Unusual presentation of more common disease/injury

A newer variant of congenital pouch colon with rectal agenesis: management strategy and review of the literature

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Summary

A 3-day-old male baby presented with clinico-radiological features of rectal atresia with colo-urinary fistula. There was no radiological evidence of congenital pouch colon (CPC), but per-operatively CPC of unusual anatomy was found. The CPC was intermediate between type I and type II of Narsimha's classification. The pouch had rudimentary appendix and proximal colovesical fistula. The distal end of CPC was free. In the first stage, diverting ileostomy was done. In follow-up, after 1 month because of rectourinary fistula baby developed hyperchloremic metabolic acidosis and uremic sepsis. First metabolic correction and control of sepsis were done. Thereafter, abdomino-posterior sagittal approach was used for excision of colovesical fistula, appendectomy, coloplasty and colorectal anastomosis. The ileostomy was left undisturbed. The infant at present is waiting for the third stage ileostomy closure.

BACKGROUND

The congenital pouch colon (CPC) is an anorectal malformation (ARM) prevalent in northern India. There is a lot of literature available regarding its clinical presentation, classification and management. The rectal atresia is also a rare entity. But, association of both these anomaly is very rare. Only three cases had been reported in the literature. Our case is the fourth one.

Our case additionally had atypical features of proximal colovesical fistula which is not yet mentioned in the literature.

Exceptionally, in spite of a very large colonic pouch, x-ray abdomen had not showed any described features of CPC. The reason was a large colovesical fistula through which all feces and air were decompressed as meconium and pneumatocoele.

Figure 1 Perineum showing normal anus and perineum.

Figure 2 Cross-table x-ray with Hagar dilator in anal canal showing high anorectal malformation.

We also thought that ligation of colovesical fistula should be a mandatory procedure at the first stage of surgery, because if left untreated, it had complications which significantly increases the morbidity and mortality of the baby.

CASE PRESENTATION

A 3-day-old full term male baby, weighing 2.1 kg presented with pneumaturia, meconiumuria and non-passage of stools through anus since birth. Abdomen was soft and non-distended. Perineum and anus were normal, but red rubber catheter could not be passed through anus beyond 3 cm as resistance was found beyond it (figure 1). After clinic-radiological diagnosis of rectal atresia, laparotomy was done. On exploratory laparotomy CPC of abnormal anatomy was found. Diversion ileostomy was done. In postoperative period, baby was unable to micturate because of diversion of urine into the pouch through large colovesical fistula. Baby was catheterised and discharge with it in situ.

INVESTIGATIONS

Haemogram and blood biochemistry were normal. X-ray erect abdomen did not show any features of CPC. Contrast enema revealed rectal atresia (non-passage of dye beyond 2 cm). The cross-table prone x-ray view with Hagar dilator in anal canal showed high ARM with 2 cm gap between intraluminal gas and the dilator (figure 2). Thus, the radiography supported the clinical diagnosis of rectal atresia. Echocardiogram and ultrasonography of the abdomen did not show any other anomaly. Postoperatively to find out the cause of urinary retention, voiding cystourethrogram (VCUG) was done, but it showed large colovesical fistula only (figure 3).

TREATMENT

Because baby was of low-birthweight, so staged surgery was planned. After adequate resuscitation and nasogastric tube placement, exploratory laparotomy was done under general anaesthesia, through left lower quadrant transverse incision. There was no evidence of normal colon, instead whole colon was replaced by $15 \times 10 \times 6$ cm³ pouch. The ileum was opening directly into the CPC. Adjacent to ileal opening a $2 \times 0.5 \times 0.53$ cm appendix like structure was arising from the pouch. There was no evidence of cecum. A large colovesical fistula was found proximally adjacent to ileal opening. The fistula was opening at the urinary bladder trigone. The distal end of pouch was free (figure 4). The pouch had minimal meconium and gas, because it was decompressed completely as meconiumuria and pneumaturia through a large proximally situated colovesical fistula. The pouch colon had very short mesentery and poor vascular arcade.

In the first stage, diverting loop ileostomy was done. In the second stage, through abdomino-posterior sagittal anorectoplasty (A-PSARP) approach; colovesical fistula excision, appendectomy, coloplasty and colorectal anastomosis were done. The specimen of appendix was confirmed by histopathological examination. The baby is waiting for ileostomy closure.

OUTCOME AND FOLLOW-UP

In postoperative period, as the urinary catheter was removed, the baby was not able to micturate per urethra, because through a large colovesical fistula urine diverted into colonic pouch. This was confirmed by passing catheter into distal stoma through which significant urine was obtained and VCUG. The baby was re-catheterised and

Figure 3 Peroperative photograph showing rudimentary appendix (left lower horizontal arrow), ileum opening in pouch (right horizontal arrow), proximal colovesical fistula (upward directed vertical arrow), urinary bladder (downward directed vertical arrow), distal blind ended pouch colon (left upper arrow).

Figure 4 Micturating cystourethrogram showing colovesical fistula. Antero-posterior view (a), lateral view (b).

parents were trained for pouch wash through the distal stoma.

In follow-up, after 2 weeks catheter was removed. One month later, the baby presented to us with tachypnea,

dehydration, refusal of feed, decreased urine output and failure to thrive. On re-catheterisation gross pyourea was found. The urine culture showed heavy growth of the *Escherichia coli* and fungal elements. Arterial blood gas

analysis suggested hyperchloremic metabolic acidosis. Baby was resuscitated. As the metabolic derangements and sepsis were settled, the second stage surgery was done in same sitting of admission. The baby is now waiting for ileostomy closure.

DISCUSSION

The first description of CPC given by Spriggs (1912).¹ Narsimha Rao *et al* in 1984 suggested the term CPC syndrome.¹ Anatomical details and classification of CPC were given in details by Wakhlu, Chadha, Narsimha Rao *et al*.¹⁻³ Mostly the incidence of rectal atresia is 1–2% incidence. But in some southern part its incidence was reported as high as 14%.⁴

The proposed aetiology of CPC was early intrauterine insult of inferior mesenteric artery. This theory also explains association of rectal atresia with CPC, as the rectum develops normally because of its supply by branches of internal ileac artery.^{1,5}

The CPC is usually associated with anorectal agenesis, but its association with pure rectal agenesis is very rare. Only four cases of this association have been reported in the literature.⁵⁻⁷ We have reported the fifth case of this association, which additionally has newer variants of the CPC.

The CPC appears in x-ray abdomen as a single large loop of bowel with single air fluid level occupying more than half width of the abdomen and displacing the small bowel to one side (usually right). These features were not evident on x-ray because air and meconium were decompressed through proximal fistula as pneumatua and meconium.

The pouch was transition between type 1 and type 2 of Narsimha's classification; as the cecum could not be differentiated, but small rudimentary appendix was arising from the pouch.¹ To the best of our knowledge and extensive search of the literature, the unusual features present in our case (rudimentary appendix, proximal colovesical fistula and distal blind end) had never been reported in the literature.

Because of proximal colovesical fistula, short mesentery and poor vascular arcade of CPC conventional coloplasty described by Chiba *et al*, could not be done.⁴ To combat that, the nature had provided the baby well-formed anal canal, so coloanal anastomosis was done easily through Pena's posterior sagittal approach.

Mathur *et al* (2002), Kazaz *et al* (2009) and Parelka (2010) had reported about the association of CPC with the rectal atresia. In none of these cases proximal urinary fistula or the rudimentary appendix were found.⁵⁻⁷ The pouch found in these cases were type II, and IV, respectively; but our case could not be classified in any category. The first case had intraoperative cardiac arrest.⁵ The second case underwent single stage abdomino-transanal endorectal pull-through. In the second case, the pouch was excised, with transanal endorectal mucosectomy and endorectal coloanal anastomosis. In this case, sacropexy of colonic tube was also done.⁶ In the third case, through A-PSARP approach, pouch was excised and coloanal anastomosis was done. In the third case, there was no colovesical fistula.⁷

Surgical options described in the literature to manage this association are either primary or staged procedure. The primary procedure is fistula ligation, coloplasty and abdomino-transanal endorectal pull-through and coloanal anastomosis.⁵⁻⁷ The staged repair proposed were excision of colonic pouch with ileostomy; window colostomy; end colostomy; transverse colostomy; ligation of fistula procedure alone or combined with coloplasty in the first stage followed by definitive surgery as abdomino-PSARP or endorectal pull-through and coloanal anastomosis preserving native anal sphincter complex.^{2,5-7} In low-birthweight baby and in the developing countries where resources and expertise are still deficient, staged surgery is better than primary procedure. The author suggested that colovesical fistula ligation should always be done in the first stage to prevent the lethal complications such as urinary track infection, hyperchloremic metabolic acidosis.⁸

Learning points

- ▶ Staged surgery is always better than primary procedure, especially in low-birthweight baby to increase their survival.
- ▶ During colostomy in anorectal malformation during the first stage surgery, entire abdomen exploration should be done to plan the next definitive surgery.
- ▶ Colo-urinary fistula should be ligated at the first stage to decrease the risk of hyperchloremic metabolic acidosis and urosepsis, as both these entity increases the morbidity and mortality of the neonates.
- ▶ The Pena's posterior sagittal approach is a good alternative of transanal endorectal repair for rectal atresia.
- ▶ The CPC colon is a challenging entity for the paediatric surgeons. Its management is further complicated by anatomical variations. So, this field still requires newer thoughts and efforts for the better management.

Competing interests None.

Patient consent Obtained.

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