



LAPAROSCOPIC LADD'S PROCEDURE FOR INTESTINAL MALROTATION IN ADULTS- A RARE PRESENTATION WITH REVIEW OF LITERATURE

General Surgery

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ABSTRACT

Malrotation of gut is a congenital developmental anomaly of foetal intestinal rotation and mostly present in early childhood as acute intestinal obstruction. Progression and presentation in adults is very rare and often silent. Delayed diagnosis is a common feature in many cases due to atypical presentation and leads to more complications. We present a case of 26 year old woman who presented with repeated episodes of pain abdomen and preoperative evaluation on contrast enhanced CT Scan showed intestinal malrotation. Laparoscopic Ladd's procedure was done successfully with division of Ladd's band, adhesiolysis, widening of mesentery and appendicectomy. The patient had an uneventful postoperative recovery. Here we discuss the rarity of intestinal malrotation in adults and various treatment options available in literature review.

KEYWORDS

Adult malrotation, Ladd band, CT scan, Laparoscopy

INTRODUCTION

Intestinal malrotation is caused by partial or complete failure of 270 degree counter clockwise rotation of midgut around superior mesenteric vessels in fetal life [1]. It is a rare congenital anomaly and presentation is usually after birth. Majority present in the first month of life (60-80%) with some sort of complication and by the age of one year it is around 90% [2]. Beyond this period presentation gradually becomes rare and mostly asymptomatic. Malrotation in adult is very rare, incidence is difficult to predict and is between 0.0001% to 0.19% [2,3]. Being asymptomatic in nature, adult malrotation is now most commonly made on routine imaging done for some other unrelated conditions. Chronic nonspecific intermittent pain abdomen with features of sub acute intestinal obstruction are other less common variety of presentation. Both the rarity of presentation in adulthood associated with mild atypical symptoms make this clinical entity a late difficult diagnosis.

Delayed diagnosis has its own pitfalls and it is difficult to predict which patients will have midgut volvulus or bowel ischemia [4]. The standard treatment is Ladd's procedure, which can be performed by either open laparotomy or laparoscopy according to the surgeon's preference. Here we present a very rare case of adult intestinal malrotation in a 26 year old woman presenting with chronic abdominal pain and was successfully managed with laparoscopic Ladd's procedure. Various aspects of adult intestinal malrotation with management options were discussed in detail with literature review.

Case Presentation

A 26 year old woman presented to our department with history of chronic intermittent abdominal pain for last 2 years. The pain was diffuse all over the abdomen, colicky in nature, mild to moderate in severity, on and off aggravated on taking food with nausea and was relieved on medication. On general appearance patient was thin built, emaciated and vital parameters were within normal limit. Abdomen was normal in shape and size with mild tenderness in the periumbilical region.

All the routine hematological parameters were within normal limit with hemoglobin at 10.2 gm% and normal white cell count. Both x ray abdomen and chest was unremarkable. But ultrasound abdomen suggested the altered position of superior mesenteric artery (SMA) and vein (SMV) with no evidence of bowel obstruction. Finally contrast enhance CT scan of abdomen confirmed that SMA and SMV relation is altered with SMA on the right side with clockwise rotation of SMV over SMA [Fig 1].

Laparoscopic Ladd's procedure was done based on the diagnosis of adult intestinal malrotation. On laparoscopy the small bowel loops

were lying in a peritoneal pouch on the right side obscuring the colon and fibrous band seen compressing the duodenum keeping DJ flexure on the right side. The procedure included division of Ladd's band [Fig.2] adhesiolysis with Lengthening of duodenum [Fig.3], widening of mesentery [Fig.4] and appendicectomy [Fig.5]. The postoperative period was uneventful and patient was discharged on 4th day. On follow up at 1 month, 6 month and 9 month, Patient was pain free and doing well.

DISCUSSION

The normal development of midgut occurs in three stage rotation process between the 4th and 12th week of embryonic period. With this normal process taking place, the duodenum is fixed retroperitoneally at the ligament of Treitz in the left and caecum on the right lower abdomen. Malrotation of the gut is due to complete or partial failure of 270° of counter clockwise rotation of the midgut around the superior mesenteric pedicle. It results in anomalous position of small bowel loops in the right side with absence of ligament of treitz and cecum, appendix lying on the left side. As a result small bowel mesentery develops a narrow vertical attachment putting it at the risk of volvulus, vascular compromise and gangrene [5]. A new peritoneal fibrous band known as Ladd's band also develops and fix the duodenum and cecum to the posterior wall, also can compress the duodenum causing duodenal obstruction.

Intestinal malrotation is a disease of newborn as it frequently manifest in the first month of life. Adult manifestation is very rare and difficult to predict because of its asymptomatic nature. But evidence from autopsy suggest that it may affect up to 1 in 6000 [2,3]. As opposed to the more common acute obstruction in neonates, clinical presentation in adults are more variable [2,6,7] and mostly asymptomatic. Sometimes it goes undetected throughout life or ignored because of self limiting mild symptoms. Wang and Welch [3] showed in their case series of 50 adolescents and adults patients, 24 were clinically asymptomatic. In the symptomatic group, chronic presentation is more common than acute and present in the postprandial period as intermittent vomiting or abdominal pain. Malabsorption and chronic weight loss are other common features as seen in our patient. Though uncommon 10 to 15 percent of adult patients present with acute midgut volvulus [8], the most common presentation in this subgroup. Other causes are internal herniation caused by Ladd's bands and appendicitis in abnormal position often making diagnostic dilemma. Now a day's most of the diagnosis of malrotations are made on routine imaging modality.

Contrast enhanced CT scan is the investigation of choice with a diagnostic accuracy of 80% [7,9]. SMV is normally located to the right of SMA, but in malrotation what is commonly seen is vertical

orientation or reversed relative left position of SMV to the SMA. This was first described by Nicholas and Li [10], pointing as a usual indicator towards the diagnosis of malrotation. A good ultrasound examination of abdomen often shows the similar finding raising suspicion as observed in our case. Failure of rotation also results in underdeveloped or absent uncinate process of pancreas. The shortened mesentery allows the small bowel and mesentery to twist and wrap around the narrowed SMA pedicle creating a distinctive 'whirlpool' appearance, observed by Fisher in a midgut volvulus [11]. The other associated findings are small bowel loops in the right upper abdomen, a lack of visualization of the caecum in the right iliac fossa, dilatation of duodenal loops and duodenojejunal flexure to the right (corkscrew sign). Upper gastrointestinal (UGI) contrast study showing deviation of normal course of duodenum is also very much suggestive and can be combined with barium enema to show abnormal position of caecum and right colon. Mesenteric angiography demonstrates the abnormal position & patency of mesenteric vasculature. Both UGI contrast series and angiography has limited role as contrast enhanced CT scan has the overall advantage of detecting both vascular and bowel abnormality in intestinal malrotation.

The management of intestinal malrotation is now well settled with surgical intervention as it is difficult to predict which patients will develop complications of malrotation [12]. First described by William Ladd in 1936[13], Ladd's procedure is the standard treatment for intestinal malrotation both in pediatric & adult patient. It includes division of Ladd's band, lengthening of duodenum, widening of mesentery, derotation of midgut volvulus if present and appendicectomy to prevent diagnostic dilemma due to abnormal position. In some cases variation of Ladd's procedure with only division of Ladd's band and appendicectomy can be done in the absence of volvulus. The successful outcome of the Ladd's procedure is well appreciated by complete resolution of symptoms in 9 of 11 patients by Fu et al [5] and 8 of 10 patients by Dietz et al [14].

With the advancement of Laparoscopy, Ladd's procedure can be safely and effectively performed and remains a first choice [15]. Open Ladd's procedure remains the gold standard in acute setting suspected of volvulus and bowel ischemia. Literature review indicates more laparoscopic procedure in pediatric age group compared to adults. Malrotation without volvulus can be successfully treated in laparoscopy by experienced surgeons [16].

CONCLUSIONS

Intestinal malrotation is an unusual disease entity and adult presentation is still very rare. Asymptomatic and nonspecific mild symptoms often pass silently throughout life, but presentation in adult life is potentially dangerous causing midgut volvulus, bowel ischemia and gangrene. High index of clinical suspicion is the key to diagnosis adult malrotation in cases of chronic obscure pain abdomen. Contrast CT scan is the investigation of choice and also rapid use of various imaging technique has helped to diagnose more number of cases. Once diagnosed, all patients should undergo surgery and the procedure of choice is Ladd's procedure. Ladd's procedure or with some modification usually relieves the symptoms and complications of intestinal malrotation as well as recurrence.

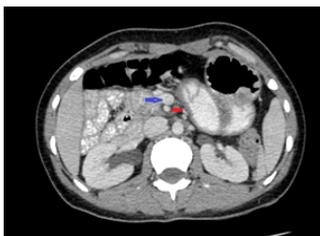


Fig.1- SMA on the right side with clockwise rotation of SMV over SMA



Fig.2- Division of Ladd's Band

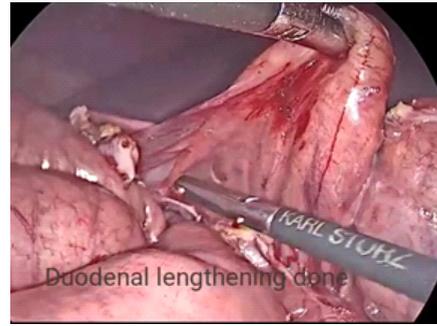


Fig.3 Duodenal lengthening

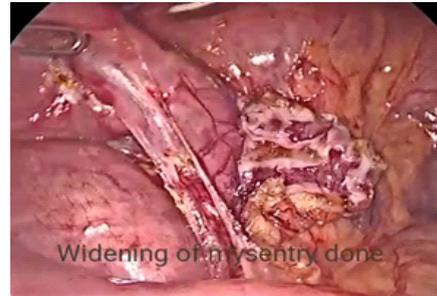


Fig.4 Widening of mesentery



Fig.5 Appendicectomy

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