

Carcinoid Syndrome: Imaging and Evaluation

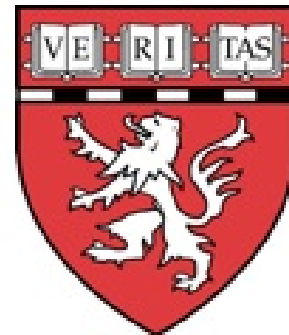
Kristin J. Nierenberg

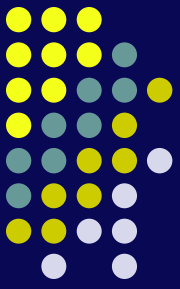
HMS III

Core Radiology Clerkship



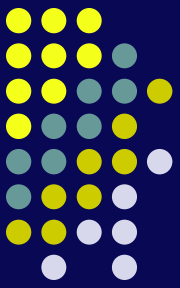
Beth Israel Deaconess
Medical Center





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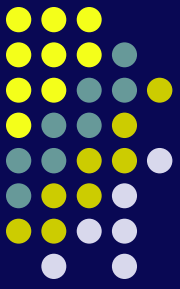
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- Basic Anatomy of Carcinoid Tumors
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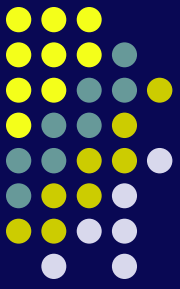
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Our patient: clinical presentation

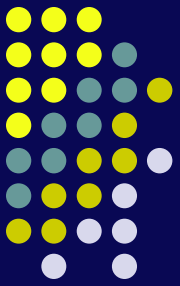


- A 46 yo F with history of celiac disease and MS has had symptoms of bloating, diarrhea, and constipation for the past 6 weeks.
- Several days before presentation developed worsening N/V, and intermittent crampy abdominal pain in epigastrium and lower abdomen.
- Denies fevers, chills or night sweats.

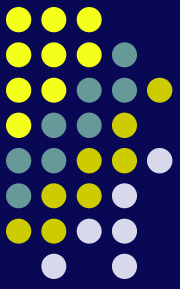
Our patient: Past Medical History



- Reports episodes of flushing for the past few years that is intermittent.
- Chronic GI symptoms from Celiac disease: on admission to hospital GI biopsy was positive but blood assays were negative.
- Has been on gluten free diet for about a year, but still reports some symptoms.
- PSH: none
- No evidence of hernias.



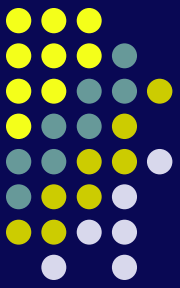
There is a wide differential diagnosis for vague abdominal pain, nausea and vomiting, especially for a complicated patient with a history of Celiac's disease. However, for the purpose of this presentation let's dive into the description of Carcinoid Tumor and Carcinoid Syndrome.



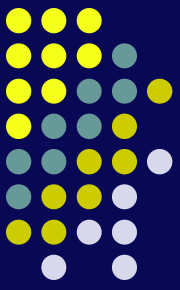
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Description of Carcinoid Syndrome



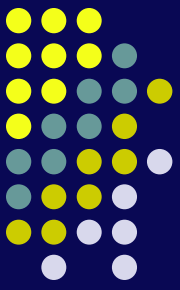
- Syndrome of symptoms caused by release of substances from carcinoid tumor:
- Bronchospasm
- Flushing
- Diarrhea
- Right-Sided Heart Failure (from valve stenoses) *Right sided failure because lungs act as filter and L. heart does not see vasoactive compounds*



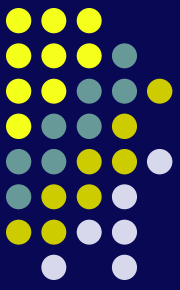
More on Carcinoid Syndrome

- 10% of patients with Carcinoid Tumor.
- Substances: serotonin and vasoactive peptides.
- Liver prevents serotonin syndrome by degrading these substances; majority of symptoms occur with metastatic disease to liver.
- Other causes: retroperitoneal disease draining to paravertebral veins and primary tumor outside of GI tract.

Carcinoid Tumor: what is it?

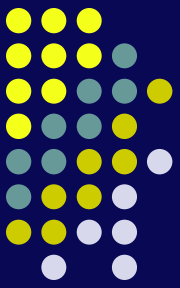


- Type of Neuroendocrine tumor (NET).
- Originates from enterochromafin cells (kulchitsky cells).
- “oid” means “resembling”, it resembles GI carcinoma but is clinically and histologically less aggressive.
- Low incidence (0.2-1.0%) but makes up 25% of all SI tumors (2nd most common).
- Most cases are asymptomatic, but can present with SBO, abdominal pain, weight loss, diaphoresis, pellagra, instussusception or carcinoid syndrome.
- SBO due to severe mesenteric fibrosis.



Carcinoid Tumor: statistics

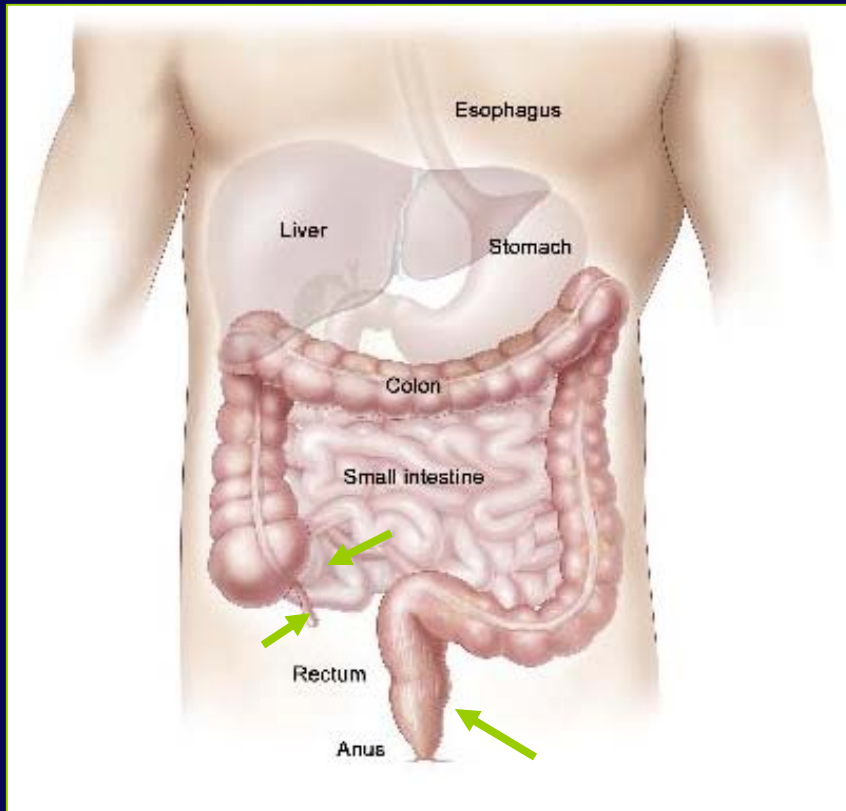
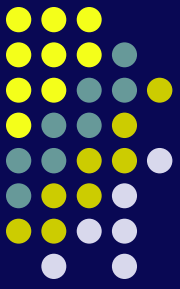
- Malignancy can only be determined by presence of metastatic disease.
- 2/3 alive at 5 years.
- Often diagnosed late, when tumor has metastasized, delay due to vague symptoms.
- Major sites of metastases are liver, lymph nodes, bone, and peritoneal cavity.
- Incidence of metastases varies with size of primary tumor: 20-30% in patients with tumor <1cm.
- Mets from small intestine primary tumors in 58-75% of patients.
- If mets are present at dx. 50% are alive at 3 years.



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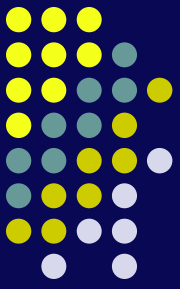
Anatomy of Carcinoid Tumor



- ~85% arise in GI tract
- 10% found in lung
- Most common sites
 - Appendix
 - Ileum
 - Rectum
 - Bronchus
- Rest found in various organs such as larynx, thymus, kidney, ovary, prostate, liver, and skin
- Ileal tumor: highest rate of metastases

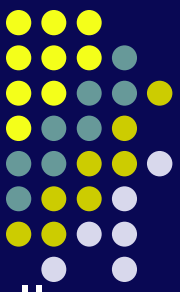
Image: <http://www.healthline.com/images/staywell/75052.jpg>

Source: Bader Till R, Semelka Richard C, Chiu Vivian CY, Armao Diane M, and Woosley John T.



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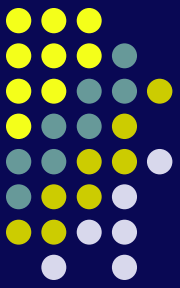
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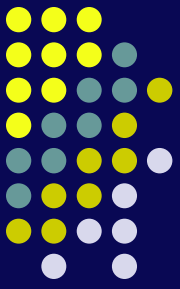
Imaging Modalities

- Diagnostic dilemmas due to overall low incidence, small size, and obscure and non-specific symptomatology.
- Evaluation includes biochemical tests, followed by anatomic and radionuclide imaging studies.
- Chest, abdomen, and pelvis **CT** are part of initial workup, offer excellent technique to demonstrate mesenteric spread and liver involvement.
- **MRI** helpful in cases where CT findings are equivocal.
- >90% of carcinoid tumors contain high concentrations of somatostatin receptors and are imaged with **OctreoScan**.
- **MIBG** and **PET** scan more recent imaging modalities.
- Can also be imaged using barium enema, upper GI series w/ SB follow through, colonoscopy, and EGD.

Companion Patient #1: A contrast enhanced CT



- CT showing typical carcinoid findings of mesenteric and liver metastases.
- Mesenteric metastasis presents as nodular, spiculated mass.
- Majority of liver mets are hypervascular.

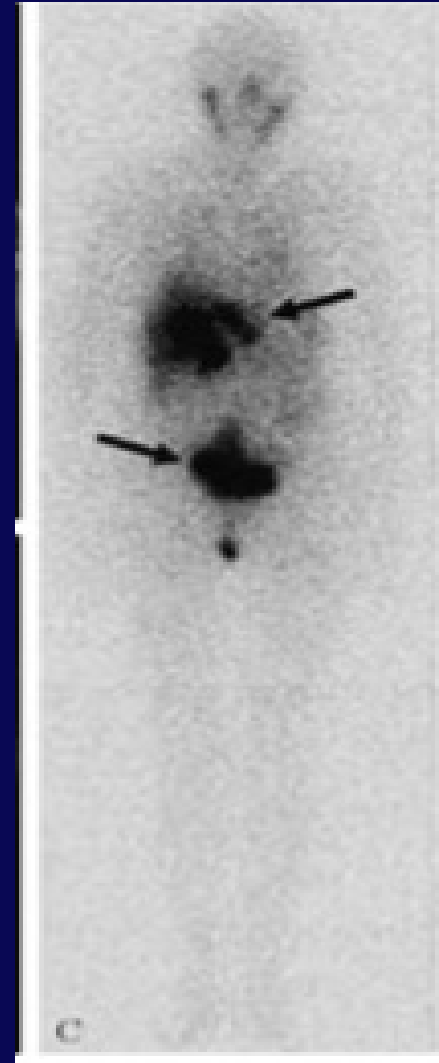


Metaiodobenzylguanidine (MIBG)

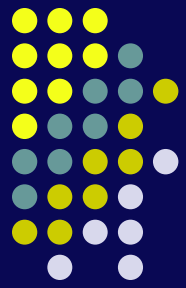
- Catecholamine analogue that is accumulated intracellularly by amine precursor uptake mechanism.
- May be incorporated into vesicles or neurosecretory granules in the cytoplasm by tissues arising from neural crest cells (I.e. carcinoid tumors).
- Labeled with radionuclide of iodine for purpose of diagnostic imaging of tumors that demonstrate MIBG uptake.
- Can be utilized for treatment using internal irradiation.
- MIBG can provide significant symptomatic relief to patients with metastatic carcinoid tumors.

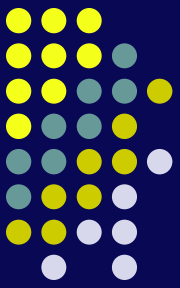
Companion Patient #2: An abnormal MIBG

- On normal MIBG whole body scan, organs that are visualized are salivary glands, liver, myocardium, and urinary bladder.
- Liver due to catecholamine metabolism.
- Bladder due to renal excretion of MIBG.
- Salivary glands and myocardium have uptake mechanism for MIBG.



Atypical MIBG showing increased focal activity in lower abdomen and multiple foci of increased MIBG Uptake in liver

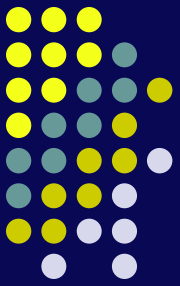




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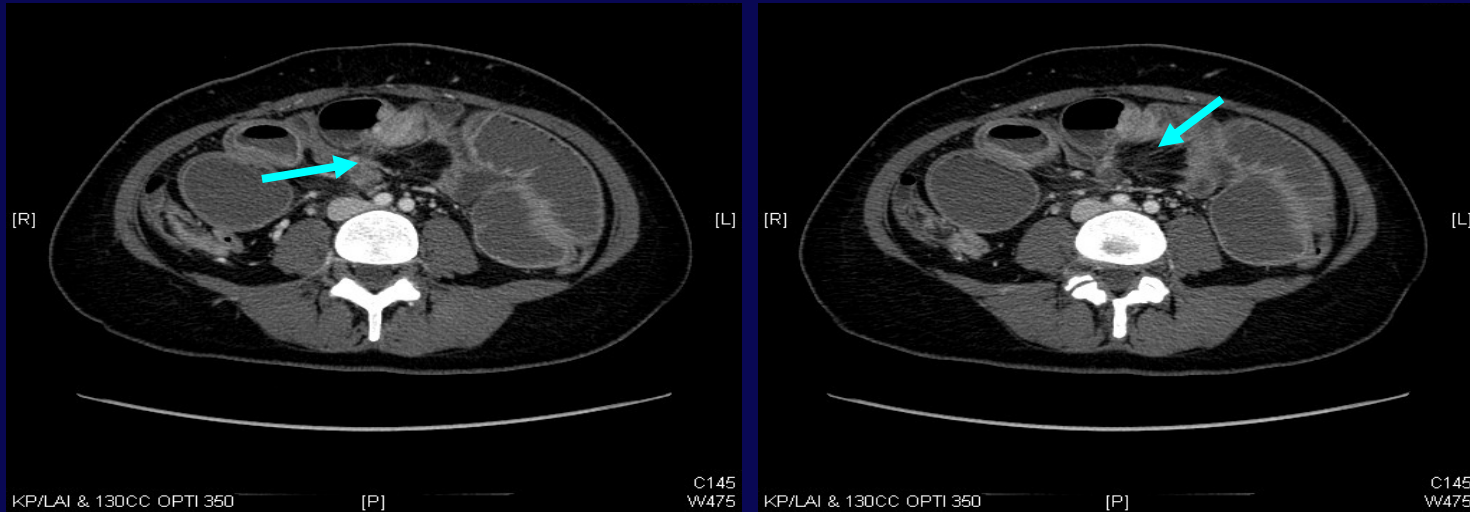
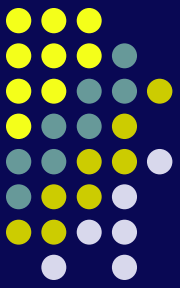
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Evaluation of our Patient



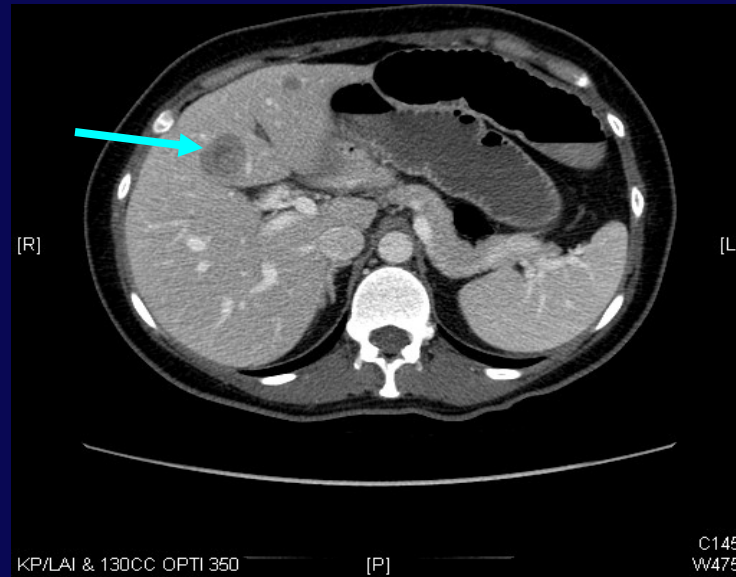
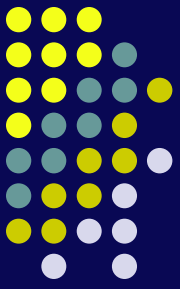
- Our patient received **CT** scan w/ & w/o contrast to evaluate for her etiology of symptoms (abd pain, n/v/d).
- This imaging showed...

Our patient: an axial CT cut of the lower abdomen



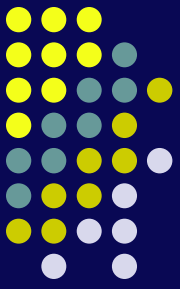
- Heterogeneous mass w/in mesentery near aortic bifurcation.
- Tethering of small bowel loops around mass.
- Transition point located in vicinity of mass.
- Multiple adjacent lymph nodes.

Our patient: an axial CT cut of the liver



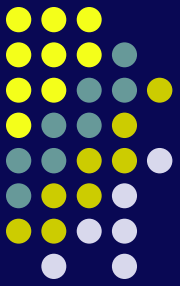
- Multiple hypoattenuating liver lesions.
- Largest lesion measuring 2.5 cm.
- Findings concerning for Carcinoid tumor with liver metastases.

Our patient: An evaluation of her CT imaging



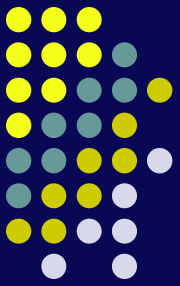
- Findings include small bowel obstruction with multiple dilated fluid filled loops of bowel.
- Transition point located in right midabdomen.
- No evidence of ischemia (pneumatosis, or bowel wall enhancement).
- Heterogeneous mass w/in mesentery.
- Multiple liver lesions.

Octreotide scan (OctreoScan)



- Most carcinoid tumors contain high concentrations of somatostatin receptors and can be imaged with OctreoScan.
- Used for dx of primary and metastatic carcinoid tumors.
- Reported sensitivity varies from 80-100%.
- Also important for assessing suitability for radiolabeled somatostatin analog therapy and for monitoring response to therapy.
- Not all carcinoid tumors express somatostatin receptors and MIBG can detect those that are negative on OctreoScan.

Our patient: An OctreoScan of the abdomen

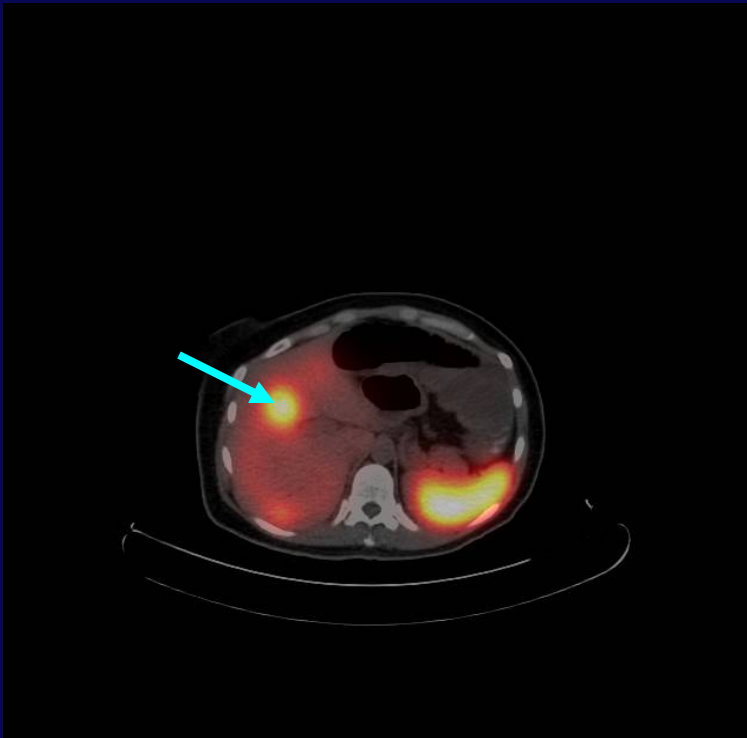
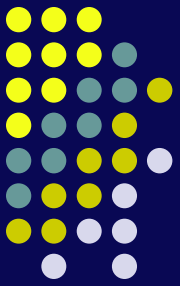


- Multiple increased tracer uptake areas in liver correlating w/ abnormalities in CT.
- Mesenteric mass adjacent to aortic bifurcation also shows increased tracer uptake.
- Superolateral to mass, area of eccentric, focal small bowel wall thickening with increased tracer uptake.
- **Appearances concerning for small bowel lesion.**



An OctreoScan of our patient showing multiple lesions in the liver as well as increased tracer uptake in the abdominal region consistent with a mesenteric mass.

Our patient: an OctreoScan of the liver

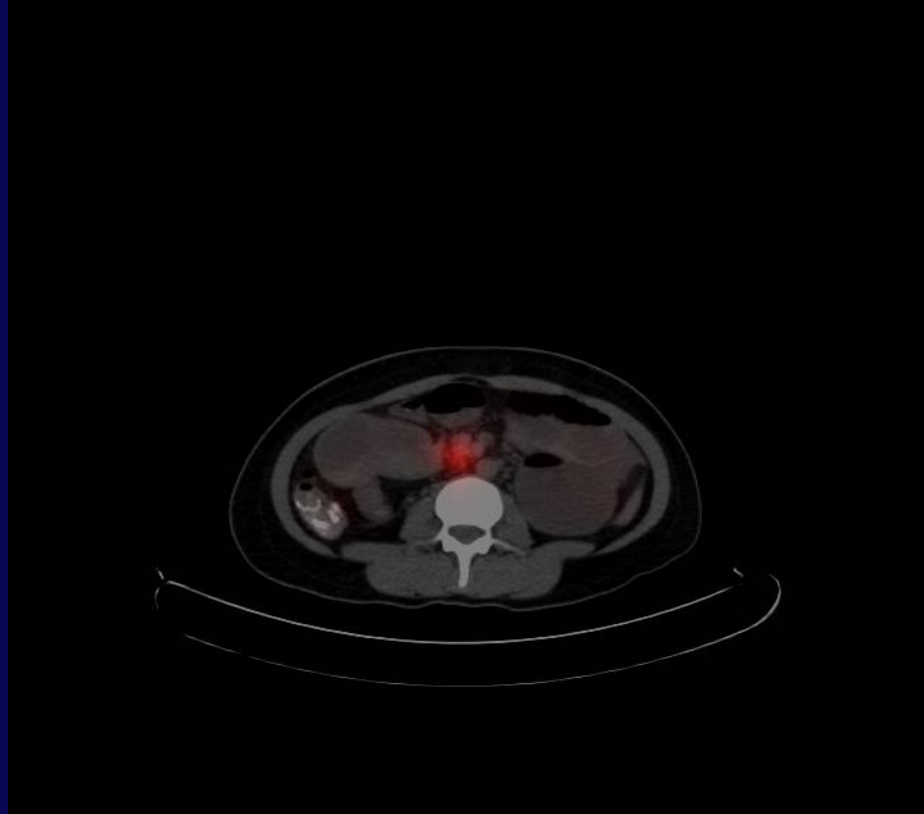
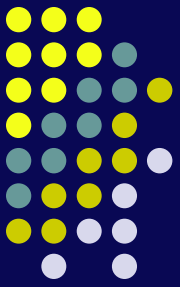


Axial CT cut of the liver of our patient matched
To her OctreoScan

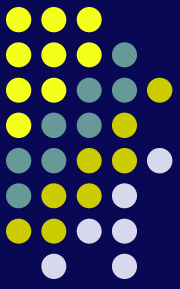


OctreoScan of our patient's liver

Our patient: a coupled axial CT to an OctreoScan

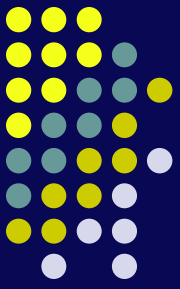


An axial CT of the lower abdomen coupled to the patient's OctreoScan showing increased uptake in the mesentery



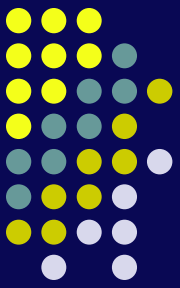
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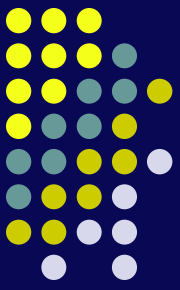
Treatment Modalities

- Surgery
- Long-acting Somatostatin analogues
- Radionuclide therapy with somatostatin analogs and MIBG
- Conventional radiotherapy
- (Chemo or Radio) embolization
- Alcohol sclerotherapy
- Radiofrequency ablation
- Cryosurgery of liver metastases
- Chemotherapy



Our patient: treatment

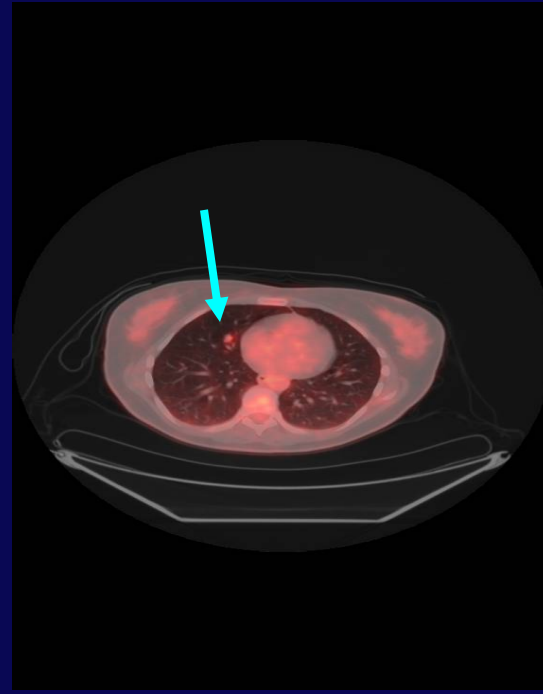
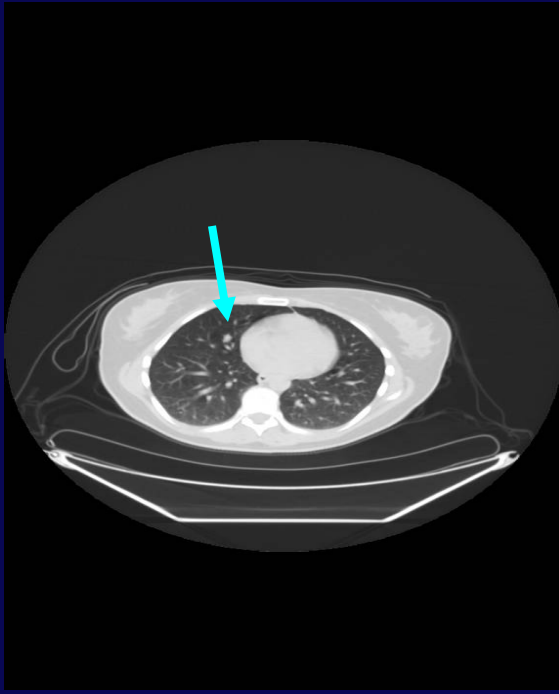
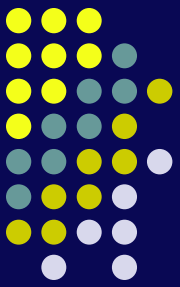
- **Surgery**
- Exploratory laparotomy
- Small bowel resection
- Wide mesenteric lymphadenectomy
- Liver disease was too extensive for resection; possible systemic therapy or chemoembolization of liver lesions in future.



Post-operative Imaging

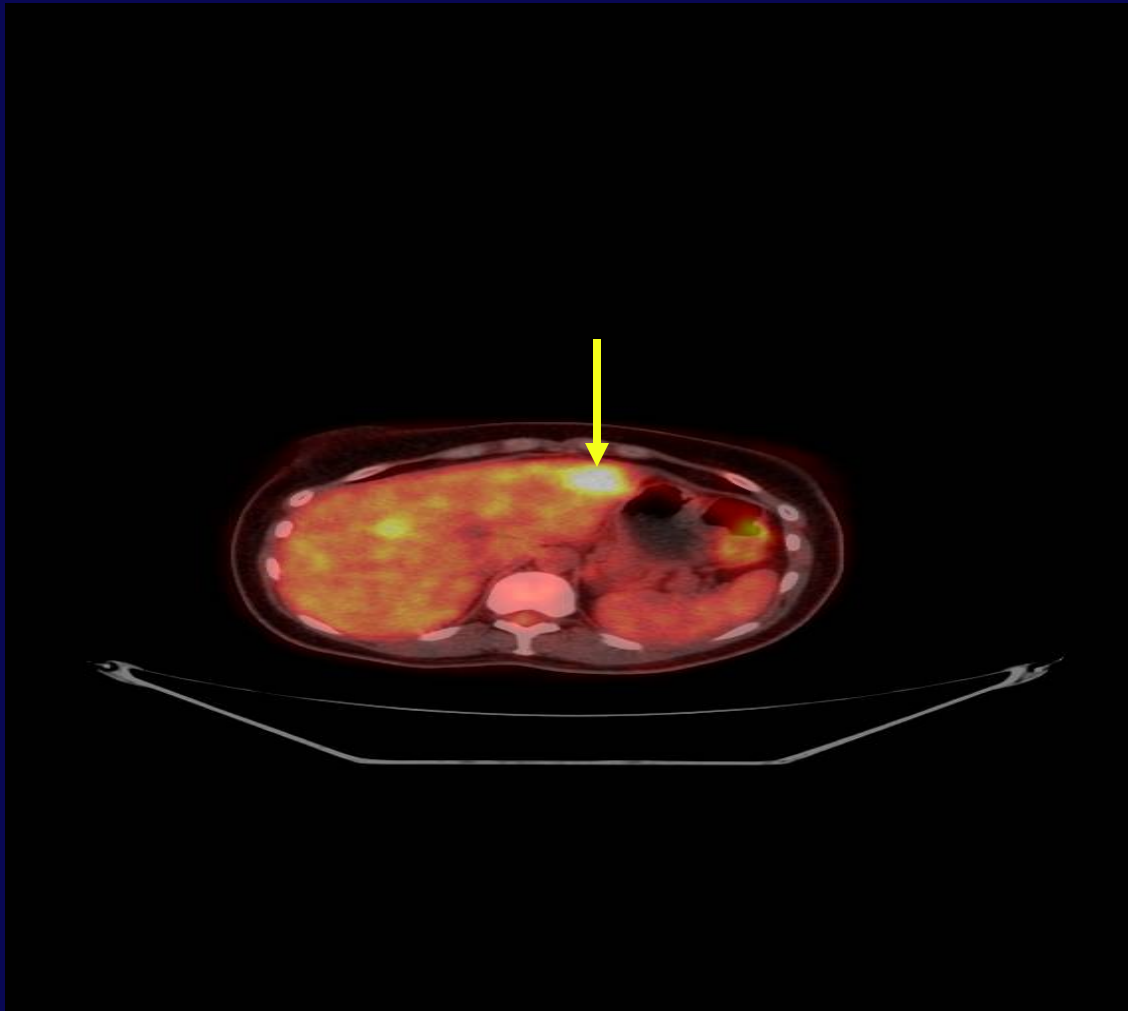
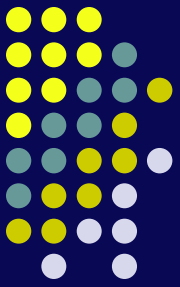
- **PET** scan using FDG (18-fluorodeoxyglucose).
- Generally malignant tumors have an increased accumulation of FDG compared with normal tissues.
- PET used to differ malignant from benign lesions and to stage malignant tumors.
- PET scan is better at detecting poorly differentiated carcinoid tumors.

Our patient: PET post-op imaging of the lungs

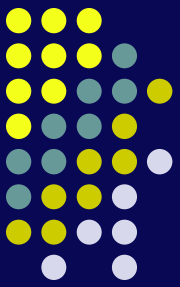


- Comparison made to previous OctreoScan.
- 2 solitary irregularly marginated FDG-avid pulmonary nodules located in middle lobe.
- Nodules represent possible metastatic foci.

Our patient: PET post-op imaging of the liver



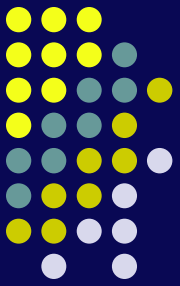
Several areas of focal increased radiotracer uptake within the liver compatible with known metastatic disease.



Take Home Points

- Carcinoid tumors are rare, but make up a large proportion of SI tumors.
- Carcinoid tumors often present late due to vague and non-specific symptomatology: thus metastases are often already present at time of diagnosis.
- There are many imaging modalities useful for diagnosis and staging including CT, OctreoScan, and PET scan.

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Acknowledgements:

- Gillian Lieberman
- Maria Levantakis
- Aarti Sekhar
- Erica Gupta

