Journal of Cardiovascular Disease Research

ISSN: 0975-3583,0976-2833 VOL15, ISSUE 08, 2024

Primary Mucinous lesions of Appendix: A Case series

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Received: 20-06-2024 / Revised: 30-06-2024 / Accepted: 08-08-2024

ABSTRACT

Background
Primary neoplasms of the appendix are rare, present in less than 2% of surgical appendectomies. They are frequently misinterpreted in histopathological examination due to their relative rarity. This study focuses on understanding the disease burden of appendiceal mucinous lesions in our tertiary centre.
Methods
A retrospective analysis of all non neoplastic and neoplastic mucinous lesions in appendicectomy specimens were done from January 2021 to May 2024, which were received in the Department of Pathology in our tertiary care center at Wayanad. All the specimens and slides available were retrieved from the archives. The clinical presentation, radiological findings, gross and microscopic findings were collected from the HIS (Hospital Information System) and records available in the Department of pathology.
Results
From the 3 years and 5 months data of all appendicectomy specimens, 7 cases of mucinous appendiceal lesions were identified. 4 out of these were non neoplastic, simple mucocele. Almost all of which were clinically diagnosed as acute appendicitis. 2 cases were LAMN (Low Grade Mucinous Appendiceal Neoplasm) and 1 was HAMN (High Grade Mucinous Appendiceal Neoplasm).
Conclusion
Mucinous neoplasms of appendix are rare tumours with challenging clinical management. Staging of LAMN and high grade tumours is necessary to determine treatment options and prognosis. Mucoceles and Low-grade tumours may be surgically treated while high-grade tumours may require other options including debulking surgery and chemotherapy.

Keywords: Mucinous lesions of appendix, Appendiceal Mucinous Neoplasms, Primary tumours of appendix, Mucocele of appendix, LAMN, HAMN.

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INTRODUCTION

Appendiceal Mucinous Neoplasms (AMN) represent a rare and heterogeneous pathology which is poorly understood. It is present in less than 0.3% of all appendectomies and many a times an incidental finding[1,2]. In the general population, the incidence is low with an average age of diagnosis between 50-60 years of life and a slight predominance in females [1,3]. Appendiceal mucinous lesions are classified histologically as either non-neoplastic or neoplastic epithelial lesions and are subcategorized by histologic subtypes. Accurate histological diagnosis requires a rigorous evaluation of the entire appendiceal specimen. Clinically the patients are often asymptomatic or presents with right lower quadrant (RLQ) abdominal pain which mimics appendicitis in most cases and also has nonspecific laboratory findings [4].

Non neoplastic mucinous lesions of appendix most commonly consists of simple mucocele. It is also called retention cysts of appendix as the aetiology is believed to be related to chronic luminal obstruction. A secondary cystic expansion of the appendix occur distal to the level of obstruction due to continued mucin secretion. The lining epithelium lacks dysplasia. A flattening secondary to increased intraluminal pressure or abundant intracellular mucin content may be present. Increased intraluminal pressure also may lead to mucin extrusion into or through the wall resulting in mucin pools with no lining epithelium [5,6,7].

Mucinous appendiceal neoplasms are mucinous tumours exhibiting dysplasia of the epithelial cells. These tumours have pushing tongues of epithelium which dissects into the muscularis mucosa. However they are confined by the muscularis propria. They lack an infiltrative growth pattern or a stromal desmoplasia. These tumours can be classified as either low-grade appendiceal mucinous neoplasms (LAMNs) or as high-grade appendiceal mucinous neoplasms (HAMNs) based on their cytologic features [6]. The World Health Organization classifies the majority of non-invasive epithelial appendiceal lesions as LAMNs [7].

Mucinous adenocarcinomas of the appendix are frankly infiltrative tumours. The features include tumour budding (discohesive single cells or clusters of up to five cells) and/or small, and irregular glands. A stromal desmoplastic reaction is typically seen. Depending on the cellular differentiation they are categorised into well, moderately or poorly differentiated mucinous adenocarcinomas. The presence of signet ring cells in these is a feature of poorly differentiated category [6,7].

MATERIALS & METHODS

We report a case series of 7 primary mucinous lesions in appendix after a retrospective analysis of all appendicectomy specimens which were studied in our tertiary care centre, DMMC, Wayanad. All the cases were diagnosed in our institution, over past 3.5 years, from January 2021– May 2024. The clinical and radiological findings were collected from the hospital information system. Histopathological evaluation of the archived slides were done in our department of pathology.

CASE PRESENTATIONS

Mucocele of Appendix

Case 1

A 24 year old male presented with sudden onset of abdominal pain, progressive in nature and radiation of pain. There were 3 episodes of non bilious, non blood stained, vomiting. No history of fever, constipation, abdominal distension or dysuria was there. On per abdominal examination, abdomen was soft with tenderness elicited in the RIF(right iliac fossa). Rebound tenderness was present without

ISSN: 0975-3583,0976-2833 VOL15, ISSUE 08, 2024

any guarding or rigidity. His vitals were stable. USG (ultrasonography) of Abdomen and Pelvis, an aperistaltic non compressible tubular structure with a blind end, measuring >8.2cm in length and upto 16mm in diameter, filled with mixed echoic content. Minimal fluid present in the RIF. An impression of Mucocele of appendix was made radiologically. A CECT also confirmed the same. An emergency laparoscopic appendicectomy was performed. Specimen of appendix received in the department of pathology measured 8cm in length. External surface was congested and blackish. Cut section showed luminal dilation filled with mucinous and hemorrhagic material. Microscopy of the same had dilated lumen with mucosal ulcerations, acellular mucin pools, a transmural neutrophilic infiltrates and serosal congested vessels. No epithelial proliferation or dysplasia was noted. A diagnosis of Acute appendicitis with Mucocele of Appendix was made.

Case 2

27 year old young male visited the surgical OPD with complaints of right sided lower abdominal pain of 1 month duration. The pain was insidious in onset and gradually progressive in nature. There was history of on and off fever. No history of vomiting or loose stools or constipation was there. The patient had underwent laparoscopic emergency appendicectomy about 2 months back. Per abdomen was soft with tenderness over the RIF was elicited. Bowel sounds were normal. Laboratory findings were unremarkable. Sliced MDCT scan of abdomen and pelvis showed a tubular structure arising from the caecum, showing luminal fluid in RIF with extensive fat stranding, abutting the caecum and terminal ileal loops. In view of post appendicectomy and with significant intervening mesenteric fat stranding, the features were radiologically favouring stump appendicitis with early mass formation. A laparoscopic stump appendicectomy was followed and specimen sent for histopathological evaluation. The specimen of appendix was enlarged and measured 6cm in length. Cut surface had luminal obliteration and showed a mucinous, jelly like material. The wall appeared thickened. Microscopic examination revealed appendix with focally preserved lining mucosa. Lumen had pools of acellular mucin in multiple sections. The mucin was seen dissecting the layers of appendix including muscularis propria. Mucin was also seen on the serosal surface (artefactual). No infiltration into the wall of appendix. No atypia of the lining epithelium was seen. No atypical cells were seen floating within the mucin pools. A histopathological diagnosis of Simple Mucocele was made.

Case 3

A 75 year old elderly lady with complaints of abdominal pain of 4 weeks duration visited the surgical OPD. The pain was colicky, progressive and non radiating in nature. There was no associated fever or vomiting. Patient had a history of total thyroidectomy 15 years back. She was a hypertensive and was on regular medication. Per abdomen was soft and tender. Blood investigations showed a mild neutrophilic leucocytosis. CT scan of abdomen and pelvis was done and showed features of appendicitis with significant peri appendiceal inflammation with possible early mass formation. A clinical diagnosis of recurrent appendicitis was made. A laparoscopic interval appendicectomy was performed. The specimen received for histopathological evaluation measured 6 cm in length., having congested external surface. Luminal degeneration noted on cut section with dilated distal end. Microscopic examination of the distal dilated end showed dilated lumen containing mucin with flattened epithelium. Cells with apical mucin was seen. Rest of the appendix showed fibrosis and adipose tissue. The final diagnosis made was Mucocele of tip of appendix with proximal fibrous obliteration.

Case 4

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A 49 year old lady had complaints of abdominal pain and discomfort of about 1 month duration. Pain was insidious and progressive with no associated vomiting. On examination tenderness was present in the RIF with rebound tenderness. No laboratory investigations were available. CT scan of abdomen were suggestive of acute appendicitis. Appendicectomy specimen was received in the department of pathology with a clinical and radiological suspicion of acute appendicitis. Gross examination showed a tubular and cystic structure measuring 8x3x2.5cm. wall appeared thickened. On section mucinous material drained. No solid areas were seen. Microscopy revealed appendix with flattened and thinned out epithelium lined by mucinous cells. Wall congested with inflammatory infiltrates. No epithelial dysplasia identified. No atypical cells seen within the extracellular mucin. A diagnosis of Mucocele of appendix was made.

LAMN- Low Grade Appendiceal Mucinous Neoplasia

Case 5

A 49 year old male presented with acute onset of lower right abdominal pain which was non radiating and of progressive nature. He had on and off abdominal pain for past 5 months. There was no associated fever or vomiting. No history of constipation. On examination his vitals were stable. Per abdomen soft with no guarding or rigidity. A tenderness was present in the right iliac fossa. Laboratory investigation were unremarkable. CT scan of abdomen and pelvis showed tubular structure with a blind and dilated end. Tip having a diameter of 10 mm. Minimal fluid in the RIF. An impression of acute appendicitis was made radiologically. An open appendicectomy was done and specimen received in the pathology department. Appendix with peri appendiceal fat measured 6x2x1.5cm. Tip of the appendix showed a nodularity. Cut surface showed a nodular thickening at the tip of the appendix measuring 0.5cm across. No areas of perforation was noted. Wall of the appendix appeared thickened. On microscopy appendix showed a neoplasm composed of villous proliferation of mucosa lined by columnar epithelium having apical mucin. The cells seen to exhibit a low grade nuclear atypia. The lymphoid tissue appeared mildly atrophied. Lumen showed mucinous material. The muscularis propria showed no atypical cells or mucin. The serosal surface also appeared unremarkable. Margins of appendix were free of the neoplasm. A final diagnosis of Low Grade Appendiceal Mucinous Neoplasm (LAMN) was given. No extra appendiceal mucin. Margins appear free of neoplasm. Rest of the appendix showed features of acute appendicitis.

Case 6

A 53 year old male had complaints of abdominal pain in the right side of lower abdomen since 2 weeks. There was associated fever and 3-4 episodes of vomiting. The pain was colicky in nature and gradually progressing. On attending the OPD, per abdominal examination showed a soft abdomen with tenderness in the right iliac fossa. A USG was done in this patient which showed a thickened mass lesion in the RIF. A CT scan was done which showed a thickened appendix with obliterated lumen. Two appendicoliths each measuring 2.5 mm were noted in the base of appendix. Tip of the appendix was not visualised. A peri appendiceal fat stranding was noted. An emergency laparoscopic appendicectomy was done and specimen sent for histopathology. On gross examination appendix measured 8x3x2cm. external surface, thickened and congested. Cut section showed thickened wall, mucinous in appearance. Lumen narrowed and filled with mucinous material. On microscopic examination lumen showed extensive villous proliferation and back to back arranged glands. The lining cells showed abundant mucinous cytoplasm with apical mucin. Nuclear stratification with mild atypia seen. The lumen showed mucinous material. No infiltration into muscularis propria or serosa seen. No high grade nuclear features or peri serosal mucin seen. Margins appeared free of neoplasm.

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A diagnosis of Low Grade Appendiceal Mucinous Neoplasm was given. Serosa and margins free of neoplasm.

HAMN – High Grade Appendiceal Mucinous Neoplasm

Case 7

A 56 year old male presented with cramping type of abdominal pain of 2 day duration. He gave history of constipation since 2 weeks. No history of nausea, vomiting or fever. He is a known case of diabetes on Oral hypoglycemic drugs. Personal and family history were unremarkable. Per abdomen examination showed right iliac fossa tenderness and rebound tenderness. His vitals were stable. USG Abdomen report suggested inflammatory pathology in right iliac fossa, most likely appendicular pathology. CT abdomen and pelvis showed perforated acute appendicitis with collection in right iliac fossa. Inflamed appendiceal base seen flush with caecum. Emergency open appendicectomy was done. Per operatively perforated appendix with mass formation was identified. The gross specimen received in the department of Pathology measured 2.8cm in length. Outer surface showed exudates. Cut section showed mucinous and grey white areas. Microscopy of the appendix showed a neoplasm with villous pattern of glands lined by mucinous columnar cells. The cells contain mucinous cytoplasm with pleomorphic and vesicular nuclei. The neoplasm was seen restricted to the mucosa without breach in the basement membrane. A diagnosis of High grade mucinous neoplasm (HAMN) of appendix was made.

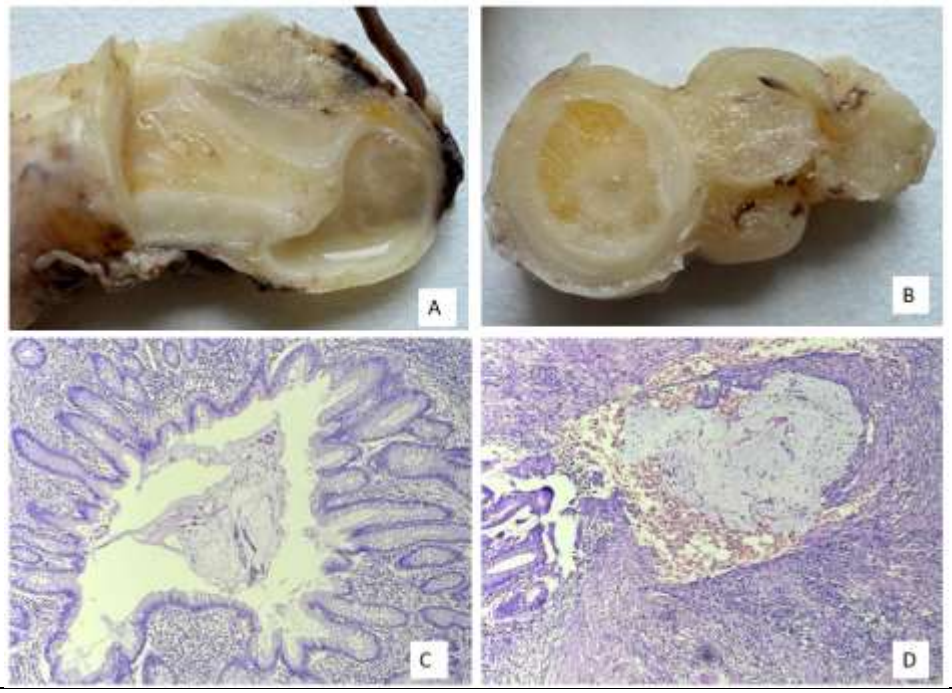

Figure 1 A: Dilated tip of appendix tip, lumen filled with mucinous material, B: Cross section of appendix with thickened lumen filled with mucin.C: Microphotograph of appendix lumen with mucin present in lumen, D: Appendix with mucin infiltrating the wall surrounded by chronic inflammatiory infiltrates.

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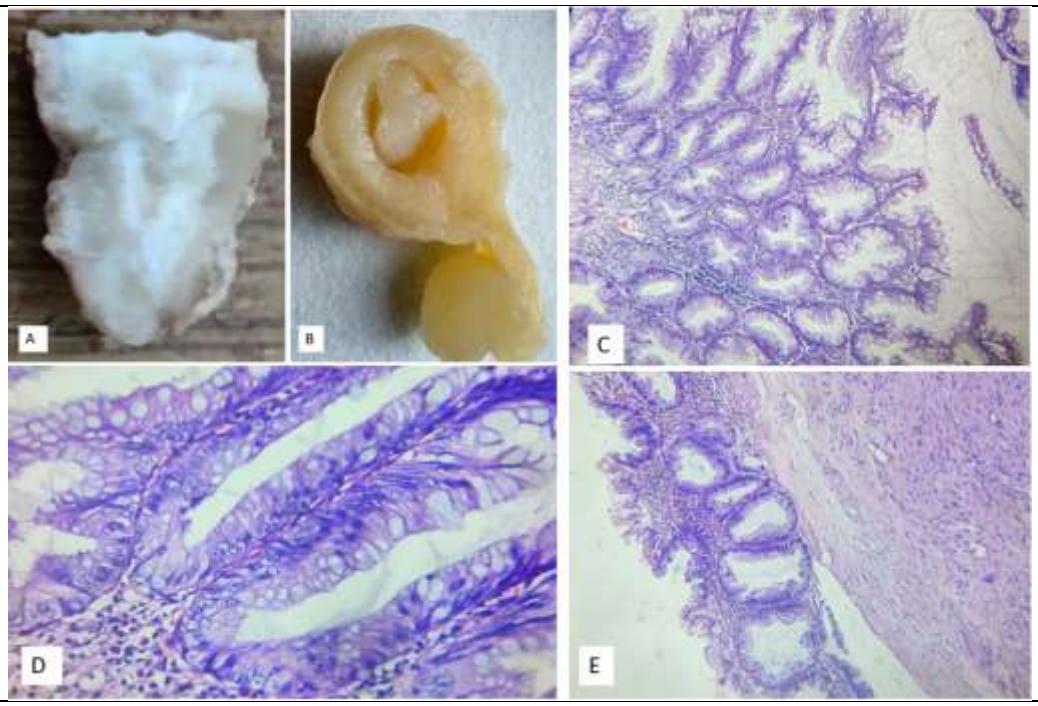

Figure 2A: LAMN (Low grade appendiceal mucinous neoplasm) of appendix, B: Appendix luminal mucinous growth, C: Villous pattern of proliferated luminal glands with luminal mucin, D: LAMN, villous glands having low grade nuclear atypia, E: Luminal proliferation of glands with low grade nuclear atypia.

RESULTS

A retrospective study was done on appendicectomy specimen from January 2021 to May 2024. We had 7 cases of primary appendiceal mucinous lesions. Out of which, 4 cases were mucocele accounting for 57%. Of these 4 patients with mucocele, two were females and other two males. In our study 2 of these cases had acellular mucin pools within the lumen and wall of appendix, limiting to the muscularis propria. One case also had mucin on the serosal surface, however no mucin was found in the muscularis propria or in the sub-serosal plane. Hence it was considered as artefactual serosal mucin. Two cases were diagnosed as LAMN (28.5%) in which mucosal lining had a low grade atypia with mucin pools seen dissecting the wall of appendix. Both patients with LAMN were males. 5 out of 7 cases (71.4%) of these mucinous lesions occurred in age group 49 years or above and 2 patients with simple mucocele were young, less than 30 years. All these patients had presented with similar clinical features with symptoms suggestive of acute appendicitis, having abdominal pain, discomfort, with or without fever and with or without vomiting. We had one patient with HAMN which accounts for 14.2% of mucinous lesions of appendix at our centre. This case was clinically and radiologically suspected as acute appendicitis with features of perforation and appendiceal mass formation. In one of the patient whose histopathological diagnosis was LAMN, CT abdomen showed a thickened appendix with luminal obliteration. One of the cases, diagnosed as mucocele had an early mass formation identified in CT. All other cases had radiological features suggestive of acute appendicitis with or without peri appendiceal inflammation. The results are represented in table 1.

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Sl. No.	Biopsy no.	Age	Sex	Clinical features	Radiological findings	Histopathology Diagnosis
1.	B/196/21	56	M	Crampy abdominal pain	Perforated acute appendicitis	HAMN
2.	B/1598/22	53	M	Abdominal pain	Thickened appendix with obliterated lumen	LAMN
3.	B/2482/23	49	M	Abdominal pain	Acute appendicitis	LAMN
4.	B/2047/24	49	F	Abdominal pain, discomfort	S/o Acute appendicitis	Mucocele
5.	B/2507/24	75	F	Colicky abdominal pain.	S/o acute appendicitis, peri appendiceal inflammation , early mass formation	Mucocele
6.	B/2821/24	27	M	Abdominal pain, post appendicectomy	Stump appendicitis with early mass formation	Mucocele
7.	B/2989/24	24	M	Abdominal pain, Vomiting, RIF tenderness	Mucocele of appendix	Acute appendicitis with mucocele of appendix

Table 1: Age, Sex, Clinical, Radiological findings and Histopathological diagnosis

RIF- Right iliac fossa, LAMN- Low Grade Appendiceal Mucinous Neoplasm, M-male, F-female.

DISCUSSION

Appendicectomy is a one of the most common surgical procedure done with a clinical diagnosis of acute appendicitis. However histopathological examination is the gold standard for diagnosis of lesions of appendix. It not only confirms the diagnosis but also reveal incidental pathologies which has a significant impact on further management of patients[8]. Primary appendiceal tumours are unusual, the majority being neuroendocrine tumours which accounts for 65% of all appendiceal tumours[9]. The other malignant tumours of the appendix include mucinous epithelial neoplasms, goblet cell or mixed carcinoid, lymphomas, adenocarcinomas and lymphoid or mesenchymal sarcomas. Adenocarcinomas which includes mucinous, signet ring or non-mucinous, constitute approximately 20% of these tumours[10]. Accurate histological diagnosis requires a thorough evaluation of the entire specimen. Examination of the entire appendiceal wall for evidence of any epithelial invasion is necessary, a feature that differentiates adenocarcinomas from all other mucinous tumours[6,11,12]. In our study the incidence of LAMN was 2 cases over the past 3 years. One case of HAMN was identified. A case of metastatic deposits in appendix was also received, but since the study was focussed on primary mucinous lesions, the case was excluded. Carr et al.[13] described mucinous neoplasm with uncertain malignant potential in appendix with the characteristics of low-grade tumours having acellular mucin in or beyond the appendiceal wall[14]. Appropriate categorization and nomenclature of these neoplasms remained problematic, with various pathologic grading systems[15,16,17].

Pseudomyxoma peritonei (PMP) is a diffuse collection of gelatinous material in the abdomen and pelvis, and mucinous implants on the peritoneal surfaces[18]. Mucinous appendiceal adenocarcinoma has the potential to progress to Pseudomyxoma peritonei (PMP), which may require extensive surgical intervention and chemotherapy. Studies revealed that the majority of PMP arise from the appendix and represent local spread into the peritoneal cavity. Therefore, it has been

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recommended that the term PMP be limited to describing the clinical entity of mucinous ascites and it should not be used as a histologic diagnosis[19]. AMNs can be associated with colonic polyps and masses and therefore studies have recommended endoscopic investigations in these patients[20]. The treatment of appendiceal mucinous neoplasm is not well defined. There are controversies regarding the extent of surgery, the role of chemotherapy, including early postoperative intraperitoneal chemotherapy (EPIC) and hyperthermic intraperitoneal chemotherapy (HIPEC). However grade of the tumour plays a critical role in choosing cytoreductive surgery/HIPEC versus systemic chemotherapy options[21].

CONCLUSION

Appendiceal mucinous neoplasms (AMNs) are a rare and heterogeneous disease for which clinical management is challenging. Treatment is based on stage and histology. Mucoceles and Low-grade tumours are treated surgically with resection of the primary lesion in early stage disease. A peritoneal debulking and HIPEC (Hyperthermic intraperitoneal chemotherapy) is offered to patients with advanced stage disease. Treatment of high-grade tumours requires further prospective trials, and options include debulking surgery and HIPEC, with or without preoperative chemotherapy.

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