Complete tubular duplication of colon presenting as rectovestibular fistula: A case report

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ABSTRACT

Background: Duplication of the gastrointestinal tract is a rare congenital disorder in pediatric patients. Complete resection in case of a total tubular duplication that requires total or subtotal colectomy is undesirable in children.

Case Presentation: This case presented a simple surgical technique for treating complete colonic duplication without colonic resection. A 2 months old female baby with abdominal distention and stool complaint came out from her vaginal vestibular and normal anus. A digital rectal exam showed a palpable mass in the posterior region. Abdominal CT Scan showed a rounded mass with size 2x2 cm in the left posterolateral of rectum region. The initial diagnosis was suspected sacrococcygeal tumor type 4 with rectovestibular fistula. Intraoperative findings showed duplication of the caecum, appendix, total colon (ascending to sigmoid) and rectum (one directed to the vagina and the other to anus). The ileostomy was done 20 cm from the ileocaecal junction as a temporary treatment to relieving distention with definitive repair planned in the following months. The second stage repair was performed after 1-year-old with posterosagital anorectoplasty followed by distal separation of the duplicated colon using a 12 cm linear stapler. For this case, the third stage repair was done one year after the previous surgery to close the remaining ileostomy. The patient’s outcome showed good results with normal defeating function and no complication.

Conclusion: Complete tubular duplication is a rare case without specific clinical symptoms, but the assessment of the diagnosis can be assisted by radiology such as abdominal CT and barium enema. We hope this article could illustrate management for complete tubular duplication of colon presenting as rectovestibular fistula, thus reflect how important clinical judgment and initial diagnostic in our health care system.

Keywords: Colon Duplication, Surgery, Repair.

INTRODUCTION

Colon duplication is a congenital disorder that occurs in 0.2% of the pediatric population. Malformations are rarely found in the large intestine (only 5-6%) and can occur along the gastrointestinal tract, with a complete tubular form which is very rare. Clinically, colonic duplication is usually asymptomatic, making it difficult to diagnose before surgery. Therefore this malformation can only be confirmed during a laparotomy. The clinical features of colonic duplication vary depending on the location, mucosal layer involved, and the lesion’s extent. More than 80% of cases present before two years of age as acute stomach or intestinal obstruction, but it can occur at any age. However, malformations in adulthood are increasingly difficult to diagnose because there is no specific clinical presentation.

The treatment option for colonic duplication is complete resection of the duplication in the long tubular colon. The procedure also requires total or subtotal colectomy because of the joint blood supply of the native bowel and duplication. The sphincter complex may need reconstruction, but this procedure is not used in cases involving children. Simple surgical techniques for the treatment of complete colonic duplication are described.

We report a case of complete tubular duplication of the colon presenting as a rectovestibular fistula with abdominal distension. Duplication starts from the caecum, showing two caeca and two appendixes and two complete tubular colonic formations. There is one normal anus and rectovestibular fistulae. Given the low incidence and unique clinical presentation of this anomaly, we hope publishing our case will increase diagnostic precision and improve surgical treatment.

CASE REPORT

A 2 months old female baby was admitted into the Emergency Room (ER) with complaints of abdominal distention, and stool came out from her vaginal vestibular and normal anus. No history of same condition in family members. Physical examination revealed signs of an acute abdomen with a large accumulation of fluid between the bowels. A digital rectal exam showed a palpable mass in the posterior region. The abdominal x-ray did not show any conclusive findings. Abdominal CT Scan showed a rounded mass with size 2x2 cm in the left posterolateral of rectum region. Other examination within the normal limit. The initial diagnosis was...
suspected sacrococcygeal tumor type 4 with rectovestibular fistula. A precise preoperative diagnosis could not be established, so an urgent laparotomy was performed.

The patient underwent exploration laparotomy. Intraoperative findings showed duplication of the caecum, appendix, total colon (ascending to sigmoid) and rectum (one directed to the vagina and the other to anus). The ileostomy was done 20 cm from the ileocecal junction as a temporary treatment to relieving distention with definitive repair planned in the following months.

One year later, the second stage repair was performed with posterosagital anorectoplasty followed by distal separation of the duplicated colon using a 12 cm linear stapler. At the next year, the third stage repair was done to close the remaining ileostomy. The patient’s outcome showed good results with normal defecating function and no complication at clinic visit after 1 month after last surgery.

DISCUSSION

Colonic duplication is the most unusual type of intestinal tract duplication, and the complete tubular form is even less common. Several studies have shown a high association between colonic duplication with prostate rectal fistulas, anorectal or scrotal abnormalities, and myelomeningoceles present in the genitourinary or lower vertebral column.\(^9\),\(^10\) The pathogenesis of these malformations is unclear but is said to be multifactorial concerning molecular genetics.\(^11\)

From a morphological perspective, the most common type of anomaly is cystic duplication along the colon, followed by partial tubular duplication, and very rare literature shows cases of complete cecoappendicular duplications such as in this patient.\(^9\),\(^11\) The most interesting aspect of this case was its clinical presentations: acute abdominal distension and rectovestibular fistula, which are very rare because they are usually asymptomatic in children and found in adulthood. The lack of specific clinical features of these malformations remains a serious diagnostic dilemma.\(^9\),\(^12\)

Digestive tract duplication is very difficult to diagnose at a preoperative time, but ultrasound can be used to help make a preoperative diagnosis as well as a screening tool to treat 10-20% of multiple lesions. Imaging using a chest or abdominal CT and barium enema can also help in making the diagnosis. A barium enema examination can be used to assist in the diagnosis of duplication of the tubular colon, with opacification of two colons being the diagnostic sign.\(^6\),\(^7\)

Several kinds of literature describe various surgical techniques for the management of colonic duplication. Riedel operated on Grohe’s case by dividing the septum between the two rectum and the tissue between the anal canal and reconstructing the perineum.\(^13\) Another study performed a lateral anastomosis between two sigmoids, then the sigmoid leading to the fistula is divided under the anastomosis by leaving the intestinal sac to allow the mucus to be expelled through the fistula.\(^14\) A similar technique was used in the case described by Sarpel et al. as recently as 2005.\(^15\) Most of the literature suggests that complete excision of duplicate channels is the preferred surgical technique.\(^10\)

In this case, we performed a distal separation from the colonic duplication using 12 cm linear stapling. After dividing the septum between the normal and duplicate distal colon, the rectum acquires a single lumen, and making evacuation easier. Large communication between the duplicated colon and the normal colon is required to ensure the emptying of the duplicated stool and ensure the passage of the stool, thereby relieving constipation.\(^7\) The study by Yucesan prefers to construct a common channel, using mechanical sutures for common wall resection to avoid accumulation feces in the duplicated intestine.\(^16\) Complete Tubular Colon Duplication has low mortality of around 4-8%. Mortality is generally associated with severe malformations or malignant transformations. Duplication of the total tubular colon treated before complications occurs generally has a good prognosis, so early diagnosis is a very important aspect.\(^12\)

CONCLUSION

Complete tubular duplication is a rare case without specific clinical symptoms, but the assessment of the diagnosis can be assisted by radiology such as abdominal CT and barium enema. This article illustrates a two months old female baby with complaints of abdominal distention, and stool came out from her vaginal vestibular and normal anus. The patient received 3 stage repairs. The first repair is ileostomy. The second stage repair is posterosagital anorectoplasty followed by distal separation of the duplicated colon, and the third stage repair to close the remaining ileostomy. We hope this article could illustrate management for complete tubular duplication of colon presenting as rectovestibular fistula, thus reflect how important clinical judgment and initial diagnostic in our health care system.

CONFLICT OF INTEREST

All of the authors declare that there were no conflicts of interest in this study.
CASE REPORT

PATIENT CONSENT

The patient’s parents had agreed and signed informed consent regarding publishing the case in an academic journal without exposing his child’s identity.

FUNDING

None.

AUTHOR CONTRIBUTION

Substantial contributions to conception and design, or acquisition of data, or analysis and interpretation of data: Muntadhar M. Isa, Amir Thayeb, Maria Meildi, Muhammad Bayu Z. Hutagalung

Drafting the article or revising it critically for important intellectual content: Muntadhar M. Isa, Maria Meildi, Muhammad Bayu Z. Hutagalung

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