A case of reversed intestinal rotation presenting as duodenal ileus

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SUMMARY

An unusual case of intestinal obstruction associated with reversed intestinal rotation is presented. The ileus was developed due to a fibrous periduodenal band of congenital origin, binding the duodenum to the mesocolon and small bowel mesentery. The embryology, clinical presentation and treatment of this extremely rare developmental anomaly of intestinal rotation are discussed.

Key words: Duodenum ileus, intestinal embryology, reversed intestinal rotation.

INTRODUCTION

Although congenital anomalies of the intestinal rotation are relatively often seen in infancy and childhood, they are very uncommon in adults. Reversed intestinal rotation (RIR) is the rarest of the rotational abnormalities of the intestine, constituting approximately 2-4% of all reported cases of malrotation.^{1,2} RIR, as well as the other types of anomalous intestinal rotation, is not usually clinically evident throughout adult life and many cases are incidental autopsy findings or are discovered at a laparotomy for another abdominal pathologic condition.^{1,2} However, some patients may suffer chronic unexplained and atypical symptoms, due to the abnormal position of the viscera, while an undefined percentage of RIR cases will experience an acute episode of duodenal or intestinal obstruction, typically in infancy or early childhood.^{3,4}

We herein report on such a case of RIR, emphasizing on the surgical importance and the life-threatening potential of this extremely rare congenital deformity.

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CASE REPORT

The patient, a previously healthy 55-year-old man, had a seven-day history of midabdominal colicky pain, epigastric bloating and copious bilious vomiting. The symptoms became progressively more severe, his general condition worsened and the patient was transferred to our hospital in a mild lethargic condition.

On physical examination the principal finding was severe dehydration, while apart from tenderness in the epigastrium there were no abnormal findings from the abdomen. The heart rate was 110 per minute and arterial pressure was 100/50 mmHg. Routine laboratory investigations revealed a mild elevation of leukocyte rate (12.220/ μ L). Blood urea was estimated to be 402 mg/dL.

The abdominal plain films revealed a distended stomach as well as the presence of a few air-fluid levels in the right subco-stal area. The gastric distention was also confirmed by ultrasonography, which did not reveal any other abnormal finding. Barium meal showed duodenal bulb with a striking "double bubble sign" (Fig. 1), suggestive of duodenal obstruction. Endoscopic examination confirmed the presence of a grade III esophagitis, while the stomach was found to contain a considerable amount of bilious fluid.

In view of the clinical picture, X-ray, ultrasound and endoscopic findings the patient was taken to the operative room with a prospective diagnosis of small intestinal obstruction.

The peritoneal cavity was approached through an upper midline incision. The normal relationship of the duodenum and the transverse colon relative to the superior mesenteric artery was reversed, with the colon passing through the root of mesentery behind the superior mesenteric artery and the duodenum lying in front of it. The right colon was completely unattached to the posterior abdominal wall and the freely mobile cecum was



Figure 1. Barium meal showing a striking "double bubble sign", suggestive of duodenal obstruction.

found in the subhepatic region (Fig. 2). The duodenum, in addition to lying anteriorly to the superior mesenteric artery, was very distended, with a diameter of about 8 cm, because of a constricting dense fibrous band causing a near-total obstruction of the duodenojejunal junction (Fig. 3). The fibrous band was lysed. An ascending colopexy was then performed and the mesentery of the small intestine was fixed to the posterior abdominal wall. Finally a typical appendectomy was performed.

The patient recovered uneventfully from the operation and his general condition quickly improved. He left the hospital on the 7th postoperative day and he remains without gastrointestinal symptoms for 5 years.

DISCUSSION

At about the end of the 11th week of intrauterine life the midgut, which was present in the extraembryonic celom, finishes its return to the celomic cavity, completing a rotation of 270 degrees counterclockwise. During



Figure 2. Drawing demonstrating the findings at laparotomy.



Figure 3. The constricting dense fibrous band which caused a near-total obstruction of the duodenojejunal junction.

this stage, the prearterial segment of the midgut, that eventually becomes the duodenum, reduces into the abdomen first, passing posterior to the superior mesenteric artery. The postarterial portion follows, coming to lie anterior to the superior mesenteric artery. Thus, the entire process of intestinal rotation, begun at the 4th week, results in a duodenum lying behind the superior mesenteric artery with the transverse colon in front of it.^{1,3,5}

At this stage, four types of anomalies may occur, as a result of faulty rotation: nonrotation, malrotation, paraduodenal hernia and reversed rotation. Reversed rotation constitutes a very rare malformation, the rarest among this group.^{1,6} The midgut, instead of going through a counterclockwise movement of 270 degrees, rotates in a reversed direction through an arc of 90 degrees, resulting in a transverse colon that lies posterior to the superior mesenteric artery and a duodenum that lies anterior to this vessel.¹⁻⁶ Associated anomalies due to deficient peritoneal fixation are common in RIR patients, occurring usually in the cecum and the ascending colon.^{1,7}

Although it has been suggested that some form of intestinal obstruction will invariably develop in patients with RIR,⁷ the actual percentage in which this condition becomes clinical evident is unknown. Because of the extreme rarity of this entity and the lack of extended studies, the physical history of RIR is not well defined.¹ However, it has been stated that most patients will present early in life with volvulus of the ileocecal segment on its narrow mesenteric attachment, while in some of them colonic obstruction may occur at the retroarterial segment of the transverse colon.⁵⁻⁷ Very rarely the obstruction may be seen at the duodenojejunal junction, due to either herniation into the retroarterial "tunnel" or by periduodenal bands.^{3,7,8} Such peritoneal bands of fibrous tissue may form along the anomalous duodenal course, binding the duodenum to the mesocolon and small bowel mesentery, as observed in our patient. Their origin is congenital and they are exceedingly rare in adult life.⁸

A number of operations for the surgical correction of reversed intestinal rotation have been described, including bypass of the transverse colon, transection and displacement of the transverse colon anterior to the superior mesenteric vessels and right hemicolectomy with ileotransverse anastomosis.⁹⁻¹⁰ Our experience with the patient presented here makes us cautious of the usefulness of these complex operations, an opinion which also has been stated by other aut,hors.⁹

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