AMSER Case of the Month November 2021

40-year-old female with obscure GI Bleed and three-year unrevealing workup



Doug Spaeth-Cook, MS4 -- Ohio University HCOM



Alex Christie, MD – Cleveland Clinic



Ruchi Yadav, MD – Cleveland Clinic (Preceptor)



Patient Presentation

 A 40-year-old otherwise healthy female with history significant for three episodes of severe acute anemia in the past three years with recurring bloody stools and several extensive unrevealing GI workups. She was admitted during her most recent episode in March 2021. Colonoscopy, EGD, push enteroscopy, and CT were all unable to localize the source of bleeding. Additional negative testing included a tagged RBC scan, video capsule endoscopy, and Meckel scan. Patient is now presenting to gastroenterology seeking a second opinion.

History

- PMHx: Iron deficiency anemia
- PSHx: Push upper endoscopy (3/2021), small bowel fluoroscopy, flexible sigmoidoscopy x3 (3/2021,2/2021,9/2018), EGD (3/2021), ERCP, EUS, VCE (3/2021, 8/2018)
- FamHx: No known problems in 1st degree relatives
- Social: Never smoker, etOH 1-2 drinks monthly, balanced diet
- Meds: Daily ferrous sulfate, folic acid, pantoprazole

Pertinent Labs

- Labs and vitals stable following 3/2021 hospitalization for acute anemia
 - FOB positive
 - Ferritin 362, iron 87, TIBC 299, % Sat 29
 - Hb 10.7, Retic % 4, MCV 92, B12 572, haptoglobin 57
 - INR 1.1
 - Negative immunological workup including: ANA; transglutaminase IgA/IgG; anti-gliadin, anti-endomysial & anti-reticulin IgA

What Imaging Should We Order?



Select the applicable ACR Appropriateness Criteria

Variant 5:

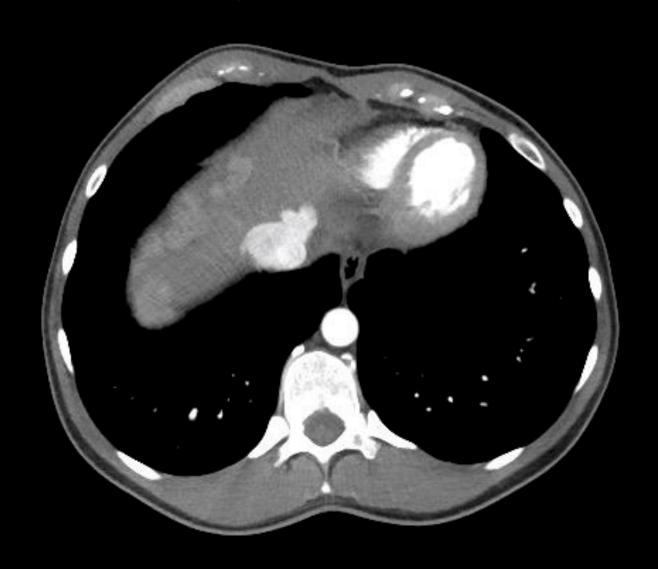
Lower gastrointestinal tract bleeding. Obscure (nonlocalized) recurrent bleeding in a hemodynamically stable patient (assumes a prior negative adequate colonoscopy and upper gastrointestinal endoscopy). Next procedure or intervention.

| Procedure | Appropriateness Category |
|---|-----------------------------------|
| Capsule endoscopy | Usually Appropriate |
| CT enterography abdomen and pelvis with IV contrast | Usually Appropriate |
| MR enterography | May Be Appropriate |
| Push enteroscopy | May Be Appropriate |
| RBC scan abdomen and pelvis | May Be Appropriate |
| RBC scan with SPECT or SPECT/CT abdomen and pelvis | May Be Appropriate |
| Transcatheter arteriography/embolization | May Be Appropriate (Disagreement) |
| Fluoroscopy small-bowel follow-through | Usually Not Appropriate |
| Surgery | Usually Not Appropriate |

These imaging modalities were ordered by the gastroenterologist.

Results: Repeat capsule endoscopy showed a distal polypoid mass in small bowel. CT enterography was then ordered.

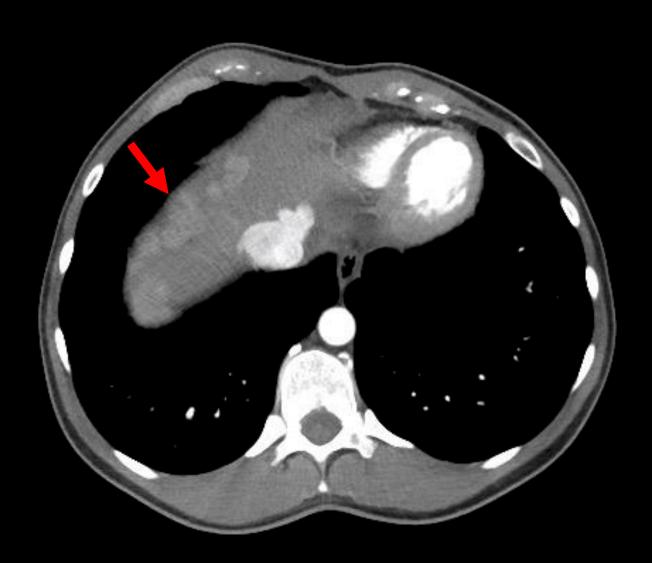




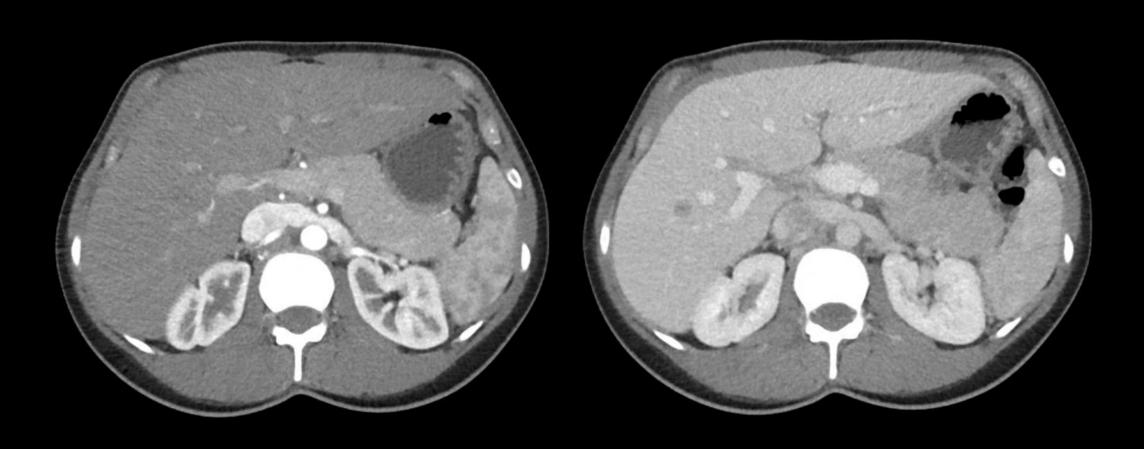


Arterial Phase CTE

Multiple, clustered right subdiaphragmatic perihepatic hypervascular serosal/peritoneal deposits



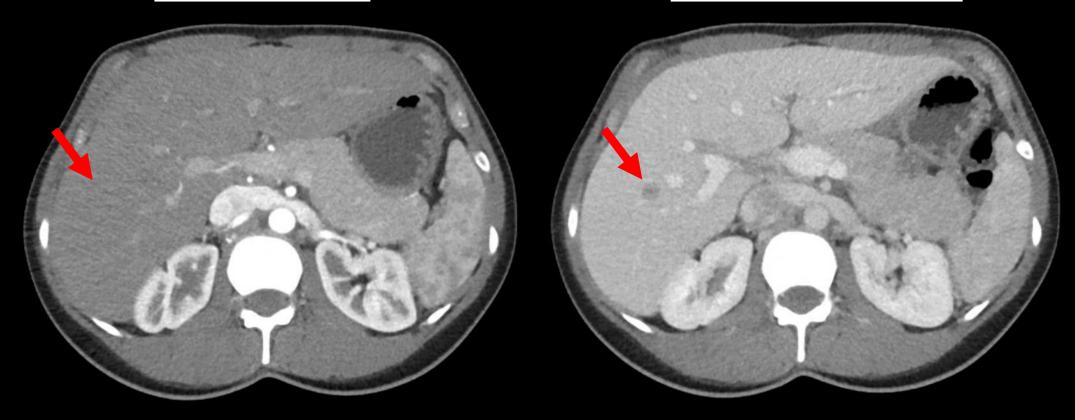






Arterial Phase CTE

Portal Venous Phase CTE



One of several subcentimeter, hypoattenuating, hepatic lesions on portal venous phase without definite correlate on arterial phase.







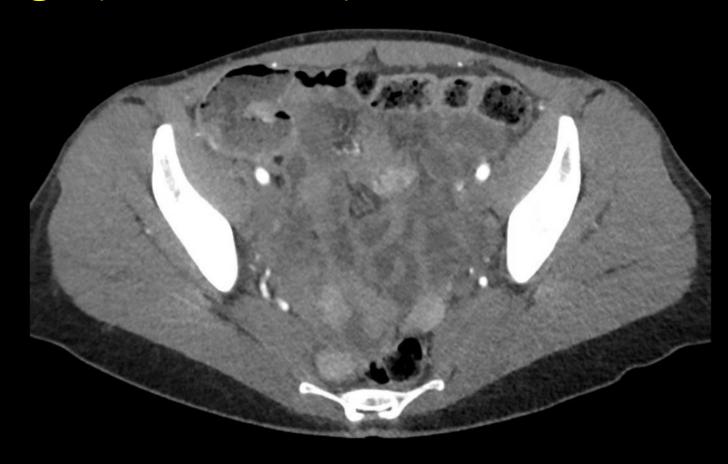
Arterial Phase CTE

Hypervascular, 1.6 x 1.7 cm, mesenteric node in the midline upper pelvis



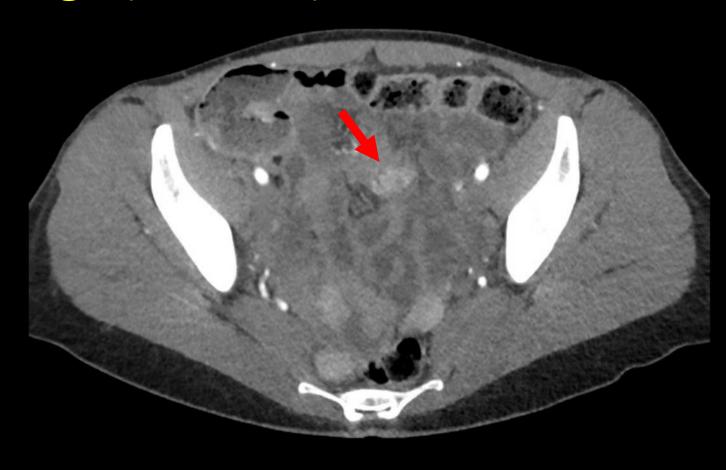






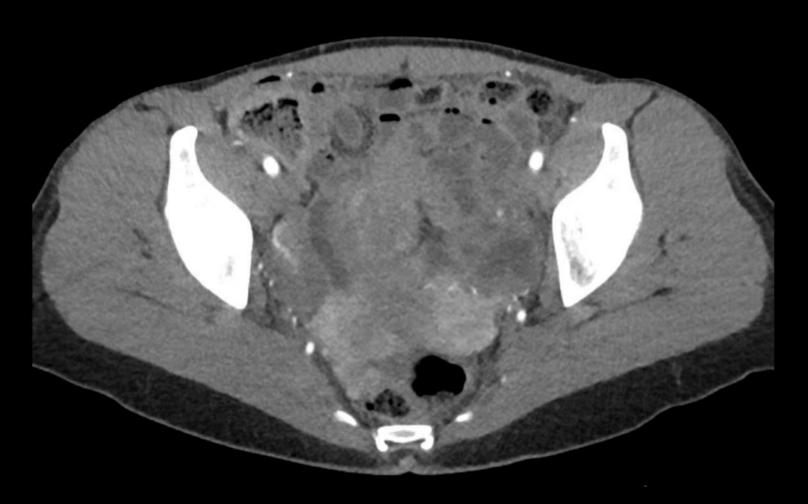






<u>Arterial Phase CTE</u> (L: Coronal, R: Axial) – 1.5 x 2.7 x 2.2 cm, polypoid, avidly enhancing, distal small bowel mass.







Arterial Phase CTE

Multiple hypervascular lesions in the posterior pelvis

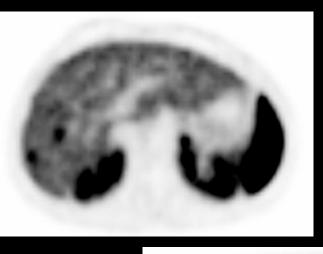




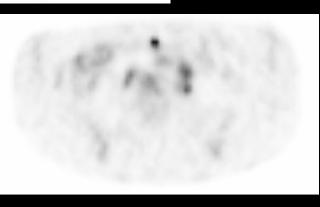
Patient Course

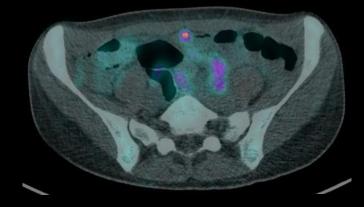
- Discussed at multidisciplinary tumor board
- Baseline chromogranin A 4,182
- Serotonin Serum 1,122
- Partial small bowel resection performed
- Intra-op biopsies: diaphragm, small bowel, peritoneum
- Ga-68 DOTATATE PET/CT ordered

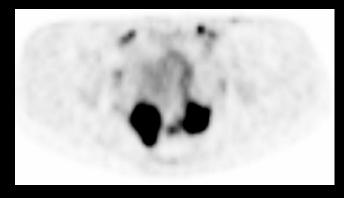


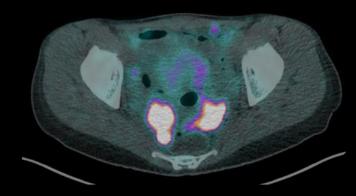






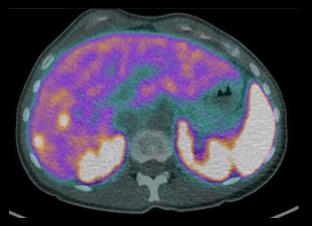




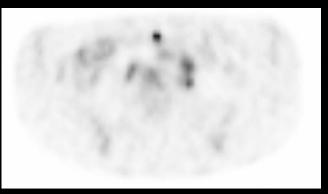


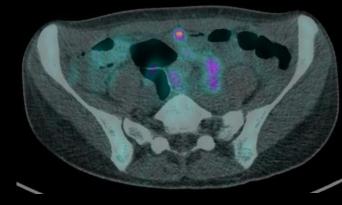






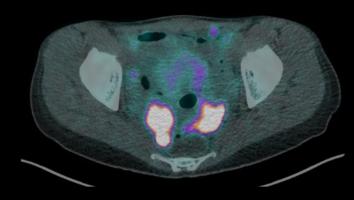






68Ga DOTATATE PET/CT demonstrated increased tracer uptake in the liver and pelvis -> confirming sites of neoplasm/metastases with high concentration of somatostatin receptor expression.







Final Dx

Metastatic, well-differentiated, neuroendocrine tumor of terminal ileum, intermediate-grade (G2); Stage pT4N2M1b

Clinical Significance

An intermediate-grade, midgut, neuroendocrine tumor (NET) presented with rare finding of peritoneal carcinomatosis, not observed on radiographic or endoscopic imaging over a three-year time course.



Case Discussion: Gl Neuroendocrine Tumors

| Site of Origin | Frequency* |
|---------------------|------------|
| Foregut | |
| Thymus | 1-2 |
| Bronchopulmonary | |
| tract | 10-25 |
| Esophagus | <1 |
| Stomach | 2-30 |
| Duodenum | 2-5 |
| Pancreas | <1 |
| Hepatobiliary tract | <1 |
| Midgut | |
| Jejunum | 1-2 |
| Ileum | 15-20 |
| Appendix | 19-35 |
| Ascending colon | 1-5 |
| Hindgut | |
| Transverse colon | 1-5 |
| Descending colon | 2-5 |
| Rectum | 10-12 |
| Ovary or testis | <1 |
| Unknown | 10 |

- Formerly known as carcinoid tumors
 - "Carcinoid Syndrome" anatomy correlation
- Well-differentiated NETs grades 1 and 2 are slower growing
- Usually Somatostatin receptor 2 (+)
 - Binds octreotide/lanreotide
 - Useful for treatment and DOTATATE PET/CT for staging and progression
- GI-NETs metastasize to liver via portal system
- Rarely present with peritoneal carcinomatosis
 - Scattered case reports only
- Recruit arterial blood supply -> suitable targets for arterial embolization

Case Discussion: CTE (CT enterography)

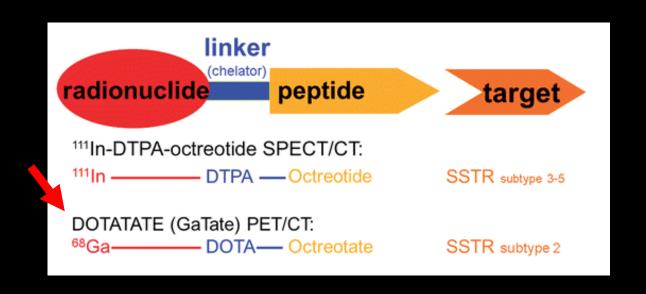
- Indications: Crohn disease, celiac disease, postoperative adhesions, radiation enteritis, scleroderma, small bowel malignancies, and polyposis syndromes
- Pt drinks neutral enteric contrast to distend the small bowel lumen. This helps increase contrast resolution between bowel mucosa and lumen.
- IV contrast is given to image arterial phase and portal venous phase

| Clinical Indication | Type of Oral Contrast Agent | Imaging Phase and Body Part |
|--|--------------------------------|---|
| Tumor staging and follow-up | Water | Arterial phase: liver Portal venous phase: chest, abdomen, pelvis |
| Identification of the primary tumor | Mannitol | Mucosal phase: abdomen and pelvis Portal venous phase: chest, abdomen, pelvis |
| Resectability assessment | Water or mannitol | Arterial phase: abdomen and pelvis Portal venous phase: chest, abdomen, pelvis |



Case Discussion: 68 Ga DOTATATE PET/CT

- Functional imaging harnessing differential somatostatin (SS) receptor (SSTR) expression in NETs
- Octreotide (ligand) connected to radionuclide via a linker (DOTA)
- Old gold-standard was octreoscan SPECT/CT
- ¹⁸F FDG PET is also used, but does not rely on SS receptor





Case Discussion: Radiographic Characteristics of midgut neuroendocrine tumors

- Appear as an arterially-enhancing, intraluminal or extraluminal mass with nearby desmoplastic reaction and vascular encasement.
- Smaller solitary lesions are more difficult to identify than large polypoid.
- Mesenteric fibrosis may occur even in the absence of metastatic spread.
- The secondary features may be easier to recognize than the primary lesion.
- Calcification of tumor is associated with worse prognosis.
- Difficult to differentiate from sclerosing mesenteritis given shared features with the desmoplastic response.
- Tissue diagnosis can be challenging due to fibrosis in the surrounding area.



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