Congenital pouch colon anorectal atresia combined with appendix duplication complicated by intestinal volvulus

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**A B S T R A C T**

Congenital pouch colon (CPC) is a rare abnormality with few reports in Europe, frequently detected in the Indian Peninsula, where it constitutes 15% of all anorectal malformations (ARM). This report presents a congenital pouch colon case combined with appendix duplication (AD) further complicated by midgut volvulus and intends to place emphasis on their concurrence.

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Anorectal atresia is a rare congenital malformation which can be classified as high or low, with or without a fistula and its type can be easily diagnosed even by simple observation [1,2]. Congenital pouch colon is a very rare form of anorectal malformation which has been described in a small number of cases in literature mostly occurring in North India (a supposition involves environmental factors with deficiency of iodine or vitamin B, pesticides) [3] and was classified as a rare regional variant of anorectal malformations, at the 2005 Krickenbeck meeting [4]. Rectal atresia combined with congenital pouch colon and appendix duplication has also rarely been described in literature [1,5,6]. Appendix duplication represents approximately 0.006% of the reported appendectomy series worldwide and is usually accompanied by intestinal, genitourinary and vertebral malformations or combinations of these [5]. The aim of this case report is to emphasize on the concurrence of other life-threatening anomalies, such as malrotation, and to discuss the preoperative diagnosis and surgical approach of congenital pouch colon and appendix duplication.

1. Clinical report

A full-term first-born 3650 g male neonate was delivered by caesarian section. He was of Greek origin with parents born in Zhambyl, a south eastern region of Kazakhstan less than 1000 km from the North Indian Peninsula. During prenatal 2nd trimester echo screening, an abdominal mass was detected and a possible diagnosis of a mesenteric cyst was made. Clinical examination in the delivery room revealed anorectal atresia with dilatation of the left abdomen, therefore the neonate was referred to the neonatal unit of our hospital. During hospitalization, the anomaly was confirmed as rectal atresia with a normal rectal imprint, scrotal-perineal ribbon and a well-formed middle buttock line. The rectal reflex was produced typically at the location of the imperforated anus. Meconium and gas passed through his urethra indicating a fistula between the bowel and the bladder, despite the fact that the clinical and X-ray findings suggested low-type atresia. The ultrasound examination revealed high atresia with great dilatation of “the rectum and sigmoid colon” a solid substance inside the bladder and a normally formed urinary tract and coccyx.

The neonate was prepared for surgery with the indication “high atresia” and a disconnecting sigmoidostomy was planned to be performed. The abdomen wall was accessed through a 3 cm left lower quadrant Lanz incision which was extended to 4 cm due to findings. That mini laparotomy was adequate to complete the operation at that time. The cecum was mobile and a pouch, part of the dilated large intestine, 8 x 8 x 10 cm in size, was found at the left lower abdomen (Fig. 1).

This dilated part of the colon was very difficult to be mobilized due to its poorly developed mesentery as it was supplied mainly by an arcuate extension of the superior mesenteric artery.

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The proximal end was linked to 3 cm of normal ascending colon while the peripheral part communicated with the dome of the bladder through a 4-mm-wide fistula (Fig. 2).

The cecum was normal and two appendices were present. Both were well formed, separate and symmetrical to the ileocecal valve with different mesoappendices (Fig. 3).

The dilated colon was excised, an ascending colostomy was performed 2 cm distal to the ileocecal valve, and both appendices were preserved. The bladder was closed in two layers and a 6 Fr urethral catheter was inserted. The postoperative period was uncomplicated and ventilator weaning was achieved some hours after surgery. The urethral catheter was removed two weeks later and the child was discharged with instructions to return to hospital at the age of 3 months to have his intestine continuity restored. A month later, when his body weight had increased to 5560 g, he developed a crisis of acute abdominal pain, refusal of feeding, abdominal dilation and vomiting. The child was admitted to hospital with symptoms of acute abdomen requiring urgent laparotomy. A midgut volvulus was found. The blood supply of the involved intestine was provided by the superior mesenteric artery, as the inferior mesenteric artery was absent. The only viable intestine extended 130 cm distal to the Treitz ligament and it was preserved. The other part till the colostomy including the cecum was excised. An end-ileostomy was performed. Postoperatively he developed malabsorption as a consequence of the short gut syndrome and he received combined oral and total parenteral nutrition. Intestinal element nutrition gradually increased with a parallel decrease of the parenteral nutrition which eventually ended several months later.

2. Discussion

Congenital anomalies of the appendix are rare and the majority of them are either appendix duplication or absence [5]. Appendix duplication is usually detected during imaging tests or during laparotomy [1,5,7]. Symptoms appear in cases of inflammation or perforation [5]. The types of appendix duplication were first described by Cave in 1936 [5]. The Cave-Wallbridge classification describes three types, our case belonging to the “bird-like” type B1 [1]. Congenital pouch colon is also classified in four groups based on the length of the remaining well-formed colon [1,7–9]. Our case belongs to type II. This finding is consistent with the few similar cases in literature in which appendix duplication was found mostly in type II congenital pouch colon [1,5]. The appendix is formed...
during the descent of the ascending colon and is a residue of herbivorous animals [10]. The pathogenesis of appendix duplication is unclear [5]. Embryological anomalies of the appendix may appear once in one’s career. More specifically, types B1 and C are related to other malformations of the intestinal and genitourinary systems as they are in our case [5]. Yanar et al. explain the possible relevance between anorectal-genitourinary malformations and appendix duplication due to cloaca separation [11]. The rectovesical fistula is the result of a developmental inability of the genitourinary system to be separated [10]. Imaging and clinical findings of a rectovesical fistula with air-fluid level inside the bladder indicate high atresia, which, as Alberto Pena contends, cannot be combined with a lower defect. Nevertheless, in our case the clinical findings (well-formed perineum, bottom and rectal imprint) came to antithesis with the presence of the rectovesical fistula and the radiologic investigation, which revealed high atresia. The surgical approach and strategy of restoring bowel continuity depend on the type of anomaly and is still a matter of discussion in literature [4,12]. In our case, excision of the pouch was performed instead of pouch colostomy, pouch-plasty and abdominoperineal pull-through described by other authors. This method was preferred due to the size of the pouch, its inability of mobilization and the lack of normal peristalsis that literature describes [4]. The rarity of malrotation presence in congenital pouch colon, 13 cases out of a total 470 reported with congenital pouch colon worldwide [4], cannot support the need for extensive exploration during the management of atresia. The option for wide laparotomy should be reserved for cases where radiologic or operative findings indicating malrotation are present. In conclusion this rare case indicates that when radiologic and clinical findings are not in accordance a wide laparotomy gives a better surgical aspect for diagnosis and treatment. Rare cases of anorectal malformations should also be part of the diagnostic range. When found randomly during surgical operation should be treated with minimal invasive techniques that secures patient’s viability but also allows a reoperation after reviewing the literature for similar cases.

Compliance with ethical standards

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. Informed consent was obtained from all individual participants included in the study.

Conflicts of interest

None.

References