Case Report

Congenital Pouch Colon with Segmental Dilatation of Ileum: Congenital Pouch Colon Type 6

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Congenital pouch colon (CPC) is classified on the basis of anatomic morphology into five types. Congenital segmental intestinal dilatation associated with anorectal malformation (ARM) is very rare. We are adding two neonates of CPC type 2 associated with segmental dilatation of the ileum to single similar case described in the literature till date. Temporary stomas with excision of either of the pouches should be considered. “Defective mesenchyme formation during organogenesis” due to abnormal mesenchymal precursor cells is hypothesized as the cause for both ARM and different types of CPC.

Keywords: Congenital pouch colon, ileal pouch, type 6

Abstract

Congenital pouch colon (CPC) is classified on the basis of anatomic morphology into five types. Congenital segmental intestinal dilatation associated with anorectal malformation (ARM) is very rare. We are adding two neonates of CPC type 2 associated with segmental dilatation of the ileum to single similar case described in the literature till date. Temporary stomas with excision of either of the pouches should be considered.

Case Report

Case 1

A 5-day-old full-term male neonate weighing 2140 g, presented to us with complaints of absent anal opening, progressive abdominal distension, and vomiting. On examination, the general condition of the child was poor. There were visible veins, periumbilical redness, and visible bowel loops with marked abdominal distention. The perineum was flat, absence of anus with pigmentation at anal site. Abdominal radiograph (antero-posterior erect view) showed a large air-fluid level suggestive of CPC. Laboratory investigations reveal raised renal functions with serum urea 74 mg/dl and serum creatinine 0.94 mg/dl, unconjugated hyperbilirubinemia (total bilirubin 14 mg/dl), and raised qualitative CRP.

At exploratory laparotomy, double pouch was present, i.e., segmental dilatation of the terminal ileum (ileal pouch) with CPC type 2 as per Saxena–Mathur (SM) classification [Figure 1]. The vascular supply of distal pouch was from the superior mesenteric vessels. The middle colic and left colic vessels were absent and replaced by an abnormal arborizing vessels supplying the pouch colon and the intervening colon. The colovesical fistula from the distal pouch was divided and ligated. It was decided to excise the major portion of distal pouch with preservation of its small portion and ileal pouch; end pouchostomy (proximal part) was performed. Postoperatively stoma did not function, and the patient developed abdominal distention with features suggestive of peritonitis. On re-exploration, there was small perforation in the ileal pouch which was resected, and end ileostomy was performed. The histopathological examination of the specimen confirmed CPC.

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baby did not recover from sepsis, and the outcome was unfavorable.

**Case 2**

A 1-day-old preterm female baby, weighing 2100 g, born by normal vaginal delivery, presented with absent anal opening along with meconuria. On physical examination, the baby was hemodynamically stable. The abdomen was distended with palpable lump in the lower abdomen. Perineum examination revealed meconuria, small perineum, and absent anal opening. A preliminary diagnosis of persistent cloaca was made.

Laboratory investigations reveal physiological jaundice (total bilirubin 5.76 mg/dl) and normal qualitative CRP; abdominal radiographs suggested CPC. After preoperative optimization, laparotomy was performed with left hockey stick incision which revealed double pouch, i.e., segmental dilatation of the terminal ileum (ileal pouch) with CPC type 2 as per SM classification [Figure 2]. The vascular supply of the pouch colon was from abnormal arborizing vessels arising from the superior mesenteric vessels. There was duplication of the Mullerian structures along with hydrometrocolpos (both uteri); both the fallopian tubes were dilated and inflamed [Figure 2].

The fistula from the distal pouch was divided and ligated. Learning from the previous experience, both the pouches were resected and end ileostomy was performed. Tube vaginostomy was performed to decompress the hydrometrocolpos. Postoperatively stoma started functioning and the patient tolerated the feeds well. The histopathological examination of the specimen confirmed CPC. The outcome was favorable; the patient is waiting for definitive surgery.

**DISCUSSION**

CPC is a rare form of high ARM. The majority of cases of CPC have been reported from North India. CPC is an unusual abnormality in which a pouch-like dilated shortened colon of a varying degree is associated with high ARM. In males, the pouch usually terminates in a colovesical fistula just proximal to the bladder neck. In girls, the terminal fistula opens either into the urethra or in the vestibule or into the vagina or cloacal chamber.

CPC was originally divided into four subtypes (types I–IV) by Narasimharao’s classification, based on the length of the normal colon proximal to the colonic pouch. According to SM classification, CPC is divided into five types on the anatomic morphology as per the length of colon involved. In type 1 CPC,
the normal colon is absent, and the ileum opens into a pouch colon. In type 2 CPC, the ileum opens into a normal cecum that in turn opens into the pouch colon. Type 3 CPC is characterized by a normal ascending colon with the transverse colon opening into the pouch colon. Type 4 CPC (most common) is normal colon up to the level of the rectosigmoid pouch. Type 5 was first described by Mathur et al.[3] It was first reported by Chadha et al., who described it as segmental dilatation proximal to CPC.[4] Type 5 or double pouch colon, the rarest form, has been described as pouch-like (segmental) dilatation of the colon with intervening normal colon of varying length between the two pouches, the distal pouch opening into the genitourinary system with a fistula.[1,2]

Congenital segmental intestinal dilatation is a rare entity with approximately 150 cases reported in the literature.[6] Swenson and Rathauzer were the first to describe this entity (colon) in 1959.[6] It can involve any part of the bowel with ileum most commonly, followed by the colon, jejunum, and duodenum.[3,6] It often manifests as partial or total intestinal obstruction in the neonatal period and early infancy. Laparotomy reveals an isolated, dilated bowel segment without any evidence of obstruction or abnormal neural innervation.[6] The criteria for its diagnosis are (a) limited bowel dilatation with a 3–4 fold increase in size, (b) an abrupt transition between dilated and normal bowel, (c) no intrinsic or extrinsic barrier distal to the dilatation, (d) clinical picture of intestinal obstruction, (e) a normal neuronal innervation (presence of ganglion cells), and (f) complete recovery after resection of the affected segment.[6] The histology of the dilated segment confirms normal neurological innervation with a hypertrophied or thin muscle layer.[6] Both our neonates were having all these features.

Congenital segmental intestinal dilatation can be detected as an intraabdominal cystic mass on ultrasonography. Abdominal radiographs may reveal a large dilated bowel shadow with or without air-fluid level.[3] Both cases were diagnosed intraoperatively. About half of these cases have associated gastrointestinal tract anomalies, e.g., Meckel’s diverticulum, omphalocele, intestinal atresia, duplication cyst, and intestinal malrotation.[3,6] Its association with ARM is very rare, and there are only a few reports of segmental dilatation of colon associated with an ARM (including rectal atresia and CPC).[3] Segmental dilatation of colon associated with CPC is referred to as CPC type 5 or double pouch colon. The earlier single reported case in the literature describes segmental dilatation of the ileum associated with CPC type 4, whereas in our cases, CPC type 2 were present.[3] This association should be labeled as CPC type 6, a modification of SM classification (anatomical classification). In earlier case, it was categorized into CPC type 5,[3] which in contrast is the presence of two colonic pouches (SM classification).[3]

The treatment of congenital segmental intestinal dilatation is resection of the dilated segment and end-to-end anastomosis.[3] Temporary stomas with excision of either of the pouches should be considered in critical neonates with associated CPC.[3]

Etiopathogenesis of congenital segmental intestinal dilatation mentioned in the literature is (a) intrauterine vascular accidents, (b) external compression to the fetal bowel,[6] (c) ganglionic dysplasia, (d) localized vacuolization of the intestinal smooth muscle, and (e) disorders of the interstitial cells of Cajal as a contributing factor for localized myopathy.[6] The etiology of type 5 CPC has been hypothesized due to “vascular insult” with the formation of both proximal and distal pouch. This “vascular insult” theory cannot explain the simultaneous occurrence of congenital segmental intestinal dilatation and CPC with an intervening normal segment on the basis of vascular theory alone. This theory also fails to explain the presence of other associated malformations.

Characteristic features of congenital segmental intestinal dilatation such as abrupt transition to normal bowel (both proximal and distal), absence of tenia coli and haustration (colon), presence of ganglion cells, and abnormal vascular pattern are similar to pouch colon malformation, suggesting common pathology.[1,2,6] The embryological basis of different types of CPC and the presence of congenital segmental intestinal dilatation may be explained by “defective mesenchyme formation due to abnormal mesenchymal precursor cells during organogenesis,” leading to abnormal segment which would dilate to form the colonic pouch and congenital segmental intestinal dilatation.[2] Our theory may also explain the presence of multiple segmental dilatation of colon associated rectal atresia (ARM) in one of the reported cases.[3] This explains the presence of disorganized muscles in the muscularis layer of pouch colon, thinned out musculature (normal ganglionicosis in colonic tissue), hypertrophied mucosa, and abnormal vascular pattern of the pouch (histopathological features).[2] Thus, “defective mesenchyme” due to abnormal mesenchymal precursor cells is hypothesized as cause for both ARM and different variants of CPC, resulting in defective organogenesis at multiple sites in the hindgut and primitive cloaca.[2]
In conclusion, congenital segmental dilatation of the ileum with CPC is an extremely rare entity in neonates. Favorable outcome is present when pathological segments are excised and stoma is created. The embryological basis of congenital segmental intestinal dilatation with CPC may be explained by “defective mesenchyme formation during organogenesis,” which may also explain all the variants (associations).

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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