Intestinal Malrotation in Adolescent: A case report

Introduction:
Congenital malrotation of the mid-gut often presents within the first month of life. The overall incidence of malrotation, however, is unknown because some patients will present years later or remain asymptomatic for life. Because presentation is nonspecific and the index of suspicion for malrotation progressively decreases in the older population, the clinical diagnosis is usually not considered in the initial evaluation. We hereby sought to present a case of a 14 year old girl came with history of right upper abdominal pain. No history of fever or vomiting was noted. Ultrasound performed was inconclusive. Patient was referred for CT scan of the abdomen.

CT Imaging findings revealed:
1. Alteration in the relation between SMA and SMV, with the vein being on left side of the artery.

2. Appendix was seen in the infraumbilical area in midline and was normal in calibre

3. The small bowel loops were seen on right side of the abdominal cavity. Large bowel loops visualized in mid and left side of abdomen.

4. The third part of duodenum did not cross the midline and was seen on right side of abdomen.

5. The uncinate process of the pancreas was underdeveloped.

Discussion: Malrotation results not only in the malposition of the bowel but also in the malfixation of the mesentery. The normally broad mesenteric attachment is shortened to a narrow pedicle that predisposes the patient to the complication of midgut volvulus. Internal hernia related to abnormal peritoneal fibrous bands of Ladd that attach to the right colon is another complication of malrotation seen in adults.
Imaging studies:
1. Upper gastrointestinal barium series: shows that the duodenal–jejunal junction fails to cross the midline and lies below the level of the duodenal bulb. An abnormal junction in an adult should not be dismissed as a normal variant. Contrast enema examination usually shows malposition of the right colon, but the caecum may assume a normal location in up to 20% of patients. This normal location may cause malrotation to be missed on this type of study.

2. CT scan: CT not only shows the intestinal malpositioning seen on barium studies but also depicts associated extra-intestinal findings like deviation from the normal relationship between the superior mesenteric artery (SMA) and superior mesenteric vein (SMV). Normally the artery is towards the left side of the vein. In most patients with quiescent malrotation, the SMA and SMV will assume a vertical relationship or show left–right inversion. Abnormalities of SMA–SMV orientation are not entirely diagnostic, however, because some patients with malrotation will have a normal relationship, and a vertical or inverted relationship can also be seen in patients without malrotation. Therefore, isolated detection of such an abnormality should warrant closer examination of the bowel to rule out malrotation.

Examination of pancreas in malrotation will reveal underdevelopment or absence of the uncinate process. In patients with abdominal abnormalities unrelated to coexisting malrotation, the altered anatomy may result in an atypical clinical presentation. A prime example is appendicitis, which may present with symptoms that are more left-sided.

Complications: Midgut volvulus is a complication of malrotation in which clockwise twisting of the bowel around the SMA axis occurs because of the narrowed mesenteric attachment. This life-threatening condition is a clear indication for emergent surgery. The clinical diagnosis of midgut volvulus in adolescents and adults is difficult because the presentation is usually nonspecific and malrotation is rarely considered. Recurrent episodes of colicky abdominal pain with vomiting over a period of months or years are typical and may eventually lead to imaging. Upper gastrointestinal examination shows the typical cork screw appearance of the proximal small bowel. However, in older patients with acute symptoms, CT is generally performed instead of a barium examination.

The CT whirl or whirlpool sign describes the swirling appearance of bowel and mesentery twisted around the SMA axis. A similar appearance can be seen on sonography. Additional CT findings include duodenal obstruction, congestion of the mesenteric vasculature, and evidence of underlying malrotation. The presence of intestinal ischemia or necrosis is an ominous sign.

Conclusion: The clinical diagnosis of malrotation after childhood is usually not considered; this oversight underscores the importance of recognizing this unsuspected condition.

Regards,

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N.B: This case is authentic and from the archives of Radiance Diagnostics. For any queries/suggestions/feedback write to us at radiance@radiancediagnostics.in. Case of the month can also be accessed anytime online at VIEW BOX at www.radiancediagnostics.in.