Surgical Management of Neuroendocrine Tumors of the Gut

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Sites of GI Carcinoid Tumors

- Small intestine: 44%
- Rectum: 21%
- Colon: 15%
- Appendix: 7%
- Stomach: 7%

Pathology of Carcinoids

“Typical”
- Well differentiated
- Slow spread
- Trabecular, glandular patterns

“Atypical”
- Poorly differentiated
- More rapid spread
- Nuclear atypia. Focal necrosis, etc
- “Neuroendocrine carcinomas”

Likelihood of malignancy is strongly correlated with size of tumor
## GASTRIC CARCINOIDS

<table>
<thead>
<tr>
<th>BENIGN (80%)</th>
<th>MALIGNANT (20%)</th>
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</thead>
<tbody>
<tr>
<td>&lt; 1 cm</td>
<td>&gt; 3 cm.</td>
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<tr>
<td>Non-functioning</td>
<td>Functioning</td>
</tr>
<tr>
<td>Setting of CAG</td>
<td>Sporadic</td>
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<tr>
<td>Highly differentiated</td>
<td>Less differentiated</td>
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</table>
TYPE 1 GASTRIC CARCINOIDS (75%)

Develop in Setting of CAG
- Body and fundus
- Small (< 1.0 cm.)
- Multifocal
- Indolent

Management
- Surveillance
- Endoscopic removal
- Antrectomy?

Kulke and Mayer
NEJM 1999
TYPE 1 GASTRIC CARCINOIDS

Borch et al (Ann Surg 2005) reviewed 65 gastric carcinoids from 24 different hospitals in Sweden. 51 of the 65 were Type 1. Median age = 65 yrs.

Treatment recommendations:

< 5 small tumors --- endoscopic removal

> 5 tumors or large (> 1 cm) tumor --- antrectomy and excision of large tumors
Associated with ZES/MEN-1 Syndrome

Genetic predisposition
Malignant potential - low grade

Management
Endoscopic removal
Long acting octreotide?
TYPE 3 GASTRIC CARCINOIDs - SPORADIC
20%

Large (> 2 cm)
Aggressive - deep invasion
Carcinoid syndrome (10-20%)

Management
Gastrectomy with lymph node dissection
SMALL BOWEL CARCINOIDS

1/3 of all small bowel tumors
- mainly ileum
- may be multi-centric

Vague symptoms
- lag time to diagnosis often years
- usually metastatic when diagnosed

5-7% have Carcinoid syndrome

Associated fibrosing mesenteritis
- stranding on CT scan a clue
- local complications: obstruction, infarction
SMALL BOWEL CARCINOIDS

5 YEAR SURVIVAL

Complete resection ....................... 60%
Incomplete resection .................... 38%
Liver mets .................................. 21%

Best/only chance of cure is surgical resection of tumor and nodal mets
CT Scan – Small Bowel Carcinoid
Surgery for small intestinal carcinoids

• Usually unexpected finding at laparotomy
• Resect the tumor(s) + as much of the involved mesentery as is feasible
• Surgical cure is rare
The surgical problem with small bowel carcinoids
Appendiceal Carcinoids

- Most common tumor of the appendix
- Found in 0.3% of all resected appendices
- Asymptomatic or associated with appendicitis
- Often diagnosed on final path

If > 2 cm, 1/3 will have nodal metastases. Therefore, do a right colectomy.
Surgery for appendiceal carcinoid tumors

< 2 cm and not at base/margin - simple appendectomy

> 2 cm or at base/margin - proceed to right colectomy
Adenocarcinoids
(aka goblet cell carcinoid, mucinous carcinoid)

Mixed tumors
May stain for chromogranin A
Usually appendiceal origin

Can be aggressive
Distant mets present at discovery - 11%
Overall mortality 20-30%
- related to size/spread at discovery
Synchronous/metachronous colon cancers

Right colectomy is generally recommended
COLON CARCINOIDS

- Predominantly R sided
- Does not cause Carcinoid syndrome
- Late detection
  - Average tumor size 5 cm
  - Mets present in 2/3
- 5 year survival
  - Localized............... 70%
  - Regional............... 44%
  - Distal............... 20%
Surgery for colonic carcinoid tumors
RECTAL CARCINOIDs

• Usually asymptomatic - discovered at flex sig
  Yellow, subcutaneous “collar button”
• Carcinoid syndrome exceedingly rare
• Probability of mets correlates with size

Management

< 1 cm: Local excision
1-2 cm: Controversial
> 2 cm: Radical resection
RECTAL CARCINOIDS

5 Year Survival

Local disease........................ 81%
Regional met’s....................... 47%
Distant met’s....................... 18%
Surgery for Carcinoid tumors of the luminal GI tract

Pre- vs Post-op diagnosis

- **Stomach, colon and rectum** - Can usually be diagnosed pre-operatively and appropriate resections performed.

- **Small bowel and appendix** - Usually are not diagnosed pre-operatively. Appropriate surgery may not be done or may need to be done at a second operation.
Surgery for Carcinoid tumors of the luminal GI tract

The location matters

- **Stomach, appendix, colon, and rectum** - Surgical resection is a standard technical approach.

- **Small bowel** - Surgical resection is technically different and more demanding than the standard operation.
Surgery for Carcinoid tumors of the luminal GI tract

Take home messages

- **Stomach** - They are very rare - must distinguish good and bad actors. Bad tumors should be treated just like adeno-CA, others can be treated with endoscopy.
- **Small bowel** - rare to make pre-op Dx. Radical resection is best chance at cure.
- **Appendix** - Usually an incidental finding. Right colectomy for tumors > 2 cm or at margin, otherwise simple appendectomy.
- **Colon** - Standard radical resection offers best chance of cure.
- **Rectum** - small tumors without LV invasion can have local excision - others should get radical resection.
Thank You