Nonrotation of the Intestines in a 14-Year-Old Male

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ABSTRACT
We describe a 14-year-old male with nonrotation of the intestines, proteinuria, and marfanoid habitus. Rotation anomalies occur in about 1 in 500 live births and have been described in 0.5% of autopsies. Affected individuals can be asymptomatic at any age, but many remain asymptomatic throughout life. The diagnosis is usually incidental to laparotomy or when abdominal imaging is done for a different pathology. Patients can present with chronic recurrent vague abdominal pain, back pain or vomiting. It can also present acutely with intestinal obstruction and volvulus. Clinically significant complications of rotational anomalies are potential volvulus, intestinal obstruction and atypical clinical presentation of appendicitis (e.g. lower back pain). Management of nonrotation of the intestines without volvulus is controversial. Asymptomatic patients have been observed or undergone elective surgical repair. Symptomatic patients warrant laparoscopy, Ladd’s procedure and appendectomy.

CASE PRESENTATION
A 14-year-old male with past medical history of Attention Deficit Hyperactivity Disorder was admitted to the general pediatric service due to acute gastroenteritis and dehydration. He complained of midabdominal pain and lower back pain. He also reported tactile fever, nausea, vomiting, diarrhea, anorexia, and tingling sensation in his four limbs. On physical examination, his vital signs were within normal limits, he weighed 50 kilograms (42nd percentile) and his height was 176 centimeters (91st percentile). He had marfanoid habitus and grade 2/6 systolic ejection murmur. The rest of the systemic examination was unremarkable.

His initial complete blood count and comprehensive metabolic panel were normal. His urine analysis and microscopy showed protein and trace bacteria, but his urine culture showed no bacterial growth. In view of his unexplained backache and proteinuria, he was tested for Anti-Nuclear Antibodies which was negative. Renal ultrasound showed normal size kidneys with no evidence of renal pathology. He was referred to nephrologist for follow up. In view of his marfanoid features he was tested for homocysteine and fibrillin1 gene mutation, both were negative. Heart echocardiogram was normal.

Because of his persistent abdominal pain, abdomen ultrasound was obtained, in which appendix could not be visualized. Therefore, CT scan of the abdomen and pelvis with contrast was done which showed the colon located on the left side of the abdomen and pelvis, and the majority of the small intestine was located on the right abdomen, the superior mesenteric artery and vein were interposed in position (figure 1), and the appendix was normal. These findings were consistent with congenital bowel rotational anomaly, likely nonrotation of the intestines. The diagnosis was confirmed by upper GI (figure 2) and small bowel follow-through barium studies.

Figure (1): CT abdomen with contrast showing the superior mesenteric vein is located to the left of the superior mesenteric artery.

Figure (2): Upper GI barium study showing the small intestine located in the right side of the abdomen.

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Pediatric surgery was consulted, and after discussion with the patient and his mother about the implications of nonrotation, the family elected to have Ladd's procedure performed. Exploratory laparoscopy, Ladd's procedure, and appendectomy, were done at day four of admission.

During laparoscopic examination, the small intestine was located on the right side of the abdomen, the cecum was located in the midline, and the entire large bowel was on the left side of the abdomen (like it was seen before on the CT scan of the Abdomen). There were some bands between the small intestine and the colon. The tip of the appendix was inflamed and injected. Pathology report confirmed appendicitis later on. The patient had uneventful postoperative course and was discharged home on day seven.

DISCUSSION

Nonrotation of the intestine results from midgut anomaly during embryologic development. The midgut receives blood supply from the superior mesenteric artery, and undergoes rotation around the same artery’s axis between the 4th and 10th weeks of gestation. In the first stage of rotation, the midgut undergoes a 90-degrees counter clockwise rotation from the sagittal to the horizontal plane. This causes the midgut to herniate out of the abdominal cavity through the umbilical ring. By the end of the 8th week, the midgut begins to return to the abdominal cavity. Failure of the midgut to return to the abdominal cavity results in omphalocele. Second stage of rotation begins at the end of 10th week of gestation, when the midgut rotates additional 180-degree counterclockwise bringing the duodenal “C” loop behind the superior mesenteric artery, thus completing complete 270-degree counterclockwise turn. Errors in midgut rotation in the second stage result in incomplete rotation, hyperrotation, reversed rotation and intestinal nonrotation. The third stage of midgut development is fixation and fusion of the mesentery. The cecum further descends into the right lower quadrant, and there is fixation of the ascending and descending colon to the posterior abdominal wall and a retroperitoneal fixation of the duodenum. The ligament of Treitz is now located in the left upper quadrant, and the small bowel mesentery fuses with the posterior retroperitoneum. Errors in this stage may lead to volvulus and hernias.

The incidence of intestinal rotation anomalies has been reported to be 1 in 6000 live births. Nonrotation of the intestine occurs in about 1 in 500 live births. True incidence remains unknown because many remain asymptomatic. Complete nonrotation have been described in 0.5% of autopsies.

Patients with nonrotation can become symptomatic at any age, including adulthood. Symptoms can arise from acute or chronic intestinal obstruction that may be caused by the presence of abnormal peritoneal bands (Ladd’s bands). 3 It can present as chronic recurrent abdominal pain, back pain and vomiting. Abdominal pain is often intermittent, colicky, and is relieved by the vomiting. Complications of intestinal nonrotation are related to short fixed root of the small bowel mesentery, which may result in volvulus and bowel ischemia. The abnormal anatomy may lead to misdiagnosis of diseases of the appendix, cecum, and colon. The feared complication of nonrotation is volvulus with intestinal necrosis, which presents with severe abdominal pain and bilious emesis. 3, 4

The diagnosis is often established by upper gastrointestinal barium studies. A diagnostic study will reveal the duodenal-jejunal junction to be located below the level of the duodenum bulb instead of horizontally crossing the midline to the left upper quadrant. Abdomen CT scan with contrast will show abnormal interposed positions of the superior mesenteric artery and vein. A superior mesenteric artery located to the left of the superior mesenteric vein usually suggests nonrotation of the intestines. Abdomen CT without contrast is falsely negative in 25% of cases.

Laparoscopic Ladd procedure can be performed safely in selected patients with no increase in complications. Short-term results are superior to open Ladd procedure and can be achieved without any increase in operative duration. 5

Ladd’s procedure consists of lysis of duodenal, cecocolic and small bowel adhesions. This increases the small bowel mesentery base. The cecum and ascending colon are placed in the left upper quadrant. Modifications include fixation of the cecum to the left lateral wall, fixing the small-bowel mesentery to the right peritoneal reflection and suturing the duodenum to the right kidney. There is no consensus regarding elective surgical intervention to decrease the risk of midgut volvulus in asymptomatic or minimally symptomatic patients. 6, 7

Because of the altered anatomy of the appendix it may be difficult to locate the appendix and hence appendicitis could be missed in the future and lead to bad outcome. Therefore, elective appendectomy can be a preventive measure and done at the same time of Ladd’s procedure.

REFERENCES


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