



Case Report

Duplicate Appendix With Acute Ruptured Appendicitis: A Case Report

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Duplication of the appendix is a rare congenital anomaly that, in adults, is most often found incidentally during surgery for other reasons. Appendicitis in the duplicated appendix is very rare and has been reported less than 10 times in the medical literature. We describe a 33-year-old woman with worsening periumbilical pain, nausea, vomiting, and fever. Physical examination showed localized peritonitis in the right lower quadrant. She had an elevated white blood cell count with neutrophilia. Computed tomography showed acute ruptured appendicitis. Diagnostic laparoscopy showed 2 appendices attached via separate bases to a single cecum with no other concurrent anomalies. Both appendices were removed laparoscopically. Histopathology confirmed normal appendiceal tissue in one and severe acute transmural appendicitis in the other. Awareness of appendiceal duplication and a thorough intraoperative inspection are critical to assess the presence of significant associated anomalies and avoid life-threatening complications.

Key words: Ruptured appendicitis – Appendiceal duplication – Acute abdomen – Cave-Wallbridge classification

Acute appendicitis is one of the most common causes of emergent abdominal surgery in the United States (US).¹ The lifetime risk for acute appendicitis is 8.6% in males and 6.7% in females in

the US.² Anatomic anomalies complicating its diagnosis and management are rare. The anomalies known to complicate management include high-lying appendix, malrotation, situs inversus, and the

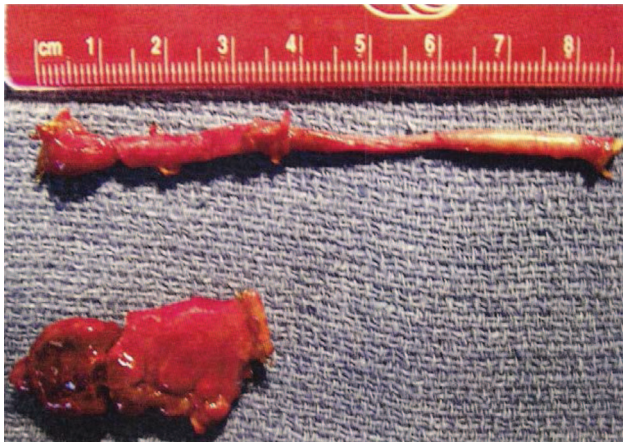


Fig. 1 Normal appearing appendix (specimen A on top); inflamed appendix with congested surface, fibrinopurulent exudate on serosa, and prominent vessels (specimen B on bottom).

duplication of hindgut structures, including the cecum and appendix. Less than 100 appendiceal duplications have been documented in the literature to date,³ of which less than 15 are cases complicated by acute appendicitis, including the case we present herein.

Case Report

A 33-year-old African-American female presented to the Emergency Department with migratory right lower quadrant pain of 4 days duration associated with nausea and vomiting. Clinical evaluation showed that she was febrile and had a tender right lower quadrant with rebound tenderness and guarding. Her white blood cell count was elevated with neutrophilia. Computed tomography scan of the abdomen showed possible acute appendicitis with perforation and purulent fluid collection in the pelvis. Diagnostic laparoscopy revealed 2 appendices attached via separate bases to 1 cecum. The

anterior appendix was inflamed with a gangrenous tip while the posterior appendix was grossly normal (Fig. 1). Both appendices were located on the taenia coli. No other intra-abdominal anomalies were discovered. Subsequently an emergent laparoscopic appendectomy was performed. Histopathology confirmed normal appendiceal tissue in one specimen and severe acute transmural appendicitis in the other (Fig. 2). There were no postoperative complications. The patient was discharged home in stable condition after 3 days in the hospital.

Discussion

Duplication in the digestive tract is a rare complication in adults since more than 80% of cases present as an acute abdomen or bowel obstruction before the age of 2.⁴ Appendiceal duplication is extremely rare with an incidence of 0.004% to 0.009% in appendectomy specimens.^{5,6} Appendiceal duplication can be isolated or associated with cecal duplication. Appendiceal duplications were first classified in 1936 by Cave,⁷ updated in 1962 by Wallbridge,⁸ and then again by Bierman in 1993.⁹ They classified the major kinds of anomalies into Types A, B, C, and D. Type A has a single cecum with various degrees of incomplete appendiceal duplication. Type B has a normal appendix that arises from the cecum and is subdivided into 4 separate types depending on where the second appendix is located. Type B1 (avian-type appendix) arises symmetrically from the other side of the ileocecal valve; this resembles the arrangement in birds. Type B2 (tenia coli type) arises anywhere along the lines of the tenia coli. Type B3 arises from the hepatic flexure,⁹ and Type B4 arises from the splenic flexure.⁶ Type C is a duplication of the cecum, each containing an appendix. Type D, the most recently described type by Mesko *et al* in 1986,¹⁰ is a horseshoe appendix—a single appendix

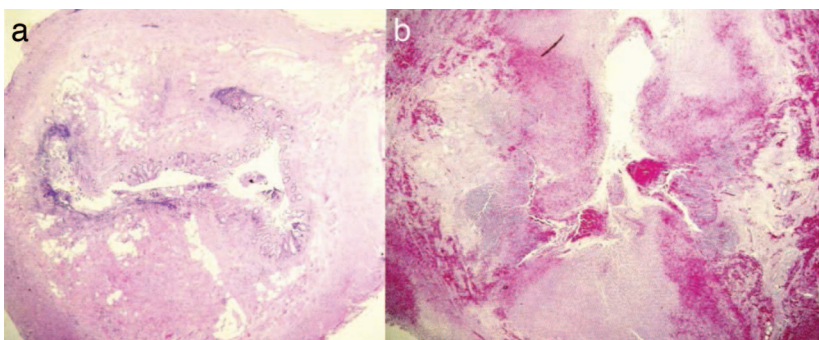


Fig. 2 Histologic sections of appendices. (Left) Specimen A normal appendiceal tissue. (Right) Specimen B severe acute transmural appendicitis.

Table 1 Modified Cave-Wallbridge duplicate appendix classification scheme. Modified from Coker *et al*¹²

	Alias	Cecum	Appendices	Embryology	Associated anomalies
Type A		Single	(1) Partial duplication	Partial fusion of transient appendix-like structure with the normal appendix precursor	None
Type B1	"Bird-like" or "avian type"	Single	(2) Completely separate	Failure of proper differentiation of the cloaca	Anal and/or colonic atresia, ectopic bladder, anomalies of the external genitalia, characteristic communication between the most distal small bowel and bladder
Type B2	"Taenia-coli type"	Single	(2) Completely separate	Persistence of transient appendix-like structure	None
Type C		Double	(2) One per cecum	Partial twinning of the hindgut	Hindgut duplication which can involve terminal ileum, double colon, anus, uterus, vagina, external genitalia, bladder, lower vertebral column
Type D	"Horse-shoe appendix"	Single	(1) Two openings into common cecum		

with 2 openings into the cecum. A case of appendiceal triplication has also been reported.¹¹ Coker *et al*¹² suggested an embryologic etiology for these anomalies (Table 1).¹³

Suspicion of a duplicated appendix should prompt further investigation into the possibility of other congenital anomalies, including duplications or anomalies of the gastrointestinal tract or genitourinary tract,¹⁴ gastroschisis,¹⁵ and vertebral anomalies,^{5,16} especially in type B1 and C cases.¹⁷ Type B2 duplication is not known to be associated with any other congenital anomalies (Table 1).¹⁷

The case described above falls under the Type B2 classification as there were 2 separate appendices attached via separate bases onto the cecum. The duplicate appendix was located over the taenia-coli. Type B2 is the most frequently reported duplication of the appendix.¹⁸

Reports of appendicitis in patients with a duplicated appendix are rare in the literature, however, it is likely that many cases are missed simply because the second appendix was never identified. Retrocecal appendices and type B duplications are most likely to remain unnoticed.¹⁷ Identification of the anomaly can be further confounded when the clinical presentation mimics other conditions, such as adenocarcinoma of the colon,¹⁹ small bowel obstruction, volvulus, or intussusception.²⁰ In cases where appendiceal du-

plication is suspected, histopathologic examination must show a complete and separate inner circular and outer longitudinal muscle layer and the appropriate amount and arrangement of lymphoid tissue to differentiate a duplicate appendix from an appendiceal or solitary cecal diverticulum.²¹

Appendiceal duplication should be in the differential diagnosis when a clinical diagnosis of acute appendicitis does not correlate with the operative finding of a normal looking appendix. Although a very rare entity, ensuring its absence would prevent diagnostic ambiguity. It is recommended that if multiple appendices are located, all of them should be removed to avoid any diagnostic ambiguity in the evaluation of abdominal pain in future. This would also avoid any medicolegal ramifications of a "previously removed" appendix.²²

Although appendiceal duplication occurs very rarely, awareness of this congenital anomaly and thorough intraoperative inspection are critical to avoid the potential consequences of missing a second appendix, as well as any associated congenital anomalies, and to minimize confusion with other intra-abdominal structures.

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