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ORIGINAL RESEARCH

Low grade mucinous neoplasm of appendix presenting as Mucocoel appendix and systematic review of management of Mucocele appendix

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ABSTRACT

Introduction: Low-grade mucinous neoplasms (LGMNs) of the appendix are rare entities, representing a spectrum of neoplastic disorders characterized by the production of mucin. These neoplasms can lead to the formation of a mucocele, an abnormal dilation of the appendix filled with mucinous material. Appendiceal mucoceles often present nonspecifically, ranging from asymptomatic presentation to mimicking acute appendicitis. Advanced imaging techniques such as CT and MRI play a crucial role in preoperative diagnosis. Surgical resection remains the cornerstone of treatment, with approaches varying based on mucocele size, mucinous dissemination extent, and presence of complications. **Case Presentation**: A 54-year-old female presented with intermittent lower abdominal pain for one year, gradually worsening over time. Examination revealed a soft, non-tender abdomen with a palpable lump in the right iliac fossa. Imaging showed a distended appendix, suggestive of a mucocele. Exploratory laparotomy confirmed a grossly distended appendix which was excised in toto. Histopathological examination revealed a low-grade appendiceal mucinous neoplasm with extensive calcifications. The patient had an uneventful postoperative course and no remnant malignancy at two-month follow-up. **Discussion**: Appendiceal tumors are increasingly recognized, with rising incidence reported in recent studies. The classification and terminology of appendiceal tumors has evolved, with the PSOGI Consensus of 2016 providing a comprehensive framework. Mucocele of the appendix can arise from both benign and malignant causes. Surgical resection, ranging from appendectomy to cytoreductive surgery combined with HIPEC, is essential for management . Accurate diagnosis and appropriate surgical intervention are crucial for improving patient outcomes and minimizing recurrence risk. **Conclusion**: The rising incidence of malignant appendiceal Neoplasms necessitates a refined approach to classification, diagnosis, and management. Surgical resection remains pivotal, tailored to individual patient characteristics and disease extent, with advanced techniques improving outcomes.

Keywords: Low-grade mucinous neoplasm, Appendix, Mucocele, Appendectomy, Cytoreductive surgery, Hyperthermic intraperitoneal chemotherapy (HIPEC), Appendiceal tumors, Pseudomyxoma peritonei

INTRODUCTION

Low-grade mucinous neoplasms (LGMNs) of the appendix are rare entities, representing a spectrum of neoplastic disorders characterized by the production of mucin. These neoplasms can lead to the formation of a mucocele, an abnormal dilatation of the appendix filled with mucinous material. While mucoceles of the appendix can result from a variety of causes, including non-neoplastic processes like obstruction and inflammation, the majority are associated with neoplastic processes such as LGMNs [1].

The clinical presentation of appendiceal mucoceles is often nonspecific, ranging from asymptomatic cases discovered incidentally during imaging, for other reasons, to acute presentations mimicking acute appendicitis or other abdominal pathologies. In some instances, patients may present with chronic right lower quadrant pain, a palpable mass, or gastrointestinal symptoms such as nausea and vomiting [2]. Advanced imaging techniques, particularly computed tomography (CT) and magnetic resonance imaging (MRI), have proven invaluable in the preoperative identification and characterization of these lesions.

Management strategies for mucocele of the appendix, particularly those caused by LGMNs, have evolved

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significantly over the past few decades. Surgical resection remains the cornerstone of treatment, with the approach being dictated by factors such as the size of the mucocele, the extent of mucinous dissemination, and the presence of complications like rupture or pseudomyxoma peritonei [3]. Laparoscopic appendectomy is generally preferred for localized mucoceles without evidence of spread, while more extensive surgical interventions, including right hemicolectomy or cytoreductive surgery combined with hyperthermic intraperitoneal chemotherapy (HIPEC), may be warranted in cases with peritoneal involvement [4].

A systematic review of the management of mucocele appendix reveals a nuanced approach that balances the risks of surgical intervention against the potential for malignant transformation and recurrence. Recent studies emphasize the importance of complete surgical excision and vigilant postoperative monitoring to detect and manage recurrence promptly. Additionally, advancements in minimally invasive surgical techniques and perioperative care have improved patient outcomes, reducing morbidity and hospital stay [5].

The current study presents a case of mucocele of the appendix, identified post histological examination as low-grade appendiceal mucinous neoplasm.

CASE PRESENTATION

A 54-year-old female patient presented to the surgical emergency department with complaints of lower abdominal pain, occurring intermittently over the past year. The pain was described as dull and gradually increasing in intensity with each episode until it reached a constant level.The pain was not associated with any diurnal or postural variations and was alleviated by pain medications prescribed by a local practitioner, though no medical records were available. The initial episodes of pain were accompanied by low-grade fever, which subsided with medication.

The patient reported a regular bowel and bladder habit, without associated nausea or vomiting. There was no history of evening rise in temperature, significant weight loss, or loss of appetite. Additionally, there was no history of melena, altered bowel habits, burning micturition, or vaginal discharge. The patient was a known case of type 2 diabetes mellitus for the past 2.5 years and underwent a total abdominal hysterectomy in 2015. She had no history of addiction, followed a non-vegetarian diet, and maintained adequate appetite and sleep.

Examination

On examination, the patient was conscious, cooperative, and oriented to time, place, and person.

- **Vitals:**
  - Blood Pressure: 104/68 mmHg
  - Pulse Rate: 77/min
  - SpO2: 98%
  - Temperature: Afebrile
- **General Examination:**
  - Pallor: Absent
  - Icterus: Absent
  - Cyanosis: Absent
  - Clubbing: Absent
  - Edema: Absent
  - Lymphadenopathy: Absent
- **Systemic Examination:**
  - **CNS:** No neurological deficits were elicited.
  - **CVS:** S1 and S2 heard, with no added sounds.
  - **Chest:** Bilateral equal air entry present and adequate; no adventitious sounds heard.
  - **Per Abdomen:**
    - Inspection revealed a slightly distended abdomen with a centrally placed umbilicus and a midline transverse scar mark present in the lower abdomen. No visible peristalsis was observed.
    - On palpation, the abdomen was soft and non-tender. A lump was palpable in the right iliac fossa, non-tender, with no appreciable mobility.
    - Digital rectal examination revealed no significant findings.

Investigations

- Hemoglobin: 11.4 g/dL
- Total Leukocyte Count: 4.26 x $10^3/\mu L$
- C-Reactive Protein: 28 mg/L
- **Ultrasound Examination:** Revealed a tubular, hypo-echoic, non-compressible, aperistaltic structure arising from the caecum with a diameter of 3 cm.
- **Contrast-Enhanced CT Whole Abdomen:** Reported a grossly distended appendix with a length of 8.8 cm and diameter of 3.5 cm, with mild peripheral mural calcification and no surrounding stranding. These findings were suggestive of an appendicular mucocele with peripheral mural calcification.

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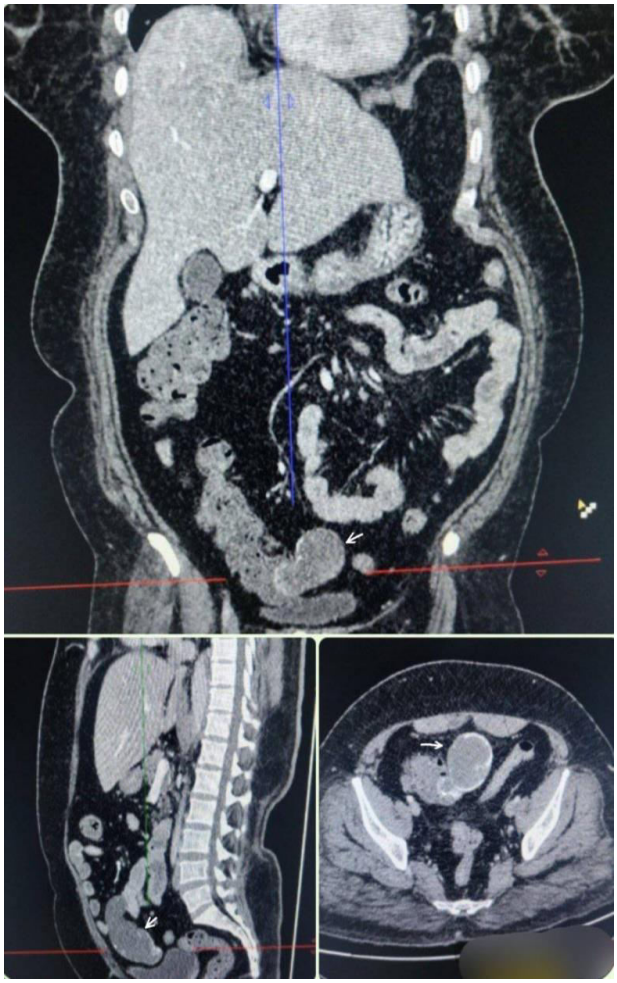

Figure 1

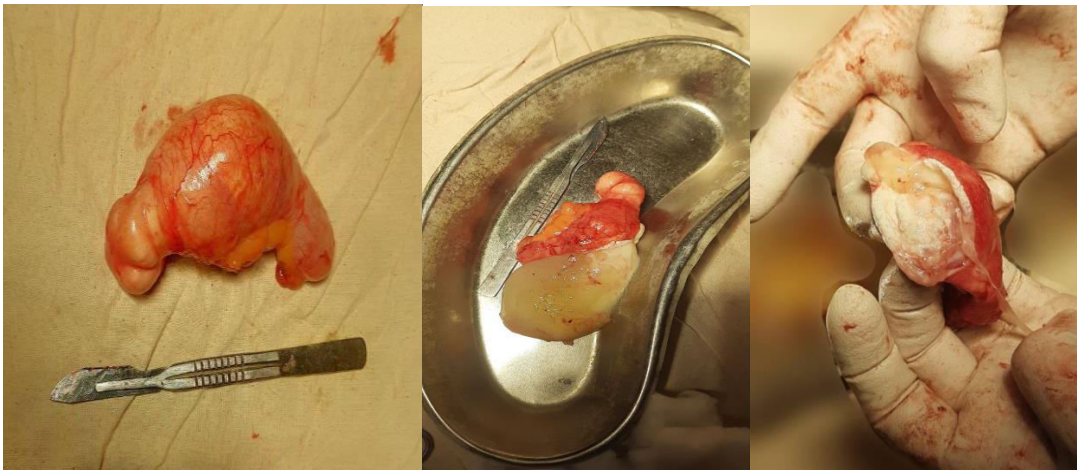

Figure 2

INTERVENTION

The patient was planned for an exploratory laparotomy. Intraoperative findings revealed a grossly distended appendix, which was excised in toto, including the adjacent mesoappendix, without any breach. The abdomen was thoroughly examined for evidence of intraperitoneal mucin or significant mesenteric lymphadenopathy.

Gross examination of the cut section of the appendix revealed mucinous material within, with the wall being thinned out and the mucosa appearing grayish-white and velvety in some areas. The specimen was sent for histopathological examination (HPE).

Microscopic analysis showed epithelial cells with elongated nuclei and low-grade nuclear atypia, with extensive mucin dissecting the wall. Extensive dystrophic calcification was also observed. These findings were suggestive of a low-grade appendiceal

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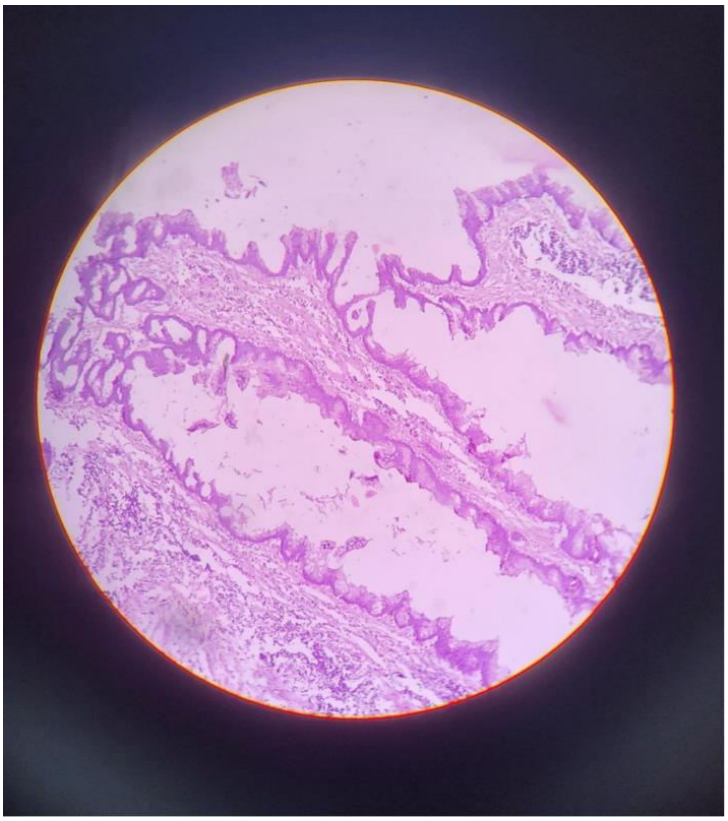
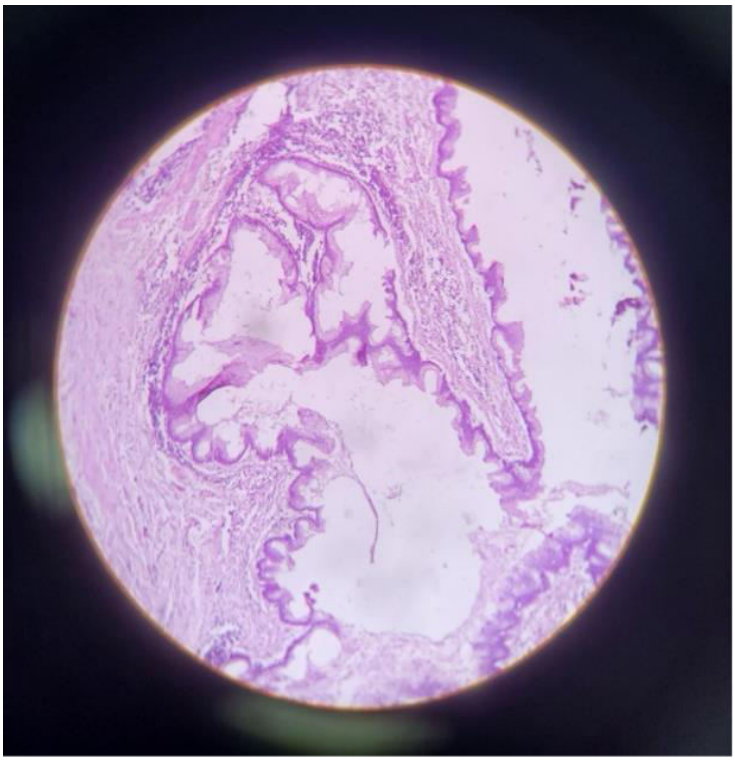
mucinous neoplasm with extensive calcifications.

The postoperative course was uneventful, and the patient was discharged with advice for follow-up. A two-month follow-up CT scan revealed no features suggestive of any remnant malignancy.



Slide1. Shows mucin dissecting in the epithelial layers.



Slide2. shows the epithelium resting on mural fibrosis with the muscularis mucosa being obliterated in certain areas.

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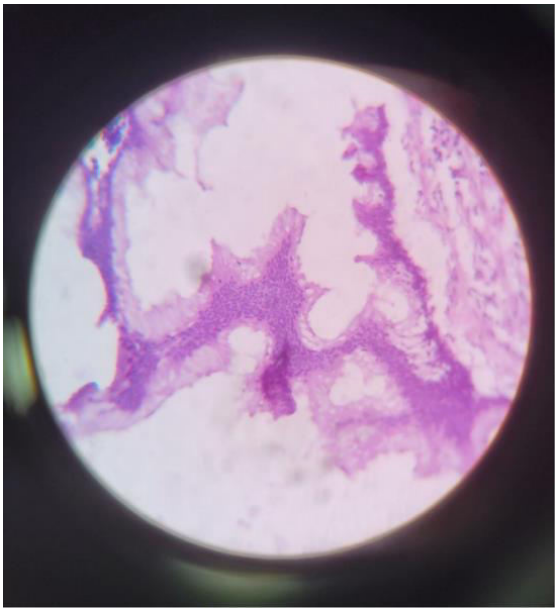
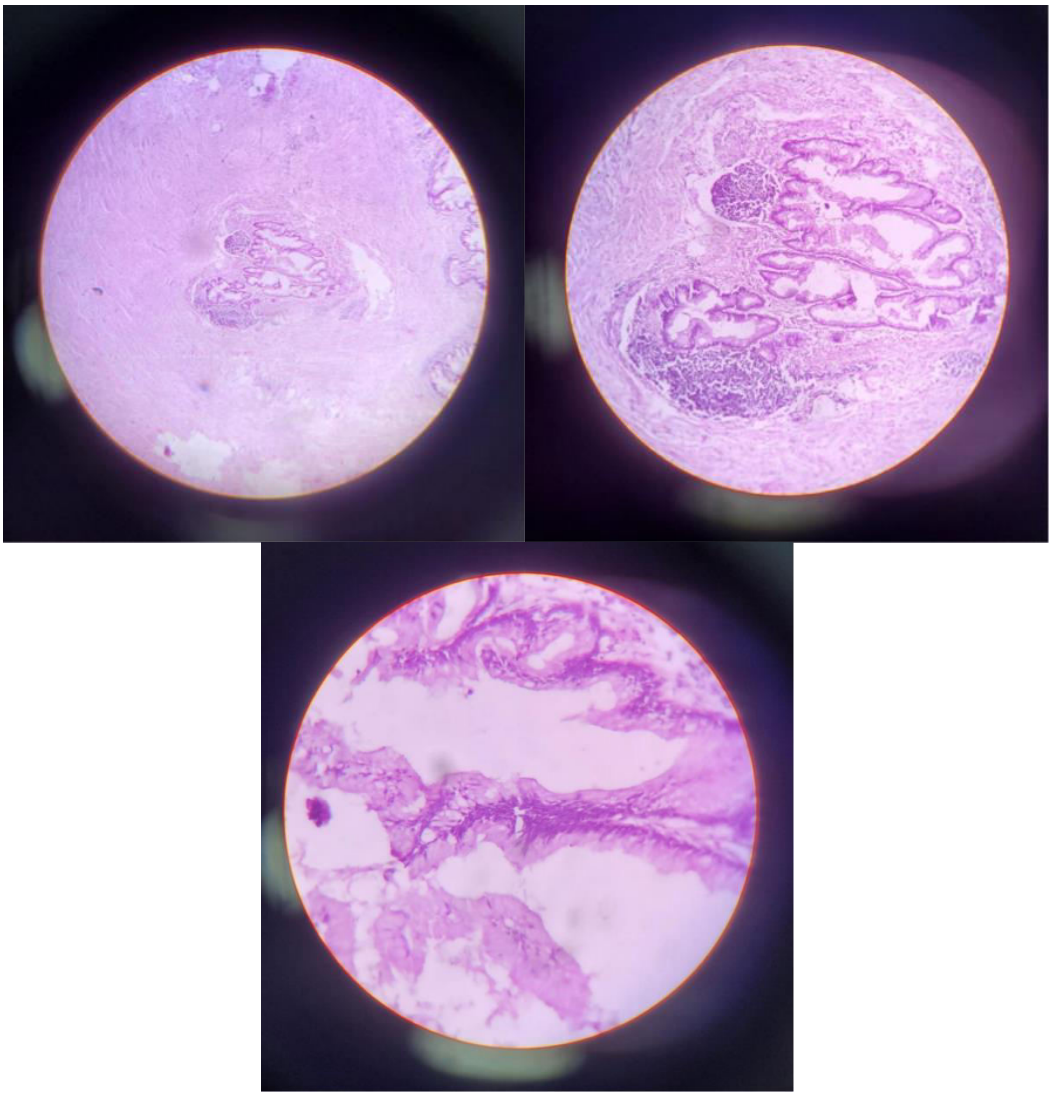


Slide3. shows a zoomed in section showing low grade nuclear atypia.



Slides 4,5 and 6 show 4x, 10x and 40x magnifications with tall mucinous cells in a villus configuration and dysplastic cells.

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DISCUSSION

Appendiceal tumors were historically considered rare causes of abdominal malignancy. However, recent studies from North America [6] and Europe [7] indicate a rising incidence of malignant appendiceal neoplasms. The overall incidence of appendiceal tumors increased from 0.3 per 100,000 to 1.6 per 100,000 over the study interval [7]. Due to the increasing incidence and significant potential morbidities, the classification and terminologies related to appendiceal tumors have evolved and continue to be refined.

The widely accepted Peritoneal Surface Oncology Group International (PSOGI) Consensus of 2016 classifies appendiceal neoplasms as follows:

- **Epithelial neoplasm - Mucinous epithelial neoplasm:**
  - Serrated sessile polyp (with/without dysplasia)
  - Low-grade appendiceal mucinous neoplasm (LAMN)
  - High-grade appendiceal mucinous neoplasm (HAMN)
  - Mucinous adenocarcinoma
- **Non-mucinous epithelial neoplasm:**
  - Adenoma
  - Adenocarcinoma
- **Epithelial neoplasm with neuroendocrine features:**
  - Goblet cell carcinoid
  - Neuroendocrine tumors
- **Mesenchymal neoplasm**

The recent TNM staging guidelines are provided in the AJCC 8th edition [8].

Mucocele of the appendix refers to the cystic dilation of the appendix filled with an abnormal amount of mucinous content. This condition was first described by Rokitansky in 1842 and later by Feren in 1976 [9]. The etiology of mucocele development ranges from benign to malignant causes. Benign causes include chronic obstruction from repeated acute appendicitis attacks, extrinsic compression, or obstructing appendicoliths, generally resulting in swellings less than 2 cm in diameter. Histopathologically, these show degenerative epithelial changes without mucosal hyperplasia or dysplasia, including simple mucous retention cysts and inflammatory mucoceles. Malignant causes include, but are not limited to, sessile serrated polyps (with or without dysplasia), low-grade or high-grade appendiceal neoplasms, and adenocarcinomas.

Studies have reported varying incidences of mucocele due to different etiologies. Woodruff et al. found the incidences to be 18% for simple retention cysts, 20% for hyperplasia, 32% for appendiceal neoplasms, and 10% for mucinous adenocarcinomas [10]. Stocchi et al., in a study of 135 patients with appendicular mucocele, found that 48% were simple mucoceles, 16% were due to appendiceal mucinous neoplasms, and 36% were due to adenocarcinomas [11]. Abreu Filho et al. reported that simple and hyperplastic mucoceles constituted 5%-25% of cases, mucoceles due to appendiceal mucinous neoplasm constituted 63%-84%, and mucinous adenocarcinomas constituted 11%-22% of cases [12].

The clinical presentation of appendiceal mucocele is generally nonspecific, ranging from asymptomatic (51%) to abdominal pain (37%), nausea and vomiting, acute appendicitis (8%), changes in bowel habits, unexplained anemia, and obstipation. Kabbani et al. found that 70% of mucinous carcinomas presented as pseudomyxoma peritonei, 4% as appendicitis, and 9% as a right lower quadrant mass [13]. Ruiz Tovar et al. reported that pain in the right iliac fossa (66%) was the most common presentation for appendicular mucocele, followed by a mass in the right iliac fossa (17%), weight loss, anorexia, and lower gastrointestinal bleeding (3% each) [14].

Following a thorough history, a complete physical examination including a per rectal examination, is essential. Abdominal examination may reveal a palpable lump in the right lower quadrant or palpable dilated small bowel loops, depending on the pathology's localization or generalization and the presence of obstructive features. Per rectal examination findings can range from no significant findings to a large appendicular mass or a boggy consistency in the rectovesical pouch, as seen in pseudomyxoma peritonei. If suspicion arises, the clinical findings are further corroborated with imaging findings.

Imaging modalities like ultrasound and CT scan are regularly utilized to clinch the diagnosis. Typical ultrasonographic findings include cystic structures with thin walls, internal echoes and septation, and complex masses with acoustic enhancement [15]. The "onion skin" sign, due to layered and repeated sedimentation of mucin, is highly suggestive of mucocele appendix. Kameda et al. concluded that the sensitivity, specificity, and accuracy of the onion ring sign for appendiceal mucocele were 63%, 100%, and 99%, respectively [16].

Typical CT scan findings include a low-attenuating, well-encapsulated round or tubular cystic lesion in the right lower quadrant. Differentiating between appendicitis and appendicular mucocele is essential, as both conditions may present similarly. Genevieve L. Benett et al. concluded that discriminating features include maximum luminal diameter, mural calcification, and cystic dilation of the appendix [17]. Marota Bradley et al. found that a combination of findings, such as focal distal dilation, absence of peri-appendiceal fat stranding, diameter greater than 2 cm, and mural calcification, had a positive predictive value approaching 100% for neoplastic causes [18].

Many cases of appendicular mucocele are asymptomatic, and appendiceal masses are often incidentally detected on imaging. Appendicular mucoceles are incidentally found in 9.7% of patients with right lower quadrant complaints, with 16%

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having radiological features suggestive of neoplasm [19]. Radiological features indicative of neoplastic origin include:

- Mucocele diameter greater than 2 cm, suggesting a tumor. This is in contrast to its benign counterpart, due to luminal obstruction, which is known as a mucus retention cyst and is generally small.
- Dilated appendix filled with low attenuation material with or without mural calcification.
- Pseudomyxoma peritonei, visible as low attenuating deposits in typical locations like the omentum, pericaecal mesentery, and rectovesical space. “Scalloping of the solid organs” might also be seen as the mucus displaces the solid viscera.

Tumor markers such as CEA, CA125, and chromogranin A are more utilized after a definitive diagnosis for post-treatment surveillance and prognostication [20]. A colonoscopy is also warranted in suspected cases of appendiceal neoplasm, as synchronous tumors are present in the colon in 13% to 42% of cases. Colonoscopic findings may include a simple rounded mass protruding into the caecal lumen or a mound-like elevation of the appendicular lumen with exudate oozing from it, known as the “Volcano sign” [21].

A definitive diagnosis is generally made post histopathological examination, as intraoperative findings may be obscured by features of acute inflammation. A thorough examination of the appendix, bilateral ovaries, and the rest of the GI tract must be done if an intraoperative isolated mucin pocket is found, and the mucin sent for histopathological examination without further spillage. This examination is of great importance as an acellular mucinous collection has a better prognosis than a highly cellular mucinous collection.

If an intraoperative finding of appendiceal mucocele is noticed, an appendicectomy should be done for all mucoceles greater than 2 cm to obtain a histopathological examination for both diagnostic and therapeutic purposes. Major surgeries should be avoided without a pathological diagnosis. Santiago Gonzalez-Moreno and Paul H. Sugarbaker suggested a concept of Radical Appendicectomy as an alternative to right hemicolectomy, ensuring an adequate negative resection margin and allowing the examination of resected lymph nodes. Even a single lymph node found positive for neoplastic cells warrants a right hemicolectomy[22].

Intraoperative spillage of mucinous content must be avoided. Further surgical interventions depend on intraoperative findings, achieving a clear resection margin, and preoperative imaging findings suggesting a more generalized disease. An intraoperative frozen section is challenging due to the complex pathology, and a single frozen section cannot reliably diagnose the lesion [20].

For mucocele cases due to serrated polyps or other non-neoplastic lesions, a simple appendectomy is definitive, and no further treatment or surveillance is necessary. For primary mucinous neoplasms, management guidelines consider the neoplasm grade, T stage, disease extent, involvement, and microscopic findings of any extravasated mucinous material.

The grade of an appendiceal neoplasm is defined by cellular atypia observed on microscopy. Low-grade appendiceal mucinous neoplasm (LAMN) is defined by low cellular atypia and any of the following features per the PSOGI consensus, 2016 [23]:

- Loss of muscular mucosa
- Fibrosis of submucosa
- Pushing invasion or diverticulum-like growth
- Dissection of acellular mucin in the wall
- Undulating epithelial growth
- Rupture of the appendix
- Mucin or cells outside the appendix

High-grade appendiceal mucinous neoplasm (HAMN) is characterized by high-grade cellular atypia with the above findings. This relatively new category was introduced by the PSOGI consensus, 2015, and was earlier referred to as cystadenocarcinoma [23]. HAMN and LAMN share high rates of GNAS and KRAS co-mutations, with additional mutations like TP53 or ATM potentially driving progression to higher cellular atypia [24].

The T stage of LAMN differs from that of HAMN, as per the AJCC 8th edition recommendation in 2018. Gonzalez et al., in 2022, reported no significant difference in the aggression of HAMN compared to LAMN. Further studies with larger sample sizes are needed to better understand the behavior of HAMN [25].

Localized extra-appendiceal mucin, once considered clinically insignificant, is now a major prognostic factor. Rhounda K. Yatis et al. found that 4% of patients with acellular peri-appendicular mucin developed diffuse peritoneal disease compared to 33% of those with cellular mucinous components [26]. Pai et al. noted that 1 out of 14 patients with LAMN with acellular peritoneal mucin developed disease recurrence, with no deaths observed [27].

Negative margin resection is crucial for managing appendiceal mucinous tumors. The extent of resection needed for a negative margin is debated. Thomas Arnason et al. found no difference in survival rates between patients with positive margins undergoing a second procedure and those who did not [28]. Gonzalez-Moreno et al. suggested that the type of surgical procedure impacts patient survival, with appendectomy alone having a median survival of 18 years compared to 10 years for right hemicolectomy. He hypothesised that the entrapment of tumour cells in the site of right hemicolectomy was the cause of recurrence and hence Hyperthermic Intraperitoneal Chemotherapy must be given with right hemicolectomy to prevent the retroperitoneal spread of the disease and increase the anastomotic line recurrence risk [29].

In 2020, a panel of worldwide experts provided

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recommendations for managing appendiceal tumors using the Delphi technique [30].

Table 1: The interventions along with the strength of recommendation by the panel[30].

	Intervention			
	Right hemicolectomy		Adjuvant CRS+HIPEC	
LAMN with no post operative residual disease				
Non perforated (pT_{is-3},N_x,M_0)	×(Strong negative)		×(strong negative)	
Perforated				
Acellular mucin (pT_{4a-b},N_x,M_0)	×(Weak negative)		√ (weak positive)	
cellular mucin (pT_{4a-b},N_x,M_0)	×(Weak negative)		√ (weak positive)	
Metastatic(any T, any N, M_1)				
acellular (M_{1a})	× (Strong negative)		√ (weak positive)	
cellular (M_{1b})	× (Strong negative)		√ (weak positive)	
HAMN and Mucinous Adenocarcinoma				
	Right hemicolectomy		Adjuvant CRS+HIPEC	
	HAMN	Mucinous Adeno Ca	HAMN	Mucinous Adeno Ca
Non perforated	√(Weak positive)	√(Strong positive)	√(Weak positive)	√(Weak positive)
Perforated	√(Strong positive)	√(Strong positive)	√(Strong positive)	√(Strong positive)
Metastasis with cellular mucin	√(Strong positive)	√(Strong positive)	√(Strong positive)	√(Strong positive)

Cytoreductive surgery followed by hyperthermic intraperitoneal chemotherapy (CRS + HIPEC) is the recommended treatment modality in certain cases as mentioned above. HIPEC involves delivering high-temperature chemotherapeutic agents into the peritoneal cavity following cytoreductive surgery, with different regimens with oxaliplatin or mitomycin-C. Extensive small bowel serosal involvement and mesenteric involvement causing retraction are absolute contraindications to CRS.

CONCLUSION

Low-grade mucinous neoplasms of the appendix, though rare, present significant clinical challenges due to their potential to form mucoceles and mimic other abdominal pathologies. Early and accurate diagnosis using advanced imaging techniques is crucial for effective management. Surgical resection remains the cornerstone of treatment, with the extent of surgery tailored to the individual patient's disease characteristics. The rising incidence of appendiceal neoplasms underscores the need for ongoing research and refinement in classification, diagnosis, and management strategies to optimize patient outcomes and minimize recurrence risk.

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