American Journal of Case Reports

e-ISSN 1941-5923
© Am J Case Rep, 2026; 27: e952870
DOI: 10.12659/AJCR.952870

Received: 2026.01.20
Accepted: 2026.08.19
Available online: 2026.09.02
Published: 2026.XX.XX

Duplicated Appendix With Mucocele Mimicking an Appendiceal Neoplasm in a 45-Year-Old Woman: A Case Report

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Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
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Financial support: None declared
Conflict of interest: None declared

Patient: Female, 45-year-old
Final Diagnosis: Duplicated appendix with mucocele
Symptoms: Pain abdomen
Clinical Procedure: Right hemicolectomy
Specialty: Gastroenterology and Hepatology • Surgery
Objective: Rare coexistence of disease or pathology

Background: Appendiceal duplication is an extremely rare congenital anomaly, with an incidence of 0.004% to 0.009%. An association with an appendiceal mucocele is even rarer. It can complicate the diagnosis and management of right iliac fossa pathology and remains difficult to identify preoperatively. Failure to recognize it can result in recurrent symptoms, missed pathology, intraoperative challenges, and medicolegal consequences. This report describes the case of a 45-year-old woman with history of a loculated peri-appendiceal abscess managed with percutaneous drainage and intravenous antibiotics, subsequently found to have a duplicated appendix with mucocele identified after right hemicolectomy.

Case Report: A 45-year-old woman presented with right lower quadrant abdominal pain and fever that began 2 months ago. Contrast-enhanced computed tomography demonstrated ruptured appendicitis with a loculated peri-appendiceal abscess, successfully managed with percutaneous drainage and intravenous antibiotics, resulting in clinical improvement. She re-presented 2 months later with recurrent abdominal pain. Diagnostic laparoscopy revealed a dilated, firm appendiceal tip and a mass-like ileocecal inflammatory complex, raising suspicion of malignancy. Due to the distorted anatomy and inability to exclude an underlying neoplasm, a right hemicolectomy was performed. Histopathological examination revealed a Type B2 (Cave–Wallbridge) duplication of the appendix, one of which showed mucocele formation and features of resolving appendicitis with no evidence of malignancy.

Conclusions: Appendiceal duplication is classified according to the Cave–Wallbridge system and is rarely associated with appendiceal mucocele. Diagnosis requires a high index of suspicion and careful intraoperative assessment. Extended resection may be required when anatomy is distorted or malignancy cannot be excluded, to prevent missed pathology and recurrence.

Keywords: Anatomical Variation • Appendiceal Diseases • Appendix • Case Reports • Gastroenterology • Mucocele

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/952870

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Introduction

Duplication of the appendix is a rare congenital anomaly, with a reported incidence of 0.004%-0.009% in appendectomy specimens [1]. Many authors have classified appendiceal duplication, but the earliest and commonest was done by Cave in 1936. Cave originally classified appendiceal duplication into 3 anatomical types based on the relationship of the appendices to the caecum [2]; this system was subsequently modified by Wallbridge in 1962 to give the widely used Cave–Wallbridge classification [2,3]. Type A (partial duplication) describes a single cecum with a single appendix showing varying degrees of partial or incomplete duplication, in which both appendices share a common base. Type B (complete duplication) describes a single cecum bearing 2 completely separate appendices, and is further subdivided by location: Type B1 (avian type), in which the 2 appendices are positioned symmetrically on either side of the ileocecal valve, and Type B2 (taenia coli type), in which one appendix occupies the normal anatomical position while the second arises along the taenia coli of the caecum or ascending colon. Type C (double cecum) describes complete duplication of the cecum, with each cecum bearing its own individual appendix [2,3].

Several theories have been proposed to explain the embryological origin of duplex appendix, including cloacal differentiation failure, the split notochord theory, and the temporary appendix theory. The danger with appendix duplication lies in the missed appendix during the primary surgery, leading to persistent or recurrent appendicitis from the second appendix, which is often misdiagnosed as stump appendicitis [3,4]. Preoperative detection is uncommon because imaging modalities like computed tomography (CT) or magnetic resonance imaging (MRI) rarely identify appendiceal duplication, and intraoperative diagnosis is common [4,5].

Appendiceal duplication has also rarely been reported in association with an appendiceal mucocele distension of the appendiceal lumen by accumulated mucus, most commonly resulting from chronic luminal obstruction [6,7]. When mucin is identified intraoperatively or on imaging, it can be difficult to distinguish a benign, inflammatory mucocele from a low-grade appendiceal mucinous neoplasm (LAMN) on gross and radiological assessment alone [6,7]. Several recently published cases have described a duplicated appendix presenting with a range of clinical scenarios, but without an associated mucocele [8-12]; the combination of duplication and mucocele formation seen in our patient appears to be very uncommon in the reported literature.

This report describes the case of a 45-year-old woman with a history of a loculated peri-appendiceal abscess managed with percutaneous drainage and intravenous antibiotics, who was subsequently found to have a duplicated appendix, with mucocele identified after right hemicolectomy performed for suspected appendiceal neoplasm.

Case Report

A 45-year-old woman presented to our hospital with a history of pain in the right lower abdomen that began 2 months ago. The pain became severe and there was associated intermittent fever. Clinical examination revealed severe tenderness in the right iliac fossa with rebound tenderness and localized rigidity. A complete blood count revealed a total leucocyte counts of 20 000 cells/μL. Contrast enhanced computed tomography (CECT) revealed a large heterogeneous thick-walled loculated collection in the right iliac fossa measuring 7 × 5.9 × 7.2 cm, with a volume of 163 cc. Few hyperdense calcified foci are seen with in it. The appendix could not be separately visualized. A percutaneous drain (PCD) was placed and the patient was started on culture based intravenous antibiotics. Subsequently, the PCD was removed after a week and she was discharged home. However, the patient presented again 2 months later with pain in the right iliac fossa for 7 days. It was not associated with fever, nausea, or vomiting. The patient was re-evaluated with a CECT of the abdomen, which showed distended tubular structure measuring 2.5 cm in diameter with subtle perifocal fat stranding in retrocaecal region, suggesting mucocele of the appendix. No intra-abdominal collection was identified (**Figure 1**). The patient subsequently had a diagnostic laparoscopy, showing dense adhesions between the intestinal loops and the parietal wall (**Figure 2**). The appendix was inflamed, oedematous, and dilated, measuring ~ 2.5 cm with a lump near its base. Because of the dilated, firm appendiceal tip and the mass-like clumping at the base, which raised concern for an underlying neoplasm that could not be safely excluded, the intraoperative decision was made to proceed to right hemicolectomy rather than a limited appendicectomy. Laparoscopic right hemicolectomy with extracorporeal ileo-transverse anastomosis (side-to-side, hand-sewn in 2 layers) was performed. Her postoperative course was uneventful and she was discharged on postoperative day 5. Macroscopic examination of the specimen showed 2 distinct tubular structures arising separately from the cecum, each communicating independently with the cecal lumen (**Figure 3A, 3B**). The native appendix had a maximum luminal diameter of 1.2 cm and the second parallel structure measuring 2.8 cm in length with a maximum luminal diameter of 1.5 cm, having a smooth inner lining, irregular outer surface, and mucoid intraluminal content, with no communication between the 2 structures. Microscopically, both structures were sampled entirely and showed entirely separate wall architecture, with no shared muscle layer or subserosa at any level (**Figure 3C**). The native appendix had a dilated lumen, flattened-to-focally denuded epithelium, intact muscularis

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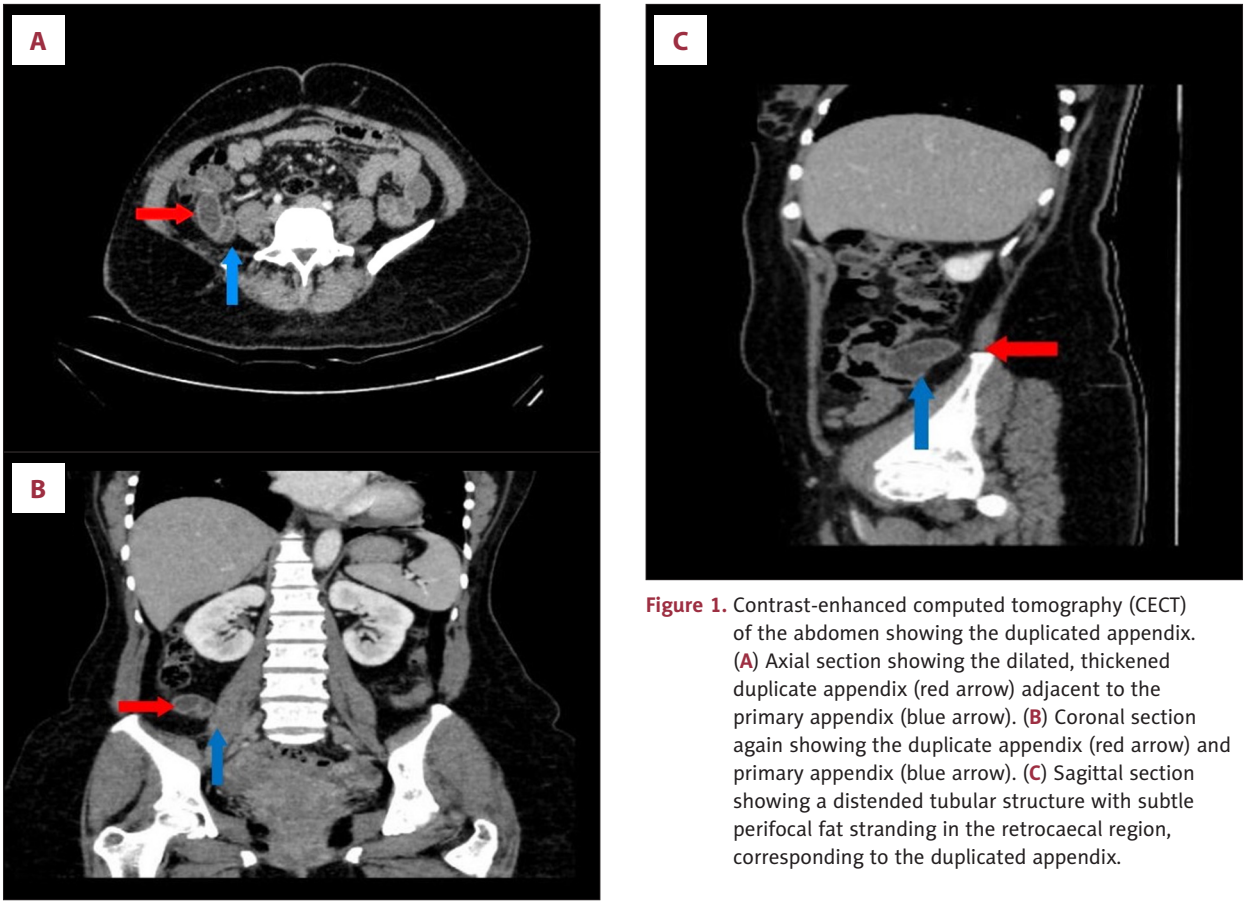

Figure 1. Contrast-enhanced computed tomography (CECT) of the abdomen showing the duplicated appendix. (**A**) Axial section showing the dilated, thickened duplicate appendix (red arrow) adjacent to the primary appendix (blue arrow). (**B**) Coronal section again showing the duplicate appendix (red arrow) and primary appendix (blue arrow). (**C**) Sagittal section showing a distended tubular structure with subtle perifocal fat stranding in the retrocaecal region, corresponding to the duplicated appendix.

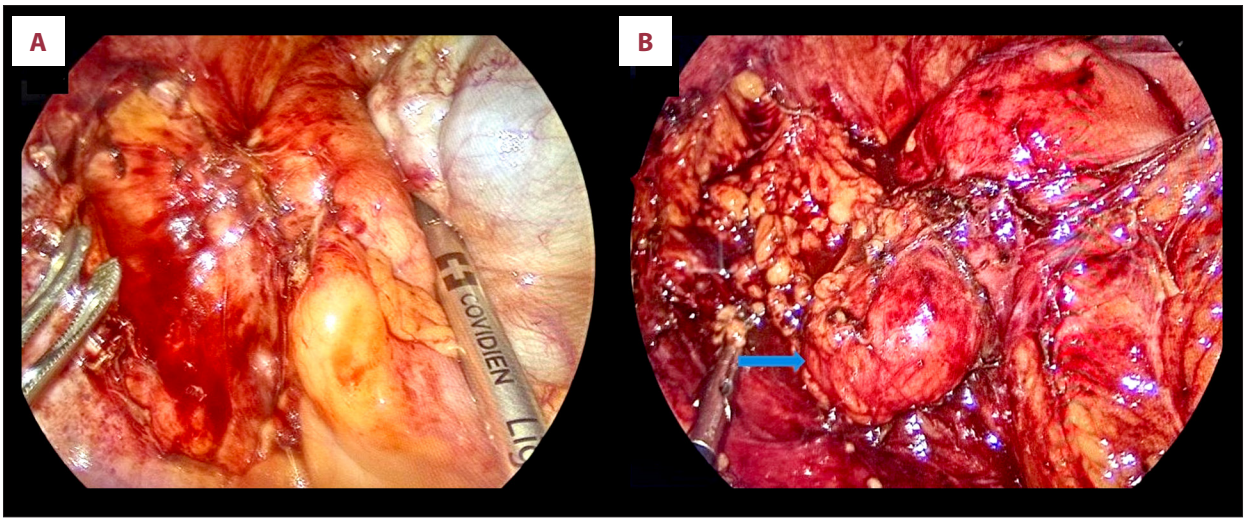

Figure 2. (**A, B**) Intraoperative photo showing adhesions between the cecum and the body wall, with a dilated appendix in the second photo.

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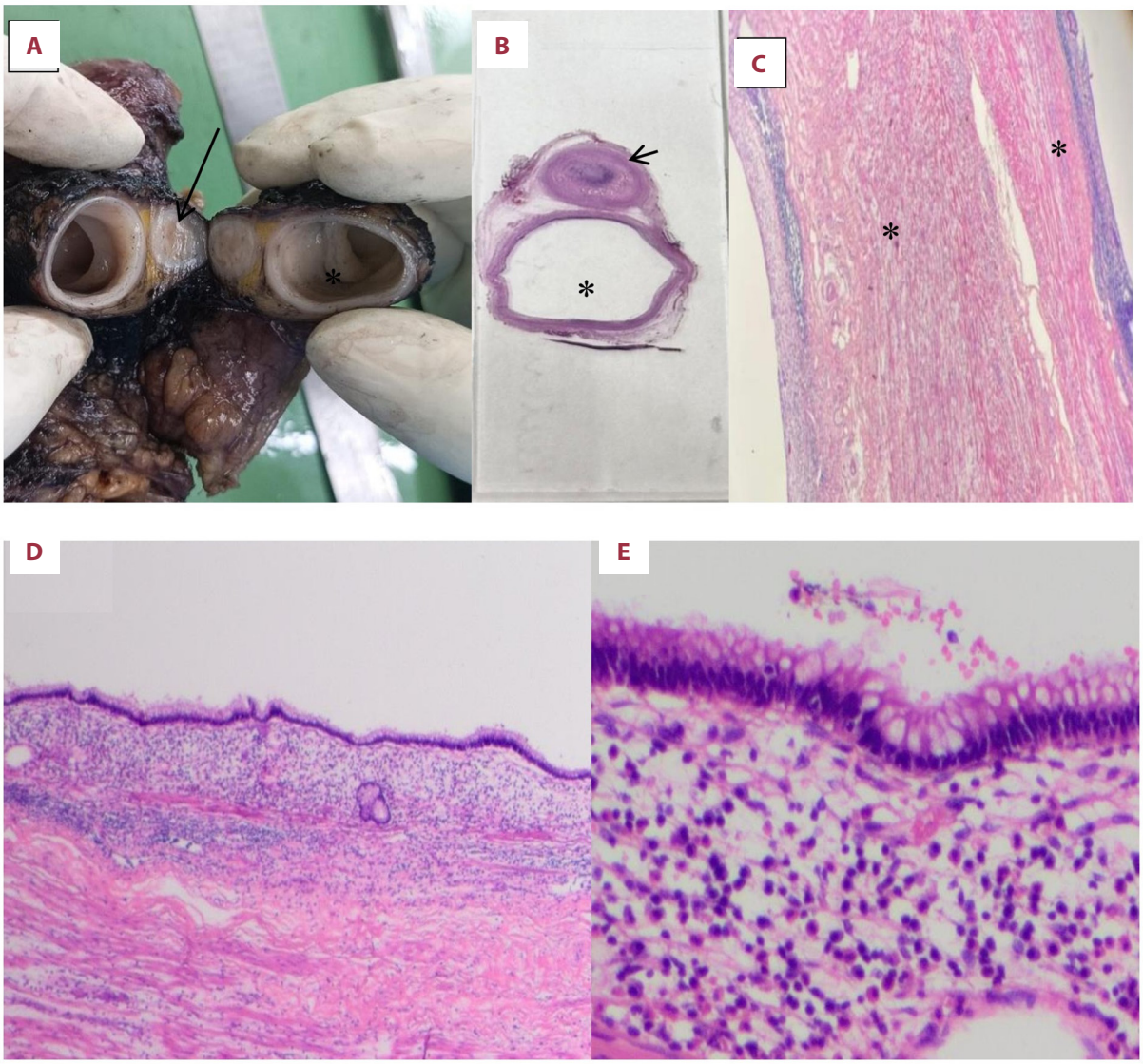

Figure 3. (**A, B**) Gross cross-section image of the resected specimen, highlighting 2 well-demarcated luminal structures. The appendix shows a narrowed lumen (arrow), while the duplication exhibits a dilated lumen (asterisk). (**C**) Low-power view showing 2 luminal structures, with closely opposed but well-demarcated walls, each with its own muscularis layer (asterisk) (hematoxylin and eosin stain, 2 ×). (**D, E**) Medium-power view showing the native appendix (**D**) with flattened-to-focally denuded lining epithelium and an intact muscularis mucosae layer, along with scattered chronic inflammatory infiltrates including eosinophils. The duplicated appendiceal structure (**E**) is lined by tall columnar mucinous epithelium and demonstrates a well-organized wall with a well-delineated muscularis mucosae, submucosa, and muscularis propria. All 3 layers show scattered chronic inflammation, mucin extravasation, and focal histiocytic response (H&E, 10 × and 20 ×).

mucosae, chronic inflammation with eosinophils (**Figure 3D**), and mural mucin extravasation extending to the subserosal layer. There was acellular mucin at the base focally reaching the muscularis propria of the cecum with a foreign body giant cell response. The duplicate structure showed well-organized wall architecture comprising distinct mucosa, muscularis mucosae, submucosa, and muscularis propria, lined by tall columnar mucinous epithelium with focal crypt paucity, lymphocytic infiltrate, chronic inflammation, mucin extravasation, histiocytic response, and areas of fibrosis (**Figure 3E**). The cecal mucosa, ileum, remaining colon, and examined lymph nodes were histologically normal. Histopathological examination thus confirmed a Type B2 (Cave–Wallbridge) duplication of the appendix, with mucocele formation classified as a simple retention-type lesion secondary to chronic luminal obstruction; there was no histopathological evidence of mucosal hyperplasia,

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Table 1. Comparison of published cases of appendiceal duplication with present case.

Author and year	Age/ sex	Type	Presentation	Imaging findings	Histopathology	Management type	Outcome
Our case	45/F	B2	Abdomen pain	CECT: duplicated appendix with appendicitis and mucin formation	Acute appendicitis with mucocele formation	Laparoscopic right hemicolectomy	Discharged, no complications
Cardona et al 2025 [9]	26/M	B2	Pain in RLQ	CT: duplicated appendix with inflammation and appendicolith	Acute suppurative inflammation	Laparoscopic appendicectomy	Discharged, no complications
Deflaoui et al 2024 [10]	27/M	B1	Pain in abdomen	CT: latero-caecal appendix with 2 stercoliths at its base	Acute suppurative inflammation, normal second appendix	Appendicectomy	Discharged, no complications
Shibata et al 2023 [8]	69/M	A	Positive occult blood test	CT: focal wall thickening of the appendix	Acute inflammatory changes with duplication	Laparoscopic ileocecal resection and anastomosis	Discharged, no complications
Gul et al 2013 [11]	19/M	A	Penetrating injury to abdomen	No imaging done	Histopathology diagnosis of duplication	Open appendicectomy	Discharged, no complications
Christodoulidis et al 2012 [12]	23/F	B2	Pain abdomen and vomiting	USG: acute appendicitis	Acute suppurative inflammation, normal second appendix	Open appendicectomy	Discharged, no complications

CECT, contrast-enhanced computed tomography; CT, computed tomography; USG, ultrasonography.

cystadenoma, or cystadenocarcinoma (**Figure 3**). When the previous imaging was reviewed, a duplicated appendix with 2 lumens could be identified (**Figure 1**). She is currently doing well in subsequent follow-ups.

Discussion

This case shows that a duplicated appendix can present as a mucocele, which may obscure its recognition on both preoperative imaging and intraoperative assessment. A high index of suspicion for a possible neoplasm should therefore be maintained in such presentations, as an isolated appendicectomy alone, without adequate resection, can leave the duplicated appendix behind and risk missed pathology or recurrence.

Duplication of the appendix is a very rare anomaly of the gastrointestinal tract. It can occur in isolation or form part of intestinal duplication [13]. It is usually asymptomatic but may predispose to recurrent right iliac fossa pain, recurrent appendicitis, and mucocele formation. The majority of diagnoses are missed preoperatively and discovered incidentally during surgery or after histopathology. They may also be missed intraoperatively, leading to serious medico-legal implications [13,14].

We document a rare case of a mucocele in a duplicated appendix that initially presented as a rupture appendix and was managed with percutaneous drainage in keeping with standard practice [14,15]. However, on recurrence of pain, CECT demonstrated a distended tubular structure in the retrocecal region, and the patient was taken up for surgery. Intraoperatively, dense adhesions and an inflammatory complex raised suspicion of an underlying neoplasm, and a right hemicolectomy was performed; postoperative histopathology revealed a duplicated appendix [6]. Although a duplicated appendix can be diagnosed using axial imaging modalities such as computed tomography (CT) and magnetic resonance imaging (MRI), it requires a high degree of suspicion [4-6,13-15].

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Our patient may be classed into Type B2 (taenia coli type), in which there is one appendix at the cecum and another one elsewhere along the taenia coli [2,3]. Even though mucin formation may be concerning for a low-grade appendiceal mucinous neoplasm, the chronic inflammation, obstruction, and pressure changes in the appendix lumen can also result in mucin accumulation within the appendix [7,8]. However, a low-grade appendiceal mucinous neoplasm (LAMN) is usually larger in size, with mucin seen beyond the mucosa and increased risk of pseudomyxoma peritonei (PMP). Appendectomy or limited resection is appropriate for appendiceal mucocele and LAMN. However, right hemicolectomy is the treatment of choice when there is uncertainty about the extent of the disease or when there are dense adhesions or mass-forming features on the cecum/appendix [7]. These entities can be difficult to differentiate intraoperatively, which made appendicectomy or limited resection unsafe in this case, in which a right hemicolectomy was done due to dense adhesions, mass-forming feature, and uncertainty about the extent of disease [7,16].

Table 1 compares our case with previously published reports of appendiceal duplication complicated by acute appendicitis or presenting with a range of clinical scenarios [8-12]. In most of these reports, the duplication itself was confirmed only intraoperatively or on histopathology, and no reports describe an associated mucocele. The management described ranged from open or laparoscopic appendicectomy to a limited ileocecal resection, generally with an uncomplicated outcome [8,11,12].

Our patient's uneventful postoperative course and 1-year disease-free survival reflect the adequacy of resection and absence of a neoplasm. Importantly, the surgical team's decision to perform hemicolectomy avoided the risk of incomplete removal of duplicated structures—one of the well-documented pitfalls leading to future complications or medico-legal challenges [17,18]. This case reinforces the principle that when encountering atypical anatomy, surgeons must maintain a high index of suspicion for congenital anomalies and consider extended resection when the pathology is unclear or involves the cecal wall. Table 1 compares previously published reports on duplicated appendix presenting as acute appendicitis.

Conclusions

Appendiceal duplication is a rare but clinically important cause of right iliac fossa pathology that can present atypically, including with mucocele formation and persistent symptoms after apparently adequate management of an appendiceal abscess. Thorough intraoperative assessment is essential, and extended resection such as right hemicolectomy may be necessary when a safe appendicectomy alone is not feasible because of inflammation, anatomical uncertainty, or suspicion of malignancy. Awareness of this anomaly among clinicians, radiologists, and pathologists may improve surgical and diagnostic outcomes, and a negative appendicectomy in the presence of persistent symptoms should prompt a search for a duplicated appendix.

Institution Where Work Was Done

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Patient Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. The patient consented to the publication of her clinical information with the understanding that her identity would not be disclosed. All reasonable efforts have been made to preserve patient anonymity.

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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