MEN1 are gastric/duodenal ulcerations** from Zollinger-Ellisson Syndrome (ZES), which occur in 25% as initial presenting symptom.[1]
- Bleeding from gastric carcinoids as in this case is much less common.[2]
- **Type 2 carcinoid tumors** develop **in up to 37% of patients with ZES and MEN-1**[3]
- The importance of **hypergastrinemia** is demonstrated by the fact **that MEN-1 is not associated with carcinoid unless ZES is present.** [4]

1. Roy PK, et al. Medicine. 2000.
2. Nikou and Angelopoulos. Gastroenterology Research and Practice. 2012
3. Norton JA, et al. Surgery. 2004.
4. Jordan PH, et al. JACS. 2004.

Thank You