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Diagnostic Challenge of Sequential Appendicitis in a Patient With Appendiceal Duplication: A Case Report

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Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Patient: Male, 33-year-old
Final Diagnosis: Appendicitis
Symptoms: Nausea • Pain
Clinical Procedure: —
Specialty: Surgery

Objective: Congenital defects / diseases

Background: Appendiceal duplication is an exceptionally rare congenital anomaly, with a reported prevalence of less than 0.003%, and remains poorly represented in the contemporary evidence-based surgical literature. Recent Cochrane reviews and PubMed-indexed meta-analyses (2024 onward) have refined diagnostic pathways and management strategies for acute appendicitis; however, congenital anatomical variants such as appendiceal duplication are largely excluded from these analyses. Among the described subtypes, type B2 duplication according to the Cave–Wallbridge classification is clinically significant due to the presence of 2 completely independent appendices arising from opposite teniae coli. This configuration carries a substantial risk of delayed diagnosis, missed intraoperative recognition, and sequential appendicitis, particularly in patients with a history of prior appendectomy.

Case Report: A 33-year-old man with a previous open appendectomy and histopathological confirmation of acute appendicitis presented 3 years later with recurrent right lower quadrant pain and signs of localized peritonitis. Cross-sectional imaging demonstrated free intraperitoneal fluid but failed to identify a definitive etiology. Exploratory laparotomy revealed a second, previously unrecognized retrocecal appendix located along the omental tenia, consistent with type B2 appendiceal duplication, complicated by acute inflammation and perforation. A retrograde appendectomy was performed. Comparative histopathological analysis confirmed acute appendicitis in both specimens, establishing the diagnosis of sequential appendicitis secondary to appendiceal duplication.

Conclusions: Appendiceal duplication, although exceedingly rare, is a clinically relevant cause of recurrent abdominal pathology after appendectomy. Careful intraoperative inspection of the cecum and its teniae is essential to avoid missed diagnoses. Increased awareness and reporting of this anomaly are necessary to integrate rare anatomical variants into evidence-based surgical practice.

Keywords: Anatomic Variation • Appendicitis • Appendix • Congenital Abnormalities

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/953106

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Introduction

Appendiceal duplication is an exceptionally rare congenital anomaly of the gastrointestinal tract and remains an underrecognized cause of diagnostic error in acute abdominal pathology. Acute appendicitis is the most common abdominal surgical emergency worldwide, with a lifetime risk of 7% to 8% [1]. A comprehensive review by Nageswaran et al further contextualizes this anomaly, describing fewer than 100 reported cases worldwide and emphasizing its considerable anatomical variability, most commonly classified under the Cave–Wallbridge system. The authors note that preoperative diagnosis remains exceedingly uncommon, as clinical presentation typically mimics standard acute appendicitis and imaging findings are often inconclusive. Consequently, most cases are identified intraoperatively, frequently after unexpected findings during exploration. The review also highlights that failure to recognize a duplicated appendix during the initial surgery can result in persistent or recurrent symptoms, necessitating reoperation and carrying potential medicolegal implications [2]. Recent evidence-based guidelines and meta-analyses published from 2024 onward have refined diagnostic algorithms and management strategies for acute appendicitis; however, congenital anomalies such as appendiceal duplication are largely excluded from these analyses, leaving a gap in evidence-based surgical guidance [3]. The reported prevalence of appendiceal duplication is estimated to be below 0.003%, based on data from large autopsy series and surgical pathology reviews of appendectomy specimens [4]. As a result, the current body of evidence remains largely limited to case reports and small case series, with a paucity of systematic investigations.

Despite its rarity, this condition carries significant clinical relevance because duplicated appendices are anatomically independent structures, each capable of developing inflammation, perforation, or other complications. Failure to recognize a duplicated appendix can result in persistent or recurrent symptoms following appendectomy, delayed diagnosis, increased risk of perforation, and avoidable morbidity [5].

From an embryological standpoint, appendiceal duplication is believed to arise from aberrant cecal development during early midgut rotation, leading to the persistence of more than 1 appendiceal outpouching [6]. Clinical manifestations are often indistinguishable from typical acute appendicitis, and preoperative diagnosis remains uncommon, achieved in fewer than 15% of reported cases, even with contemporary imaging modalities [7,8]. Recent case reports and small series continue to demonstrate that duplicated appendices are frequently identified intraoperatively, underscoring the importance of heightened surgical vigilance.

Given the absence of high-level evidence addressing this anomaly, increased reporting and awareness of appendiceal duplication are essential to improve recognition, optimize surgical management, and reduce the risk of missed pathology.

Case Report

A 33-year-old man presented with a 3-day history of right iliac fossa abdominal pain associated with nausea, vomiting, fever, and malaise. He had undergone an open appendectomy 3 years earlier via a Rocky–Davis incision, with histopathological confirmation of acute appendicitis at that time.

On physical examination, he was hemodynamically stable and in an antalgic position. A well-healed Rocky–Davis scar was noted. Abdominal examination revealed tenderness to both superficial and deep palpation in the right iliac fossa, with clear signs of localized peritoneal irritation. No palpable masses were identified.

Abdominal ultrasound demonstrated approximately 200 cc of free fluid in the right iliac fossa. Computed tomography revealed fluid collection in the pelvis and right iliac fossa without a clearly identifiable source. Laboratory evaluation showed leukocytosis and elevated acute-phase reactants.

Given the clinical signs of peritonitis, an exploratory laparotomy was performed. Intraoperatively, free fluid and localized contamination were observed, along with dense post-surgical adhesions surrounding the cecum. After performing Cattell's maneuver to mobilize the right colon, a second retrocecal, subserosal appendix was identified along the omental tenia coli, with a perforation in its middle third. A retrograde appendectomy was completed. The intraoperative identification of the duplicated appendix is shown in **Figure 1**.

Gross pathological comparison between the specimen obtained during the first appendectomy and the duplicated appendix resected during the second intervention is presented in **Figure 2**, demonstrating 2 anatomically independent appendices. Histopathological examination of both specimens confirmed transmural acute appendicitis, with perforation identified in the second appendix.

Preoperative imaging is shown in **Figure 3**, with axial contrast-enhanced pelvic CT demonstrating a large cystic lesion occupying the pelvic cavity. Initial laparoscopic visualization is presented in **Figure 4** prior to conversion, demonstrating a markedly distended appendix with inflammatory changes.

The postoperative course was uneventful. Comparative macroscopic and microscopic analysis definitively established the presence of 2 independent appendices, consistent with type B2 appendiceal duplication according to the Cave–Wallbridge classification.

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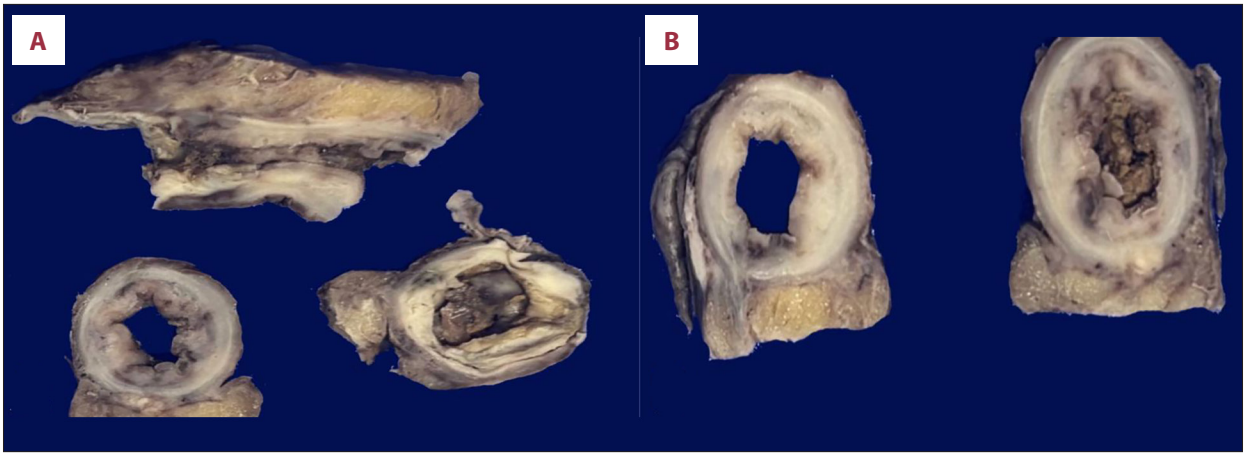

Figure 1. Sequential appendicitis in type B2 appendiceal duplication. (**A**) Macroscopic cross-sectional view of the appendix obtained during the first intervention (initial open appendectomy), demonstrating luminal dilation and transmural inflammatory changes consistent with acute appendicitis. (**B**) Macroscopic and sectional views of the second appendix resected during the subsequent intervention, identified retrocecally along the omental tenia coli. The specimen shows wall thickening, luminal obstruction, and perforation in the middle third, confirming acute appendicitis in the duplicated appendix.

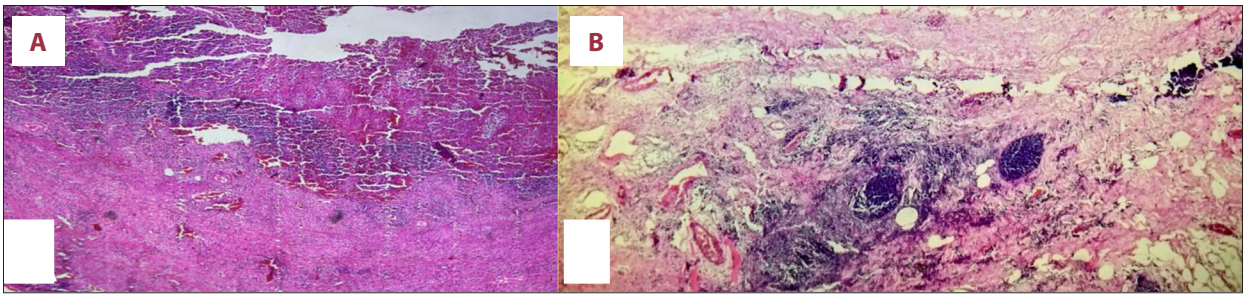

Figure 2. Histopathological confirmation of sequential acute appendicitis. (**A**) Hematoxylin and eosin-stained section of the appendix obtained during the first intervention, demonstrating extensive mucosal necrosis with loss of surface epithelium and diffuse acute inflammatory infiltrate predominantly composed of neutrophils. The inflammatory process involves the mucosa, submucosa, muscularis propria, and serosa, consistent with transmural acute appendicitis. (**B**) Histological section of the second appendix resected during the subsequent intervention, revealing similar transmural acute inflammatory changes with marked serosal congestion, fibrinopurulent exudate, and focal disruption of the appendiceal wall consistent with perforated acute appendicitis.

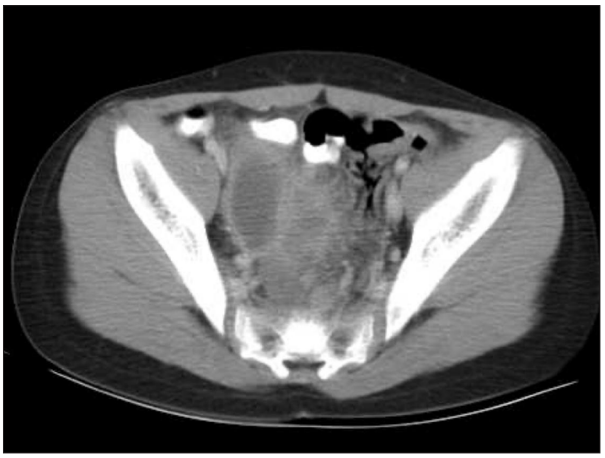

Figure 3. Axial pelvic CT with intravenous and oral contrast showing a large pelvic fluid collection with homogeneous low attenuation, consistent with inflammatory fluid or early abscess formation, causing mass effect on adjacent structures. Preoperative imaging is shown in Figure 3, demonstrating a large pelvic fluid collection consistent with an inflammatory process, most likely representing complicated free fluid or evolving abscess formation secondary to perforated appendicitis. These findings correlate with the intraoperative evidence of perforation and localized contamination.

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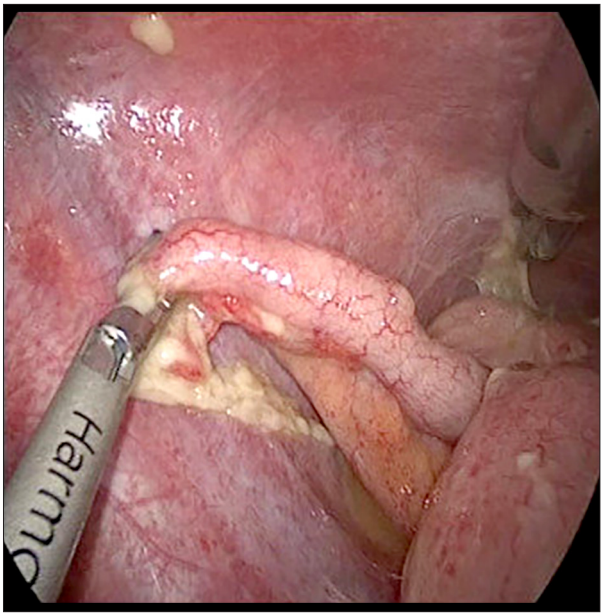

Figure 4. Laparoscopic image demonstrating a distended appendix with inflammatory changes. Laparoscopic view demonstrating a markedly distended appendix with thickened walls, congestive changes, and surrounding inflammatory tissue adherent to adjacent structures.

Discussion

Appendiceal duplication is an uncommon congenital anomaly with important diagnostic and surgical implications, particularly in patients presenting with recurrent right lower quadrant pain after previous appendectomy. Nageswaran et al emphasize that the main clinical challenge is not only its rarity but also the high risk of intraoperative oversight, especially in the presence of atypical anatomical locations or adhesions, which can obscure the accessory appendix. Because standard clinical scoring systems and conventional imaging algorithms are designed for normal appendiceal anatomy, rare variants can remain undetected preoperatively. In patients with prior abdominal surgery, adhesions and altered cecal orientation further limit radiologic interpretation, often restricting findings to indirect inflammatory signs rather than direct visualization of the anomaly [2,9]. In our case, these factors contributed to delayed diagnosis until surgical exploration was undertaken.

The Cave–Wallbridge classification remains the most widely accepted system for categorizing appendiceal duplication. It divides anomalies into type A (partial duplication of a single appendix) and type B (2 separate appendices arising from the cecum, with subtypes according to implantation site), and type C (duplication associated with double cecum). Type B2, identified in our patient, is characterized by a second appendix arising separately along the tenia coli in addition to the normal appendiceal origin [10]. This subtype is particularly relevant because the accessory appendix can be retrocecal, subserosal, or concealed by inflammatory adhesions, increasing the likelihood of being overlooked during the initial procedure.

The embryological independence of duplicated appendices explains the occurrence of sequential appendicitis, in which inflammation develops in the remaining appendix after prior appendectomy. Each appendix possesses independent luminal continuity, vascularization, and lymphoid tissue, allowing obstruction, infection, and perforation to occur separately [11]. This mechanism accounts for the perforated inflammatory process observed during the second intervention in our patient. Preoperative identification remains uncommon, although recent reports suggest that high-resolution computed tomography with multiplanar reconstruction can improve detection in selected cases [12].

From a surgical perspective, complete cecal inspection remains essential whenever operative findings appear disproportionate to the clinical presentation. Identification of the taeniae coli and adequate mobilization of the right colon may reveal accessory appendices hidden in atypical positions or obscured by adhesions. Failure to identify appendiceal duplication can lead to recurrent disease, repeat surgery, and avoidable diagnostic confusion [13].

Histopathological examination remains definitive for distinguishing true appendiceal duplication from stump appendicitis. In our case, independent pathological confirmation of 2 anatomically separate appendices with acute transmural inflammation established the diagnosis conclusively. Awareness of this anomaly is therefore critical for surgeons and radiologists when evaluating patients with persistent abdominal symptoms despite previous appendectomy.

Conclusions

Appendiceal duplication type B2 is a rare but clinically significant congenital anomaly that can manifest as sequential appendicitis, even years after a correctly performed appendectomy. This case highlights the diagnostic limitations of conventional imaging and clinical scoring systems when anatomical variants are present, reinforcing the need for heightened clinical suspicion in post-appendectomy patients with recurrent right lower quadrant pain. Thorough intraoperative evaluation of the cecum and all teniae coli remains essential to prevent missed pathology. Comparative histopathological confirmation of independent inflammatory processes is critical to differentiate true duplication from stump appendicitis. Systematic reporting of such cases is fundamental to integrating rare anatomical variants into contemporary evidence-based surgical practice and improving patient outcomes.

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Informed Consent

Written informed consent was obtained from the patient.

Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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