Case presentation

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History

- 51 yo male c/o diffuse abd pain x 2 days
  - Nausea, Vomiting x 4 episodes
  - Last BM day prior
- PMHx: Crohn’s dz (15 yrs)
  - SBO x 4 (cons. Rx)
- PSHx: colonoscopy 2007 (normal)
- Meds: Pentasa
  - Previously on steroids
Physical Exam

- Vitals: 96.8, 108/74, 78

- Abdomen: soft, mild distention, diffuse tender, no rebound or guarding

- Labs: WBC 10.9
Imaging

- CT scan: small bowel obstruction with possible transition point in mid-abdomen
Hospital course

- NG tube and IV fluid resuscitation
- + flatus/BM on HD #1
- UGI series: Normal transit of contrast material into the cecum
Hospital course

- HD #2: Tolerated clears
- Capsule endoscopy: Polyp lesion in mid jejunum possibly causing intussusception
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<th>Patient Information</th>
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<td>Capsule No.:</td>
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<td>Referring Physician:</td>
<td>R. Bookooe</td>
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<td>Presenting Problem:</td>
<td>Abdominal pain and SBO</td>
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<td>Differential Diagnosis:</td>
<td>Polypoid lesion in mid jejunum</td>
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<td>Summary:</td>
<td>Polypoid lesion in mid jejunum</td>
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Examination Date: 4/5/2016

Signature: ________________________________
Operative course

- Laparoscopic assisted small bowel tumor resection
  - Polyp lesion in mid jejunum
  - Segmental resection & primary anastomosis
  - No other lesions noted
Pathology

- Carcinoid tumor
  - 1.5 x 1.2 x 0.4 cm
  - Low grade neuroendocrine tumor
  - No necrosis
  - Margins negative
  - Ki-67 immunostain negative
Management of Small Bowel Tumors
Small bowel neoplasms

- Benign
  - Adenomas
  - Fibromas
  - Lipomas
  - Hemangiomas
  - Lymphangiomas
  - Neurofibromas

- Malignant
  - Adenocarcinomas
    - 35 - 50%
  - Carcinoid
    - 20 - 40%
  - Lymphomas
    - 10 - 15%
  - GIST
    - 15%
  - Melanoma (mets)

Majority asymptomatic
Incidental on EGD

5th or 6th decade
Related diseases

- Crohn’s disease
- Celiac sprue
- HNPCC
- FAP
- Peutz-Jeghers syndrome
Pathophysiology

- 90% GI tract mucosal surface
  - 1.1 - 2.4% of all GI malignancies
- Proposed explanations:
  - Dilution of carcinogens in liquid chyme
  - Rapid transit limits mucosal contact
  - Low bacterial concentration
    - Decreased metabolic carcinogenic products
  - Mucosal protection by IgA & benzpyrene hydroxylase
  - Efficient apoptotic mechanisms
Pathogenesis

- Adenocarcinomas
  - From pre-existing adenomas
    - Tubular, villous, and tubulovillous
      - Villous usually large
      - 2nd portion of duodenum
      - 45% present malignant degeneration at Dx
    - FAP = 100% cumulative lifetime risk
      - Duodenal cancer: leading cause of death in pts s/p colectomy
  - Peutz-Jeghers: hamartomatous polyps
    - Adenomatous foci undergo malignant transformation
  - Crohn’s: distal ileum
Pathogenesis

- Carcinoid tumors
  - Enterochromaffin cells
    - GI tract and mainstem bronchi
  - Secretion of biologically active substances
    - Serotonin
    - Amine, tachykinins, peptides, prostaglandins

Most common endocrine tumor of GI tract
Appendix > Jejunoileum > Rectum
Clinical presentation

- Mostly asymptomatic
- Bowel obstruction most common
  - Lumen narrowing
  - Intussusception
- Hemorrhage
- Obstructive jaundice (periampullary)
- Cachexia
- Ascites
Carcinoid syndrome

- Small intestinal carcinoids
  - More aggressive than appendiceal
    - Larger than 1 cm = higher metastatic potential
  - 25 - 50% of related metastatic liver tumors
    - Diarrhea
    - Flushing
    - Hypotension
    - Tachycardia
    - Endocardial and right heart valves fibrosis
Soft tissue tumors

- Lymphomas
  - Disseminated systemic disease
  - Primary small intestinal most common in ileum
    - Highest concentration of lymphoid tissue
- GIST
  - Small intestine: 2nd most common (gastric)
    - 25 - 30%
    - Propensity for overt hemorrhage
- Sarcomas
  - Pain with abdominal mass
  - Bleeding
  - Extramural extension (no luminal obstruction)
Diagnosis

- Rarely diagnosed pre-op
- Non-specific labs
  - 5-HIAA (carcinoid)
  - CEA (in the presence of liver mets)
- Contrast Radiography
  - Enteroclysis: 90% sensitive
  - UGI series w/SBFT: 30-40% sensitive
- CT scan: useful for staging
- Angiography or Bleeding scan if hemorrhagic
Diagnosis

- Endoscopy
  - EGD w/biopsy for duodenal tumors
  - EUS to determine degree of invasion into wall
- Capsule endoscopy
  - Less invasive
  - Unable to obtain tissue diagnosis
  - Capsule can cause obstruction
- Double balloon or “push” enteroscopy
  - Entire length of small bowel
  - Previous surgery with long afferent limbs
Therapy - Duodenal tumors

- Incidental duodenal tumors
  - Endoscopic biopsy
- Polyps or adenomas:
  - Less than 1cm: endoscopic removal
    - Surveillance endoscopy
  - More than 1 cm: surgical resection
    - Transduodenal
    - Segmental resection
- Periampullary tumors
  - Pancreaticoduodenectomy
Therapy - FAP related

- Duodenal adenomas
  - Screening EGD in 2nd or 3rd decade of life
  - Endoscopic removal
    - Surveillance EGD in 6 months and yearly after
  - Pancreaticoduodenectomy
    - Multiple and sessile
    - Periampullary
    - Localized resections = high recurrence rates
    - Classic Whipple
Therapy - Jejuno/Ileal tumors

- Wide local resection
  - Mesenteric excision
  - Regional lymphadenectomy
- Locally advanced or metastatic dz
  - Palliative resection
  - Bypass
Therapy - Carcinoid

- Localized tumors
  - Segmental resection
  - Regional lymphadenectomy
    - Nodal mets common in > 3cm tumors (75-90%)
- 30% cases = multiple carcinoids present
  - Pre-operative small and large intestine evaluation recommended
- Metastatic dz: Tumor debulking
  - Improves survival
- Chemotherapy: doxorubicin, 5FU, streptozocin
Therapy - Lymphoma

- Segmental resection w/ mesentery
- If diffuse...chemotherapy
Therapy - GIST

- Segmental resection
  - If known diagnosis...can avoid lymphadenectomy
- Imatinib (Gleevec)
  - 80% beneficial on pts w/ unresectable or metastatic dz
  - 50-60% w/ reduction in tumor volume
  - Neo-adjuvant therapy also beneficial
Outcome

- Complete surgical resection
  - Duodenal tumors
    - 5 year survival rate: 50-60%
  - Jejuno-ileum tumors
    - 5-30%
  - Carcinoids
    - 75-95%
    - If liver mets: 20-54%
Outcome

- Lymphomas
  - Localized: 60%
  - Overall: 20-40%

- GIST
  - 35-60%
  - Smaller tumor size and low mitotic index = better prognosis
Capsule Endoscopy

- Multicenter European Study
  - 5129 pts underwent VCE
  - 124 (2.4%) had small bowel tumors
    - 112 primary
    - 12 metastatic
- Indications
  - Obscure GI bleed
  - Abd pain
  - Suspected neoplasm
  - Diarrhea w/ malabsorption
- 54 pts had 2 or more negative prior imaging

Capsule Endoscopy

- Single lesions: 89.5%
- Multiple: 10.5%
- GIST: 32%
- Adenocarcinoma: 20%
- Carcinoid: 15%
- Melanoma: 66% of metastatic tumors
- Surgical treatment: 95% cases