Case #42

NAME Educational Activities Committee
Case provided by:

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1. A 12-year-old boy with no previous medical history complained of abdominal pain, intractable nausea, and vomiting and was sent by urgent care to the hospital with concerns for dehydration. Imaging of the abdomen was remarkable for ascites, mesenteric lymphadenopathy, thickening of the cecal and ileal walls in the setting of diffuse thickening of the small and large bowel, and a poorly visualized appendix. He remained afebrile, and stool cultures were unable to be obtained as he had no bowel movement while admitted. On the third day of admission, he developed altered mental status, shortness of breath, and tachycardia followed by seizure activity and cardiopulmonary arrest.

The photos from the autopsy most likely represent:

- ileocecal intussusception
- Lymphoma
- Trauma with intestinal perforation
- Volvulus
- Hamartomatous polyp
Answer...
A. Ileocecal intussusception (CORRECT RESPONSE, 63.62% responses)

The photograph depicts the ileum telescoping within the cecum at the ileocecal junction. Intussusception occurs when peristalsis propels one segment of bowel into the immediately distal segment. Most cases are seen during the first 5 years of life, with over half occurring in the first year. In older children and in adults, intussusception is frequently the result of a pedunculated intraluminal tumor, such as lipoma, GIST, inflammatory polyp or malignant lymphoma. More than 90% of cases of childhood intussusception begin at the ileocecal valve. Appendiceal intussusception is particularly rare with an incidence of 0.01% according to a pathologic review of 71,000 human appendix specimens. Progressive compression of the mesentery and blood supply of the invaginated bowel causes edema, hemorrhage, and eventually ischemic necrosis. In this case, the acutely inflamed appendix (note the hyperemic and purulent appearance of the associated serosa and mesentery) was within the telescoping segment of ileum. The tip of the appendix is barely observed in the image.
Other responses:

B. Lymphoma (11.75% responses)
Malignant lymphoma may serve as a lead point in cases of intussusception in adults and older children. Burkitt lymphoma is the most common GI lymphoma of childhood and can arise in the submucosal lymphoid tissue of the ileocecal region and extend transmurally to involved local mesenteric lymph nodes and form a bulky tumor mass. The erythematous luminal mass in the photograph represents the edematous segment of ileum telescoping within the cecum, rather than a malignant mass.

C. Trauma with intestinal perforation (5.76% responses)
Small bowel perforation due to blunt abdominal trauma is relatively uncommon and is characterized by a delayed onset of symptoms following traumatic injury such as a motor vehicle collision. Findings include mucosal necrosis, granulation tissue, transmural edema, and ischemic changes.

D. Volvulus (8.02% responses)
Volvulus can lead to sudden death and is characterized by the complete twisting of a loop of bowel about its mesentery, and generally occurs in redundant loops of sigmoid colon or cecum but may also involve transverse colon, small bowel, or stomach. It is commonly seen in infancy associated with malrotation and may result in occlusion of the blood supply and infarction. In this short segment of bowel, there is no evidence of twisting along the mesentery. See attached EAC case #9

E. Hamartomatous (juvenile) polyp (10.85% responses)
Grossly, juvenile polyp is classified as a hamartomatous polyp and has an irregular, granular, red surface, and is often pedunculated. Juvenile polyposis is an autosomal dominant syndrome characterized by the development of multiple hamartomatous GI polyps (between 5 and 100 polyps). Juvenile polyps are also associated with Peutz-Jeghers polyposis syndrome.
References: