Case Report

Congenital Absence of Jejunum and Ileum: A Case Report and Literature Review

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Abstract

We report an extremely rare finding 'congenital absence of jejunum and ileum' during explorative laparotomy of a 16-day-old female neonate. The dilated duodenum was terminating blindly, and the next segment of intestine was a peanut-sized cecum followed by microcolon. On an extensive survey of literature this type of intestinal atresia is not reported in living babies.

Keywords: Absence of jejunum and ileum, duodenocolic anastomosis, intestinal atresia

Introduction

Jejunoileal atresias are one of the common causes of intestinal obstruction in neonates. It is believed to be due to intrauterine vascular insult.[1] Current classification describes four types of intestinal atresia,[2] and they are also commonly encountered in clinical practice. We faced a rare case of intestinal atresia, where there was total absence of jejunum and ileum. This prompted us to carry out an extensive literature search, and this type of intestinal atresia is no where mentioned in living human babies. We operated the patient successfully, and the patient was discharged on breast feeds.

Case Report

A 16-day-old female baby presented to us with bilious vomiting and non-passage of meconium. Antenatal ultrasonography was not performed and there was no history of maternal drug intake during pregnancy. Baby was born by normal vaginal delivery at home and was 1.3 kg, sick and dehydrated at admission. Respiratory rate was 38/min, pulse 150/min and blood pressure was 80/60 mmHg. There was upper abdominal distention, but abdomen was soft without any rigidity. Anal opening was normally placed. Although haemogram was normal, serum urea and creatinine were moderately raised.

Erect abdominal X-ray revealed two gas-filled bowel loops in upper abdomen and paucity of gas in pelvis [Figure 1]. Clinical picture and plain X-ray abdomen suggested the diagnosis of proximal intestinal atresia. The baby was resuscitated with intravenous fluids, antibiotics and nasogastric (NG) tube was passed into the stomach. Laparotomy done by right supraumbilical transverse incision revealed grossly dilated stomach and duodenum. Duodenum was about 8 cm long and terminating blindly. There was total absence of jejunum and ileum. Cecum was about the size of a peanut and appendix very small. The microcolon was lying over the dilated duodenum [Figure 2] and followed till rectum.

Cecum and appendix were excised. Saline was flushed into unused colon and milking of saline done in microcolon down to the rectum, confirming colonic patency. Duodenum was opened at distal dependent part, and single layer end-to-back anastomosis was done with ascending colon using 5-0 polyglactin suture. Baby had an uneventful post-operative recovery. NG aspiration gradually decreased and became non-bilious. NG tube was removed, and breastfeeding started on the 7th post-operative day. Baby tolerated breast milk and was discharged on the 10th post-operative day. Parents attended outpatient department with the baby for the first follow up after 1 week and were advised about future plans.

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Figure 1: Erect abdominal radiograph demonstrating two dilated gas-filled bowel loops in upper abdomen and paucity of gas in pelvis

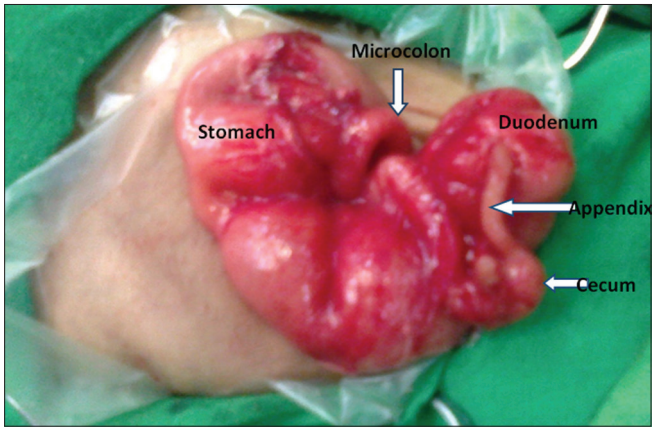

Figure 2: Intraoperative photograph showing dilated stomach and duodenum, a peanut-sized cecum followed by microcolon

Discussion

The classification of intestinal atresia is based on observations made by Louw and Barnard in 1955.[3] He reported three varieties of jejunoileal atresia. Type I was mucosal or membranous atresia. Type II was described as two blind ending atretic segments connected by fibrous cord along the edge of an intact mesentry. Type III was described as two separated segments of bowel with a V-shaped mesenteric defect. Later on 'apple peel' or 'Christmas tree' variety of proximal jejunal atresia and multiple atresias were added to the classification. The present classification system described by Grosfeld *et al*. is widely accepted.[2]

Here, the Type III variety described by Louw and Barnard is referred to as Type IIIa and the 'apple peel' variety is added as Type IIIb atresia. Multiple intestinal atresia is called Type IV variety. However, total/complete jejunoileal atresia is not mentioned in any of the classification systems. On extensive search of literature, we could not find the complete congenital absence of jejunum and ileum reported in living human babies. In 1884, Thomas reported post-mortem finding of a premature (7 months) baby with complete absence of jejunum, ileum and greater part of colon.[4] Duodenum was ending blindly, and the large bowel was terminating in vagina. The baby was not operated and died after 3 days. It was an autopsy report and photographs were not published. In 2013, Sham and Singh published a neonate with near total jejunoileal atresia, who survived for 3 months.[5] There was 2–3 cm jejunum distal to duodenum and 3–4 cm of ileum proximal to ileocecal junction. In our case, there was total absence of jejunum and ileum, so we have performed an end-to-back duodeno-colic anastomosis. One of the common post-operative complications in intestinal atresia is functional obstruction at the site of anastomosis. But here, bowel function started without much delay, and feeding started.

The currently accepted theory of intrauterine vascular insult as the cause of jejunoileal atresia was first confirmed by Louw and Barnard.[3] They demonstrated intestinal atresia in animal models by ligating mesentric vessels late in gestation. Processes such as volvulus, intussusception and internal hernias can cause intestinal atresia secondary to intrauterine vascular accident.[6] Familial forms of jejunoileal atresia can occur due to the developmental disorder of superior mesenteric artery and its branches. Shorter *et al*. proposed a classification system for familial cases of gastrointestinal atresia.[1] In the original vascular accident model, there is loss of pre-existing vessel. The novel classification model by Shorter *et al*. points towards failure of the vessel to develop, that is, embryogenic malformation. Hence although rare, embryologically there is a possibility for the occurrence of complete jejunoileal atresia due to an extensive vascular insult early in gestation.

Prognosis in jejunoileal atresia is significantly improved over the past two decades, but long gap intestinal atresia is still a management challenge. Due to the extreme rarity of total or near total jejunoileal atresia, a precise treatment protocol is yet to develop. Intestinal plication and proximal tapering enteroplasty procedures are utilised to prevent short bowel syndrome. Longitudinal intestinal lengthening and tailoring operation as proposed by Bianchi and serial transverse enteroplasty procedure advocated by Javid *et al*. are for patients with some length of available small intestine.[7,8] However, in our baby without any existing jejunum or ileum, intestinal transplant, which is available only in few centres of the world may be the sole salvage option in long-term.

Although congenital absence of jejunum and ileum is reported in one preterm human neonate as post-mortem finding, it is not yet reported in living human babies as an intraoperative finding. We have overcome the initial management challenge; however, long-term outcome is awaited. This case report which is first of this kind and the literature review will lead to a management guideline for these extremely rare neonates, to be addressed in future.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have

given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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