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Case Report

Congenital colonic stenosis in a term newborn, a case report and literature review

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Colonic stenosis is a rare anomaly and a type of colonic atresia. The most frequent clinical manifestations are chronic constipation and obstructive episodes in newborn or infant. Contrast enema helps to establish primary diagnosis. The colonic stenosis was characterized by dilated proximal intestine and a narrow transitional zone. It is diagnosed during laparotomy and needs to be differentiated from several colonic obstructive disease. We share this case and review of contemporary management. A term female infant, with progressive abdominal distention, vomited after feeding and defecated colorless mucous. A retrograde contrast enema revealed small caliber of the rectum and marked distention of the proximal colon. Laparotomy revealed isolated stenosis in sigmoid colon. After segmental resection and primary anastomosis, an uneventful postoperative course was followed. The child has remained symptom-free at subsequent follow-up. Differential diagnoses in a neonatal intestinal obstruction is well reviewed in literature. Roentgenogram combine with contrast enema is compatible in both tentative diagnosis and initial treatment. Although these may subside spontaneously, most of the diagnostic delay are associated with unsatisfactory outcomes. Colonic stenosis is generally treated by surgical resection as reported in this case. And, as improvement in colorectal surgery, up-to-dated management of colonic atresia and stenosis yielded less morbidity and mortality. We collected pathological description in articles of congenital colonic stenosis and made a comparative table.

Keywords: colonic stenosis, retrograde contrast enema

1. Introduction

The first colonic atresia was reported in 1673 and the first congenital colonic stenosis in 1968. According to the definition of Louw and Barnard in 1959, occlusions conform to four types: type 1 atresia, a diaphragm or membrane; type 2 atresia, blind ends joined together with a cordlike remanent; type 3 atresia, disconnected blind ends by a gap in the mesentery. And the colonic stenosis belongs to type IV atresia [1]. In article reviews, the occurrence of colonic atresia was about 1.8-15% of intestinal atresia. And it was about 1 of 40,000 newborns per year in the United State [2]. Colonic stenosis, including both acquired and congenital, is an extremely rare cause of low-intestinal obstruction. It was mostly recorded in neonates, and in elder children or adults, there were different clinical findings as reported. Rectal stenosis or atresia are considered within the spectrum of imperforate anus and were discussed separately from colonic atresia in published articles. Almost all the reported patients with colonic stenosis have well outcomes. And with the improvement of contemporary colonic surgical technique, prognosis in atresia patients is generally good and mortality was

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mostly attributed to associated anomalies or difficult comorbidities. The etiology of congenital colonic atresia was long under debate. There were the near-contemporary hypothesis of the epithelial plug and the classic one of the intrauterine vascular accident, which was experienced in the animal model. In the articles of the latest decade, the genetic mechanism of Fgf10 (fibroblast growth factor 10) and series of overexpression of TGF-β (transforming growth factor beta) were elucidated and under development [8].

2. Case report

We present this case of a one-day-old term female infant with progressive abdominal distention after birth. Vomit after feeding and colorless mucous defecation were noticed. A retrograde contrast enema performed the first day after birth revealed a small caliber of the sigmoid and rectum and marked distention of the proximal colon. Some contrast media passing the transitional zone into the sigmoid colon was also observed (Fig.1). The rectal suction biopsy shows submucosal nerve plexuses with the presence of ganglion cells. And under the impression of colon obstruction with sigmoid colonic transitional zone, an exploratory laparotomy was performed on the fifth day after birth. We conducted laparotomy through transverse, supra-umbilical position. The obstructing lesion was resected along with a compromised dilated proximal intestine (Fig.2). In macroscopic findings, a 10.2cm in length and 1cm diameter segment of dilated, ulcerative, congested sigmoid colon, followed by a 3cm long segment of intestinal stricture lesion. The diameter of the stenosis was 0.3cm (Fig.3). Segmental resection and primary end-



Figure 1. The retrograde contrast enema showed small diameter of the sigmoid and rectum and marked distention of the proximal colon (arrow). Also, some contrast media passing the transitional zone into the sigmoid colon was observed.

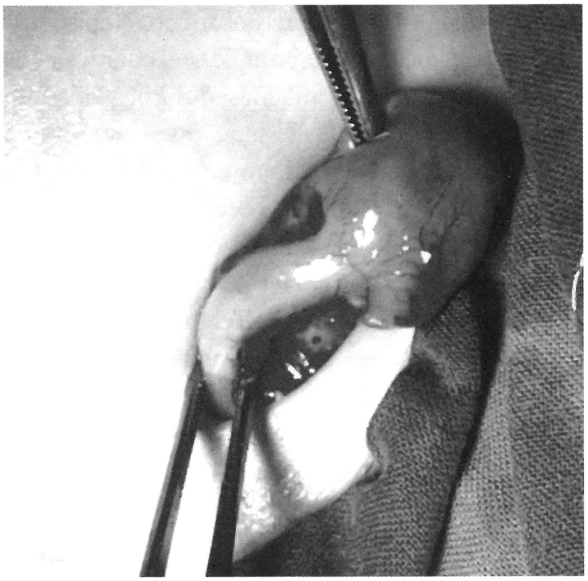


Figure 2. The laparotomy incision was through transverse and supra-umbilical position. The segment of dilated sigmoid colon was followed by the transitional zone of stricture lesion (arrow).

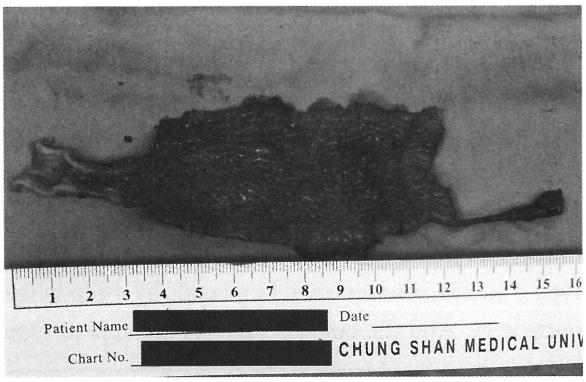


Figure 3. The segmental resection of dilated proximal intestine, 10cm in length and 1cm in diameter, was an ulcerative and congested sigmoid colon. The distal 3cm long segment of intestinal stricture lesion was stenosis with a 0.3cm caliber (arrow).

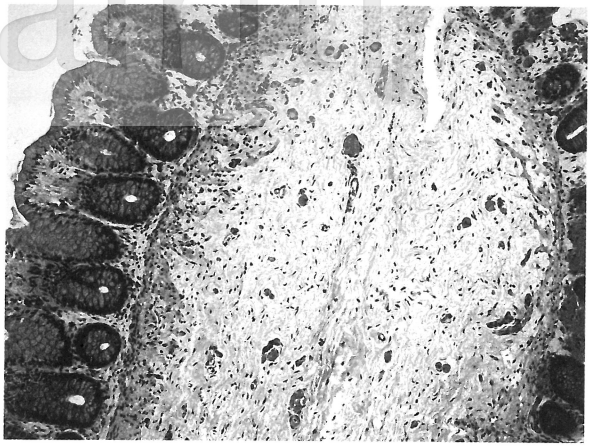


Figure 4. The hematoxylin and eosin stain of resected sigmoid colon segment showed mucosa ulceration, vessel congestion, and submucosa infiltrated with lymphoplasmacytic cells and neutrophils (arrows). (Magnification: 100X)

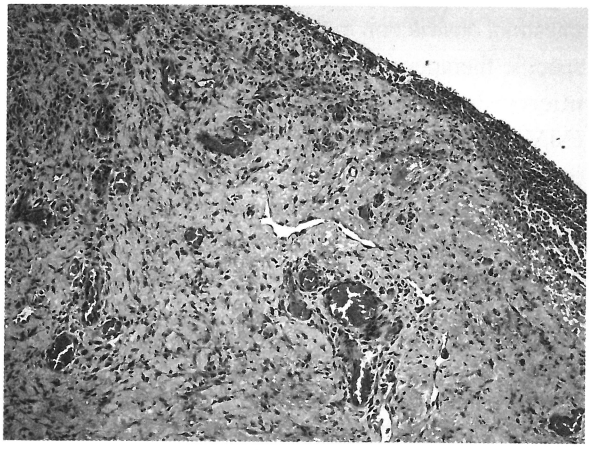


Figure 5. Subserosa edema and lymphoplasmacytic cells and neutrophils infiltration was also observed in hematoxylin and eosin stain (arrows). (Magnification: 100X)

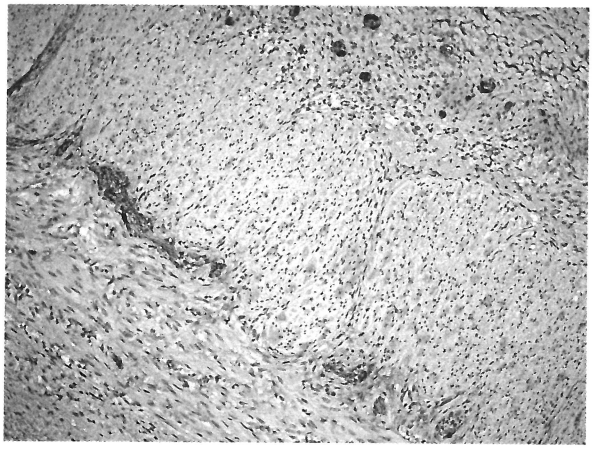


Figure 6. The calretinin immunostaining highlights ganglion cells in the submucosal plexus and myenteric plexus (arrows). (Magnification: 100X)

to-end anastomosis were conducted. No uneventful postoperative course was followed. The infant has remained symptom-free at subsequent follow-up. Microscopic examination of resected sigmoid colon segment shows mucosa ulceration, vessel congestion, subserosa edema, submucosa and subserosa infiltrated with lymphoplasmacytic cells and neutrophils (Fig.4 and Fig.5). There are ganglion cells in the submucosal plexus and myenteric plexus. The calretinin immunostaining highlights small intrinsic nerves of muscularis mucosae and lamina propria (Fig.6).

3. Discussion

The cause of pediatric colonic obstruction is diverse. In colonic atresia or stenosis, there were no known racial, maternal, neonatal or gender predilection. "Acquired" colonic stenosis is one of the two division of colonic stenosis related to inflammation, infection, traumatic, neoplastic processes in infants and children. Neonatal necrotizing enterocolitis was reported as the most common causation [1]. It is remarkable that Hirschsprung disease was reported related to at least 2 % of colonic atresia [2]. The mechanism is through vascular insult before retroperitoneal fixation of colon, and interrupted caudal migration of myenteric neurons. Thus, in the scene of imperatively rule-out Hirschsprung disease related atresia, suction rectal biopsy was largely suggested in published articles.

Before the classic etiology, an animal-experienced hypothesis of intrauterine vascular insufficiency during organogenesis [3], there were an "epithelial plug" hypothesis. It was based on the observation of failed to resorb of solid stage of embryonic precursor. But the hypothesis is not suitable to explain non-membranous type [4]. Based on the classic hypothesis of vascular accident, articles presented mechanical defect implicated in intestinal atresia including incarceration or strangulation of hernia, volvulus, intussusception [5] and maternal reason including cocaine and nicotine usage [6].

With the discovery of fibroblast growth factor 10 (Fgf10), genetic mechanism for bowel atresia was elucidated as a contemporary hypothesis of a new possible etiology and histopathology of intestinal atresia. In an article published in 2011, Nichol et al. arouse a new round of questions to provoke

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genetic investigation of atresia by the models of mouse [7]. One of them is the time point when did atresia occur? It is probably on days 33th to 48th in equivalent to data based on murine models. Another one is to figure out what genes are involved in mouse models. Some signaling pathway genes were excluded in mice, and FGF10 / fibroblast growth factor receptor 2 (Fgfr2) signaling axis was found associated with duodenal stenosis in mouse model and humans. These not-well-defined downstream signal pathways and morphogenetic events were considered a result in the improper development of the affected intestinal segment. Transforming growth factor beta (TGF-beta) / Smad7 (mothers against decapentaplegic) cascade, another promising signaling pathway implicating bowel tissue fibrosis, maintained intestinal hemostasis and mucosal immune response [8]. The mechanism of TGF-beta signaling in intestinal fibrosis is still not fully understood [9].

Although clinical symptoms including failure to advance oral feeding in neonate, bilious vomiting, colicky pain, and abdominal distention are typical, it is difficult to differentiate from other intestinal obstruction [10]. When the small intestinal dilation was not observed until 24 hours after birth and failed to pass meconium until 48 hours after birth, we put atresia into consideration [11]. Other two major findings in elder children are: fail to gain weight and history of off-and-on constipation since birth.

Some articles suggested algorithms based on image pattern. The algorithm started from abdomen radiograph [12]. Lower gastrointestinal obstruction is then differentiated by using of contrast enema, which is well recognized to establish the diagnosis of colonic atresia versus stenosis in contemporary articles. Classic pattern of colonic atresia in contrast enema are a "cut-off" point, a small caliber in distal colon and distended proximal colon with air fluid level [13]. Colonic stenosis is characterized by limited proximal contrast filling, which delineate both unused distal colon and distended proximal portion. But there are some shortages of this procedure. Barium enema was reported with initially normal image findings and delayed presentation of stenotic or atretic segment [14]. And the retention of colonic contrast may worsen peritonitis in case of perforation [15].

Nevertheless, contrast enema is a suggested and reliable clinical differential tool, it is never a conclusive diagnosis of colonic obstruction. Congenital colonic stenosis likewise could only be diagnosed during laparotomy [16]. In the work-up of clinical differentiation, it is important in identifying other cause of obstruction. Since the operation of colonic atresia/stenosis should not be delayed due to the risk of perforation and segmental volvulus, on-time diagnosis and appropriate management guaranteed good overall prognosis. Because of the observed disease-association with Hirschsprung disease, a suction rectal biopsy to evaluate for ganglion cells is strongly suggested. Colonic obstructions have varied pathogenesis, some preferred nonsurgical approach. Functional colonic obstruction is also a part of differential diagnosis of pediatric colonic obstruction. Severe functional intestinal obstruction in the neonatal period and had no specific therapeutic intervention. Neonate megacystis-microcolon-intestinal hypoperistalsis syndrome (MMIHS) is rare and severe congenital condition is associated with non-obstructed urinary bladder, microcolon and decreased intestinal peristalsis. There is no specific therapeutic intervention [17]. Meconium plug syndrome is relative common, and rectal stimulation including contrast enema are therapeutic. Meconium ileus is obstruction of the distal ileum due to abnormally impacting meconium. And most cases feature of cystic fibrosis association. Both nonoperative and operative therapies can be relieving [18]. Small left colon syndrome is partial obstruction mostly involving descending colon. Unknown etiology related to insulin and glucagon were reported. Contrast enema is diagnostic and maybe therapeutic. The syndrome develops in association with gestational diabetes, and it often recovers spontaneously with improvement in 1–2 weeks [19]. Other congenital malformation may lead to obstruction including volvulus, peritoneal bands, and hindgut duplication. Partially obstructing intraluminal membrane was also reported a cause of colonic stenosis [20].

The laparotomy is stated classically through supraumbilical position and transverse incision. The obstructive segment was resected along with compromised dilated proximal intestine. Similar to other colon surgery, surgeon may adapt primary anastomosis or proximal ostomy with staged repair

Table 1. Part A. Acquired colonic stenosis

	gender	GA	age	segment	length	antecedent	Mucosa	submucosa	muscle	serosa	other
1970 Erskine	M		8 w	D	6 cm	erythroblastosis fetalis	granular, eroded mucosa, ulceration extend to muscle layer		granulation tissue with plasma cells and lymphocyte, ganglion cells in myenteric plexus	fibrosis, especially in mesentery vessels	
1998 Takahashi	F	term	3 y	T	0.7 cm	necrotizing enterocolitis	mucosal folds converged toward stenotic center, proximal side edematous lesion	absence of submucosal layer	focal thickening muscle layer		ganglion cell throughout the specimen
2013 Pelizzo	M	28 w	19 d	T		norovirus infection	hyperplastic vessels with thicken wall			hyperplastic vessels with thicken and fibrosis wall without necrosis	
	M	30 w	44 d	Rt colon							
2018 Xie	M	38 w	20 d	DS / T	3.5 cm / 4cm	Clostridium difficile infection	fibrosis of lamina propria in the narrow segments.				normal ganglion cells
2019 Naito	M	30 w	30 d	A		acute neonatal appendicitis	necrotic granulation limited within lamina muscularis mucosae and submucosa			old hemorrhage, subtle calcification, few foreign body granuloma in the area	
2020 Upadhyaya	M	term	25 d	D		secondary to Hirschsprung disease					Granulation tissue, capillaries and mixed inflammatory cells (neutrophils, lymphocytes, histiocytes) at site of stenosis
	M	term	32 d	DS			denudation of lining epithelium with mild reactive atypia in places, inflammatory cells (histiocytes), thinned out mucosa				
	F	term	40 d	SF				congested vascular channels			marked increase in lymphomononuclear inflammatory cell infiltrate
	M	term	96 d	D							Granulation tissue, capillaries and mixed inflammatory cells (neutrophils, lymphocytes, histiocytes) at site of stenosis

Abbreviation: A, ascending colon. D, descending colon. d, day(s). DS, descending-sigmoid colon iunction. F. female. GA, gestational age. M, male. Rt colon, right colon. S, sigmoid colon. SF, splenic flexure of colon. T, transverse colon. w, week(s). y, years(s).

Table 1 (cont'd). Part B. Congenital colonic atresia

	gender	GA	age	segment	length	mucosa	submucosa	muscle	serosa	other
2010 Baudet	F		32 y	SF	2 cm	normal mucosa with calcified fecaloma		hypertrophy muscle layer		no inflammatory, ischemic, neoplastic changes, or changes in the nerve plexuses
2010 Montaño	M		3 y	T	3 cm			lymphatic cells		
2013 Lim	M		6 m	T	2 cm		increased collagen deposition			normal ganglion cells ; Trichrome stain, hematoxylin and eosin stain: collagen deposition in colon
2017 Bharti	M	term	5 m	S	2 cm					ulceration, perforation and granulation tissue, capillaries and mixed inflammatory cells (neutrophils, lymphocytes, histiocytes)
2017 Natarajan	F		4 m	T		non-specific inflammation of mucosa				thick intra-luminal membrane (web) within central hole ; ganglion cells presence
2019 Shah	F	term	1 m	DS					abundant mixed inflammatory infiltrate with serositis and thick walled blood vessels	
2021 Gupta	M		5 d	A	4.5 cm		hematoxylin and eosin stain: nonspecific inflammation along with an area of fibrosis			Mason stain: fibrous tissue presence ; Orcein stain: areas of elastin fiber deposition
	M		15 d	S	2 cm					
	M		2 m	A	1 cm					
	F		4 m	T	3 cm					
	M		7 m	T	5.5 cm					
	F		11 m	DS	3 cm					
	M		2 y	A	8 cm					
2022 Liao	F	term	1 d	S	3 cm	ulceration, vessel congestion	edema, ganglion cells in submucosal plexus	ganglion cells in myenteric plexus	subserosa edema and lymphoplasma cells, neutrophils infiltration	

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in complex colonic atresia. Also, a primary repair similar to pyloroplasty was reported in a relative benign and early detected colonic stenosis [20]. Notwithstanding the debate on following surgical planning according to the atresia of left or right part of colon, in recent case reports, segmental resection and primary anastomosis, which is not related to particular segment of the colon, is preferred and warranted. The clinical outcome and long-term survival rate of primary anastomosis was showed to be very good in the latest decade. But exception was still noticed in comorbidities of children with concurrent complicate anatomical anomaly like abdominal wall defect or illness like sepsis at presentation. And there were more than 10% of complication in colonic atresia including late adhesive bowel obstruction, prolonged ileus, stoma prolapse, and wound infection [21]. Another emphasis of caliber mismatch between two ends leading to morbidity were seen in some case series before. The current surgeons reestablished colonic continuity with end-oblique anastomosis to minimize the mismatched lumen.

Submucosal fibrosis and varying maturation of intestinal scar are seen on resected stenotic specimens. A case report observed the submucosal granulation and fibrosis were compatible with several weeks of duration [22]. However, the significance of pathological findings such as submucosal collagen deposition is not known. We collected narrative pathological reports from published articles of both acquired and congenital colonic stenosis since 1970 (Table 1) [14,16,22-32]. Most of the stenotic segment were abundantly infiltrated with mixed inflammatory cells (neutrophils, lymphocytes, histiocytes) and fiber deposition in different layers from mucosa to serosa. The muscle layer was reported hypertrophy and thickening muscle layer. Hyperplastic capillaries were noticed with thicken vessel wall. Nerve plexus and ganglion cell were noticed throughout the specimens.

4. Conclusion

Colonic stenosis is generally surgically treated. As improvement in colorectal surgery, up-to-dated management of colonic atresia and stenosis yielded less morbidity and mortality. In the article, we collected pathological description from articles of congenital colonic stenosis since 1970. We presented the data in a comparative table. Infiltration of mixed inflammatory cells and fiber deposition in different layers of stenotic segments. Nerve plexus and ganglion cell were noticed throughout the specimens.

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