Case report

Adult Midgut Malrotation with Duodenal Stricture: Diagnostic Dilemma and Its Management - A Case Report

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Abstract:
Intestinal malrotation is a rare congenital disease due to abnormal intestinal rotation and fixation of the intestinal tract in the early embryonic state. Adult cases are very rare. Midgut malrotation typically presented during the first few months of life but may sometimes appear later in life, causing difficulties and mistakes in diagnosis. An unusual presentation of this condition has led to diagnostic dilemma. Diagnosis is delayed because respective symptoms were not adequately considered in adults. We present a case report of 35 years old Muslim male presented with history of gradually increasing vague abdominal pain at upper and mid abdomen for 5 months with history of vomiting after heavy meal. On examination we found that patient was anxious and ill looking, Body stature average but patient was malnourished and dehydrated. Mild tender epigastric region. No palpable lump. No organomegaly. Succussion splash absent. Endoscopy of upper GIT was normal and full colonoscopy found also normal. Barium follow through x-ray revealed stricture at 3rd part of Duodenum with malrotation of gut. Open Ladd procedure with gastrojejunostomy under G/A was performed.

Keywords: Malrotation, Duodenal stricture, Ladd procedures, Congenital Ladd's band.

Introduction:
Midgut malrotation is an anomaly of fetal intestinal rotation that usually presents in the first month of life. It is rare in adulthood.[1] There is either lack of or incomplete rotation of the fetal intestines around the axis of the superior mesenteric artery during fetal development.[2] Most patients present with bilious vomiting in the first month of life because of duodenal obstruction or a volvulus. The true incidence in adults is difficult to estimate because most patients remain asymptomatic and their conditions are, therefore, never diagnosed. Approximately 90% of patients with malrotation are diagnosed within the first year of life, of whom 80% are diagnosed within the first month of life.[3] The frequency of occurrence has been reported as 1/6000 of all live births, and most cases are present in the first month of life and 90% within the first year.[4,5] The number of cases in which symptoms appear in adults is small, comprising only 0.2 to 0.5% of overall cases.[5] Surgical therapy remains the mainstay of treatment regardless of age at presentation. The most commonly used approach is the Ladd procedure, which involves counterclockwise reduction of the volvulus if present, division of any Colo-duodenal bands (Ladd's band), widening of the mesenteric base to prevent repeated volvulus, and prophylactic appendicectomy.[6] Laparoscopic approach has become more common since the report by van der Zee and Bax.[7] Here we document the case of a midgut malrotation with duodenal stricture an adult male.

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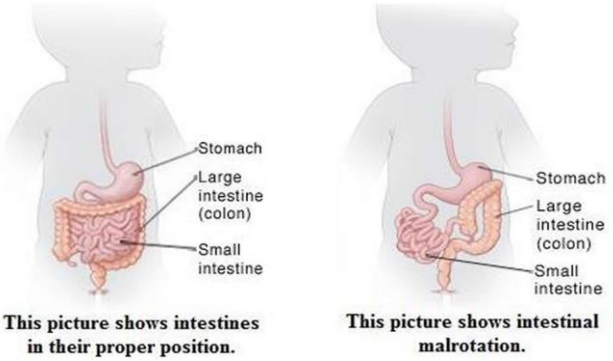

Figure-I: Normal position of intestine and malrotation of intestine.

Case report:

A thirty five years old Muslim male, born in Jhinaidah presented to the Outpatient Department with complaints of abdominal pain for 5 months involving upper and mid abdomen (Lt Lumber- Lt Hypochondrium- Epigastric- Rt Hypochondrium- Rt Lumber region), gradual onset, colicky in nature. Pain was aggravated by food intake and patient felt fullness after eating. Patient can take liquid diet without any problem. Patient do not have any history of heart burn and acidity. Pain was associated with vomiting, anorexia and constipation. Vomiting occurred after solid meal, induced, contained partially digested and undigested food particle, sour in taste, yellowish green in color, foul smelling. He occasionally uses laxatives for constipation relieve. Actually, no accurate measurement of weight loss but patient felt losing weight during last 5 months. According to his mother, he suffered abdominal pain (whole abdomen) with anorexia and constipation from the age of 2 years. His parents took him to local doctors, but there was no diagnosis. Pain was relieved by analgesics. He suffered similar kind of pain till 12 years of age. After that he was relatively healthy (occasional pain ignored as thought about PUD pain). There is no other significant past medical or surgical illness. He used to take antacid and some other medication, couldn't mention the name. He is a smoker taking 10-15 sticks per day in last 16 years. He is non-alcoholic and non-betel nut chewer. There is no known allergy to medications or food.

On examination, he was anxious and ill looking. Body stature average but malnourished. He was mildly anemic, mildly dehydrated. Shape of the abdomen was normal. Flank was not full. Umbilicus centrally placed, inverted. Hair distribution was normal. Skin condition was normal, no scar mark. Engorged vein was absent. Visible peristalsis and visible pulsation were absent. There was no obvious swelling and hernial orifices are intact. Mild tenderness present in epigastric region but other quadrants are nontender. No palpable lump. Liver, spleen not palpable. Kidney not ballotable. Succussion splash absent. There were no significant general examination findings except mild anaemia and dehydration.

Ultrasonography was done and showed normal findings. Endoscopy of upper GIT was normal. Full colonoscopy was also normal. Tumor marker CA-125 and CA-19.9 was within in normal range. Barium follow through x-ray revealed suggestive of stricture at 3rd part of duodenum with malrotation of gut [Figure-II]. Other investigations showed, RBS- 9.3 mmol/L, FBS- 5.37 mmol/L, Serum Creatinine- 1.21mg/dl, CXR P/A view- Normal, ECG- Normal, Echocardiography- Good LV function LVEF 63%., Blood group- B+ve, HBsAg- negative, Anti HCV- negative, Serum Albumin- 3.5 gm/dl, MT- negative, Rapid antigen test for covid 19- negative.

Optimization and preparation of patient for operation done by plenty of liquid diet for dehydration improvement, Correction of electrolyte imbalance by Inf. 5%DNS + Inj. KCl and Gastric lavage: Three days preparation with wide bore nasogastric tube, Irrigation with 200ml normal saline every 4 hourly until clear fluid comes out, In between the lavage, only plain water or liquid diet was allowed. Patient underwent laparotomy and open Ladd procedure. The Ladd procedure is the operation of choice for rotational anomalies of the intestine.

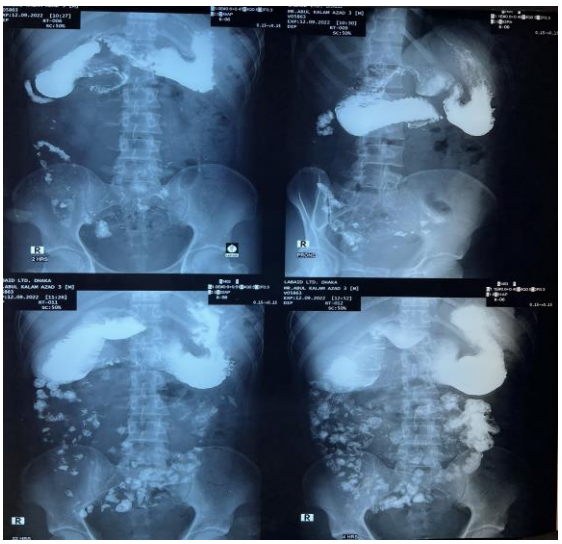

Figure-II: Barium follow through x-ray revealed suggestive of stricture at 3rd part of duodenum with malrotation of gut.

Conventional Ladd Procedure includes (Figure-III)

a. Twisted bowel was eviscerated.
b. The bowel is de-rotated in a counterclockwise fashion.

a. The peritoneal attachment between the cecum and retroperitoneum (Ladd band) was divided.
b. The base of the mesentery was widened, and an appendicectomy was performed.[8]

We had done midline laparotomy and malrotation of gut found. Caecum and ileum found into left side and descending colon is found in the right side of abdominal cavity. Ladd's band is found at caecum to duodenum, encircling duodenum adherent to liver and gall bladder. De-rotation of gut is done. Ladd's band was divided. Appendicectomy was done.

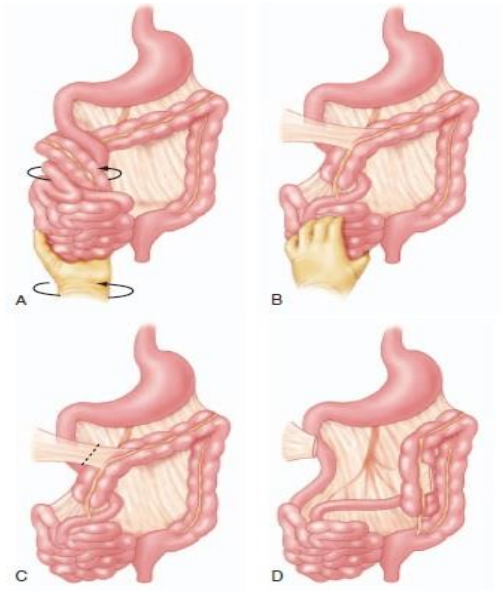

Figure-III: Ladd Procedure.

Gastrojejunostomy was done because of stricture at duodenum. 2 drain tube is given one in the hepatorenal pouch and another in the pelvis. After securing all the bleeding points, abdominal cavity is closed in layers. Patient was recovered in HDU. In the post operative period, patient developed electrolyte imbalance and superficial surgical site infection. Patient was discharged after treating electrolyte imbalance and surgical site infection. Patient was advised to avoid strenuous activity. After 14 days of secondary suture, stitches were removed as the wound was healthy

Figure-IV: Photograph showing abnormal location of appendix and congenital Ladd's band.

Discussion:

Rotational abnormalities of the intestine occur when the normal embryologic rotation and fixation of the mesentery fail to take place.[9] Intestinal malrotation, especially in adults, is rare.[5,10] So a single surgeon is likely to encounter only a small number of surgical cases or surgeons may never experience it. In adults, it may cause chronic, but indistinct symptoms that are often difficult to diagnose. Adult presentation of malrotation is a difficult diagnosis because of the low incidence of the disorder. Patients with intestinal malrotation who were not diagnosed until adulthood may present with a variety of chronic symptoms, including nausea, vomiting, diarrhea, vague abdominal pain, early satiety, bloating, dyspepsia, and peptic or duodenal ulcer disease. Unfortunately, many patients never receive surgical referral and are instead labeled with functional or psychiatric disorders.[11]There may be a significant number of patients with malrotation who were undetected in the neonatal period either because they were asymptomatic, or because their symptoms were mild and misinterpreted. As these patients grow into adolescence and adulthood, they may continue to have misinterpreted symptoms, remain asymptomatic, or present with new onset of acute or chronic symptoms later in life, as did our patient. Adults, however, present with vague symptoms such as vomiting (bilious or non-bilious), weight loss, and recurrent or colicky abdominal pai(often postprandial).[12-14] Intestinal obstruction, diarrhea, malabsorption, peritonitis, and septic shock also

have been reported in the adult group.[13] Timing and frequency of the pain also can be variable.[15] It is these vague symptoms along with the relative rarity of adult presentation of malrotation that often lead to diagnostic dilemma. Barium follow through x-ray still plays an important role in the diagnosis of duodenal disorders. For the best management of duodenal diseases, barium studies in combination with cross-sectional imaging modalities may offer detailed evaluation of the duodenum and its surrounding organs. However, CT, Ultrasound and MRI all can provide excellent cross-sectional anatomic orientation, which allows accurate pre-operative evaluation.[16] The treatment for intestinal malrotation is generally the Ladd procedure. Important considerations regarding Ladd procedure include the following: release of any midgut volvulus, resection of the abnormal adhesive retroperitoneal band (Ladd's band) between the duodenum and the right colon mesentery, opening the base of the mesentery including the SMA, performing prophylactic appendicectomy, and rearranging the intestinal tract.[17] We suggest that resection of the abnormal band relieves compression of the duodenum or jejunum. The opening of the base of the small intestinal mesentery is especially important to create sufficient space for the small intestine and the colon, removing one of the causes of malrotation. In our cases, the clinical symptoms were thought to have been caused by the duodenum being compressed by the abnormal retroperitoneal band and the right colonic mesentery. The band is connected to the adhesion between the pre pancreatic fascia and the colonic mesentery. This adhesion is considered to be more severe in adult cases than in infant cases because of the longer period of illness. Opening the base of the mesentery is also thought to be important. The narrow base of the mesentery could lead to the patient having subsequent midgut volvulus and obstruction with potential vascular catastrophe.[18] Opening the base of small intestinal mesentery is therefore crucial to reducing the likelihood of midgut volvulus.[19] As there was stricture at the 3rd part of duodenum, we did gastrojejunostomy in addition to Ladd procedure.

Conclusion:

Intestinal malrotation should be considered as a differential diagnosis of abdominal disorders in older children and adults presented with vague abdominal pain and vomiting. The diagnosis of intestinal malrotation associated with duodenal obstruction secondary to Ladd's bands should be considered in adult patients presenting with duodenal obstruction and malrotation of the small intestine with the cecum in the medial position. We believe that barium follow through x-ray is still the method of choice for the diagnosis of such malrotations in resource limited setting. Laparotomy and laparoscopy are alternative and feasible techniques with low rates of complications for the treatment of intestinal malrotation in adults.

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