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Total Colorectal Duplication in an Infant Presenting with Fecaluria as the Initial Symptom

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Introduction

Alimentary tract duplication can occur anywhere throughout its entire length. Colonic duplication is a rare subtype, constitutes about 17% of cases (1) and may present in a wide variety of ways. The diagnosis is often a challenge thus, some patients can remain undiagnosed or misdiagnosed for years (2). We report a case of an infant with total colorectal duplication and rectourethral fistula who manifested with a unique presentation of fecaluria. The diagnosis, however was not made until later. He presented again at 11 months of age with progressive abdominal distension that prompted further evaluation before arriving at the correct diagnosis and management.

Case Report

An 11-month old boy presented with two-month history of progressive abdominal distension. He was hospitalized during the neonatal period for orchitis when he developed testicular swelling and scrotal redness with evidence of urinary tract infection. At that time, there was a concern of a possible rectourethral fistula after he was noticed to pass frank faeces per urethra. He underwent ultrasound of the abdomen and pelvis followed by contrast enema and voiding cysto-urethrogram (VCUG) but did not show any obvious abnormality. He was discharged and planned for elective examination under anaesthesia and cystoscopy later. However, his parents were not keen to proceed afterwards as he had spontaneous resolution of the symptom.

He became asymptomatic until about 9 months of age when he started to develop progressive abdominal distension. He could still pass motion regularly but reduced in frequency. There was no recurrence of fecaluria or vomiting.

On presentation, his abdomen was grossly distended. Rectal examination revealed a single anal opening with a smooth, firm mass anterior to rectum that pushed the rectum posteriorly. He underwent ultrasound of the abdomen followed by a repeat contrast enema and VCUG that showed presence of large filling defect in between the bladder and sigmoid colon, compressing on these structures. There was no obvious urinary fistula seen. He then underwent CT of the abdomen and pelvis which revealed a long tubular structure from splenic flexure to anorectal junction in close relation to the native colon, grossly dilated and filled with fecal material which was highly suggestive of colorectal duplication.

He underwent laparoscopic-assisted fenestration and stapling of the distal-most common wall of the duplicated rectum the next day. Intra-operatively, there was a long tubular duplication of the entire colon and rectum, arising from mesenteric border. It was filled with faeces and the native rectum was displaced postero-laterally. No obvious fistula was visualized. The blind ending, obstructed duplicated bowel was then opened at its distal-most point, evacuated and irrigated. The common wall between the distal part of the duplicated rectum and the native rectum was then divided with a linear stapler-cutter to form a common lumen.

He recovered well after surgery, discharged after two days and followed up as an outpatient. Up until two years after surgery, he has remained asymptomatic.

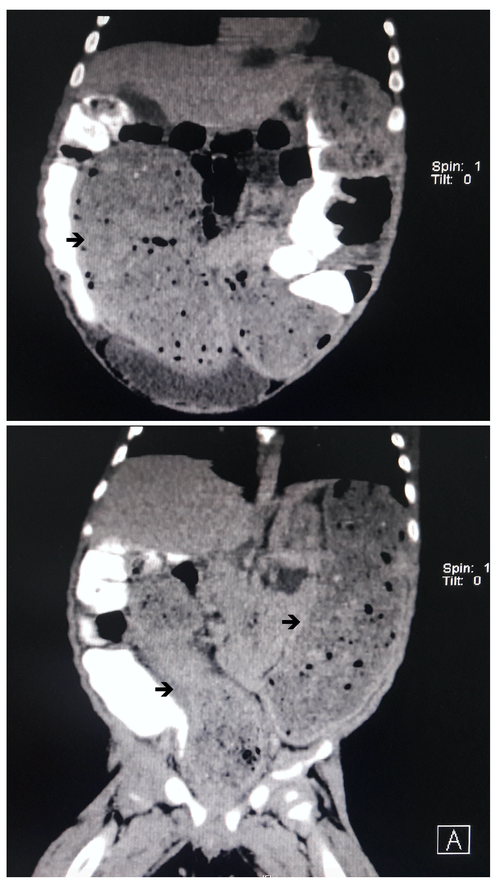


Figure 1 & 2 CT abdomen and pelvis (coronal image) with rectal contrast opacifying part of large bowel. There is a long tubular structure from splenic flexure until anorectal junction filled with fecal material (black arrow), compressing the native colon and pushing the rectum posterolaterally to the right. No demonstrable contrast was seen within the lesion thus unable to demonstrate communication to the native bowel or urinary system. Both kidneys and bladder (not shown in this image) were normal.

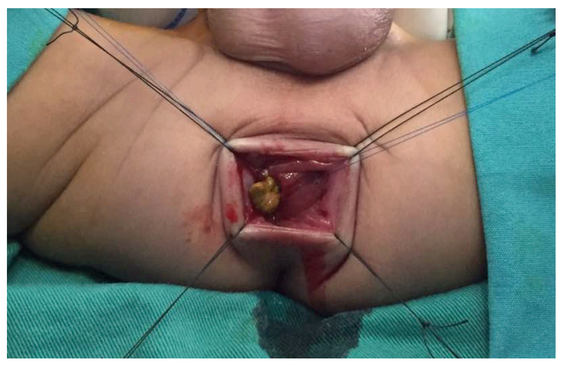
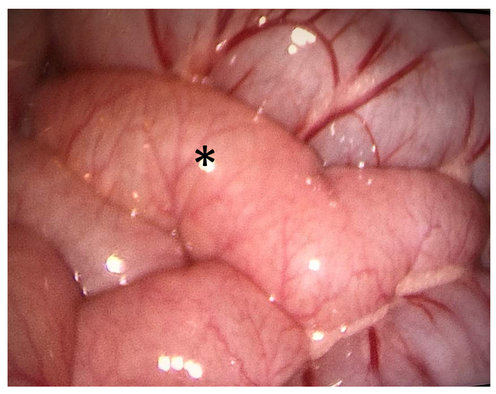


Figure 3 Intra-operative finding of soft tissue mass anterior to rectum which prolapsed through anus on suprapubic compression. Incision at the distal part of the blind-ending mass exposed fecal material that had been retained.



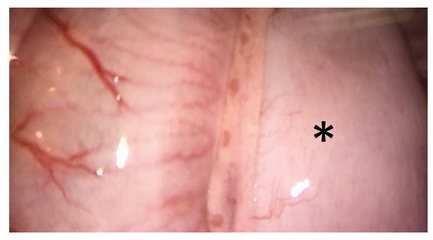


Figure 4 & 5 Laparoscopic view showed colonic duplication at the mesenteric border adjacent to the native colon. Duplicated bowel (asterisk) appeared to be less vascular with no haustration.

Discussion

The presentation of patients with colonic duplication varies depending on the size, location, and mass effect. Unlike some cases of gastric and small bowel duplication which can be detected by antenatal ultrasound (3), patients with colonic duplication usually present in the postnatal period with vague abdominal pain, constipation, failure to thrive or rectal bleeding when associated with ectopic gastric mucosa (1). The diagnosis is not usually straightforward and often confused with other gastrointestinal pathology.

Its association with genitourinary fistula has been described in the literature. Among the earliest descriptions was an analysis by Yousefzadeh et al. in 1983 (4). From their study, 50% of patients were reported to have some form of genitourinary fistula. Despite this high incidence, the number of patients who presented with initial symptoms of fecaluria or pneumaturia are uncommon (1). The cause of the development of fistula is not well described, however it is believed to be related to the long-standing pressure of the duplicated segment on the bladder wall or urethra caused by fecal impaction (5). However, this theory is not applicable to our patient as he presented in the early neonatal period with fecaluria, before the onset of the long-standing fecal impaction. This presentation suggests that the development of fistula took place early in the course of illness.

Other authors described their series of patients with similar condition who underwent abdominal-perineal exploration in order to locate and ligate the fistula (6–9). However, we opted for a less invasive approach. As he no longer had fecaluria after the neonatal period with negative pre-operative VCUG, we postulated that there had been spontaneous resolution of the fistula, or that the fistula was too small to be demonstrable. In the latter circumstance, the fistula was likely to be at its distal most part, in this case the rectum (4). Therefore, the act of uniting common wall would allow evacuation through the common anus and eventually divert the fecal stream away from the fistula. As alimentary tract duplication commonly associated with complications, surgical intervention is recommended (10). Simple short segment duplication warrants surgical excision. However, complete resection for patients with long tubular duplications would require significant colectomy due to the shared blood supply and common wall (1). Therefore, division of the distal part of the common wall to form common lumen (11) as performed in our patient, is an acceptable surgical option in order to preserve bowel length.

Conclusion

Diagnosis of colonic duplication may be challenging as the presentation can be confused with other gastrointestinal pathology. This case report highlights the importance of having a high index of suspicion for colorectal duplication in patients presenting with isolated fecaluria. We would also like to highlight the less invasive option of distal fenestration and limited division of the common wall as a viable alternative to a major, potentially morbid procedure of total colectomy.

References

1. Lund DP. Chapter 90: Alimentary tract duplications. In: Pediatric Surgery (7th Edition). 2012. p. 1155–63.
2. Puligandla PS, Nguyen LT, St-Vil D, Flageole H, Bensoussan AL, Nguyen VH LJ. Gastrointestinal duplications. J Pediatr Surg. 2003;38(5):740–4.
3. Laje P, Flake AW, Adzick NS. Prenatal diagnosis and postnatal resection of intraabdominal enteric duplications. J Pediatr Surg. 2010;45(7):1554–8.
4. Yousefzadeh DK, Bickers GH, Jackson JH Jr BC. Tubular colonic duplication: Review of 1876-1981 literature. Pediatr Radiol. 1983;13(2):65–71.
5. Payne CE, Deshon GE, Kroll JD SJ. Colonic duplication: An unusual cause of enterovesical fistula. J Urol. 1995;46(5):726–8.
6. Kokoska ER,Steinhardt GF, Tomita SS WT. Prostatorectal fistula associated with tubular colorectal duplication. J Pediatr Surg. 1999;34(10):1546–8.
7. Sengar M, Gupta CR, Jain V, Mohta A. Colorectal duplication with prostatorectal fistulae. J Pediatr Surg. 2013;48(4):869–72.
8. Karkera PJ, Bendre P, D'souza F, Ramchandra M, Nage A PN. Tubular colonic duplication presenting as rectovestibular fistula. Pediatr Gastroenterol Hepatol Nutr. 2015;18(3):197–201.
9. Costa EC, Ferreira CT, Salle JLP, Fraga JC. Diagnosis and management of congenital rectourethral fistula in a child with long tubular duplication of the colon and Klippel-Feil syndrome. J Pediatr Surg. 2011;46(11): 2184–6.
10. Temiz A, Oğuzkurt P, Ezer SS, Ince E, Gezer HÖ, Hiçsönmez A. Different clinical presentations, diagnostic difficulties, and management of cecal duplication. J Pediatr Surg. 2013;48(3):550–4.
11. Nolan HR, Wengler C, Hartin CW, Glenn JB. Laparoscopic excision of an ascending colon duplication cyst in an adolescent. J Pediatr Surg Case Reports. 2016;4:32–4.