Case Report

DOI:10.7860/IJNMR/2017/29190.2213

Paediatric Surgery Section

Type II Colonic Atresia and High Anorectal Malformation: A Rare Coexistence and Literature Review

PRASANTA KUMAR TRIPATHY, PRADEEP KUMAR JENA

ABSTRACT

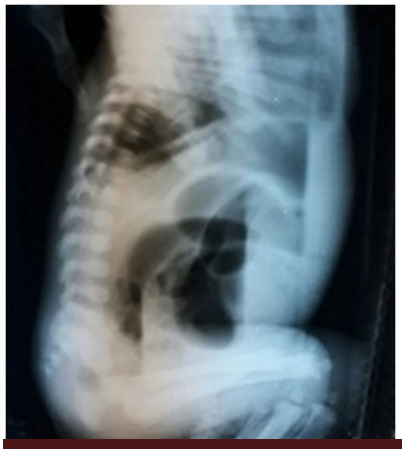
Colonic atresia and anorectal malformation are congenital anomalies affecting distal gastrointestinal tract. Individually, both the anomalies need urgent surgical and medical care for survival of the newborn. However, the association between them is reported only in a handful of cases and very little information is available regarding the management of the combined anomaly. We came across a three-day-old male neonate having Type II colonic atresia along with high anorectal malformation. Because of the rarity and unique challenges needed for management of both the anomalies, this case is being reported with review of relevant literature.

Keywords: Alimentary tract atresias, Colostomy, Posterior sagittal anorectoplasty

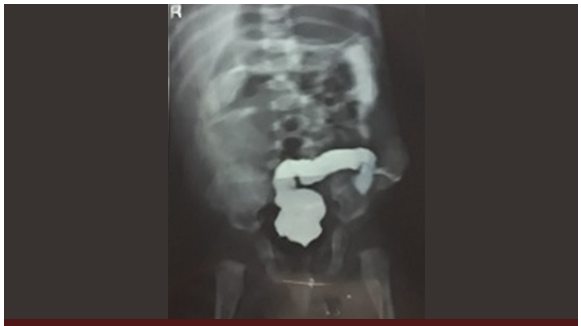
CASE REPORT

A three-day-old full-term male newborn was presented to emergency with abdominal distension, bilious vomiting and absence of anal opening. He was born by normal vaginal delivery at home and antenatal ultrasonography was not done. Mother had not taken any drugs other than those prescribed during pregnancy and there was no exposure to infectious agents. Rest of the family and maternal history is unremarkable. The baby was the first child of the family and there was no history of meconuria. At presentation he was weighing 2.2 kg and dehydrated. Pulse rate was 150/min and respiratory rate 60/min. Abdomen was distended with engorged veins over abdominal wall. There was no rigidity or tenderness. Perineal examination revealed a flat perineum. Although, anal dimple was present, anal opening was absent. No obvious anomaly was detected on sacrum, spine or vertebra. Cross-table lateral radiograph of abdomen showed high ARM [Table/Fig-1].

After adequate resuscitation the baby was taken for emergency laparotomy. Parents consent was obtained prior to the treatment. Colon was dilated up to proximal 5 cm of sigmoid colon and ending blindly. It is followed by a short fibrous cord and microcolon, the Type II



[Table/Fig-1]: Cross-table lateral radiograph showing high anorectal malformation. **[Table/Fig-2]:** Intraoperative photograph showing dilated and blind ending proximal sigmoid colon followed atretic distal colon. **[Table/Fig-3]:** Showing distal descending colostomy formation. Perineum showing anal dimple, but anal opening is absent.



[Table/Fig-4]: Distal colostogram at 3 months revealing good calibre of sigmoid colon and rectum.

colonic atresia [Table/Fig-2]. End-to-back anastomosis was done between proximal dilated and distal atretic ends of colon. Distal descending loop colostomy was performed [Table/Fig-3] and laparotomy wound closed. Postoperative period was uneventful and colostomy functioned well. Care of colostomy and distal colon wash with normal saline was explained to parents. The newborn was discharged on 4th postoperative day with breast feeds. Distal colostogram done at three month follow-up shows good calibre of distal colon and no fistulous connection with urinary tract [Table/Fig-4]. Follow-up is continuing along with distal colon wash. The infant is growing and gaining weight according to his age. Definitive procedure for ARM, Posterior Sagittal Anorectoplasty (PSARP) is planned at the age of six months followed by colostomy closure.

DISCUSSION

Anorectal Malformation (ARM) and alimentary tract atresias are important causes of neonatal intestinal obstruction. ARM is associated with esophageal atresia in 10% of cases. It is also well described with duodenal and Jejuno-Ileal Atresias (JIA) [1,2]. However, the association of ARM with Colonic Atresia (CA) is an unusual finding [3]. Similarly, CA is associated with proximal small bowel atresias in about 20% of cases and Hirschsprung's disease in 2% of cases [4]. But, its co-existence with imperforate anus is very rare [1,3,4].

The etiology of CA is suspected to be intrauterine mesenteric vascular accident similar to the pathogenesis of JIA [1,3]. The first infant with colonic atresia was survived about 250 years after the initial description of this congenital anomaly in 1673 [4,5]. But, very few cases have been reported since then and the incidence of CA is 1 in 20000 live births. It is the rarest among all varieties of alimentary tract atresias, accounting for 1.8 to 15% of cases [6,7]. Therefore, CA is not only rare, but also a fatal disease affecting the newborns. The situation is expected to become more complicated when another congenital malformation of gastrointestinal tract like small intestinal atresia, Hirschsprung's disease or high ARM is associated.

Colonic atresia is classified as per classification system described by Louw [8]. Type I represents mucosal atresia with intact mesentery and bowel wall. Type II consists of a fibrous cord separating the two bowel segments and in Type III atresia, bowel ends are separated by a V-shaped mesenteric defect. Although, Type III is the most common variety encountered in clinical practice, our case was having Type II anomaly [4]. Chouikh T et al., reported four cases of CA in 2014, out of which one case was Type I and rest were Type III variety [7]. Mirza et al., described four cases of Type I and 5 cases of Type III atresia of colon and in only one case the atresia started distal to the splenic flexure [6]. Both the authors have not mentioned Type II atresia in their series. Therefore, even among colonic atresias, type II variety and atresias starting beyond splenic flexure are uncommon. Preoperatively our diagnosis was high ARM and we have planned for colostomy on emergency basis. The additional finding of Type II colonic atresia during laparotomy prompted us for extensive search of literature. The coexistence of CA with ARM is reported only in eight cases (to the best of authors' knowledge) in English literature [1,3,4].

Colonic atresia can be managed by resection and primary anastomosis. This may be technically difficult due to size discrepancy between proximal and distal bowels and wide separation between the ends after resection of proximal dilated colon [5,7]. There is higher risk of complications due to unrecognized distal pathology. Many surgeons add a covering enterostomy after colocolic anastomosis [6]. Authors also advocate a staged approach of initial decompressing ostomy procedure followed by delayed anastomosis for atresia. This procedure is particularly needed when condition of the newborns is not stable [7].

The ideal operative management of CA with imperforate anus is a staged approach. An initial ostomy procedure proximal to the atresia is done to decompress the intestine [3,9]. Accordingly we have performed colostomy proximal to the anastomosis. Proximal stoma will protect the colonic anastomosis and will maintain the bowel function till definitive procedure for high ARM is performed. High ARM is associated with fistula to the urinary tract in 95% cases [2]. But in our patient there is no fistula to the urinary tract on distal colostogram making the indexed case extremely unusual. We are planning PSARP at the age of six months followed by colostomy closure for complete correction of the combined anomalies.

CONCLUSION

Although, rare ARM may coexist with colonic atresia and presents an unique surgical challenge. The distal bowel must be looked for any pathology during colostomy formation in high ARM. The optimal operative procedure should be individualized depending upon the location and type of CA and the general condition of the newborn.

REFERENCES

[1] Maitra S, Sarkar R. Anorectal malformation associated with small and large bowel atresias. J Neonat Surg. 2012;1(4):59.
[2] Levitt MA, Pena A. Anorectal malformations In: Coran AG, Adzick NS, Krummel TM, LAberge J, Shamberger RC, Caldamone AA. Editors. Pediatric Surgery. 7th ed. Elsevier Saunders 2012; pp. 1289-309.
[3] Derenoncourt MH, Baltazar G, Lubell T, Ruscica A, Sahyoun C, Velcek F. Colonic atresia and anorectal malformation in a Haitian patient: a case study of rare diseases. Springer Plus. 2014;3:203.
[4] Arca MJ, Oldham KT. Atresia, stenosis and other obstructions of the colon In: Coran AG, Adzick NS, Krummel TM, LAberge J, Shamberger RC, Caldamone AA. Editors. Pediatric Surgery. 7th ed. Elsevier Saunders 2012; pp.1247-53.
[5] Hsu CT, Wang SS, Houng JF, Chiang PJ, Huang CB. Congenital colonic atresia: Report of one case. Pediatr Neonatol. 2010;51(3):186-89.
[6] Mirza B, Iqbal S, Ijaz L. Colonic atresia and stenosis: our experience. J Neonat Surg. 2012;1:4.
[7] Chouikh T, Chareieg A, Mrad C, Ghobel S, Saadda S, Benkhalifa S et al. Congenital colonic atresia: 4 case reports. J pediatr Neonatal Care. 2014;3(1):00018.
[8] Louw JH. Resection and end to end anastomosis in the management of atresia and stenosis of the small bowel. Surgery. 1967;62(5):940-50.
[9] Karnak I, Ciftci AO, Senocak ME, Tanyel FC, Buyukpamukcu N. Colonic atresia: surgical management

AUTHOR(S):
1. Dr. Prasanta Kumar Tripathy
2. Dr. Pradeep Kumar Jena

PARTICULARS OF CONTRIBUTORS:
1. Assistant Professor, Department of Pediatric Surgery, SVP PG Institute of Pediatrics, Cuttack, Odisha, India.
2. Professor, Department of Pediatric Surgery, SVP PG Institute of Pediatrics, Cuttack, Odisha, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Prasanta Kumar Tripathy,
Arunodaya Nagar, Link Road,
Cuttack 753012, Odisha, India.
Email: drpktripathy555@gmail.com

FINANCIAL OR OTHER COMPETING INTERESTS:
None.

Date of Publishing: **Jul 01, 2017**