



CONGENITAL COLONIC ATRESIA PRESENTING AS PNEUMOPERITONEUM: A RARE CLINICAL ENTITY

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ABSTRACT

Introduction: Colonic Atresia is the least common type of Intestinal Atresia which occurs as a result of ischemic necrosis of a segment of large intestine. It presents with abdominal distention, bilious vomiting and failure to pass meconium. Perforation leads to peritonitis and sepsis

Case Report: We describe a 3 day old male baby presenting with Colonic Atresia type IIIa and pneumoperitoneum and his surgical management

Conclusion: In conclusion, Colonic Atresia is managed by either by colostomy or primary anastomosis. Terminal ileostomy, as in this case is done when there is Ascending Colon atresia with distal ileal perforation.

KEYWORDS : Colonic Atresia, Pneumoperitoneum, Terminal Ileostomy

INTRODUCTION:

Intestinal atresia is one of the most frequent causes of bowel obstruction in the newborn and can occur anywhere along the gastrointestinal tract. Most cases occur in the duodenum (60%). Colonic Atresia is the least common type which occurs only in 7-10 percent of all intestinal atresias [1]. The pathogenesis of Colonic Atresia is unknown but most people attribute it to vascular disruption leading to ischemic necrosis of a segment of fetal intestine. Thus, blind proximal and distal ends are formed after resorption of necrotic tissue. The majority of colonic atresias occur proximal to the splenic flexure with a distal microcolon as was found in our case. Apart from dilatation of small bowel loops, prenatal ultrasonographic findings may be normal and hence the diagnosis may be missed. Patients usually present with abdominal distention, bilious vomiting and failure to pass meconium with many patients landing up in dehydration and shock. Proximal perforation may complicate the clinical picture leading to peritonitis and sepsis.

Case Report:

A 3 day old full term male baby with a birth weight of 3kg was admitted with abdominal distention bilious vomiting and failure to pass meconium. The prenatal examination and ultrasounds of the mother were within normal limits. There was no exposure to drugs or infections during pregnancy. On examination, the baby was dehydrated and his body weight was 2.7kg. There was marked abdominal distention and subcostal retractions signifying respiratory distress. A feeding tube inserted per orally gave copious greenish aspirate. X ray examination of chest and abdomen showed pneumoperitoneum.



Fig 1. Marked abdominal distention with bilious aspirate



Fig 2. Pneumoperitoneum seen on X ray

Operative Steps:

An emergency laparotomy was done which revealed Type IIIa colonic atresia with distal microcolon. There was a 0.5x0.5cm perforation in the distal ileum 7cm from the atretic colon. The atretic colon and the perforated ileal segment along with appendix were resected and end ileostomy was done. The distal microcolon was fashioned into a mucous fistula and closure is done. Despite aggressive surgical and paediatric management, the patient's sepsis progressed and he succumbed on the third postoperative day.



Fig 3. Ascending colon Type IIIa atresia



Fig 4. Terminal ileostomy with mucous fistula

Histopathology:

The biopsies of the resected segment of colon and appendix revealed presence of ganglion cells. All the coats showed areas of haemorrhages, dilated and congested blood vessels and dense infiltration by mononuclear cells.

DISCUSSION:

Although the first case of Colonic Atresia was reported by Binniger in 1673, it was in 1922 that the first survival was documented by Gaub: a case of sigmoid atresia treated by delayed colocolic anastomosis after a primary colostomy. Colonic Atresia is a very rare entity and its incidence is 1 in 40,000 to 66,000 live births [2]. The atresia of the Ascending Colon as seen in our case is the least common location of Colonic Atresia [3]. The prognosis of Colonic Atresia depends on the presence of other complications like multiple atresias, proximal perforation and sepsis. Hirschsprung disease should be ruled out in all cases of Colonic Atresia and hence histopathological examination of all resected specimens/ intraoperative biopsies is essential. Colonic Atresia may also be associated with other malformations of the gastrointestinal tract like small intestine atresia, gastroschisis, imperforate anus, malrotation and omphalocele. Surgical management is the mainstay of treatment with left sided atresias requiring staged surgeries of colostomy followed by anastomosis and right sided atresias requiring resection anastomosis [4]. The management undergoes a variation if there is presence of small bowel perforation proximal to ascending colonic atresia as in our case wherein end ileostomy and formation of a mucous fistula would be the valid surgical option.

Extensive review of literature disclosed that even high volume paediatric surgery centres have reported to come across only one case of Colonic Atresia per year over a period of more than 10 years. Chouikh et al studied four cases of Colonic atresia between 2000 and 2012 three of which were identified as Type III and one as Type I Colonic atresia [5]. Two patients were operated for colostomy followed by anastomosis and the other two for primary anastomosis one of who died due to Respiratory Distress Syndrome post surgery. The others were uneventful on follow up. Cox S et al (2005) reviewed 14 cases of Colonic Atresia over a 38 year period and concluded that primary anastomosis would only be rarely advised [6]. However Tripathy PK et al in 2020 after operating 12 cases in 2.5 years came to a conclusion that all cases of Congenital Atresia can be successfully managed by primary anastomosis irrespective of the location [7]

In conclusion, Colonic Atresia is a rare entity with a high rate of comorbid conditions and progression to sepsis in case of proximal perforation. Early intervention by colostomy or primary repair is essential. In the rare event of Ascending Colon Atresia with distal ileal perforation, terminal ileostomy needs to be done.

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