Case of the Month - September 2013

15 year old female complaining of chronic post-prandial abdominal pain and bloating with intermittent vomiting.

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Ultrasound and Upper GI Study

Abdominal ultrasound followed by Upper GI contrast study were performed. Examine the following images:

• What are the imaging findings on ultrasound?
• What are the imaging findings on the Upper GI study?
• What is the differential diagnosis? Diagnosis?
Imaging Findings

• Abdominal ultrasound:
  – abnormal positional relationship between SMA and SMV

• Upper GI contrast study:
  – Abnormal morphology of the duodenum (lack of “C” shape, and 2\textsuperscript{nd} and 3\textsuperscript{rd} part not within retroperitoneum)
  – Small bowel isolated within the right hemi-abdomen
  – Colonic gas isolated to left hemi-abdomen
Differential Diagnosis

- Intestinal volvulus
- Small bowel obstruction
- Intestinal malrotation
- Duodenal atresia
Diagnosis

Intestinal malrotation (subtype: nonrotation)
Discussion

• Malrotation is a congenital anomaly due to arrest of normal gut rotation during embryogenesis.
• Normal rotation of bowel occurs 270° counter clockwise around the SMA axis. The duodeno-jejunal flexure then becomes left of midline at L1. Terminal ileum results in right iliac fossa.
• Depending on stage of arrest, nonrotation, incomplete rotation, and incomplete fixation (subtypes of malrotation) can occur.
• Nonrotation results in small bowel in right hemi-abdomen, large bowel on the left.
Incidence estimated at 1:500 (not all symptomatic) with M:F = 2:1 in age < 1 year.

Up to 80% present within the first month of life

Clinical presentation varies with age:
- Infant: most commonly midgut volvulus
- Older child or adult: colicky abdominal pain likely related to resolving duodenal obstruction and/or internal hernias
Discussion

• Associated with duodenal atresia, diaphragmatic hernia, gastroschisis, omphalocele, asplenia/polysplenia, situs ambiguous, and choanal atresia.

• Predisposition to midgut volvulus due to malfixation of mesentery which results in narrowed mesenteric attachment.

• Increased risk of internal hernias due to abnormal fibrous peritoneal bands called “Ladd bands”.

• Treatment for acute presentation is surgical
  – Ladd Procedure: Reduction of volvulus, division of mesenteric Ladd bands, placement of small bowel on right and large bowel on left and appendectomy
Discussion

Normal rotation and fixation of the midgut

Malpositioned bowel with narrow base of mesenteric fixation. Ladd bands indicated by blue lines.

Images from Pickhardt and Bhalla. AJR 2002;179: 1429-1435.
Discussion

• Radiologic features:
  – Plain film:
    • “Double bubble sign” – distended stomach and duodenum with little gas in the remainder of the small bowel.
    • Right sided jejunal markings or absence of gas in right-sided cecum
  – Ultrasound:
    • Inversion of SMA/SMV relationship (SMV to the left of SMA)
  – CT:
    • Corkscrew appearance of bowel/mesentery twisted around SMA axis.
    • Small bowel on right, large bowel on left
    • SMV ventral or to the left of SMA.
  – Upper GI contrast study (diagnostic):
    • Abnormal DJ location as it fails to cross midline to left side
    • 2\textsuperscript{nd} and 3\textsuperscript{rd} part of duodenum not positioned in retroperitoneum
References