

Ulcerative Colitis-Associated Appendiceal Mucinous Neoplasms: A Systematic Review

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Abstract

Background

Ulcerative colitis (UC) is a well-established risk factor for colorectal malignancy; however, primary appendiceal mucinous neoplasms (AMNs) remain a rare entity in this population. These lesions, ranging from benign cystadenomas to invasive adenocarcinomas, present significant diagnostic challenges as their symptoms often mimic UC flares or acute appendicitis. Given the scarcity of data and lack of standardized treatment guidelines, this systematic review aims to synthesize the clinical characteristics and management outcomes of AMNs in patients with UC.

Aims

This study aims to review the characteristics and management of appendiceal mucinous neoplasm in ulcerative colitis.

Methods

Inclusion criteria were adults with ulcerative colitis that presenting with appendiceal mucinous neoplasm under the histological confirm. Exclusion criteria were patient not affected by ulcerative colitis, not adults, lack of abstracts, articles that is not in English. We screened PubMed, Medline and Cochrane Library databases up to January 2026. Total 20 articles were included.

Results

This systematic review analyzed 29 patients (mean age 52 years; 55.2% male) with concurrent UC and AMNs. The majority had long-standing UC (mean duration 11.1 years among the 18 patients with available data), and 52% exhibited active colonic inflammation at the time of diagnosis. While abdominal pain was the primary symptom (48%), 14% of patients were asymptomatic with incidental findings. All patients underwent surgical resection, with procedures ranging from simple appendectomy (21%) to colectomy or proctocolectomy (38%). Histopathologically, benign mucinous cystadenomas were predominant (52%), while LAMN and mucinous adenocarcinomas each accounted for 27.6% and 20.7% respectively. These findings highlight that while most lesions are benign or low-grade, a significant proportion represents malignancy, necessitating high vigilance and careful surgical planning in UC patients.

Conclusion

Concurrent AMN and UC are rare. Diagnosis is challenging due to overlapping symptoms and nonspecific endoscopic findings. While most lesions are benign cystadenomas, the rate of malignancy (20.7%) and risk of pseudomyxoma peritonei necessitate rigorous preoperative imaging, and definitive surgical resection to ensure optimal outcomes.

1. Introduction

Appendiceal mucocoele is an uncommon clinical entity characterized by the distension of the appendix due to abnormal mucus accumulation, which can often be associated with neoplastic transformation. While the first case report of an appendiceal mucocoele associated with ulcerative colitis (UC) was published in 1949[1], such occurrences remain exceedingly rare. The coexistence of these two conditions is not yet well understood. It has been suggested that chronic appendicular inflammation related to UC, or an associated appendicular neoplasm, may lead to luminal obstruction and subsequent mucocoele formation[2]. More recently, a case-control study has suggested that these lesions may instead represent a neoplastic complication of long-standing colitis [10]. Given the scarcity of reported cases and the absence of any

consolidated analysis, we systematically reviewed all published cases of appendiceal mucinous neoplasm occurring in patients with ulcerative colitis, including one recently reported by our own group [22], in order to characterize their clinical presentation, diagnostic pitfalls, pathological spectrum and surgical management.

2. Methods

We followed a standardized methodology, reporting our findings in accordance with the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines. The full texts of potentially eligible articles were independently reviewed by two authors (LKC and CYL) to identify cases of ulcerative colitis-associated appendiceal mucinous neoplasms.

2.1. Inclusion/exclusion criteria

Inclusion Criteria

Studies were included if they evaluated adult patients with histologically confirmed appendiceal mucinous neoplasms secondary to, or presenting concurrently with, ulcerative colitis.

Exclusion Criteria

Studies were excluded if they involved pediatric populations, patients with Crohn's disease or without a confirmed diagnosis of ulcerative colitis, or lesions lacking histological confirmation. Narrative and systematic reviews, editorials, and conference abstracts without an accompanying full text were also excluded; correspondence and letters to the editor reporting original case-level data were retained. Reports for which the full text could not be retrieved were excluded.

2.2. Search Strategy

To identify all relevant publications addressing ulcerative colitis-associated appendiceal mucinous neoplasms, a systematic and exhaustive literature search was conducted across the PubMed, Medline, Cochrane Library. Databases were searched from inception to January 2026; searches were conducted between August 2025 and January 2026. We utilized a combination of Medical Subject Headings (MeSH) terms, exploded terms, truncated words, and free-text terms. The core search string combined keywords related to ulcerative colitis and appendiceal pathology, using Boolean operators as follows: ((((((appendiceal neoplasm) OR(mucocele))OR(appendiceal cancer))OR(appendix adenocarcinoma))OR (appendiceal cystadenoma))OR(appendiceal mucocele)))AND((ulcerative colitis)OR(inflammatory bowel disease))). Human-study filters were applied to refine the results. Additionally, the reference lists of all included articles and relevant review papers were manually scrutinized to capture any additional literature. Ultimately, a total of 20 articles met the criteria and were included in this review. For cases in which the full text could not be independently retrieved, data were supplemented from a previously published systematic review (Bonomi et al., 2023) reporting the same cases; the original case reports remain cited as the primary source.

2.3. Study Selection and Screening Process

The study selection process followed the PRISMA 2020 statement guidelines, as illustrated in the flow diagram (Fig. 1). A total of 690 records were initially identified through database searching. Prior to screening, 78 duplicate records were removed, leaving 612 unique records for title and abstract screening. During the initial screening phase, 581 records were excluded based on irrelevance to the study topic or failure to meet basic inclusion criteria. The remaining 31 reports were sought for full-text retrieval; however, 5 reports could not be successfully retrieved. A total of 26 full-text reports were subsequently assessed for eligibility. Of these, 6 reports were excluded for specific violations of our criteria: 4 studies evaluated patients with Crohn's disease (CD) rather than ulcerative colitis, and 2 studies involved a pediatric population. Ultimately, 20 eligible articles fulfilled all criteria and were included in the final qualitative synthesis.

2.4. Data Extraction

Data extraction was performed independently by two authors (LKC and CYL). The included papers were reviewed in detail to extract relevant variables, including the number of patients studied, publication year, study inclusion and observation periods, and the mean or median duration of follow-up. Discrepancies between the reviewers were resolved through discussion and consensus. All enrolled patients from the selected eligible studies were included in the final analysis.

3. Results

3.1. Baseline and Demographics Characteristics

The systematic review analyzed data from 29 patients across 21 eligible publications spanning from 1997 to 2026[2–21], including one case published by our own group[22]. Their characteristics, management and histopathology are summarized in Table 1; individual patient data are presented in Tables 2 and 3. Most reports were single case reports; one retrospective case–control series contributed six patients, and two further reports contributed two and three patients respectively. Among the cohort, there was a slight male predominance, with 16 male patients (55.2%) and 13 female patients (44.8%). The age of the patients at the time of diagnosis ranged widely from 24 to 76 years.

Table 1

Summary of clinical characteristics, management and histopathology of 29 patients with ulcerative colitis–associated appendiceal mucinous neoplasms.

Characteristic	Value
Demographics (n = 29)	
Age at diagnosis, years – mean (range)	52.3 (24–76)
21–30	2 (6.9)
31–40	4 (13.8)
41–50	6 (20.7)
51–60	10 (34.5)
61–70	4 (13.8)
≥71	3 (10.3)
Male	16 (55.2)
Female	13 (44.8)
Ulcerative colitis characteristics (n = 29)	
Disease duration, years – mean (range)¶	11.1 (0–35)
Long-standing, duration unspecified	2 (6.9)
Not reported	9 (31.0)
Disease activity at diagnosis	
Remission	10 (34.5)
Active (any severity)	15 (51.7)
Mild	4 (13.8)
Moderate	3 (10.3)
Severe	2 (6.9)
Severity not specified	6 (20.7)
Newly diagnosed UC	1 (3.4)
Not reported	3 (10.3)
Clinical presentation (n = 29)	
Symptomatic	16 (55.2)
Asymptomatic (incidental detection)	4 (13.8)
Not reported	9 (31.0)
Reported symptoms¶	
Abdominal pain	14 (48.3)
Diarrhea	5 (17.2)
Bloody stool / rectal bleeding	4 (13.8)
Fever	2 (6.9)

Characteristic	Value
Demographics (n = 29)	
Weight loss	2 (6.9)
Palpable abdominal mass	1 (3.4)
Lesion size (reported in 23 patients)	
Greatest dimension, cm – median (range)	3.5 (0.4–12)
≤2 cm	8 (34.8)
>2–5 cm	5 (21.7)
>5–10 cm	8 (34.8)
>10 cm	2 (8.7)
Endoscopic findings (reported in 17 patients)	
Appendiceal or cecal abnormality identified	11 (64.7)
Inflammatory changes only, no discrete lesion	5 (29.4)
No lesion visualized	1 (5.9)
Surgical management (n = 29)	
Appendectomy	6 (20.7)
Partial cecectomy ± appendectomy	4 (13.8)
Right hemicolectomy	8 (27.6)
Colectomy or proctocolectomy ± ileostomy	11 (37.9)
Surgical approach (reported in 16 patients)	
Open	8 (50.0)
Laparoscopic	7 (43.8)
Robotic assisted	1 (6.3)
Appendiceal histopathology (n = 29)	
Mucinous cystadenoma	15 (51.7)
Low-grade appendiceal mucinous neoplasm (LAMN)	8 (27.6)
Mucinous adenocarcinoma	6 (20.7)
High-grade appendiceal mucinous neoplasm (HAMN)	0 (0)
Synchronous colorectal neoplasia (n = 29)	
Present	8 (27.6)
Adenocarcinoma ± dysplasia	6 (20.7)
Dysplasia only	2 (6.9)
Absent	18 (62.1)
Not reported	3 (10.3)

Characteristic	Value
Demographics (n = 29)	
Values are n (%) unless otherwise stated. Percentages are calculated using the denominator stated for each block.	
☒ Calculated for the 18 patients with a quantifiable disease duration, including 2 patients with newly diagnosed UC counted as 0 years; a further 2 patients were described only as long-standing and 9 were not reported.	
☒ Categories are not mutually exclusive; individual patients could report more than one symptom.	
☒ Includes a bulging or patulous appendiceal orifice, a cecal submucosal or mass lesion, extruding mucin, extrinsic cecal compression, and a laterally spreading tumor-like lesion at the appendiceal orifice.	
☒ Classified according to the definitive resection; includes one patient who underwent completion right hemicolectomy after an initial appendectomy with caput cecectomy.	
☒ Includes one patient with adenocarcinoma in situ arising within a mucinous cystadenoma.	
☒ Dysplasia includes flat dysplasia and dysplasia-associated lesions or masses (DALMs). Two patients had extensive multifocal dysplasia with multiple synchronous invasive colorectal adenocarcinomas.	
LAMN, low-grade appendiceal mucinous neoplasm; HAMN, high-grade appendiceal mucinous neoplasm; UC, ulcerative colitis.	

Table 2
Patient and disease characteristics of the 29 included cases.

Author (year)	Age	Sex	UC duration (years)	Disease activity	Presenting symptoms
Zannoni (1997)	69	F	NR	Remission	NR
Lyda (1998)	48	M	26	Pancolitis, severity NR	NR
Lakatos (2005)	58	M	4	Moderate (proctosigmoiditis)	Bloody diarrhea; RLQ pain
Villanueva (2006)	60	F	NR	Active (left-sided colitis)	NR
Kuester (2008)	36	M	1	Severe (pancolitis)	Abdominal pain; diarrhea; weight loss
Takeda (2008)	48	M	20	Remission	RLQ pain
Noaki (2009)	33	F	5	Remission	RLQ pain
Orta (2009)	76	F	NR	NR (left-sided)	NR
Orta (2009)	34	M	NR	Active (pancolitis)	NR
Orta (2009)	24	M	NR	Active (extensive)	NR
Orta (2009)	57	F	NR	Active (left-sided colitis)	NR
Orta (2009)	28	F	NR	Active (pancolitis)	NR
Orta (2009)	75	M	NR	Active (extensive)	NR
Ghosh (2010)	55	F	NR	NR	Asymptomatic
Hernández-Ramírez (2010)	54	M	3	Moderate (proctosigmoiditis)	RLQ pain
Wong (2012)	62	F	Long-standing	Remission	Asymptomatic
Wong (2012)	34	F	Newly diagnosed	Mild (pancolitis)	Bloody diarrhea
Shen (2014)	68	M	35	Remission	RLQ pain
Tonolini (2015)	71	M	Long-standing	Remission [‡]	Rectal bleeding; increased stool frequency; abdominal pain; fever; weight loss
Nehme (2019)	42	M	10	Severe (pancolitis, Mayo 3)	RLQ pain
Davey (2020)	42	M	18	Mild (proctitis)	Abdominal pain; bloody diarrhea
Fukumoto (2021)	52	M	6	Remission	RLQ pain; fever
Abdulghaffar (2022)	48	F	Newly diagnosed	Newly diagnosed	Palpable RLQ mass
[‡] Reported as being in clinical remission on maintenance mesalazine; colonoscopy at presentation nevertheless demonstrated mild proctosigmoiditis.					
[‡] Reported as long-standing quiescent disease; colonoscopy at presentation demonstrated an acute relapse with moderately active pancolitis.					
F, female; M, male; NR, not reported; RLQ, right lower quadrant; UC, ulcerative colitis.					

Author (year)	Age	Sex	UC duration (years)	Disease activity	Presenting symptoms
Fakheri (2023)	55	F	2	Remission (Mayo 1)	RLQ pain
Fakheri (2023)	52	F	12	Moderate (Mayo 4)	RLQ pain
Fakheri (2023)	60	M	11	Mild	RLQ pain
Huang (2025)	70	F	19	Remission	Asymptomatic
Muto(2025)	59	M	24	Remission	Asymptomatic
Chang (2026)	46	M	3	Mild (Mayo 2)	Diffuse abdominal pain
☒ Reported as being in clinical remission on maintenance mesalazine; colonoscopy at presentation nevertheless demonstrated mild proctosigmoiditis.					
☒ Reported as long-standing quiescent disease; colonoscopy at presentation demonstrated an acute relapse with moderately active pancolitis.					
F, female; M, male; NR, not reported; RLQ, right lower quadrant; UC, ulcerative colitis.					

Table 3

Lesion size, endoscopic findings, surgical management and appendiceal histopathology of the 29 included cases.

Author (year)	Size (cm)	Endoscopic finding	Definitive procedure	Approach	Appendiceal histopathology	Synchronous colorectal neoplasia
Zannoni (1997)	NR	NR	Right hemicolectomy	Open	Mucinous adenocarcinoma	No
Lyda (1998)	8.0	NR	Proctocolectomy	Open	Mucinous cystadenoma, LGD	Multifocal dysplasia (4 DALMs); 4 invasive adenocarcinomas
Lakatos (2005)	11	Proctosigmoiditis with moderate activity	Partial cecectomy + mucocele removal	Open	Mucinous cystadenoma	No
Villanueva (2006)	5.0	NR	Proctocolectomy with ileostomy	NR	Mucinous adenocarcinoma	No
Kuester (2008)	NR	NR	Proctocolectomy with ileostomy	NR	Mucinous cystadenoma, LGD	Multifocal DALMs; 4 invasive adenocarcinomas
Takeda (2008)	8.0	NR	Appendectomy (emergency)	NR	Adenocarcinoma in situ in cystadenoma	No
Noaki (2009)	12	Cecal submucosal tumor, 5 cm; biopsy non-diagnostic	Partial cecectomy including appendix	Open	Mucinous cystadenoma	No
Orta (2009)	0.4	NR	Colectomy (extent not specified)	NR	Mucinous cystadenoma, LGD	No
Orta (2009)	1.3	NR	Colectomy (extent not specified)	NR	Mucinous cystadenoma, LGD	LGD (rectum)
Orta (2009)	0.5	NR	Colectomy (extent not specified)	NR	Mucinous cystadenoma, LGD	No
Orta (2009)	1.7	NR	Colectomy (extent not specified)	NR	Mucinous cystadenoma, LGD	Adenocarcinoma (descending colon); LGD (sigmoid)
Orta (2009)	1.0	NR	Colectomy (extent not specified)	NR	Mucinous cystadenoma, LGD	Adenocarcinoma (hepatic flexure); multifocal LGD/HGD
Synchronous colorectal neoplasia includes flat dysplasia, DALMs and invasive adenocarcinoma. Lesion size for the patients reported by Lyda and Villanueva was taken from the tabulated data of Orta et al.						
⌘ Completion right hemicolectomy after an initial appendectomy with caput cecectomy, when final pathology showed a T2 adenocarcinoma at the staple line.						
DALM, dysplasia-associated lesion or mass; HGD, high-grade dysplasia; IPAA, ileal pouch–anal anastomosis; LAMN, low-grade appendiceal mucinous neoplasm; LGD, low-grade dysplasia; LST, laterally spreading tumor; NR, not report						

Author (year)	Size (cm)	Endoscopic finding	Definitive procedure	Approach	Appendiceal histopathology	Synchronous colorectal neoplasia
Orta (2009)	NR	NR	Colectomy (extent not specified)	NR	Mucinous cystadenoma, LGD	Adenocarcinoma and HGD (rectosigmoid)
Ghosh (2010)	1.3	Smooth rounded cecal lesion, 2.5 cm	Right hemicolectomy	Laparoscopic	Mucinous cystadenoma, LGD	No
Hernández-Ramírez (2010)	10	Extrinsic compression of caecum; rectosigmoid inflammation	Partial cecectomy + mucocele removal	Open	Mucinous cystadenoma	NR
Wong (2012)	2.0	2-cm mass at appendiceal orifice	Appendectomy	Laparoscopic	Mucinous cystadenoma	No
Wong (2012)	2.0	Bulging appendiceal orifice; mild pancolitis	Appendectomy	Open	Mucinous cystadenoma	No
Shen (2014)	NR	NR	Right hemicolectomy (completion)¶	Laparoscopic	Mucinous adenocarcinoma (T2)	No
Tonolini (2015)	3.5	Moderately active pancolitis	Appendectomy	Laparoscopic	LAMN	No
Nehme (2019)	2.6	Severe pancolitis (Mayo 3)	Proctocolectomy with ileostomy	NR	Mucinous adenocarcinoma, invasive	Multifocal LGD
Davey (2020)	8.6	Globular mass, transverse colon (intussusception)	Right hemicolectomy + ileocecal anastomosis	Open	LAMN	No
Fukumoto (2021)	NR	Circumferential stenosis, no tumor seen; biopsy negative	Right hemicolectomy	Laparoscopic	Mucinous adenocarcinoma (stage IIIB)	No
Abdulghaffar (2022)	9.0	No mass visualized	Appendectomy	Open	LAMN	No
Fakheri (2023)	10	Bulging appendiceal orifice	Right hemicolectomy	NR	Mucinous cystadenoma	No
Fakheri (2023)	5.7	Bulging appendiceal orifice	Right hemicolectomy	NR	LAMN	No
Fakheri (2023)	NR	Inflammation in rectum and	Appendectomy	NR	Mucinous cystadenoma	No
Synchronous colorectal neoplasia includes flat dysplasia, DALMs and invasive adenocarcinoma. Lesion size for the patients reported by Lyda and Villanueva was taken from the tabulated data of Orta et al.						
¶ Completion right hemicolectomy after an initial appendectomy with caput cecectomy, when final pathology showed a T2 adenocarcinoma at the staple line.						
DALM, dysplasia-associated lesion or mass; HGD, high-grade dysplasia; IPAA, ileal pouch–anal anastomosis; LAMN, low-grade appendiceal mucinous neoplasm; LGD, low-grade dysplasia; LST, laterally spreading tumor; NR, not report						

Author (year)	Size (cm)	Endoscopic finding	Definitive procedure	Approach	Appendiceal histopathology	Synchronous colorectal neoplasia
		caecum				
Huang (2025)	3.2	Bulging orifice with extruding mucin; biopsy negative	Partial cecectomy + appendectomy	Robotic	LAMN (grade 1)	NR
Muto (2025)	5.5	LST-like granular lesion at appendiceal orifice	Total colectomy with IPAA + ileostomy	Laparoscopic	LAMN	Mucinous adenocarcinoma (transverse colon)
Chang (2026)	3.5	Cecal submucosal tumor, enlarging 1.5 → 3 cm	Right hemicolectomy	Laparoscopic	LAMN	NR
Synchronous colorectal neoplasia includes flat dysplasia, DALMs and invasive adenocarcinoma. Lesion size for the patients reported by Lyda and Villanueva was taken from the tabulated data of Orta et al.						
☒ Completion right hemicolectomy after an initial appendectomy with caput cecectomy, when final pathology showed a T2 adenocarcinoma at the staple line.						
DALM, dysplasia-associated lesion or mass; HGD, high-grade dysplasia; IPAA, ileal pouch–anal anastomosis; LAMN, low-grade appendiceal mucinous neoplasm; LGD, low-grade dysplasia; LST, laterally spreading tumor; NR, not report						

3.2. Ulcerative Colitis History and Clinical Activity

The duration of UC prior to the diagnosis of an appendiceal mucinous neoplasm varied substantially, ranging from newly diagnosed concurrent cases to a maximum history of 35 years; however, this information was not recorded for a significant portion of the cohort (9 patients, 31%). At the time of clinical presentation, 10 patients (34.5%) were in clinical or histological remission. Concurrently, 15 patients (51.7%) presented with active UC inflammation, with severity ranging from mild to severe and involving various disease extents, including pancolitis, extensive colitis, left-sided colitis, and proctitis (Table 1). Of the remaining 4 patients, one presented with a newly diagnosed UC detected concurrently during appendiceal evaluation, while disease activity was not reported in the other three. Notably, disease extent was frequently reported without a corresponding severity grade, so that six patients could be classified only as having active disease of unspecified severity. The lesion was therefore identified across the full spectrum of disease activity, and quiescent colitis did not preclude the diagnosis.

3.3. Clinical Presentations and Endoscopic Findings

Clinical symptoms were highly variable. Among the 29 patients with presentation data, the majority were symptomatic, most commonly with right lower quadrant or diffuse abdominal pain, frequently accompanied by UC-related symptoms such as bloody diarrhea, or by systemic features including fever and weight loss (Table 1). Four patients were entirely asymptomatic, the lesion being detected during surveillance colonoscopy or within a colectomy specimen resected for another indication.

Colonoscopy findings were reported in 17 patients. Endoscopic evaluation of the cecum revealed distinct abnormalities in 11 cases, most commonly presenting as a bulging appendiceal orifice, a mass at the appendix orifice (with or without extruding mucus), a submucosal tumor-like appearance, an external compression lesion, or a laterally spreading tumor–like lesion at the appendiceal orifice. Where endoscopic biopsy of a suspicious lesion was attempted and reported, it was non-diagnostic or negative for neoplasia in every instance (Table 3).

3.4. Surgical and Therapeutic Management

Surgical interventions were tailored to the extent of the appendiceal tumor, the degree of cecal involvement, and the activity of the underlying UC. Procedures were classified according to the definitive resection performed. Localized or limited resection was undertaken in 10 patients, comprising simple or laparoscopic appendectomy (n = 6) and partial cecectomy

with appendectomy or mucocoele removal (n = 4). Radical or extensive resection was undertaken in 19 patients, comprising right hemicolectomy, including laparoscopic-assisted variants (n = 8), and colectomy or proctocolectomy with or without ileostomy (n = 11) (Table 1). Within the latter group, six patients derived from a single retrospective series in which the extent of colectomy was not specified. Notably, one patient initially treated with laparoscopic appendectomy and caput cecectomy required completion right hemicolectomy after final pathology demonstrated a T2 adenocarcinoma extending to the staple line and was therefore classified according to the definitive procedure. The surgical approach was reported in only 16 patients, of whom half underwent open surgery; laparoscopic and robotic approaches were confined to reports published from 2010 onwards (Table 3).

Postoperative medical management varied. Patients with active UC or residual inflammatory burden were treated with standard UC therapies, including mesalazine (5-ASA), oral or topical corticosteroids, or biologics (adalimumab). For patients with advanced malignant histology, adjuvant interventions such as chemotherapy (e.g., modified FOLFOX6), radio-chemotherapy, or planned chemotherapy were administered.

3.5. Histological Classification

Histopathological evaluation of the resected specimens classified the appendiceal mucinous lesions into three primary categories. Mucinous cystadenoma was the most frequent diagnosis, followed by LAMN and mucinous adenocarcinoma; no high-grade appendiceal mucinous neoplasm was reported (Table 1). This predominance is driven by the series of Orta et al. [10], which contributed six incidentally detected lesions; excluding it, the three categories were comparably represented, and the malignancy rate rose to 26.1%. Synchronous colorectal dysplasia or carcinoma was identified in eight patients (27.6%), comprising invasive adenocarcinoma with or without accompanying dysplasia in six and dysplasia alone in two, while this information was not reported in three patients. The severity of the colonic disease in these cases was frequently substantial: two patients had extensive multifocal dysplasia with four synchronous invasive colorectal adenocarcinomas each, and in a further three the synchronous carcinoma was accompanied by multifocal low- or high-grade dysplasia. The relationship between lesion size and histology is shown in Fig. 2. All eight lesions measuring ≤ 2 cm were cystadenomas, five of which were incidental microscopic findings within colectomy specimens. Above that range size offered no discrimination: the four largest lesions, measuring 10–12 cm, were all cystadenomas, whereas LAMN clustered between 3.2 and 9.0 cm and the three adenocarcinomas with a reported size measured 2.6, 5.0 and 8.0 cm.

4. Discussion

Appendiceal mucocoele is a rare pathological entity, accounting for less than 0.3% of appendectomy specimens, and its association with ulcerative colitis is infrequently reported. The cause of these two disease may be due to inflammation and lumen stricture[1]. Research shows that 82.6% of patients with IBD and appendiceal mucocoele are symptomatic, with 61% showing moderate-to-severe activity[23]. However, our case was detected via routine screening colonoscopy[22], underscoring the importance of close clinical monitoring even in asymptomatic patients. In cases of UC with moderate-to-severe activity, the inflamed environment creates significant "visual noise" that makes endoscopic differentiation difficult[17].

Three factors converge to make endoscopic recognition unreliable in this setting. First, an AMN is fundamentally a submucosal or extramural lesion; the overlying cecal mucosa is frequently intact, so the endoscopic signature is indirect — a bulging or patulous appendiceal orifice, a smooth submucosal elevation, extruding mucin, or simply extrinsic compression of the cecal wall. Second, forceps biopsy of such a lesion samples only the overlying mucosa and is therefore characteristically non-diagnostic. In the present cohort this was documented repeatedly: endoscopic biopsy failed to yield a histological diagnosis in the patient reported by Noaki[9], was negative for malignancy in the patient described by Fukumoto[17] whose resected specimen ultimately demonstrated a stage IIIB appendiceal adenocarcinoma and was likewise negative in the patient reported by Huang[20] who proved to have a LAMN. Third, in a colon that is actively inflamed, the same features that would otherwise raise suspicion — mucosal elevation, nodularity, granularity, and luminal narrowing — are indistinguishable from inflammatory pseudopolyps, post-inflammatory scarring, and the mucosal irregularity of active colitis itself. The consequence is that a lesion may be entirely overlooked: in the patient reported by Abdulghaffar[18], no mass was visualized

endoscopically despite a 9-cm appendiceal mucocoele, and in the patient reported by Fukumoto[17], colonoscopy showed only circumferential stenosis with reddened mucosa and no intraluminal tumor.

Conversely, aggressive or repeated biopsy of a suspected mucocoele is best avoided, given the risk of perforation and mucin spillage with subsequent pseudomyxoma peritonei. Interval growth on serial surveillance is a particularly useful discriminator, since inflammatory lesions fluctuate with disease activity whereas a neoplasm enlarges progressively; in our own case[22], the cecal submucosal lesion increased from 1.5 cm to 2.5 cm and then to 3 cm over successive examinations, which prompted definitive resection. Ultimately, as the endoscopic appearance is at best suggestive, histopathological examination of the resected specimen remains the only means of definitive diagnosis.

In patients with inflammatory bowel disease, computed tomography (CT) is a critical diagnostic tool for identifying appendiceal mucocoeles, especially when active inflammation makes clinical and endoscopic differentiation difficult. On a CT scan, an appendiceal mucocoele typically appears as a well-defined, tubular, or cystic mass in the right lower quadrant with low-attenuation content [24, 25]. Importantly, low-grade appendiceal mucinous neoplasms can progress to pseudomyxoma peritonei if rupture occurs, underscoring the need for timely surgical intervention. Right hemicolectomy is generally favored when there is suspicion of malignancy or when the lesion is large and cannot be safely removed by simple appendectomy[24–27]. These data qualify the conventional reliance on lesion size: although no LAMN or adenocarcinoma in our series measured less than 2.6 cm, above that threshold size lost all discriminatory value, the four largest lesions being benign cystadenomas of 10–12 cm. Size should therefore determine whether an appendiceal lesion is resected, not how extensively – the appendiceal base and margin status remain the operative determinants.

Synchronous colorectal dysplasia or carcinoma was present in 27.6% of our cohort, closely matching the 26.9% reported among the UC patients of Bonomi et al.[23], and the colonic disease was often advanced, with two patients harboring four synchronous invasive adenocarcinomas each. This is unlikely to be coincidental: in the only case–control study[10] addressing the question, appendiceal cystadenomas were eightfold more prevalent in IBD colectomy specimens harboring colonic dysplasia or carcinoma than in uncomplicated IBD. An appendiceal mucinous neoplasm in UC should therefore be regarded as a marker of colitis-associated neoplasia rather than an isolated appendiceal problem, and the appendix warrants deliberate sampling whenever colectomy is performed for colonic dysplasia.

In the context of UC, where patients undergo regular surveillance, the incidental detection of appendiceal pathology may offer an opportunity for earlier diagnosis. Clinicians should maintain a high index of suspicion for appendiceal neoplasms in UC patients presenting with right lower quadrant symptoms or atypical submucosal lesions. Increased awareness of this rare coexistence may improve diagnostic accuracy and guide optimal management strategies.

5. Conclusion

Appendiceal mucinous neoplasms in ulcerative colitis are rare but clinically consequential. Diagnosis is difficult because symptoms overlap with those of a colitis flare and because the endoscopic appearance is indirect. In every case in which it was attempted and reported, endoscopic biopsy failed to establish the diagnosis. Although just over half of the lesions were benign cystadenomas, one in five was an adenocarcinoma, and lesion size did not distinguish benign from malignant disease beyond the conventional 2 cm threshold. Cross-sectional imaging and a low threshold for definitive resection are therefore warranted, with the extent of resection determined by the appendiceal base and margin status rather than by lesion size alone. The frequent coexistence of synchronous colorectal dysplasia or carcinoma indicates that these lesions should be regarded not as isolated appendiceal pathology but as a marker of colitis-associated neoplasia, and that the appendix deserves explicit attention during surveillance colonoscopy and careful sampling in colectomy specimens.

Limitations

This review has several limitations. The evidence base consists almost entirely of single case reports, a design subject to publication bias favoring unusual or severe presentations, and reporting was incomplete across every domain, so that

denominators differ between analyses and follow-up was too inconsistent to assess recurrence. Six of 29 patients derive from a single retrospective series of incidentally detected lesions, which shifts the size and histology distributions towards small benign lesions. The findings should therefore be regarded as descriptive.

Table legend:

Declarations

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Nil

Author Contribution

L. K. Chang wrote the main manuscript text, and C. Y. Liao, W. C. Lin, and C. Y. Lee reviewed the references. C. W. Chang and P. H. Le discussed the findings. As the corresponding author, C. W. Chang reviewed the manuscript and prepared it for submission. All authors reviewed the manuscript.

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Figures

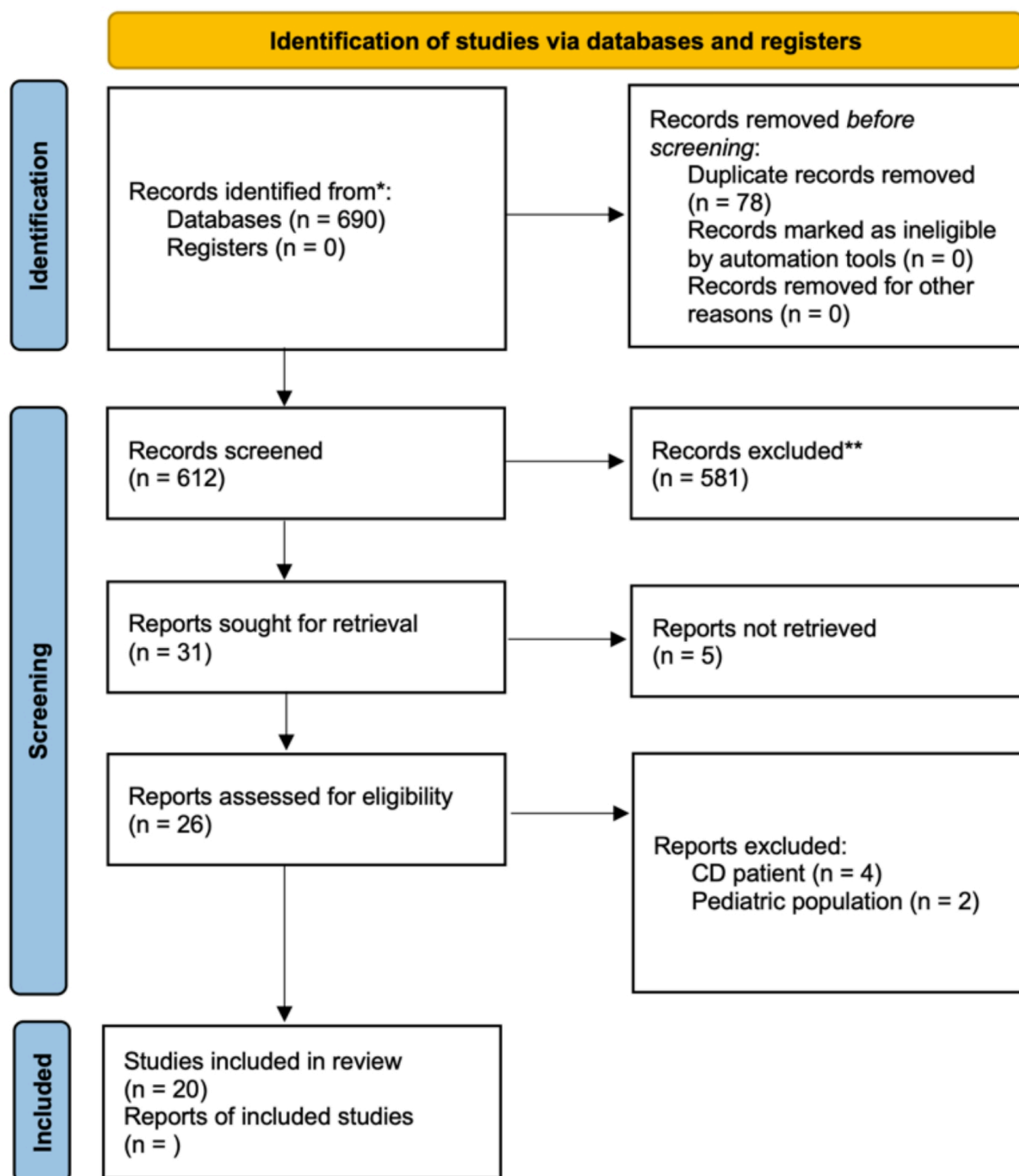


Figure 1

Flow diagram

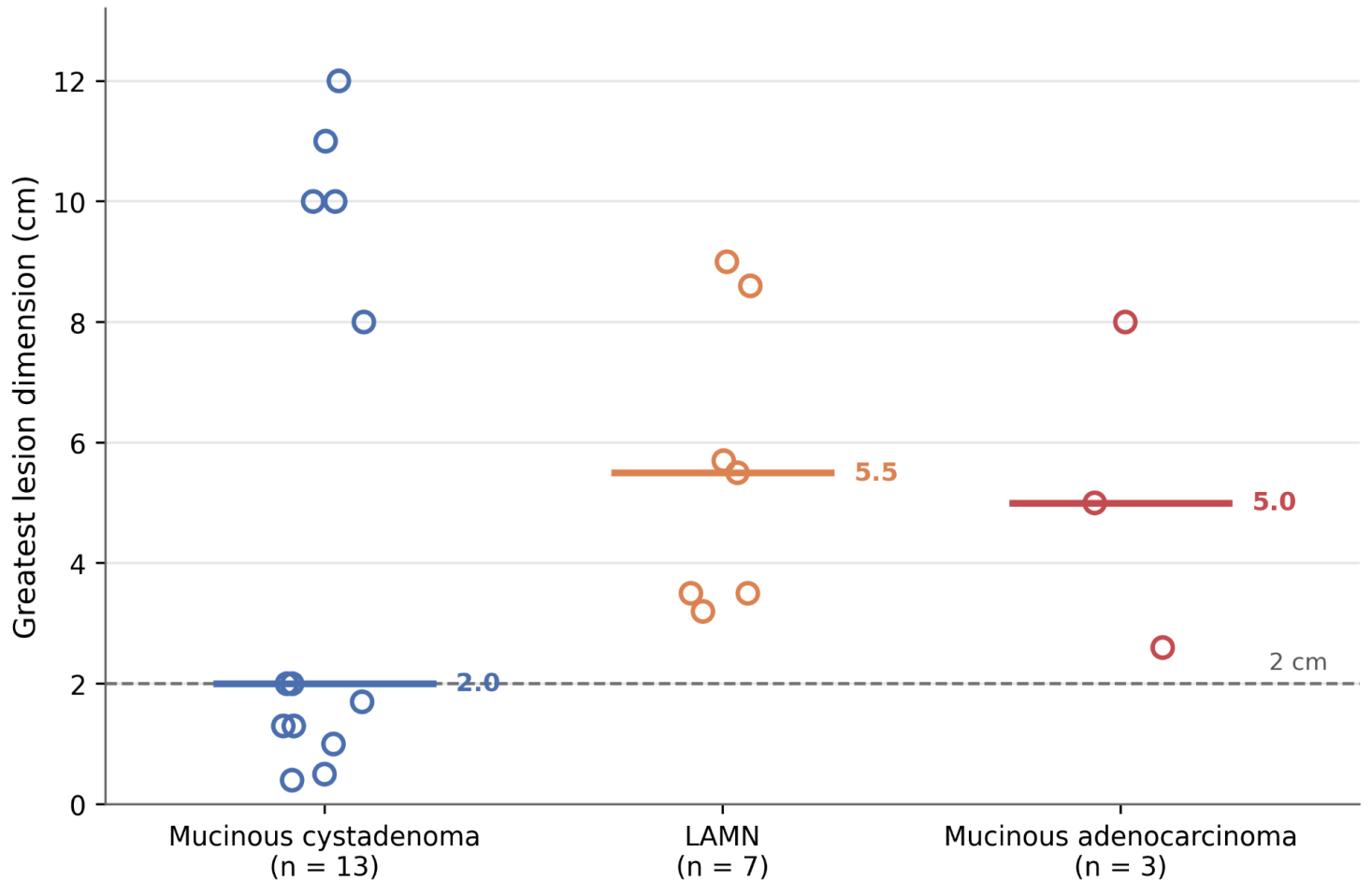


Figure 2

Greatest lesion dimension by appendiceal histopathology.

Each circle represents one patient; horizontal bars indicate group medians, which are also given numerically. The dashed line marks the 2 cm threshold above which appendiceal mucinous neoplasia is conventionally suspected. Data are shown for the 23 of 29 patients in whom the greatest lesion dimension was reported. Mucinous cystadenomas show a bimodal distribution: eight lesions measuring ≤ 2 cm were all incidental microscopic findings within colectomy specimens, whereas four were giant mucocèles measuring 10–12 cm. Because only three patients with mucinous adenocarcinoma had a reported lesion size, no formal between-group comparison was undertaken, and the figure is presented descriptively.

LAMN, low-grade appendiceal mucinous neoplasm.