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A Rare Case of Anorectal Malformation having Pouch Colon with Duplication of Vermiform Appendix

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Abstract: Duplication of vermiform appendix is an extremely rare condition, with a reported incidence of 0.004%. Abnormal development of the appendix usually takes the form of a double appendix. Accompanying intestinal, genito-urinary or vertebral malformations may be present when appendiceal duplications are detected in childhood. We are presenting a case of neonate having ARM with peritonitis which on exploration found to be a case of perforated pouch colon with duplication of vermiform appendix and rectourinary fistula.
Keywords: ARM, VACTERL, Appendicular duplication, Pouch colon.

INTRODUCTION

Anorectal malformation (ARM) is avery common congenital disorder having incidence of 1 per 5000 live births [1] which also associated with multiple other congenital anomalies for example VACTERL group of anomalies. Congenital pouch colon and duplication of vermiform appendix combined with ARM is one of the rarest associations which are found in single patient.

Duplication of vermiform appendix is a very rare condition, having incidence of 0.004% [2]. Mostly the anomalies of appendix are found incidentally during surgery.

CASE REPORT

A full term, male baby of birth weight 1.9 kg on his 3rd post-natalday was transferred from community health center to the Pediatric NICU with provisional diagnosis of "imperforate anus" having tense, distended abdomen. There was no family history of any congenital malformation, consanguinity and no history of any medication intake by mother during anti-natal period. Physical examination revealed absence of anal opening with meconium staining of diaper, suggestive of any recto-urinary fistula. Routine compete blood count and biochemical test revealed no abnormalities except slight increase in WBC counts. The plain abdominal radiograph (Fig. 1) shows dilated loop of bowel on left side with bowel gas was above to level of pubic rami and no bony deformity was present.

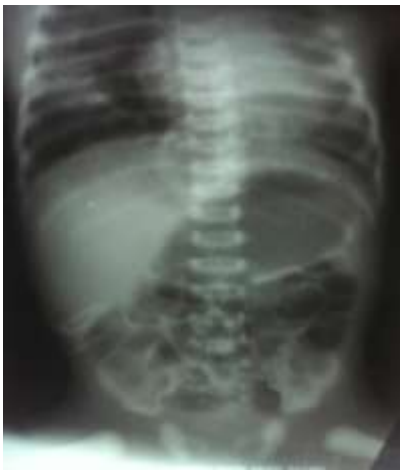

Fig. 1: Image showing plain X-Ray abdomen

Patient underwent emergencyexploration, which showed dilated, shortened and mobile, colonic segment which was perforated and with two vermiform appendix with separate base arising from caecumm (Fig. 2-4). A window colostomy by exteriorization of the perforated colon segment was done and both appendices were left as such.

On second post op day colostomy was functioning.

Patient was discharged from NICU on 10th post-operative day.

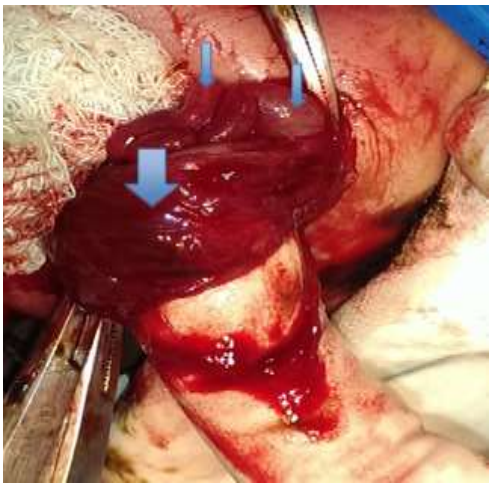

Fig. 2: Thin arrows shows appendices and thick arrow shows perforated pouch colon

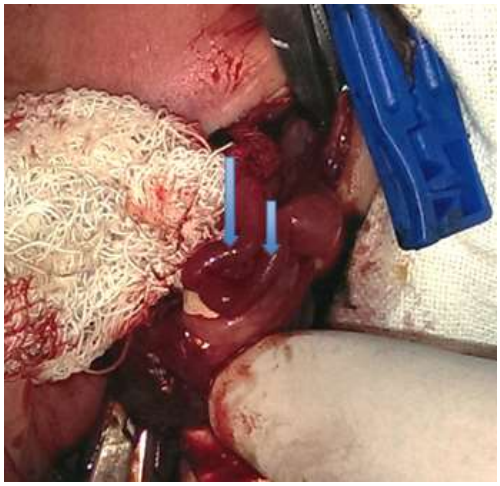

Fig. 3: Both arrows shows appendix

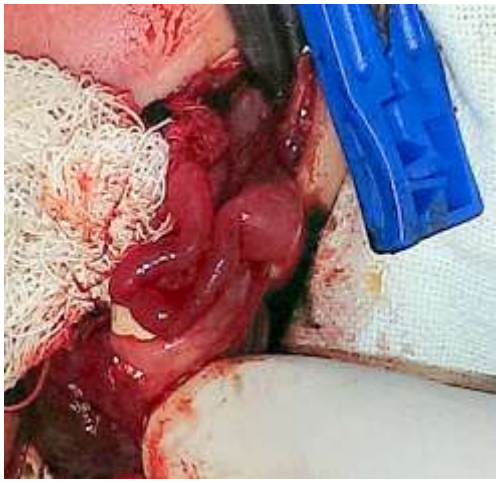

Fig. 4: Showing two appendix arising from caecum with separate base

DISCUSSION

It is observed that the condition having ARM associated with pouch colon mainly reported from Indian subcontinent and its incidence in northern India has been reported between 5-10% [3].

In this condition the colon is replaced by a pouch-like dilatation that communicates distally with the urogenital tract by a large fistula [4]. The pouch differs from a normal colon structurally, histologically and functionally. The management involves a diversion colostomy at birth with or without the excision of pouch followed by pull through [5].

Congenital anomalies of appendix are rare [6], there are two more common anomalies of appendix: congenital absence and appendicular duplication [7].

Appendicular duplication is the condition which is mostly diagnosed incidentally on laparotomy or contrast study of GI system done for other clinical situations preoperatively [6]. These patents usually present when they became symptomatic as a result of any inflammation or due to any obstruction [8]. Appendicular duplication may present as a constricting lesion of the ascending colon and mimic a colonic adenocarcinoma [9].

The Appendicular duplication first described by Picoli (1892) in a female child [2]. Appendicular duplication was classified first by Cave [7] in 1936 and modified by Wallbridge [10] in 1963 then it again modified by Biermann in 1993 as follows.

- Type A: Single caecum with one appendix exhibiting partial duplication.
- Type B: Single caecum with two obviously separate appendices.
  - B1: The two appendices arise on either side of the ileocaecal valve in a “bird-like” manner.
  - B2: Normal positioned appendix with a second, usually rudimentary, appendix arising from caecum along the lines of the taenia at a varying distance from the first.
  - B3: The second appendix is located along the taenia of the hepatic flexure of the colon.
  - B4: The location of the second appendix is along the taenia of spenic flexure of the colon.
- Type C: Double caecum, each bearing its own appendix.

Some times a horseshoe anomaly of the appendix was also described by some authors which is considered as type D anomaly [11].

Duplication of appendix treated mainly by appendectomy of the one appendix then second appendectomy was done after 2-3 years [12].

Although it is seen quite rarely, in patients with a history of appendectomy and clinical complaints suspicious of appendicitis, appendicular duplication must be kept in mind, and ultrasonography, computed tomography and when required diagnostic laparoscopy should be performed. This is important for patient health and medicolegal problems.

Chadha *et al*. [13] have described the association of appendix duplication with pouch colon and anorectal malformations in 6 patients of their series.

Kothari *et al*. [8] reported a case of appendix duplication associated with imperforated anus. But no detailed informations were given to explain the pathogenesis.

CONCLUSION

The exact pathogenesis is not known, but some point in the management of appendiculardupli cation has been clarified: (i) especially type B duplications, where the second appendix base may lie at any point along the colon, present the greatest risk of missing the second appendix; (ii) appendicular duplication may also present as a constricting lesion of the ascending colon and mimic a colonic adenocarcinoma; (iii) some cases of duplicated appendix are associated with intestinal, genitourinary, or vertebral malformations, and this needs to be considered after diagnosis.

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