Rare Anorectal Neoplasms
CT/MRI-PATHOLOGIC CORRELATION

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Disclosures

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Anorectal Neoplasms

THE MENU [15 MIN]

- Background
- Anorectal Anatomy
- Rectal AdenoCA
- Others: Anal cancer, GIST, Melanoma, Lymphoma, Carcinoid
  - CT/MRI/PATH
- Conclusions
Malignant Rectal Dx

BACKGROUND

- **Adenocarcinomas** comprise most (98%) of anorectal malignancies
  - 45,000 new cases/year in the USA
  - 17,000 deaths/year
- **Squamous cell carcinomas** arise at the ano-rectal transition area & are considered anal CA
- **Other rare cancers** may involve the anorectum
  - lymphoma (1.3%), GIST (0.3%)
  - carcinoid (0.1%), melanoma (0.1%)

*American Cancer Society: Cancer facts and figures 2007. Atlanta, GA*
"When in the anus, think squamous"

- 4,650 new cases/year; 690 deaths/year
- Lymphatic drainage to inguinal & femoral nodes
- Most caused by HPV infection (88%)
- Cigarette smoking, immunosuppression, HIV
- Tumor size most important prognostic factor
- Tx: Radiation therapy (70-90% cure rate)

American Cancer Society: Cancer facts and figures 2007. Atlanta, GA
TNM classification

- **T1**: < 2 cm
- **T2**: 2 - 5 cm
- **T3**: > 5 cm
- **T4**: adjacent organs

**T1 Anal CA**
normal rectum

ulcer

nets of malignant keratinizing (pink) squamous cells
T4 Anal CA
nets of malignant non-keratinizing (purple) squamous cells

normal anus
N3 anal CA

TNM classification
- N1: perirectal
- N2: unilateral iliac inguinal
- N3: bilateral iliac or inguinal
  OR perirectal and inguinal
The Other Malignant Dx
LYMPHOMA

- primary uncommon (0.2% of rectal cancers)
- non-Hodgkin (95%), HIV/AIDS (90%)
- male predominance (AIDS), in adults (30-70yrs)
- diffuse large B-cell
- longstanding UC
- immunosuppression
- 5-y survival 50% (surgery)


ulcer

sheets of discohesive (white border) malignant round epithelioid blue cells, prominent nucleoli
The Other Malignant Dx
LYMPHOMA

Imaging
- homogeneous
- mural large mass
- severe wall thickening
- luminal narrowing
- “aneurysmal” dilation
- minor obstruction
- fistula formation
- adenopathy

GASTRO-INTESTINAL STROMAL TUMOR

- most common mesenchymal tumor in the GI tract
- interstitial cells of Cajal (ICC) of the myenteric plexus
- 95% express cell surface receptor KIT (CD 117)
  - targets for KIT-inhibitor therapy [Imatinib (Gleevec)]

The Other Malignant Dx

GIST

- 3300-4350/year in the USA
  - 5% anorectum (250 cases)
- slight male predominance, 50yo
- high rate of recurrence & mets after local excision (62-75%)
  - liver, peritoneum
- stratified by size and mitotic rate
  - >3cm, >5 mitoses/hpf
- majority located within 10cm of the anal verge
- complete surgical excision, adjuvant Gleevec therapy

The Other Malignant Dx
GIST

Imaging

- submucosal, mural
- well-margined
- expands the rectal wall
- smooth, broad border
- hemorrhage or necrosis
- no perirectal adenopathy
- exophytic component
- cavitation
- hypervascular

Malignant GIST
The Other Malignant Dx

CARCINOID TUMOR

- uncommon (1.3% rectal tumors), small (0.6cm)
- rectum 2nd/3rd most common GI tract location
- arises from submucosal neuroendocrine cells
- no gender predilection, median age 56 years
- more common in African American population
- rarely produces serotonin (carcinoid syndrome)
- <1cm extremely low; 1-2cm: 4-30%; and > 2cm: metastases in >70%

_Hamada et al_. Pedunculated rectal carcinoid removed by endoscopic mucosal resection. _World J Gastroenterol_ 2003;9:2870-2872
<table>
<thead>
<tr>
<th>Site</th>
<th>Percentage</th>
<th>5-Year Survival (%)</th>
</tr>
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<tbody>
<tr>
<td>Stomach</td>
<td>4</td>
<td>48</td>
</tr>
<tr>
<td>Duodenum</td>
<td>3</td>
<td>–</td>
</tr>
<tr>
<td>Jejunum</td>
<td>3</td>
<td>–</td>
</tr>
<tr>
<td>Ileum</td>
<td>20</td>
<td>–</td>
</tr>
<tr>
<td>Small intestine (all)</td>
<td>29</td>
<td>55</td>
</tr>
<tr>
<td>Appendix(^1)</td>
<td>25</td>
<td>85</td>
</tr>
<tr>
<td>Colon</td>
<td>10</td>
<td>41</td>
</tr>
<tr>
<td>Rectum</td>
<td>17</td>
<td>72</td>
</tr>
</tbody>
</table>

**TABLE 21-1. Relative Frequency of Gastrointestinal Neuroendocrine Tumors by Site**
normal rectum

well-circumscribed nests of cells, submucosal in location
The Other Malignant Dx
CARCINOID TUMOR

Imaging

- small
- solitary
- smooth
- submucosal
- Octreotide+
- broad base
- polypoid
- homogeneous, hypervascular

Carcinoid recurrence
The Other Malignant Dx

MELANOMA

- anus 3rd most common source (skin and eyes)
  - 0.2%-3% of all melanomas
  - 1%-4.6% of anorectal cancers
- rectal melanomas exceedingly rare
- elderly pts with rectal bleeding and tenesmus
- immunostain S-100, HMB-45, melan-A
- pigmentation (81-90%)
- 5 year survival: 6.7-12%

Other Malignant Dx
MELANOMA

Imaging
- polypoid or fungating mass
- intraluminal protrusion
- no obstruction
- high SI T1-WI (melanotic)
- lymphadenopathy

Anorectal Pitfalls

DEEP ENDOMETRIOSIS

- most often upper rectum
- mucosal and submucosal involvement common
- fibrosis, inflammation, and smooth muscle hypertrophy common
- CD10 highlights endometrial stroma
- endometrioid adenocarcinoma rare
- **IMAGING:** mural mass, T2/T1 hypointense, enhancing, associated endometriomas, triangular, punctate T1 spots, T2 cysts

Anorectal Pitfalls

TAILGUT CYST (CYSTIC HAMARTOMA)

- anal canal mass with bleeding and pain
- multilocular, hemorrhage common
- squamous, columnar or transitional epithelium
- may become infected
- very rare cases of malignant transformation
- **IMAGING:** multilocystic, variable SI on T1/T2WI due to mucinous material, high protein content, or hemorrhage

Conclusions
IMAGING RARE ANORECTAL TUMORS

- **The Other Cancers**
  - different staging & Tx
  - specific CT/MRI features

- **Pitfalls**