

Management Congenital Colonic Stenosis

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Abstract

Background: Colonic Stenosis (C.S) is a rare entity. It has been reported either as an isolated disorder or in association with other anomalies like Hirschsprung's disease, craniofacial and musculoskeletal abnormalities. This survey was conducted to study the clinical profile and challenges in the management of this rare anomaly.

Material and methods: Retrospective analysis of 6 patients diagnosed to have colonic stenosis during 2007-2014. The diagnosis was confirmed in 5 patients during the surgical exploration.

Results: In one patient colonic stenosis was detected pre-operatively. In most cases X-ray findings included multiple air fluid levels and absent gas in pelvis. All the patients presented in neonatal period. One patient had ascending colon, one had transverse colon, 2 descending colon and 2 patients had sigmoid colon stenosis. In 5 patients resection anastomosis was done and in one stoma was formed. All the patients are on follow-up and are doing well.

Conclusion: Colonic atresia should always be kept in mind while managing cases of suspected partial /complete intestinal obstruction in a neonate.

Keywords: Congenital colonic stenosis; Colon; Obstruction

Introduction

Colonic stenosis (CS) is a very rare anomaly, and to date, only 15 cases have been reported in English literature [1]. It usually presents during early neonatal period, although some late presenting cases have also been reported [2]. We present our experience and challenges in management of congenital CS.

Case 1: A 5-day-old, full-term male baby presented with abdominal distension, bilious vomiting, and failure to pass meconium. Abdominal radiograph showed multiple dilated loops of bowel with air-fluid levels and absence of gas in the rectum. Exploratory laparotomy was performed with findings of ascending CS. This stenosed segment was resected and end-to-end anastomosis was accomplished (Figure 1).

Case 2: A 4-day-old full-term male-baby born through cesarean delivery presented with failure to pass meconium and abdominal distension. Plain X-ray of the abdomen showed multiple air-fluid levels and paucity of gas in the rectum. The baby was diagnosed with small bowel atresia, and subsequently laparotomy was performed that showed a stenosing lesion of about 2 cm in the descending colon. This stenosing segment was resected and end colostomy with colonic mucous fistula was created. Colostomy was closed after 6 weeks. The baby did well in follow-up.

Case 3: A 3-day-old preterm male baby presented with bilious vomiting, abdominal distension, and failure to pass meconium. Radiographic study with water-soluble contrast medium showed a delayed transit time and dilated bowel loop. During operation,



Figure 1: Intra-operative photograph ascending colon stenosis



Figure 2: Intra-operative picture of transverse colon stenosis

transverse CS of 3 cm was seen, and resection and anastomosis was performed with uneventful postoperative recovery. The baby was discharged on sixth day after operation (Figure 2).

Case 4: A 20-day-old female child presented bilious vomiting and abdominal distension. She had history of delayed passage of meconium. X-ray of the abdomen showed multiple air-fluid levels and absence of gas in the rectum. Decision was taken for laparotomy and the child was diagnosed as a case of congenital CS in the descending

colon. Resection and anastomosis of the stricture was performed. Histo-pathological examination (HPE) of stenosed colonic segment confirmed the diagnosis of CS. Postoperative period of the patient was uneventful, and the baby was discharged on sixth day after operation.

Case 5: A 6-day-old female baby presented with bilious vomiting, failure to pass meconium, and abdominal distension. Contrast enema showed an abrupt change in caliber in the sigmoid colon, and after ruling out other disorders CS diagnosis was made. Laparotomy was performed and sigmoid CS was confirmed. Resection and anastomosis of the stenosed segment was performed. Patient recovery was rapid and functional results were excellent (Figure 3).



Figure 3: Intra-operative picture showing sigmoid colon stenosis

Case 6: A 4-day-old, full-term male baby born through normal vaginal delivery presented with abdominal distension, failure to pass meconium, and bilious vomiting. A plain abdominal radiograph showed air–fluid levels and paucity of gas in pelvis. Contrast enema appeared to completely fill the lumen of colon up to ileocecal valve with a narrowing in ascending colon. On exploration sigmoid CS was found, and resection and anastomosis was performed. The postoperative course was uneventful, and the patient is currently in good clinical condition and has normal thriving.

Results

CS was suspected preoperatively in only one patient (case 5). In all other cases, it was detected during operation/exploration. X-ray of the abdomen was found to be a good initial investigation that can show multiple air–fluid levels and distension of both small and large gut. Furthermore, it can show pneumo peritoneum in case of colonic perforation. Contrast enema was found to fill the gut proximal to CS in case 6. Abrupt change in caliber of the colon in sigmoid stenosis was observed in case 5 and presumptive diagnosis of CS was made that was later confirmed on exploration. Paucity of gas in the rectum was observed in two patients, absence of gas in three patients, and presence of gas in the rectum in spite of multiple air–fluid levels in one patient (case 5). Salient features of CS in six patients are summarized in Table 1.

Case	Day of life/sex	Gestational kgs	maturity/Weight	Main Clinical features	Radiology	Colonic segment involved	Operative Procedure Performed
1	5/male	Term/3 kgs		Abdominal distension, Failure to pass meconium	Multiple air fluid levels, no gas in rectum	Ascending colon	Resection anastomosis
2	4/male	Term/2.5 kgs		Abdominal distension, bilious vomiting	Multiple air fluid levels, paucity of gas in rectum	Descending colon	Colostomy and mucous fistula
3	3/male	Pre-term/1.8 kgs		Bilious vomiting, Abdominal distension,	Multiple air fluid levels, no gas in rectum	Transverse colon	Resection anastomosis
4	20/female	Term /4 kgs		Constipation, abdominal distension	Multiple air fluid levels, no gas in pelvis	Descending colon	Resection anastomosis
5	6/male	Term /3.2 kgs		Bilious vomiting, Failure to pass meconium	Multiple air fluid levels , gas present in in rectum	Sigmoid colon	Resection anastomosis
6	4/male	Term/3 kgs		Abdominal distension, Failure to pass meconium	Scanty gas in pelvis	Sigmoid colon	Resection anastomosis

Table 1: Salient features of the colonic stenosis patients

CS was present in ascending colon in one patient (case 1), transverse colon in one patient (case 3), descending colon in two patients (cases 2 and 4), and sigmoid colon in two patients (cases 5 and 6). The length of the stricture was between 1 and 2 cm in these six patients. The discrepancy between the gut proximal and distal to stenosis was not more than 3:1, which could make colonic anastomosis easier in five cases (cases 1, 2, 4, 5 and 6). No associated gut anomaly was observed in these six cases of CS.

Resection and primary anastomosis was accomplished successfully in five patients (cases 1, 3, 4, 5 and 6) as the disparity between the two colonic segments was not >3:1. Only one patient (case 2) required colostomy for descending CS to decrease the operative time in view of acidosis and greater disparity between the two segments. All the babies are asymptomatic at last follow-up with normal bowel function.

Discussion

Several theories have been proposed to describe the etiology of intestinal stenosis. Failure of resorption of the solid stage of embryonic precursor was proposed by Tander [3] Theory of emboli originating in the placenta reaching the mesenteric circulation after bypassing pulmonary circuit has been proposed by some authors [4], which states that mesenteric vascular insult results in colonic atresia and stenosis. This vascular compromise could result from fetal herniation, kinks, intussusception, or primary vascular accidents. Cocaine has been implicated in neonatal gut ischemia [5].

Depending on the severity of ischemia, stenosis or atresia may occur. Santulli and Blanc [6] reported a case of sigmoid stenosis treated with colostomy. Pai and Pai [7] reported a case of recto sigmoid stenosis in a 4-month-old infant. Sax [8] described a CS in a 2-dayold, full-term newborn. Mirza et al. [9] reported four cases of CS, in transverse colon in two patients and in sigmoid colon in two patients. Among these, two were congenital and two secondary to necrotizing enterocolitis (NEC) [9]. In our study, stenosis was more common in left colon, sigmoid colon in two cases, descending colon in two, and transverse and ascending colon in one each. Intestinal atresia is classified into type I (intraluminal diaphragm), type II (separation of two lumens by a fibrous cord with intact mesentery), and type III (gap between the two colonic segments with a mesenteric defect). Intestinal stenosis was offered a fourth category by referring to narrowing of the intestinal lumen that varies from a slight reduction in diameter to almost complete occlusion [4].

Symptoms in neonates may be impossible to differentiate from other cases of intestinal obstruction such as atresia. It may present during first few days or may be apparent after several days of life [10], as was noted from the cases reported by us, where five patients presented within first six days of life. In our patients, most common presenting features were bilious vomiting (four cases), abdominal distension (five cases), and failure to pass meconium (two cases). As the symptoms of CS progress, it may result in dehydration, weight loss, or failure to gain weight. In all the reported cases of CS, there was evidence of dilated proximal intestines in radiographic images [10]. Contrast enema studies have been reported to help in preoperative diagnosis of CS. It can help differentiate between colonic atresia and stenosis. In colonic atresia, contrast will fill the distal unused colon terminating at a point adjacent to the segment that is more distended with the luminal air "the cutoff point," whereas in CS the contrast enema will delineate both unused distal colon with a small caliber followed by a distended portion of colon proximal to the stenosis [11,12]. In our survey on CS, the contrast enema has not been performed in every case. As five patients were explored with a suspicion of small gut atresia and CS was an operative finding, we believe that contrast enema should always be carried out whenever there is doubt about the location and nature of neonatal intestinal obstruction to delineate the unused colon and ascertain any colonic atresia and stenosis. Although paucity of the rectal gas was a consistent finding on radiography, it is not a specific feature for diagnosing CS.

CS is a rare anomaly, with less than 15 cases reported in literature [10]. Colon is the least common site of congenital intestinal stenosis and atresia [5]. Experience with this rare anomaly and improved postoperative care has resulted in increase in survival rate [13]. Our reviewed cases of CS were isolated, not associated with other anomalies.

Early surgical intervention is imperative if preoperative diagnosis is made. Most of left-sided CS lesions are treated by resection and primary colonic anastomosis with a covering stoma. In four of the patients with left-sided CS, primary anastomosis was performed without covering stoma. As observed in our survey, primary anastomosis is preferred by some in right-sided CS lesions [2,14]. We did form a stoma in case 2 where ascending end colostomy with mucous fistula was made and later closed after 6 weeks. In all the six cases, segment was sent from HPE that confirmed the stenosis. The significance of pathological findings such as sub mucosal collagen deposition is not known [15]. Some authors have described the overexpression of transforming growth factor- β (TGF- β) in localized collagen deposition in mouse colon that can lead to intestinal fibrosis and obstruction. Excessive collagen deposition due to mutation in the TGF- β pathway and scar formation precipitated by in utero vascular insult ultimately lead to clinical presentation of CS.

Outcome remained good in all the six reported patients with CS in our survey possibly due to the isolated nature of the lesion because many authors believe the prognosis might be bad in case CS lesion is associated with other anomalies [9].

Conclusion

To conclude, CS is a rare anomaly, diagnosed frequently during laparotomy. Pro radiological studies and contrast enema can help us to arrive closer to the diagnosis. Considering our experience and literature review, it is wise to consider CS as unusual, but possible cause of intestinal obstruction in a neonate. It should be kept in mind for avoiding diagnostic dilemma during the management of neonatal intestinal obstruction.

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