Case Presentations

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November 6, 2006
Case 1 - HPI

- 9 year old boy
- Pedi Surgery Office 10/2005
- HPI
  - Chronic intermittent rectal bleeding since infancy
  - Painless, coating and after BM’s
  - Varied from daily to q 3-4 months
  - No transfusions, but recent anemia req. iron
Case 1 - HPI

- PMH – multiple studies
- Meds – iron
- All – amoxicillin
- PE
  - Unremarkable
  - Abdomen benign
  - Anus without hemorrhoids or fissures
PMH - workup

- 1/99 – colonoscopy - proctitis
- 8/99 – colonoscopy - same
- 1/02 – colonoscopy - tortuous vascular pattern 14-20cm, rectal ulcers
  - US Abdomen – normal, incl. portal flow
- 12/02 – MRA – normal vasculature
PMH - workup

- 8/05 – CT abd – thickened SB
PMH - workup

- 8/05 – CT abd – inflamed rectum
PMH - workup

- **9/05**
- MRV abdomen
- Normal
- Nonvis. IMV
PMH - workup

- Other Tests
  - Meckels Scan – negative
  - Stool cx, h.pylori - negative
  - Dietary changes – unhelpful

- 11/05 – angiogram
  - Dilated rectal plexus of veins and superior rectal vein

- 6/06 – bleeding continues, surgery planned
Operation – 6/30/06

- Exploratory laparotomy
- Extensive venous malformation from sigmoid to distal rectum/anal verge
- Normal appearing iliac veins
Operation – 6/30/06
Operation – 6/30/06

- Rectosigmoid resection down to reflection
- End sigmoid colostomy
Pathology

- Colonic wall with numerous, irregular, dilated vessels in the submucosa and serosa.
- Thin walls most consistent with venous structures.
Postop

- **Course**
  - Bowel function and discharge POD 3
  - Minimal bleeding by POD 14

- **4 weeks postop – Proctoscopy**
  - No varices seen

- **8 weeks postop – Takedown**
  - Endorectal pullthrough with coloanal anastomosis
    - Distal mucosectomy, proximal proctectomy
Follow-up

- 3 weeks postop – Office visit
  - No problems, incontinence or major bleeding
- 4 weeks postop – ER visit for constipation
Rectosigmoid Vascular Malformation

- **Differential diagnosis / associated conditions**
  - **Klippel-Trenaunay Syndrome**
    - Skin vascular malformation, hypertrophy, venous varicosities
  - **Blue Rubber Bleb Nevus Syndrome**
    - Skin, mucus membrane, GI lesions
  - **Diffuse Infantile Hemangiomatosis**
    - Disseminated hemangiomas of skin and internal organs
  - **Osler-Weber-Rendu Disease**
    - Hereditary hemorrhagic telangiectasia
  - **Maffucci’s Syndrome**
    - Long bone and visceral vascular malformations
Rectosigmoid Vascular Malformation

- Fishman et al. JPS 2000
- 3 pts with colorectal VM
- Resection, pullthrough
- Bleeding stopped in all

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Endorectal Pull-Through Abates Gastrointestinal Hemorrhage From Colorectal Venous Malformations

By Steven J. Fishman, Robert C. Shamberger, Victor L. Fox, and Patricia E. Burrows
Boston, Massachusetts

Background/Purpose: Lower intestinal venous malformations are rare anomalies resulting from errors in vascular morphogenesis. These lesions may cause significant chronic and acute gastrointestinal hemorrhage. Venous malformations are unresponsive to angiogenesis inhibitors. Although these anomalies generally are incompletely resectable because of diffuse pelvic and mesenteric involvement, the authors sought to abate bleeding by excluding the lesion from the gastrointestinal lumen.

Methods: Three patients with circumferential transmural venous malformations of the colorectum, pelvis, and mesentery were identified. Imaging findings were similar among the patients and included circumferential septated bright signal on T2-weighted magnetic resonance imaging (MRI) contrast enhancement, and multiple phleboliths, seen best on computed tomography (CT). The lesion extended from the anus to the splenic flexure in 2 patients and throughout the entire colon in the other. Each had daily hematochezia for many years and required transfusions and chronic iron therapy. Although bleeding began in childhood in each patient, no therapy was successful until ages 7, 24, and 45.

Results: Colectomy, anorectal mucosectomy (through the pelvic venous malformation), and endorectal pull-through and anastomosis was performed (colorectal and ileocolonic in 1).

Conclusion: Colectomy with mucosectomy and endorectal pull-through should be considered for diffuse venous malformations of the colorectum before the development of large transfusion requirements.

INDEX WORDS: Venous malformation, vascular malformation, colorectal, gastrointestinal hemorrhage, pull-through.
Questions?
Case 2 - HPI

- 11 year old boy
  - 2 months of “bubble” sensation in stomach
  - 3 weeks of progressive crampy abd pain
  - Intermittent nonbilious emesis x 10 days
  - Occasional headache and back pain

- PMH noncontributory

- PE normal except P 110, BP 137/85
Labs

- UA normal
- CBC, electrolytes, lipase normal
- LFT’s normal
- Admitted from 9/27 to 10/1/06
CT
Other studies

- Bone scan
  - normal
- CT neck
  - normal
Differential Dx

- Pheochromocytoma
- Neuroganglioma
- Lymphoma
- Neuroblastoma
### Further Labwork

<table>
<thead>
<tr>
<th>Test</th>
<th>Result</th>
<th>Normal Range</th>
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<tbody>
<tr>
<td>Serum normetanephrine</td>
<td>17.8</td>
<td>&lt;0.9</td>
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<tr>
<td>Serum epinephrine</td>
<td>46</td>
<td>20-115</td>
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<tr>
<td>Serum dopamine</td>
<td>105</td>
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<td>Serum norepinephrine</td>
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<td>Urine VMA</td>
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<tr>
<td>Urine norepinephrine</td>
<td>1256</td>
<td>15-80</td>
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</table>
Outpatient

- Sent home on phenoxybenzamine
- Propranolol added
- 10/19 - Surgery Office
  - Plan for surgery
- 2 ED visits for chest pain, abd pain
- 10/26 – case cancelled due to tachycardia to 110
Pheochromocytoma

- Mayo - retrospective review
- 12 pheo, 18 paraganglioma
- Sx: HTN(64%), palpitations(53%), HA(47%)
- Malignancy(47%) risk factors:
  - Sporadic tumors, paraganglioma, size > 6 cm

Pheochromocytoma and Paraganglioma in Children: A Review of Medical and Surgical Management at a Tertiary Care Center

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The authors have indicated they have no financial relationships relevant to this article to disclose.
Pheochromocytoma

- Vanderbilt - retrospective review
- 11 children with 14 pheochromocytomas
- Avg age 15 years
- HTN seen in 82%
- Familial 2 (18%)
- Bilateral 2 (18%)
- Extra-adrenal 6 (55%)
- Malignant 3 (27%)

Questions?