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Case Report

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Congenital pouch colon with colonic atresia- An unusual embryological association: A case report

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KEYWORDS

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Vascular insult

ABSTRACT

Background: Congenital pouch colon (CPC) is a rare variant of high anorectal malformation. More and more varied associations of CPC with other entities are being added to the literature.

Case presentation: A 1-day-old male baby presented to the emergency room with marked abdominal distension and absent anal opening. On exploration, the baby was found to have CPC (Type 2) with colonic atresia. This association has not been reported in English literature.

Conclusion: This is a report of a case of type 2 CPC with colonic atresia. To the best of our knowledge, such an association has not been reported so far. The final embryological outcome is dictated both by the topography and timing of vascular insult.

INTRODUCTION

Congenital pouch colon (CPC) is a rare form of high anorectal malformation (ARM) in which a part of or the entire colon is replaced by a pouch, with a fistula to the genitourinary tract. This entity bears maximum incidence in Northern India. It is more commonly found in males with a ratio of 3-4.3:1 (male: female).[1] Colonic atresia is itself a rare entity; with an incidence of 1 in 20,000 live births; and accounts for ~ 1.8-15% of intestinal atresias.[2] Association of CPC with colonic atresia has not been reported so far. We report the first case of CPC with colonic atresia.

CASE REPORT

A 1-day-old male baby, weighing 2100 grams, birth order 1, delivered by the normal vaginal route, to non-consanguineous parents, after 39 weeks of uneventful gestation. The patient presented to us with marked abdominal distension and absent anal opening. The mother had no history of any off-labeled drug intake, gestational diabetes, and congenital anomalies in the family. The baby cried immediately after birth and had an Apgar score of 10 at 5 minutes. In view of the absent anal opening, feeds were not initiated. The mother had antenatal follow-ups but was not diagnosed antenatally.

The patient was moderately dehydrated. The apex beat was located in the 5th intercostal space, mid-axillary line anteriorly. There was no evident cyanosis and murmur. Abdomen was distended. On perineal examination, buttocks were poorly formed and the anal opening was absent. External genitalia was normal, and the tip of the penis was meconium-stained (suggestive of meconuria).

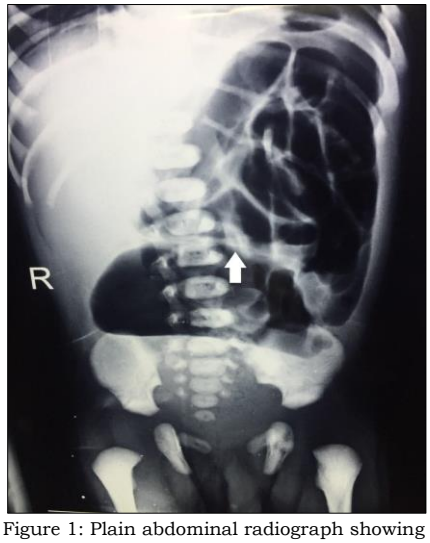

Figure 1: Plain abdominal radiograph showing dilated pouch with the apex directed towards left hypochondrium (marked with white arrow).

Routine investigations, namely complete blood counts, renal and liver function tests were within normal limits, and the septic screen was negative. The abdominal radiograph showed dilated colonic shadow, occupying more than half of the abdomen, suggestive of CPC type 2 as the apex of the pouch was directed towards the left hypochondrium (Fig.1).[3] Abdominal ultrasonography was suggestive of left hydroureteronephrosis; with normal right kidney. 2D echocardiogram was normal.

After preoperative optimization, laparotomy was performed via left "hockey stick" incision, and ileum was seen opening into caecum; which in turn opened into a colonic pouch measuring 12×10×10 cm; and the diagnosis of CPC was confirmed (Fig. 2a, 2b-shaded area). This pouch continued distally into a normal bowel (Fig. 2a, 2b-non-shaded area); which had distinct taenia coli and normal vasculature; but ended abruptly; with a V-shaped mesenteric defect (colonic atresia type III a) (Fig.2c). The proximal part of the dilated colon lacked haustrations, taenia coli and appendices epiploicae, had abnormal vasculature; and had a fistulous communication with the urinary bladder (Fig.2c). Ligation of colovesical fistula and excision of the atretic colon along with colonic pouch and ileocecal junction and; an end ileostomy was performed. The postoperative period was uneventful.

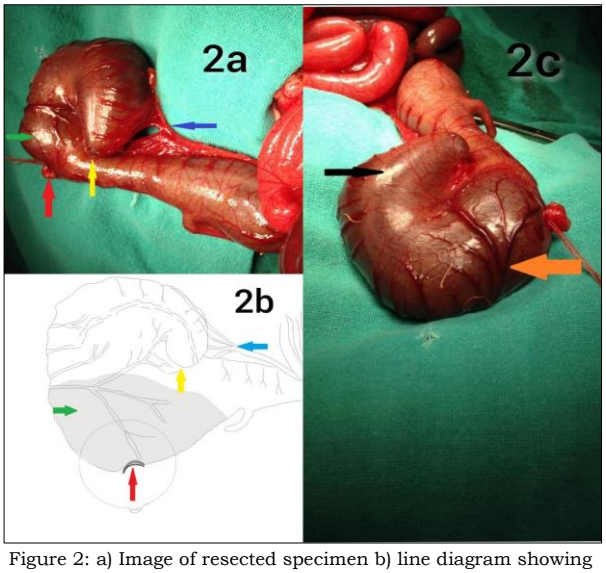

Figure 2: a) Image of resected specimen b) line diagram showing congenital pouch colon (green arrow) with divided genitourinary fistula (red arrow); terminating into an atretic colon (yellow arrow) with a mesenteric defect (blue arrow) c) image of the resected specimen showing distinct taenia coli in the atretic segment (black arrow) and; thickened and abnormal vasculature of pouch colon (orange arrow).

Histopathological examination of the resected specimen showed congested and hemorrhagic mucosa and submucosa disrupted muscularis propria, reduced number of mature ganglion cells, and congested and hemorrhagic serosa in proximal part (findings in favor of CPC) and normal mucosa and submucosa with undisrupted muscularis propria in distal part (findings in favor of colonic atresia).

Micturating cystourethrogram performed in the follow-up period; was suggestive of grade 2 left vesicoureteral reflux (VUR). Dimercaptosuccinic acid renal scan was suggestive of bilateral preserved cortical function; with no evidence of scars / pyelonephritic changes. Presently the patient is doing well over a close follow-up of 6 months and is awaiting definitive repair, and is also being managed conservatively for left VUR.

DISCUSSION

Narasimha Rao et al, are credited with giving us the most widely accepted classification of CPC.[4] They classified this entity into four subtypes based on the length of the normal colon proximal to the colonic pouch. Saxena and Mathur came up with another classification in 2008, which was based on anatomic morphology. They came up with type 5 CPC; which they described as "dumbbell-shaped", with a normal-looking intervening colon of variable length, with a distal pouch opening into the genitourinary tract through a wide fistula.[5]

More and more varied associations of pouch colon are being reported. There have been reports of Y-shaped duplication of the normal colon just proximal to the pouch.[6] There is also a report of CPC associated with an ileovesical and a colovesical fistula [7], CPC with rectal atresia [8,9], and CPC with double colovesical fistulae [6]; have also been reported. But the association of CPC with colonic atresia is unknown and to the best of our knowledge, the index case is the first to be reported. We have tried to decode the embryogenesis behind this unusual combination of malformations.

The exact embryogenesis of CPC is not known, but is best explained by combined Stephen's theory of incomplete downward growth of cranial fold of Tourneux; with failure of lateral fold i.e., Retterer fusion [10] and vascular insult theories (intrauterine obliteration of inferior mesenteric artery).[8] All these together lead to the defective formation of the mesenchymal components. This accounts for the lack of haustrations, taenia coli, and appendices epiploicae. To summarize, CPC is the result of vascular insults occurring in the early intrapartum period. On the contrary, colonic atresia could have been the result of vascular insult, in the late intrapartum period; after the organogenesis or mesenchyme formation is complete.[2] Hence, haustrations, taenia, and epiploicae are preserved in this entity.

The senior author is credited with reporting the first association of CPC with rectal atresia in literature.[7] After that, there have been sporadic case reports of

association of CPC with rectal atresia; and its embryogenesis has been ascribed to multiple vascular insults (obliteration of inferior mesenteric artery and branches of internal iliac artery).[8,9] This theory of multiple vascular insults holds true for the index case too; though at different times of the intrapartum period. The vascular accident may have occurred at the level of the right colic and middle colic artery (branches of the superior mesenteric artery) and left colic artery (branch of the inferior mesenteric artery) in the early intrapartum period. This along with defective mesenchyme formation explains the replacement of ascending, transverse and descending colon, with a pouch with colovesical fistula. We also hypothesize that sigmoid branches and superior rectal artery (branches of the inferior mesenteric artery) must have escaped insult during this period. Henceforth, leading to the growth of sigmoid colon with intact haustrations, taenia coli, and appendices epiploicae. However, it again suffered a blow during the late intrapartum period; arresting the further growth leading to colonic atresia. The presence of a mesenteric defect strongly validates this late vascular accident. We propose this hypothesis as the underlying embryological basis of the association of pouch colon with colonic atresia.

The presence of a proximal fistula has also been reported by Singh et al.[9] But this was seen in association with rectal atresia; whereas the index case is an association of CPC with colonic atresia with a proximal fistula; hence making it vividly different.

The vascular supply of the colon remains compromised with the obliteration of inferior mesenteric arteries. In other words, the type of pouch is governed by the length of the colon suffering ischemia. We also believe that if both the vascular insults would have occurred simultaneously; probably the outcome would have been different; maybe type 5 CPC.[11] In the index case, a part of the pouch escaped the vascular insult; and continued to grow; until it again suffered a blow in the late intrapartum period; resulting in CPC with colonic atresia.

With the above background, we would like to postulate that the final embryological outcome is dictated not only by the specific vessel suffering a blow; but also, by the temporal distribution of vascular insults in the intrapartum period.

In conclusion, CPC is a rare variant of high ARM. This is the first report of the association of CPC with colonic atresia. More and more atypical associations of CPC with other anomalies are being reported. The final embryological association is governed both by the topography and time of vascular insult.

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