Gastrointestinal Carcinoid Tumors: A Diagnostic Overview

Jeffrey I. Campbell, HMS III
Gillian Lieberman, MD
Outline

• Patient: A 58 year old woman with guaiac + stools
• Path/Epi: What are carcinoids? Who gets them?
• Imaging
• Treatment and outcomes
• Bronchogenic versus GI carcinoids
A 58 year old patient with...

- Vague discomfort in the left lower quadrant
- 8 lbs weight loss over past 2-3 months
- Guaiac-positive stools
- Immigrated from Thailand 2 years ago
- ...has not received routine cancer screening
GI Tumors

Esophagus:
- Squamous Cell
- AdenoCA

Stomach:
- AdenoCA

Small Intestine:
- Carcinoid
- AdenoCA
- GIST

Colon:
- AdenoCA
- Carcinoid

Rectum:
- AdenoCA
- Squamous Cell

Everywhere:
- Lymphoma
- Metastases

Our patient: Colonoscopy

Image source: PACS, BIDMC
Our Patient: Biopsy

Below: positive stain for chromogranin A

Above: sub-mucosal cellular proliferation (right arrow)

Our Patient: Sagittal MRI of the Pelvis

T2 Weighted Sagittal MRI

Image: PACS, BIDMC
Our Patient: Sagittal MRI of the Pelvis

T2 Weighted Sagittal MRI
Image: PACS, BIDMC

Rectal mass
What is a carcinoid tumor?

- “Resembling a carcinoma”
- Well- to moderately-differentiated neuroendocrine tumor (NET)
- Often secrete hormones, ex. serotonin (5HT)

Biochemical testing for carcinoids

- Break-down products of 5HT
- 5-HIAA (Tryp $\rightarrow$ 5-HTP $\rightarrow$ 5HT $\rightarrow$ 5-HIAA)
- Other hormones and breakdown products (histamine, substance P, pancreatic polypeptide, etc.)

Carcinoid: Epidemiology

Epidemiology

- Incidence: 38.4/million\(^1\)
- Appendiceal carcinoids: 1/200-300 appendectomies\(^2\)
- Most common site of metastasis: liver

Distribution (% of all carcinoids)

- Stomach: 7%
- SI: 45%
- Appendix: 17%
- Colon: 11%
- Rectum: 20%
- Other GI: 1%

Sources:

Carcinoid Syndrome

- From release of 5HT
- Almost always after metastasis to liver
- Symptoms:
  - Flushing and diarrhea
  - Carcinoid crisis...may be fatal!
- Rare: <20% of patients with carcinoid tumors

What would you expect this patient’s abdominal CT to look like?

Companion Patient 1: C+ Axial

Companion Patient 1: C+ Axial CT

Multiple hepatic metastases (examples identified with arrows)

Carcinoid Syndrome: Explanation

- 5HT is deactivated in the liver
- Venous return from most GI carcinoids must pass through the liver before entering systemic circulation
- 5HT can only reach systemic circulation if it is produced in, or after, the liver

Companion Patient 2: C+ Coronal CT

• Before arriving at the liver, carcinoid may first spread to local lymph nodes


Companion Patient 3: C+ Axial CT

- What else could this be?

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- What else could this be?

Mass in the Small Intestine

Companion Patient 3: C+ Axial CT

- What else could this be?
  - Lymphoma
  - Metastasis
  - GIST
  - AdenoCA

Mass in the Small Intestine

Companion Patient 4: Barium Enema

Companion Patient 4: Barium Enema

- Large carcinoids may also appear as filling defects in the large intestine

Companion Patient 5: T1 MRI with Gadolinium to Detect Metastasis

- Which lesion is a metastasis?

Companion Patient 5: T1 MRI with Gadolinium to Detect Metastasis

- MRI with gadolinium can help distinguish between liver metastases and cysts.

Companion Patient 6: Octreotide Scan to Detect Carcinoids

- Nuclear scintigraphy – radiolabeled hormones concentrate in carcinoid tumors
  - Octreotide (and octreotide analogs)
  - I-131 MIBG

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Abnormal enhancement

Image: courtesy of Dr. Koenraad Mortele

Companion Patient 7: Octreotide Scan to Detect Metastases


Treatment

Local resection

Debulking of metastases
  - Surgical
  - Intra-arterial embolization

Systemic therapy:
  - Somatostatin analogs
  - Cytotoxic drugs

Octreotide scan showing multiple liver metastases

Companion Patient 8: Octreotide Scan

Before and after radiotherapy ablation of hepatic metastases

Outcomes

• A paradox: carcinoids tend to grow slowly (remember, they are well- to moderately-differentiated by definition)
  – Not highly aggressive
  – But hard to find
  – Hard to treat with cytotoxic therapy

# Lung vs GI Carcinoids

<table>
<thead>
<tr>
<th></th>
<th>Bronchogenic</th>
<th>Gastrointestinal</th>
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</thead>
<tbody>
<tr>
<td><strong>Origin</strong></td>
<td>Epith endocrine cells</td>
<td>Enterochromaffin-like cells, epith endocrine cells</td>
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<tr>
<td><strong>Prevalence</strong></td>
<td>38.4/million (1)</td>
<td>2-20/million (2)</td>
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<tr>
<td><strong>Classification</strong></td>
<td>Proliferation</td>
<td>Location, differentiation</td>
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<tr>
<td><strong>Carcinoid syndrome</strong></td>
<td>Very rare</td>
<td>~8% (3)</td>
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<tr>
<td><strong>Curative treatment</strong></td>
<td>Resection</td>
<td>Resection</td>
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<td><strong>5-year survival</strong></td>
<td>74% (varies by proliferative index) (4)</td>
<td>71-88% (varies by location) (4)</td>
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Sources:
Return to Our Patient

- Underwent surgical resection
- No metastases found on octreotide scanning
- Doing well, with frequent follow-up
Conclusions

- Carcinoids are relatively rare overall, but most commonly affect the SI, appendix and rectum.
- They can present with classic carcinoid syndrome, but infrequently do.
- Imaging includes the usual suspects (barium studies, CT, MRI) and specialized scintigraphy (octreotide, MIBG).
- These are indolent tumors, and local resection is often curative.
- Aside from surgery, therapy includes octreotide and cytotoxic agents.
Acknowledgments

• Our patient
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• Dr. Jonathan Kim
• Dr. Koenraad Mortele
• Megan Garber and Claire Odom
4. Harrison’s.
Questions?

You're Blushing

Must Be Carcinoid Syndrome