Section: General Surgery

Case Series

APPENDICEAL MUCOCELE: A SERIES OF RARE CASES

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ABSTRACT

Appendiceal mucocele, an uncommon pathological condition, poses diagnostic hurdles due to its varied clinical presentations. Mucoceles arise from the abnormal buildup of mucus within the appendix, causing distension and potential complications like rupture, peritoneal pseudomyxoma, or malignant progression.[1,2] In this case series, we report on eight exceptional cases of appendiceal mucoceles encountered in the last 35 years, comprising four females and four males, falling within the age range of 50 to 75 years, who sought evaluation and treatment from the General surgery department.

Keywords: Appendicitis, Appendix, Appendiceal mucocele, Appendicectomy.

INTRODUCTION

Appendiceal mucoceles are uncommon pathological entities characterized by abnormal dilation of the appendix due to the accumulation of mucoid material within its lumen. Appendiceal mucocele (AM) was initially documented by Rokitansky in 1842, and subsequently, in 1976, Feren provided a formal definition for this condition. It is an exceedingly rare finding, appearing in only 0.2% to 0.7% of dissected appendix specimens. While appendiceal mucoceles represent a rare subset of appendiceal pathology, they pose diagnostic and therapeutic challenges due to their varied clinical presentations and potential for complications. Despite their rarity, recognizing and appropriately managing appendiceal mucoceles are essential to prevent potential morbidity and mortality associated with this condition.[2]

In this case series, we present a collection of eight rare cases of appendiceal mucoceles. Through the detailed examination of each case, we aim to provide insights into the clinical presentation, diagnostic workup, surgical management, and outcomes associated with appendiceal mucoceles. Additionally, we highlight the importance of considering appendiceal mucoceles in the differential diagnosis of patients presenting with abdominal symptoms and emphasize the significance of timely intervention to optimize patient outcomes.

CASE-1

A 70-year-old female reported to the emergency department, with complaints of pain in the right lower quadrant of the abdomen, fever, nausea and vomiting since three days. On physical examination patient was conscious, febrile (38°C) with tachycardia and hypertension. On palpation rebound tenderness in the right lower quadrant was observed. Laboratory investigation revealed Leukocytosis with left shift. The ultrasonography (USG) revealed distended loops of intestines filled with gas in the right lower quadrant of the abdomen. A provisional diagnosis of acute appendicitis was made and open surgery was performed. During the surgery, a cystic mass of appendix measuring 8.0×5.0×3.1 cm was found, with inflamed walls without perforation, located in the right iliac fossa. An appendiceal

mucocele was suspected. For confirmation of diagnosis resected specimen was sent for histopathological examination which was suggestive of hyperplastic mucocele (figure-1) showing compressed appendiceal mucosa with lumen showing acellular mucinous pool.

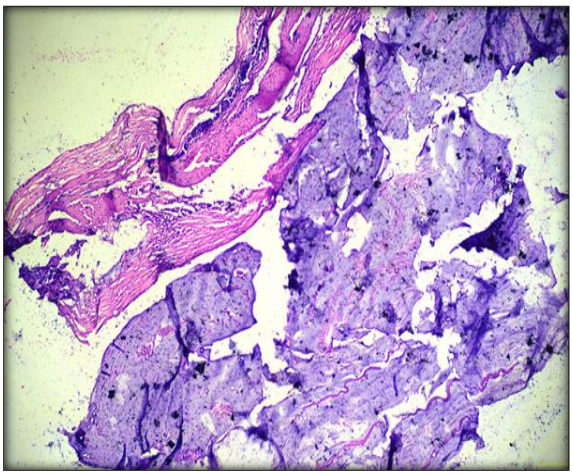

Figure 1: Compressed appendiceal mucosa with lumen showing acellular mucinous pool

The post-operative recovery was uneventful. Patient was managed with Intravenous (IV) antibiotics, analgesics, antacids and other supportive treatment.
She was discharged on oral medications after 5 days. The patient was advised to take preventive measures such as avoiding lifting heavy weights and consuming heavy fatty meals. She was instructed to report immediately if she experienced severe pain or fever. The patient now attends routine follow-up appointments with the surgeon and is in a completely stable condition with no new complaints.

CASE-2

A 66-year-old female reported to the department of General Surgery, with complaints of vague pain in the right side of the abdomen since one month along with loss of appetite and generalized weakness since last four months. On physical examination vitals were stable. On palpation mild tenderness in the right iliac fossa was noted. Laboratory investigations were within normal limits. Whole abdomen sonography was suggestive of an elongated (11cm. long and 2.3-2.6 cm. thick) hypoechoic tubular shadow with mixed echogenic appearance and linear central echoes extending in entire length medial to the cecum possibly appendiceal mucocele. Contracted gall bladder with multiple calculi was also seen on sonography. Laparoscopic cholecystectomy with appendicectomy was performed. During the surgery, a distended appendix with thickened walls was found. No discharge was found in the peritoneal cavity.

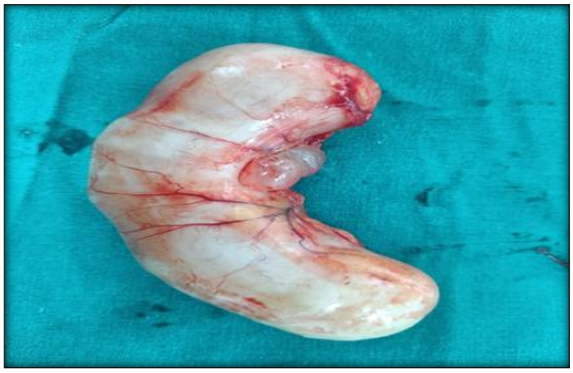

Figure 2: Appendicular mucocele showing distension and thickened walls.

Post operatively the resected specimens (Figure-2) were sent for histopathological examination to confirm the diagnosis. Features were consistent with chronic calculus cholecystitis with cholesterolosis and mucocele of appendix.
Patient stood the procedure well. The post-operative recovery was uneventful. Patient was managed with Intravenous fluids, antibiotics, analgesics, antacids and other supportive treatment.
She was discharged on oral medications after 5 days in a stable condition. The patient was advised to report back immediately if she experienced fever or severe pain. She was also advised to avoid lifting heavy weights and consume frequent small meals at regular intervals. The patient now attends routine follow-up visits with the surgeon and is in excellent health, with no new issues or complaints.

CASE-3

A 60-year-old diabetic female presented to Surgery department with complaints of right-sided abdominal pain, nausea, and unintentional weight loss over several months. Initial physical examination revealed tenderness in the right lower quadrant. Vitals were stable. Laboratory investigations were unremarkable. An ultrasound of the abdomen and pelvis revealed a mixed echogenic pelvic mass with an echogenic rim, initially suggestive of an ovarian origin. Further evaluation with a CT scan of the abdomen and pelvis demonstrated a calcified adnexal cyst measuring 7.3×2.5×3.0 cm, with no evidence of lymphadenopathy.

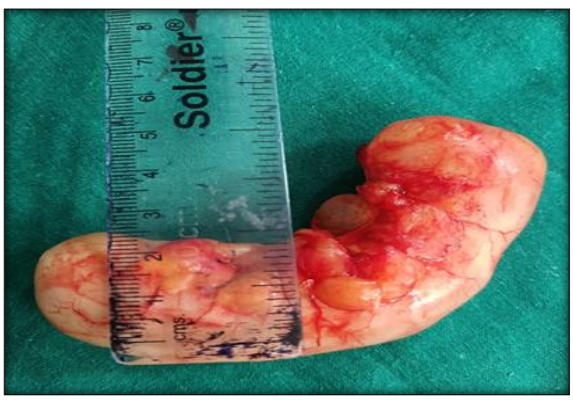

Figure 3: Resected distended appendix measuring 7.3×2.5×3.0 cm.

Given the imaging findings, the patient was referred to a gynecologist for further management. However, during exploratory laparotomy, the only pathology identified was a distended appendix (Figure-3). As a result, a routine appendicectomy was performed to address the appendiceal issue. Histopathological examination of the resected appendix confirmed the diagnosis of an appendiceal mucocele.

The patient tolerated the procedure well, and post-operative recovery was smooth without any complications. Intravenous fluids, antibiotics, analgesics, antacids, and other supportive treatments were administered as needed during the hospital stay. After three days, the patient was discharged in stable condition and transitioned to oral medications.

Upon discharge, the patient received instructions to report any signs of fever or severe pain promptly. Additionally, advice was given to avoid heavy lifting and consume frequent small meals at regular intervals to aid in recovery. The patient now attends routine follow-up appointments with the surgeon, where they are found to be in excellent health with no new complaints or issues.

CASE-4

A 52-year-old obese male (Body Mass Index-30kg/m2) was referred to the Surgical Outpatient Department with complaints of acute right lower abdominal pain, abdominal discomfort, fever, nausea and vomiting since two days. On physical examination patient was conscious, febrile (37.5°C) with tachycardia and tachypnea. On palpation of abdomen rebound tenderness in the right lower quadrant with muscle guarding and rigidity was noted, which was greatest at Mc Burney's point. Laboratory investigations showed leukocytosis. Ultrasonography (USG) revealed a dilated appendix with surrounding inflammatory changes, consistent with acute appendicitis. Computed tomography (CT) scan confirmed the findings of an enlarged appendix with peri-appendiceal inflammation, suggesting acute appendicitis.

The patient was taken for an emergency laparotomy with a preoperative diagnosis of acute appendicitis. Intraoperatively, a distended appendix with a cystic mass at its base was identified. The appendix measured 8x3x2.5cm in length and exhibited cystic dilation consistent with a mucocele. Given the suspicion of an appendiceal mucocele and no evidence of perforation, a decision was made to perform an appendicectomy. The intraoperative image of appendiceal mucocele has been shown in figure-4.

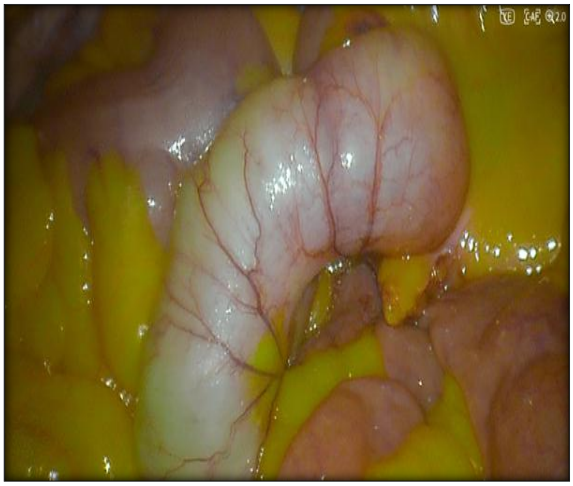

Figure 4: Intraoperative picture of appendiceal mucocele.

Histopathological examination of the resected appendix confirmed the diagnosis of an appendiceal mucocele. The appendix showed significant dilation with mucinous material filling its lumen, characteristic of a simple mucocele.

The post-operative period was uneventful. Patient was managed with Intravenous fluids, analgesics, antibiotics, proton pump inhibitors and other supportive treatment.

After a five-days hospital stay, the patient was discharged in stable condition with oral medications. Instructions were given to report immediately if experiencing fever or severe pain, and to avoid heavy lifting while consuming frequent small meals at regular intervals. The patient was also advised to walk as frequently as possible. The patient now attends routine follow-up appointments with the surgeon and remains in a completely stable condition with no new complaints.

CASE-5

A male patient aged 74 years presented to the hospital with complaint of dull pain in right iliac fossa and in inguinal region for 2-3 days. He was thereby admitted for further investigation and management. The patient then underwent whole abdomen USG.

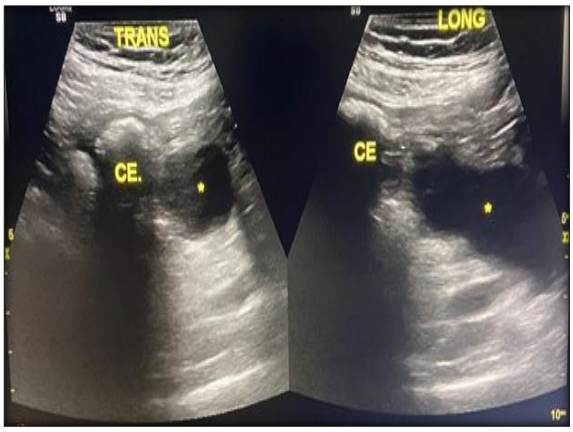

Figure 5: USG Image showing Appendiceal Mucocele

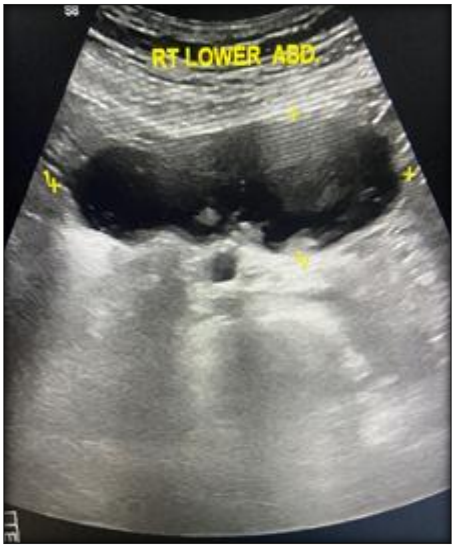

Figure 6: USG Image showing Appendiceal Mucocele

The USG report had an impression of mucocele appendix. On medial aspect of cecum there was an elongated approximately 9.7 x 3.2 cm cystic tubular focus (Figure-5&6). He was also diagnosed with B/L inguinal hernia.

After all appropriate investigations and preparations, informed consent was taken for surgery and Laparoscopic Appendicectomy was performed.

The patient stood the procedure well. Post operatively sample of appendix was further sent for biopsy.

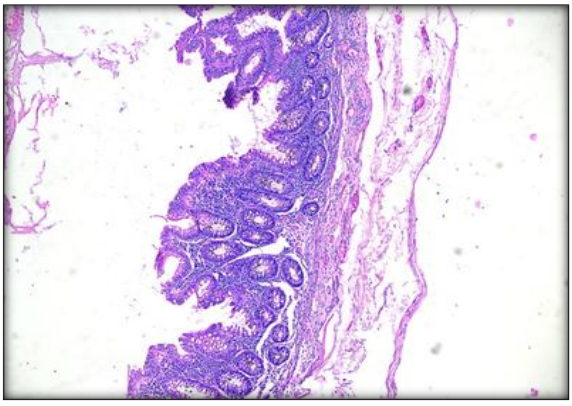

Figure 7: Microscopic view of Mucocele of Appendix

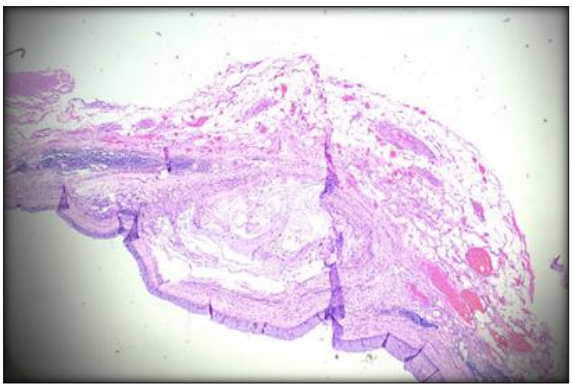

Figure 8: Microscopic view of Mucocele of Appendix

Post-operative recovery was uneventful. The patient was managed with IV fluids, antibiotic, analgesic, PPI and other supportive treatment. He was then discharged in stable hemodynamic condition with advice to report back immediately if configured with fever or severe pain. He was also advised to avoid heavy lifting till further advise.

CASE-6

A 75-year-old female was referred to the Surgical Outpatient Department with complaints of persistent abdominal pain, bloating, and altered bowel habits for the past few months. Physical examination revealed tenderness in the right lower quadrant with no palpable masses. Laboratory investigations, including complete blood count and biochemical parameters, were within normal limits.

Ultrasonography (USG) of the abdomen showed a complex cystic mass in the right iliac fossa, suggestive of an adnexal or appendiceal origin. Computed tomography (CT) scan confirmed the presence of a large cystic mass arising from the appendix, measuring 10×8cm, with enhancing mural nodules and adjacent fat stranding, indicative of a mucinous cystadenocarcinoma.

The patient underwent exploratory laparotomy, during which a large cystic mass (figure-9) arising from the appendix was identified. The mass was adherent to the surrounding tissues but appeared resectable. A right hemicolectomy with en bloc resection of the appendix and regional lymphadenectomy was performed.

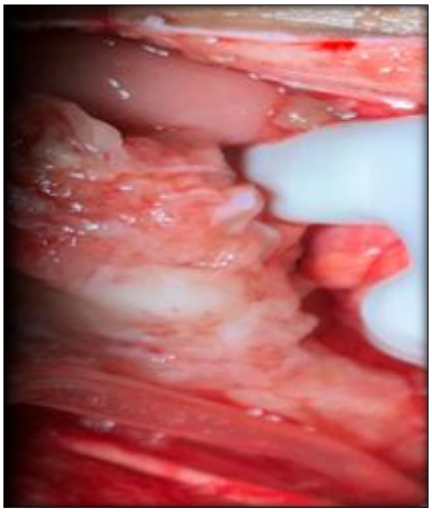

Figure 9: Intraoperative image of appendiceal mucocele.

Histopathological examination of the resected specimen revealed a mucinous cystadenocarcinoma of the appendix with invasion of the appendiceal wall and adjacent mesoappendix. The tumor exhibited areas of high-grade dysplasia and mucin production. Regional lymph nodes showed evidence of metastasis. Immunohistochemical staining was positive for cytokeratin and negative for CDX2, consistent with appendiceal origin.

The patient underwent adjuvant chemotherapy with a regimen of intravenous fluorouracil and oxaliplatin. Regular follow-up visits were scheduled to monitor for recurrence and assess the patient's overall condition. At the last follow-up appointment six months post-surgery, the patient was doing well with no evidence of recurrence on imaging studies.

CASE-7

A 69-year-old male presented to the Surgery Department with complaints of right lower quadrant abdominal pain, fever and localized tenderness since 48 hours. He also complained of increased frequency of micturition since last 6 months. On physical examination presence of tachycardia, tachypnea and tenderness in the right iliac fossa was noted. Laboratory investigations showed leukocytosis with a left shift.

Ultrasonography (USG) of the abdomen revealed a complex cystic mass in the right lower quadrant with surrounding inflammatory changes, suggestive of an appendicular abscess along with enlarged prostate with small amount of residual urine. Computed tomography (CT) scan confirmed the presence of a cystic mass arising from the appendix, measuring 8x3x2.5cm, with adjacent fat stranding and inflammatory changes, consistent with an appendicular abscess.

The patient was initially managed conservatively with intravenous antibiotics and bowel rest. However, due to persistent symptoms and concern for underlying pathology, surgical exploration was deemed necessary. During exploratory laparotomy, a distended appendix with a cystic mass at its base was identified. The appendix was adherent to the surrounding tissues but appeared intact without signs of perforation. A decision was made to perform an appendicectomy.

Histopathological examination (Figure-10) of the resected appendix confirmed the diagnosis of a simple appendiceal mucocele. The appendix exhibited significant dilatation with mucinous material filling its lumen, characteristic of a mucocele. There were no signs of acute inflammation or perforation.

Following appendicectomy, the patient's postoperative recovery was uneventful. He was discharged home with a course of oral antibiotics and scheduled for regular follow-up appointments. At the last follow-up visit three months post-surgery, the patient was asymptomatic with no evidence of recurrence on imaging studies.

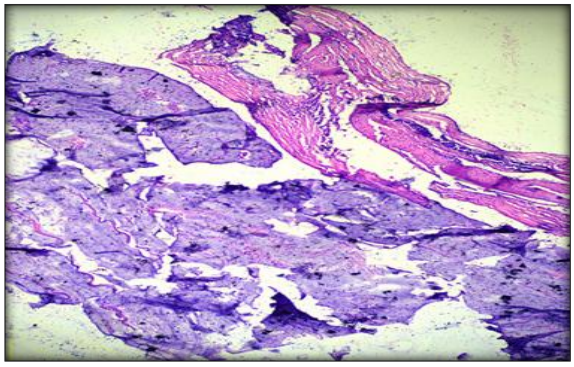

Figure 10: Significant dilatation of appendix with mucinous material filling its lumen.

CASE-8

A 74-year-old male presented to the Surgery Outpatient Department (OPD) with complaints of pain in the right lower quadrant. The patient reported experiencing intermittent pain for the past few weeks, which gradually worsened over time. There were no associated symptoms such as fever, vomiting, or changes in bowel habits.

Ultrasound (USG) and magnetic resonance imaging (MRI) were performed, revealing the presence of an appendiceal pathology along with bilateral inguinal hernias. The imaging studies indicated a dilated appendix measuring 11.5x3.5x2 cm. Considering the findings of the imaging studies and the patient's clinical presentation, a decision was made to proceed with laparoscopic stapler appendicectomy. Laparoscopic stapler appendicectomy was chosen as the preferred surgical intervention due to its minimally invasive nature and favourable outcomes. The surgery was performed successfully, addressing both the appendiceal mucocele and the bilateral inguinal hernias.

Following the surgery, the excised specimen (Figure-11), comprising a dilated appendix measuring 11.5x3.5x2 cm, was sent for histopathological examination to determine the underlying pathology.

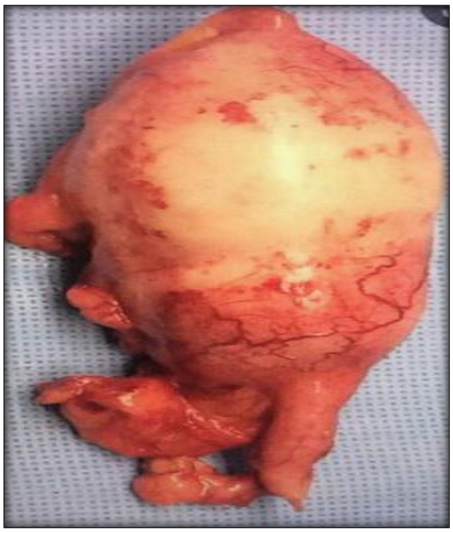

Figure 11: Dilated appendiceal mucocele measuring 11.5 x 3.5 x 2 cm

The histopathological examination revealed a low-grade appendiceal mucinous neoplasm, confirming the presence of a tumour within the dilated appendix. The dimensions of the appendix were consistent with the imaging findings, further validating the diagnosis.

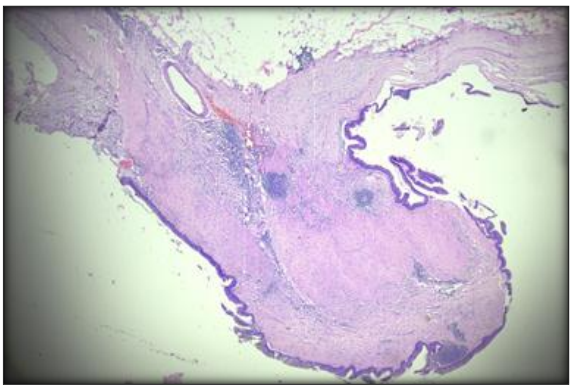

Figure 12: Low-grade appendiceal mucinous neoplasm

Patient's postoperative recovery was uneventful and was discharged home on oral antibiotics and scheduled for regular follow-up visits. The patient is being followed up since two years and has no new complaints.

DISCUSSION

Appendiceal mucoceles represent a rare subset of appendiceal pathology. In this case series, we presented a collection of 8 rare cases encountered in our clinical practice of 35 years, highlighting the diverse clinical presentations, diagnostic challenges, and management strategies associated with appendiceal mucoceles.

All patients in our case series were above the age of 50 years and the number of males and females were comparable. Singh MP (2020),[2] and Gao J et al (2022),[3] stated that appendiceal mucocele is commonly observed in individuals over the age of fifty with a slight predominance in females.

The clinical presentation of appendiceal mucoceles can vary widely, ranging from asymptomatic incidental findings to right lower quadrant pain, abdominal discomfort, nausea, vomiting altered bowel habits and weight loss, as discussed by Singh MP (2020),[2] Vyas J et al (2021),[4] Abiyere OH et al (2021),[5] BB SK & Jasuja P (2019),[6] and Demetrashvili Z et al (2012).[7] Similar findings were reported in our case series. The nonspecific nature of these symptoms often leads to delays in diagnosis and poses challenges in distinguishing appendiceal mucoceles from other intra-abdominal pathologies, such as acute appendicitis, ovarian masses, or appendicular abscesses.

The signs of appendiceal mucocele can mimic those of acute appendicitis or a tubo-ovarian mass in females upon examination as per Singh MP (2020).[2] Similarly in a retrospective study by Gao J et al (2022),[3] which included 3,071 patients with appendicitis, it was observed that out of these, 9 were diagnosed as appendiceal mucocele. Among these majority exhibited signs such as muscle guarding and rebounding pain. Louis TH et al (2014),[8] reported a similar case with maximum tenderness at Mc Burney's point on palpation. Similar observations were made in our study.

In the present case series ultrasonography (USG) and computed tomography (CT) were the investigations used for initial evaluation of the patients and further confirmation of the diagnosis was done by histopathology. Singh MP (2020),[2] discussed that preoperative diagnosis of appendiceal mucocele is challenging, even with the use of abdominal sonography or computed tomography. Histopathological examination of the dissected specimen is necessary for a definitive diagnosis. On the contrary, Vyas J et al (2021),[4] discussed that Pre-operative diagnosis is crucial for selecting the appropriate surgical procedure to prevent intra-operative complications, particularly peritoneal dissemination. Sonographic examination is often the first-line diagnostic modality that can potentially differentiate between benign and malignant mucoceles. An appendicular diameter of 15 mm or more has been established as a threshold for diagnosing mucocele, with a sensitivity of 83% and a specificity of 92%. Computed tomography (CT) scan plays a vital role in confirming the diagnosis and assessing the extent of the disease. BB SK & Jasuja P (2019),[6] similarly stated that CT is considered the most accurate diagnostic method for appendiceal mucocele. It can reveal specific signs of mucocele with high accuracy, including a dilated appendix lumen greater than 1.3 cm, cystic dilatation of the appendix, and wall calcification.

The management of appendiceal mucoceles depends on various factors. Based on the observation of our cases, appendicectomy remains the mainstay of treatment for appendiceal mucoceles, with the extent of surgical resection depending on the size, location and presence of associated complications. While benign mucoceles may require simple appendicectomy, malignant mucoceles may necessitate more extensive surgical resection and adjuvant therapies, such as chemotherapy or radiation therapy. Singh MP (2020),[2] and BB SK & Jasuja P (2019),[6] similarly opined that the mainstay of treatment is meticulous surgical resection, which aims to avoid spillage of the contents.

Histopathological examination of the resected appendix is essential for confirming the diagnosis of appendiceal mucoceles and determining the underlying pathology. This was also reported by Singh MP (2020),[2] Vyas J et al (2021),[4] Abiyere OH et al (2021),[5] BB SK & Jasuja P (2019), [6] Demetrashvili Z et al (2012),[7] Louis TH et al (2014),[8] and Gao J et al (2022).[3]

The histological subtypes of appendiceal mucoceles include hyperplastic, mucinous cystadenoma, mucinous cystadenocarcinoma, and retention cysts as per Abiyere OH et al (2021).[5] In the present case

series all subtypes have been reported except mucinous cystadenoma.

Regular follow-up and surveillance are essential for monitoring for recurrence and assessing treatment response in patients with appendiceal mucoceles. Long-term outcomes depend on factors such as the histological subtype, stage of disease, and adequacy of surgical resection.

CONCLUSION

Appendiceal mucoceles are rare entities that pose diagnostic and therapeutic challenges. By sharing our experiences and observations from this series of cases, we hope to contribute to the existing body of knowledge regarding appendiceal mucoceles and provide valuable insights for clinicians involved in the care of patients with this rare but clinically significant condition. Further research and collaborative efforts are needed to better understand the pathogenesis, natural history, and optimal management of this rare but clinically significant condition.

Informed Consent: Written informed consent was obtained from patients who participated in this case series.

Conflict of Interest: No conflict of interest was declared by the authors.

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REFERENCES

1. Santos SF, Horta M, Rosa F, Rito M, Cunha TM. Mucocele of the appendix: what to expect. Radiologia Brasileira. 2022 May 2; 55:193-8.
2. 2.Singh MP. A general overview of mucocele of appendix. Journal of Family Medicine and Primary Care. 2020 Dec 31;9(12):5867-71.
3. 3.Gao J, Chu Z, Chen J, Zheng Y, Huang X, Hu K. Retrospective analysis of 9 cases of appendiceal mucocele in 3,071 cases of appendicitis. Biomedical Reports. 2022 Jul 1;17(1):1-5.
4. 4.Vyas J, Badgurjar M, Saxena P, Parihar S, Thakor P. Prudent planning in management of mucocele of appendix. International Journal of Surgery Case Reports. 2021 Apr 1; 81:105766.
5. 5.Abiyere OH, Adewara O, Akute OO, Babatunde O. Mucocele of the appendix: case report & review of literature. Int J Surg Res Pract. 2021; 8:125.
6. 6.BB SK, Jasuja P. Appendiceal mucocele—a rare case report. International Journal of Surgery Case Reports. 2019 Jan 1; 58:21-5.
7. 7.Demetrashvili Z, Chkhaidze M, Khutsishvili K, Topchishvili G, Javakhishvili T, Pipia I, Qerqadze V. Mucocele of the appendix: case report and review of literature. International surgery. 2012 Oct 1;97(3):266-9.
8. 8.Louis TH, Felter DF. Mucocele of the appendix. InBaylor University Medical Center Proceedings 2014 Jan 1 (Vol. 27, No. 1, pp. 33-34). Taylor & Francis.