Terminal Ileum and Total Colonic Duplication Associated with a Rectovestibular Fistula in a Child

Abstract:
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Abstract
The presence of terminal ileum and complete colonic duplication associated with a rectovestibular fistula in a child presents an extremely rare diagnostic and management conundrum. We report our surgical approach to successfully correcting this anomaly.

Introduction
Complete duplication of the colon is extremely rare. Herein, we discuss one such case highlighting the diagnostic difficulties and the pros and cons of different surgical options.

Case Report
A 7 day old female of West African origin was referred with a history of passing stools via her vagina. Physical examination revealed a normal anus and a rectovestibular fistula. A rectal duplication was suspected. Abdominal ultrasonography and a micturating cystourethrogram were normal. A contrast enema and subsequent fistulogram were non-diagnostic for the nature and extent of her duplication. A loop sigmoidostomy was required at 10 months old due to an obstructing distal fecaloma. After the occurrence of a stoma prolapse, it was observed that both proximal and distal limbs of the sigmoidostomy had dual lumens. Contrast was then instilled through both distal lumens showing duplicated distal colons (Figure 1). Magnetic resonance imaging confirmed a complete colonic duplication (Figure 2) but the precise anatomy of the caecum and terminal ileum could not be outlined.

At laparotomy, the anterior colon terminated as a rectovestibular fistula and proximally had a caecal diverticulum and 5 appendixes. The posterior colon terminated as the normal anus. The terminal ileum was duplicated, each entering a duplicated caecum. After taking down the sigmoidostomy, a mucosectomy of the distal anterior rectum was performed as far down to the vestibular fistula as possible; the seromuscular layer of the distal rectum below the peritoneal reflection was then closed leaving a small rectal stump. Because both colons shared a common wall (Figure 2) and blood supply, a linear stapler was used to divide the common wall thus forming a stapled anastomosis single lumen colon. The duplicated terminal ileum, caecal diverticulum and all appendixes were excised and an ileoleostomy was created between the distal ileum and the remaining terminal ileum that entered the caecum of the posterior colon. Finally, a proximal ileostomy was fashioned. After ileostomy reversal she initially reported mucus discharging per vagina but this subsided. Apart from constipation and an episode of bowel obstruction requiring adhesiolysis she remains well at 8 years old.

Discussion
The features described in this case probably form part of the spectrum of caudal duplication syndrome, whereby structures derived from the cloaca and notochord are duplicated. Duplications of the external genitalia, genitourinary system, hindgut and vertebra have all been infrequently reported. One theory for the embryological cause is a defect in the caudal cell mass during the first trimester. Classification systems proposed for these hindgut duplications depend on the presence or absence of a communication between both colons, a perineal fistula, or a rectovestibular and rectourethral fistula in females and males respectively. A Contrast enema and a fistulogram performed concomitantly, preferably with contrast media of differing densities, should delineate both colons. It is often difficult to identify the proximal extent of the duplication; however, upper gastrointestinal contrast follow-through studies have been reported to facilitate the diagnosis. Depending on the features present, surgical options for managing hindgut duplications include a total colectomy of both colons. Proponents of this approach reference the reported malignancy risk and risk of heterotopic mucosa in intestinal duplications as justification. It remains undetermined whether there truly is an increased incidence of malignancy in duplicated colons compared to native colons; however, bleeding due to the presence of gastric mucosa in a colonic duplication has now been recently reported. A single lumen colon, as described in this case, can also be created. A mucosectomy and transection of the fistulating rectum below the peritoneal reflection is then performed transabdominally, or a posterior sagittal approach can be used to excise the fistula, this prevents mucus discharge per vagina from a residual rectal pouch. Finally, total mucosectomy or resection of the duplicated colon while retaining the native colon has been described. This option avoids the morbidity when the colonic blood supply is not intimately related to that of the duplication. This option avoids the morbidity of a total colectomy while eliminating the reported risks posed by the duplication.

In conclusion, these clinical features described should raise the suspicion of total colonic duplication. Associated anomalies should be investigated for and the outcome after a stapled anastomosis of both colons can be favourable.
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References

Comments:
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