CASE REPORT

Malrotation Masquerading as Duodenal Atresia: Case Report and Review of Literature.

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ABSTRACT

Intestinal malrotation is a relatively uncommon condition with diverse outcomes. Familiarity with variations in the presentation of malrotation is imperative as early diagnosis and prompt subsequent surgical intervention are essential to optimizing outcome. We report a rare case of a 34 weeks preterm neonate who presented with complaint of non-bilious vomiting since birth. Antenatal ultrasonography showed two dilated cystic structures with polyhydramnios, suggestive of duodenal atresia. X-ray abdomen standing after birth showed classical 'double-bubble' appearance. The contrast study was suggestive of dilated stomach and 1st part of duodenum with total absence of distal bowel gas which suggested possibility of duodenal atresia. Exploratory laparotomy was done which revealed malrotation of gut with Ladd's band with normal patency of distal bowel. Division of Ladd's band, derotation of gut, widening of base of mesentery, and appendectomy was done. Child recovered uneventfully and no recurrence of symptoms on subsequent follow-up for 3 months.

INTRODUCTION

Duodenal atresia is one of the most common sites of neonatal intestinal obstruction. The incidence of duodenal atresia has been estimated at 1 in 6000 to 1 in 10,000 live births[1]. Duodenal atresia is often discovered on antenatal sonogram. Maternal polyhydramnios and classic 'double-bubble' sign on fetal ultrasonography suggests the diagnosis. Duodenal atresia is due to embryo's developmental defect, in which the duodenum does not normally change from a solid to a tube-like structure.

We present here a rare case of actual neonatal malrotation, which masqueraded as duodenal atresia.

Neonatal intestinal malrotation is rare congenital condition caused by absence of or incomplete rotation of small bowel around the axis of the superior mesenteric artery during embryonal period[5]. The incidence in general population is one for every 200 to 500 newborns. Symptomatic cases are infrequent, occurring in one of 6000 newborns.

During normal abdominal development, three divisions of GI tract (i.e. foregut, midgut, hindgut) herniate out from the abdominal cavity, where they then undergo a 270◦ counterclockwise rotation around the superior mesenteric vessels. Following this rotation, bowels return to abdominal cavity, with fixation of duodenojejunal loop to the left of the midline and the cecum in right lower quadrant[6].

Interruption of typical intestinal rotation and fixation during fetal development can occur at a wide range of locations; this leads to various acute and chronic presentations of disease. The most common type found in pediatric patients is incomplete rotation predisposing to midgut volvulus, requiring emergent operative intervention.

CASE REPORT

A 24 hours old preterm (34 weeks), 1.9 kg weighing male child was referred to surgery from neonatal intensive care unit. It was prenatally diagnosed as a case of duodenal obstruction on ultrasonographic findings of polyhydraminos with 'double-bubble' appearance. On examination, baby was alert, active, and had no obvious external morphological congenital anomalies. There was upper abdominal fullness and nasogastric tube drained non-bilious aspirate. External genitalia and anal opening were normal.

Further evaluation by X-ray abdomen showed the classic 'double-bubble' appearance of duodenal atresia with total absence of distal bowel gas [Figure 1].

Contrast study was suggestive of dilated gas filled stomach and first part of duodenum with twisting of mesentery along with small bowel loops and mesenteric vessels which represented volvulus. No evidence of gas was seen in the distal bowel loop which suggested possibility of associated duodenal atresia. Another possibility was non-rotation or malrotation of gut with midgut volvulus.

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Figure 1: X-ray abdomen showing classical 'double bubble' appearance of duodenal atresia.

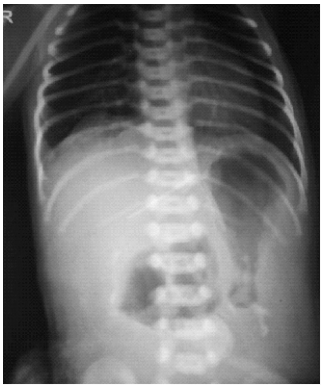
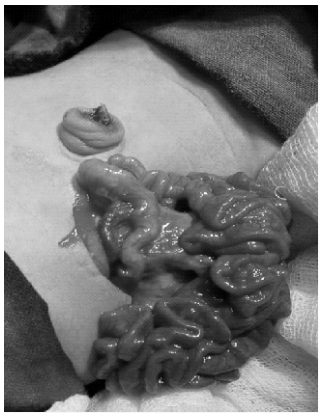


Figure 2: Intra operative picture of bowel loops.



Echocardiography didn't reveal any congenital heart anomaly. Ophthalmogical examination was normal and stromme syndrome was ruled out. Based on this information, child was taken-up for surgery.

On exploratory laparotomy stomach and 1st part of duodenum were dilated. There was malrotation of gut. There was a 'Ladd's band' obstructing duodenum. Rest of bowel was normal [Figure 2].

So division of Ladd's band, derotation of gut, widening of base of mesentery and appendectomy was done.

Postoperatively child was kept on ventilatory support for a day.

Gradually increasing nasogastric feed was started from postoperative day 3. The child was on full breast feed by postoperative day 5 and discharged from hospital.

DISCUSSION

The absence of a complete rotation of the midgut, during the embryonal period, is the key to the physiopathology of intestinal malrotation.

The duodenum does not assume its normal position, posterior to the superior mesenteric artery. Consequently, there is no fixation of the mesentery in posterior abdominal wall. This causes intestinal torsion through the superior mesenteric artery, one of the most common complications of rotation abnormalities[2].

Clinical presentation of intestinal malrotation can be unspecific. Most of intestinal malrotation patients present signs of obstruction during neonatal period and this condition should be considered in all newborns with bilious vomiting and abdominal pain[1].

The diagnosis of intestinal malrotation can be confirmed with upper GI tract contrast imaging[3][4]. That can reveal a vertical duodenum, with a right location in the abdominal cavity, and the absence of the duodenojejunal angle. These results are found in nearly 80% of the patients[4].

A double-contrast barium enema can show abnormal cecal location, just below liver, near midline, and entire colon located laterally to spine on left side[3].

CT scan can also identify these abnormal positions of small bowel and colon and opposite positioning of superior mesenteric vein, located on left side of the artery. It may also be helpful in identifying acute obstruction[3].

The Ladd procedure, initially described in 1936, is the classic surgical treatment for intestinal malrotation[2]. It is described as an association of the mobilization of the duodenum and the right colon, section of the Ladd's bands, section of possible adhesions near the superior mesenteric vessels and appendectomy.

The aim of this procedure is to reduce the risk of acute volvulus, by positioning small intestine in a non-rotating position and widening the base of the mesentery.

Appendectomy is performed due to possible difficulty in the diagnosis of future appendicitis, distant from the classic lower right quadrant position.

OUTCOME AND FOLLOW UP

The patient was discharged after it recovered uneventfully. There was no recurrence of symptoms on subsequent follow-up for 3 months.

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