



A Rare Case of Type 4 Congenital Pouch Colon without Fistula with External Anal Marking in a Female Subject

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Received: 13/05/2019

Accepted: 05/11/2019

Abstract

A 2-day-old female baby presented with radiological features of pouch colon, which was intra-operatively revealed to be type 4 congenital pouch colon (CPC) without fistula. The vestibule had two normal openings, with an external anal marking in the perineum with puckering of skin but without any caliber. She underwent excision of pouch with end stoma and is awaiting abdomino-perineal pull through when she reaches six months of age. To the best of our knowledge, no case of CPC without fistula with external anal marking and puckering has previously been reported.

Keywords: Congenital pouch colon (CPC); Anorectal malformation (ARM); Fistula

Please cite this paper as:

Kumar P, Jain R, Khatri A. A Rare Case of Type 4 Congenital Pouch Colon without Fistula with External Anal Marking in a Female Subject. *Ann Colorectal Res.* 2019;7(3):2-4. doi: .

Introduction

Congenital pouch colon (CPC) is a rare malformation as per the international Krickenbeck classification, in which the colon is partially or completely replaced by a pouch-like dilatation, opening into the urogenital tract via a large fistula. The association of CPC without a fistula and with anorectal malformation (ARM) is very rare. Rao et al. in 1984 described the following classification for CPC (1):

Type I: Absent large colon, with the ileum opening directly into the pouch.

Type II: The presence of a short segment of caecum that opens into the pouch.

Type III: The presence of a good length of colon

between the ileum and the pouch.

Type IV: The presence of a large colon with only the sigmoid and rectum being converted into the pouch.

Case Report

A 2-day-old female born of normal vaginal delivery, weighing 2.4 kg, came to us with non-passage of meconium since birth. Multiple episodes of bilious vomiting were reported, and abdominal distension was apparent on examination. The baby had signs of dehydration with a heart rate of 153/min and delayed capillary refill time. On local examination of perineum after retracting the labial folds, two openings were seen in the vestibule. There was



Figure 1: external marking of anus (without any lumen) with puckered peri-anal skin.

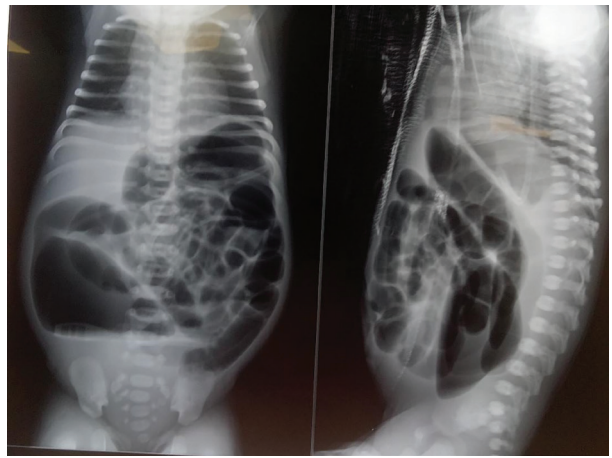


Figure 2: X ray abdomen showing large air fluid level.

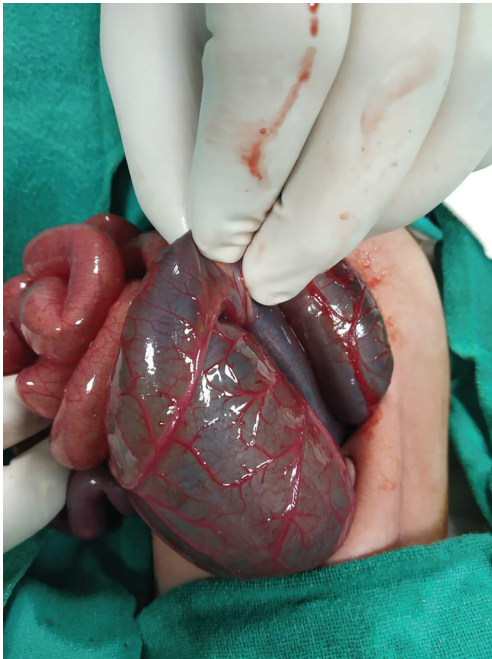


Figure 3: Sudden transition of colon with abnormal lease of blood supply



Figure 4: CPC without any fistulous communication

no septation of the vagina. There was an external marking of the anus without any lumen, slight puckering of peri-anal skin was observed (Figure 1).

A plain abdominal radiograph showed a large air fluid level on the right side of the abdomen encompassing more than half of it and displacing the small bowel to the left. There was no vertebral abnormality (Figure 2).

The baby was fluid resuscitated and then underwent exploratory laparotomy, which revealed CPC. A type 4 CPC was noted at the distal sigmoid colon with an abrupt transition and blind end without fistulous communication to the urogenital system. An abnormal lease of blood vessels from the superior mesenteric vessels could be seen. There was a single uterus with normal bilateral adnexa (Figures 3 and 4). The other abdominal viscera were normal. The pouch was excised and end colostomy was performed.

The histopathological examination revealed

absence of haustrations, appendices epiploicae and tenia coli, with congested serosa and pattern-less mucosa rugosities, along with some inflammatory and fibrotic changes [as evident on Masson's trichrome stain]. There was no muscular disarray, with normal anatomical location of ganglion cells.

Discussion

Congenital pouch colon is more common in northern India and its neighboring countries. Among the various hypotheses proposed concerning the etiology of CPC, the obliteration of the inferior mesenteric artery in early gestation causing missed or aborted hindgut development is well known. These patients always have abnormal blood supply with prominent superior mesenteric artery that supplies the whole distal bowel (1). The pouch communicates with the genitourinary system. In males, the fistulous communication is mostly at the bladder neck, and the

fistula is usually broad and thick walled, measuring up to 1cm in external diameter. In females, the fistula may be colocloacal, colovaginal or colovestibular (1-3).

Chadha et al. studied 21 female subjects of type 1, 2 and 3 CPC for various genitourinary abnormalities, out of whom 5 did not reveal any fistula, 11 had vestibular fistula, 4 had urethral fistula and 1 had perineal fistula (2). Also, all of their subjects had septate vagina and uterine didelphys. Our patient had a single uterus and a non-septate vagina, with two normal openings in the vestibule and an external marking of the anus without any lumen. In a previous study by Chadha et al., the anomalous clinical anatomy of 22 girls with CPC were evaluated, and the results were compared with various published series; the presence of fistula was reported in most cases. They also stated that the dilated pouch slowly tapered toward a fairly wide fistula accessible at laparotomy even in neonates and is not to be missed (3). The index case did not have any fistulous communication.

Udawat H et al. did a histopathological and IHC study on 49 subjects of CPC and enumerated the various findings of gross and microscopic

examinations, most of which were seen in our case (4). The pouch lacked haustrations, taeniae and appendices epiploicae. They attributed fibrotic changes in muscle wall of CPC to dysmotility issues. The inflammatory and fibrotic changes in CPC may be because of prolonged in-utero ischemia (5).

The management options for type 4 CPC include fistula division, pouchoraphy or excision of pouch with end stoma or primary pull through in a single stage. In our case, there was no fistula, and the pouch was excised and end stoma was made. The division of fistula (when present) should be the primary step in the first stage to prevent complications of uremia or hyperchloremic metabolic acidosis. Our patient is awaiting abdomino-perineal pull through at six months of age. Ghritlaharey and Budhwani reported no mortality with two-stage definite procedure (as planned in our case) in 11 patients of CPC (6).

To the best of our knowledge, there is no case reported of CPC without fistula and with external anal marking and puckering.

Conflict of Interests: None declared.

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