BC CRC Update
Unusual Colorectal Tumors

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Disclosures

• I have no disclosures
Objectives

- Neuroendocrine Tumors (Carcinoids)
  - Appendiceal
  - Rectal
- Rectal GISTs
- Melanoma
  - Small bowel
  - Anorectal
- ColoRectal Lymphoma
Quality of Evidence Poor!!!
Neuroendocrine Tumors of Appendix

• Most common appendiceal tumor
• Most are found incidentally
  • 1 per 300 appendectomies
• Overall mets 4%, distant mets 0.7%
• Size predicts metastatic potential
Size Matters!

• < 1cm
  – Rarely metastasize - Appy

• 1-2cm
  – Depends

• >2cm
  – Risk of lymph node mets 30% - right hemi
High Risk Features for 1-2cm

- Invasion into mesoappendix
- Lymphovascular invasion
- Serosal involvement
- Involved margins
- Positive lymph nodes in appy specimen
- Hi Ki 67 index (>2%)
- Goblet cell variant
Does Resection affect Survival?

• Most would probably assume so.

  – Tumor size > 2cm predicted LN mets
  – No difference in survival between R hemi and appy

Goblet Cell Carcinoid / Adenocarcinoid AKA Mixed endocrine / exocrine tumor

• Rare variant
• Aggressive
  – Often see peritoneal disease at presentation

• Treatment
  – Right hemicolecotomy regardless of size, consideration of adj chemo, consideration of peritoneal stripping & HIPEC when disseminated
• Overall 10 year survival 60%
Rectal Carcinoid (or NET of Rectum)

- Incidence increasing
  - 11% of GI NETSs & 1.5% of rectal neoplasms
  - 10 fold increase past 35 yrs

- More common in African descent
- Typically > 55 yrs of age
- Most discovered incidentally
- Prognosis size dependent
Size Matters (again)

- **< 1 cm**
  - Incidence of LN involvement 0-3%
  - Excise endoscopically, TEM, or transanally

- **1-2 cm**
  - Regional or distant disease 7-34%
  - Treatment controversial

- **> 2 cm**
  - Regional and distant spread 67%-100%
  - Radical excision indicated
<table>
<thead>
<tr>
<th></th>
<th>Nodal Involvement</th>
<th>Total, n (%)</th>
<th>P</th>
<th>Metastases</th>
<th>Total, n (%)</th>
<th>P</th>
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</thead>
<tbody>
<tr>
<td>Sex</td>
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<tr>
<td>Male</td>
<td>19</td>
<td>62 (30%)</td>
<td>0.55</td>
<td>7</td>
<td>62 (11%)</td>
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<td>Female</td>
<td>15</td>
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<td>38 (12.8%)</td>
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<tr>
<td>≤60</td>
<td>19</td>
<td>54 (35%)</td>
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<td>&lt;0.001</td>
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<tr>
<td>&gt;60</td>
<td>15</td>
<td>46 (32%)</td>
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<td>10</td>
<td>46 (21%)</td>
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<tr>
<td>Tumor size (mm)</td>
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<tr>
<td>1–10</td>
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<td>0.038</td>
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<td>25 (0%)</td>
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<td>11–20</td>
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<td>2</td>
<td>36 (6%)</td>
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<tr>
<td>&gt;21</td>
<td>17</td>
<td>29 (59%)</td>
<td></td>
<td>7</td>
<td>29 (24%)</td>
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<td>Tumor depth</td>
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<tr>
<td>T1</td>
<td>4</td>
<td>32 (13%)</td>
<td>&lt;0.001</td>
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<td>32 (0%)</td>
<td>0.029</td>
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<tr>
<td>T2</td>
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<td>1</td>
<td>25 (4%)</td>
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<tr>
<td>T3</td>
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<td>5</td>
<td>29 (17%)</td>
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<tr>
<td>T4</td>
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<td>11 (55%)</td>
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<td>3</td>
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<td>Lymphovascular invasion</td>
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<td></td>
<td></td>
</tr>
<tr>
<td>−ve</td>
<td>2</td>
<td>41 (5%)</td>
<td>&lt; 0.001</td>
<td>0</td>
<td>38 (0%)</td>
<td>&lt;0.001</td>
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<tr>
<td>+ve</td>
<td>24</td>
<td>37 (65%)</td>
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<td>6</td>
<td>36 (17%)</td>
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<td>Distance from anal verge, mm</td>
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<td>≤70</td>
<td>10</td>
<td>39 (26%)</td>
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<tr>
<td>≥70</td>
<td>17</td>
<td>47 (36%)</td>
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<td>7</td>
<td>43 (16%)</td>
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<tr>
<td>Treatment</td>
<td></td>
<td></td>
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<tr>
<td>Ante</td>
<td>12</td>
<td>41 (29%)</td>
<td>0.271</td>
<td>4</td>
<td>41 (10%)</td>
<td>0.050</td>
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<td>AR and TME</td>
<td>14</td>
<td>45 (31%)</td>
<td></td>
<td>2</td>
<td>45 (4%)</td>
<td></td>
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<tr>
<td>Hartmann’s procedure</td>
<td>3</td>
<td>7 (21%)</td>
<td></td>
<td>3</td>
<td>14 (21%)</td>
<td></td>
</tr>
<tr>
<td>APR</td>
<td>3</td>
<td>7 (21%)</td>
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</table>

APR indicates abdomino-perineal resection; AR, anterior resection; TME, total mesorectal excision.
Effect of Size on Metastases

Lymph Node Metastases

Distant Metastases

< 1 cm 1-2 cm > 2 cm
Effect of LVI on Metastases

- Lymph Node Metastases
- Distant Metastases

- neg LVI
- pos LVI
Rectal GIST

- 7% of all GISTs
- Typically over 50 yrs
- Male predominance
- Frequent positive margins on excision (40%)

- Express KIT – so can be targeted with imatinib
Rectal GIST

• Behaviour
  – Based on size, Number of mitoses per 50 high power fields
    – \(<2\text{cm} \& \text{<5 mitoses per 50 HPF}
      – No risk of metastatic disease
    – 2-5 \text{ cm} \& \text{<5 mitoses per 50 HPF}
      – Low risk of metastatic disease
    – \(>5\text{ cm or any tumor size with >5 mitoses per HPF}
      – **High risk** of metastatic disease (\(>50\%\))
Rectal GIST - Approach

• Clearly resectable, acceptable morbidity
  – Primary Surgery

• Resectable but not without significant morbidity
  – Neoadjuvant imatinib

• Unresectable
  – Neoadjuvant or palliative imatinib
Small Bowel Melanoma

- Most are metastatic lesions
- Difficult to be certain if primary or met
  - Criteria: No evidence of concurrent melanoma or atypical melanocytic lesion, absence of metastases other than regional nodes, presence of intramucosal lesion
- Worse prognosis than cutaneous melanoma
Small Bowel Melanoma

- 60% of pts who die of melanoma have GI mets
  - Only 1.5%-4% are diagnosed
- Typically see multiple polyploid masses
- Present as other SB tumors do
- Surgery indicated for symptom control, esp if no evidence of disseminated disease
- Median survival 6-9 months, 15 months if completely resected
- 5 year survival < 10%
AnoRectal Melanoma

- Very rare!
  - 0.2% of all melanoma
  - 0.1%-4.6% of all malignant tumors of the rectum & anus
  - Can be melanotic or amelanotic

- Outcome very poor
  - 5 yr disease free survival 6.7%-12%
  - Median survival 19 months
Anorectal Melanoma
Local vs Radical Surgery

• Most reports indicate no difference in outcome between local & radical excision
  » Homsi & Garrett. Dis Colon rectum. 2007; 50: 1004-1010

• One case series (MSK) suggested advantage to APR (84 pts over 65 yr period, 71 not metastatic)
ColoRectal Lymphoma

• Again very rare!
  – 0.2-0.6% of colorectal malignancies
  – Dawson’s criteria to establish primary CR lymphoma

– In our series, rectal lymphoma was associated with reduced median survival (42 vs 110 months); and pts having surgical resection had improved survival (110 vs. 56 months)
Thanks!
TABLE 3. Multiple Variable Logistic Regression Analysis of Risk Factors for Lymph Node Metastases*

<table>
<thead>
<tr>
<th>Variables</th>
<th>Odds Ratio</th>
<th>95% CI</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tumor size &gt; 10 mm</td>
<td>32.7</td>
<td>14.8–72.3</td>
<td>0.006</td>
</tr>
<tr>
<td>Lymphovascular invasion</td>
<td>19.6</td>
<td>12.3–146.0</td>
<td>&lt; 0.001</td>
</tr>
</tbody>
</table>

*Risk factors for nodal involvement calculated in patients who underwent formal surgical resection (n = 100).
**TABLE 4**

Shields, Conor; MD, FRCSI; Tiret, Emmanuel; Winter, Desmond; MD, FRCSI 
DOI: 10.1097/SLA.0b013e3181fb8df6

<table>
<thead>
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<th>Variables</th>
<th>Odds Ratio</th>
<th>95% CI</th>
<th>P</th>
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<tbody>
<tr>
<td>Lymph node metastases</td>
<td>12.3</td>
<td>1.8-84.7</td>
<td>0.033</td>
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<tr>
<td>Lymphovascular invasion</td>
<td>74.4</td>
<td>4.6-120.2</td>
<td>0.022</td>
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</table>

*Risk factors for distant metastases calculated in patients who underwent formal surgical resection (n = 100).*