



Chronic Right Flank Pain due to an Underlying Mucocele of the Appendix: A Case Report

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Abstract

The Mucocele of the appendix is a very rare clinical entity with nonspecific clinical manifestations. Right lower abdominal pain and swelling are the most common clinical manifestations. Our patient presented with pain in her right lower abdomen. Imaging studies revealed an underlying mucocele of the appendix with a wide base for which we performed laparotomy with appendectomy and limited right hemicolectomy. The patient had a very good outcome after surgery and recovered well with no signs of malignant disease. Early surgical resection with delicate intraoperative handling of the tumor is recommended for all appendiceal mucoceles to keep the disease benign and free of complications.

Introduction

The Mucocele of the appendix is a morphological description of an appendix that has a distended lumen filled with mucus. It is a very rare clinical entity and can be the result of retention, epithelial proliferation, inflammation, or obstruction [1]. The clinical presentation of appendiceal mucocele is nonspecific and is often an incidental finding during surgery for acute appendicitis [2]. Hereby, we present the case of a lady who presented with chronic right flank pain caused by an underlying mucocele of the appendix.

Case Presentation

Our patient is a 49-year-old woman who came to the clinic with a five-year history of insidious-onset, dull pain in her right lower abdomen. The pain was on and off, gradually progressing in severity with each recurrence, and has been continuous for the last 10 days. She also gives a history of increased frequency of micturition and has to wake up multiple times at night to pass urine. She does not give any history of nausea, vomiting, difficulty in passing stools, blood-mixed stool, loss of appetite, night sweats, fever, burning micturition, vaginal discharge, or bleeding. She denied any significant weight changes. On abdominal examination, there were no significant findings present; the abdomen was non-tender and non-distended; no abnormal mass was palpable over the abdomen.

On ultrasound of the abdomen, a cystic lesion was seen in the right inguinal fossa measuring 8.6 cm × 5.8 cm × 3.6 cm without any septation, calcification, or solid components. Contrast-enhanced CT scan of the abdomen (Figure 1) revealed a well-defined non-enhancing cystic tubular lesion measuring 8.5 cm × 4.8 cm × 4.1 cm (12 HU) with a homogeneously enhancing wall arising from the ileocecal junction region in the right iliac fossa.

A diagnosis of mucocele of the appendix was made at this point. Due to the wide and distended appendix and a significantly increased risk of rupture, it was not possible to perform a simple appendicectomy. So, an exploratory laparotomy with the possibility of limited right hemicolectomy was planned.

Intraoperatively, a cystic mass arising from the appendix was noted. It was free from the terminal ileum but attached to the caecum. There was no evidence of ascites or gross peritoneal metastases and mesenteric lymph nodes could not be palpated. The mucocele (Figure 2) was grossly enlarged, extending to the base of the appendix. Our assessment concluded that any attempt to tie the base could result in the rupture of the tumor and, therefore, the spread of the neoplasia to the peritoneal cavity. So, we performed an appendectomy with limited right hemicolectomy. The specimen containing the appendix with the caecum and terminal ileum was sent for histopathological examination.

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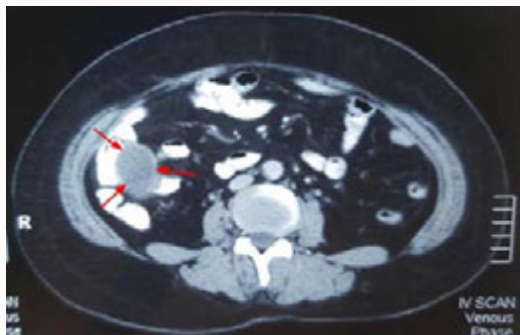


Figure 1: Axial view of the contrast-enhanced CT scan of the abdomen showing a well-defined non-enhancing cystic mass (red arrows) with a uniformly enhancing wall in the right iliac fossa.



Figure 2: Appendiceal mucocele extracted along with terminal ileum and caecum.

Histopathological examination revealed that the neoplasm was involving the appendix diffusely, the wall was thinned out and the dilated appendiceal lumen was filled with mucin. Signet cells could be identified in the mucosal lining. A histopathological diagnosis of Low-grade Appendiceal Mucinous Neoplasm (LAMN) with mucocele was made with pathological staging (pTNM) of pTis(LAMN)N0.

The patient was discharged after an uneventful postoperative period and has been regularly following up with us in the clinic. She has had a very good recovery and is satisfied with the care she received. She and her family believe that they should have come for medical help the first time she developed abdominal pain.

Discussion

Appendiceal mucocele is an uncommon disorder with an estimated incidence of 0.2% to 0.3% of all appendectomies performed and 8% to 10% of all appendiceal tumors [3]. They can be divided into four pathological types: Simple retention cyst, mucocele with mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma [4].

The clinical manifestations are atypical and nonspecific. Right lower abdominal pain or a right lower abdominal mass is the most common manifestation, and up to half of the cases are asymptomatic and are found accidentally during physical examination or other

operations [4].

Although the definitive diagnosis can be made only postoperatively by histopathological examination, the preoperative diagnosis of appendiceal mucocele is essential for the selection of an adequate surgical method. CT is the initial imaging modality for the evaluation of the right lower quadrant or diffuse abdominal pain, which may lead to the detection of appendiceal tumors. On CT scan, a typical appendiceal mucocele will manifest as a homogenous hypoattenuating material that has Hounsfield values similar to water filling the lumen of the appendix. The presence of curvilinear calcifications on the wall of the appendix is highly suggestive of a mucocele [5].

Ultrasound can be very useful in demonstrating secondary signs of malignancy such as appendiceal mucocele, regional adenopathy, or ascites and allows differentiation of appendiceal from ovarian masses, as well as appendicitis from a mucocele of the appendix [6].

Early surgical resection is recommended for all appendiceal mucoceles to exclude mucinous neoplasms and to prevent spontaneous rupture in the future [7]. The goal of the surgery should be removing the tumor without peritoneal dissemination of the neoplasm with minimal intraoperative and postoperative complications, with low to no chance of needing repeat surgery. Intact mucocele is considered a benign disease and it does not pose any threat to the patient.

The selection of surgical methods is essential. Careful intraoperative handling is paramount for extracting the intact appendix and preventing iatrogenic rupture. An open approach is preferred to laparoscopic surgery to ensure proper handling and avoid tumor rupture [2].

Dhage-Ivatury et al. suggested an algorithm for the management of the mucocele of the appendix. Patients may need anywhere from appendectomy to the right colectomy, including cytoreductive surgery, heated intraoperative intraperitoneal chemotherapy, and early postoperative intraperitoneal chemotherapy [8]. We performed an appendectomy with limited right hemicolectomy. The prognosis depends upon histology and the presence and extent of peritoneal spread and invasion which determine the recurrence. After appendectomy, 5-years survival rate for simple appendiceal mucocele is 91% to 100% [9]. This case emphasizes the importance of suspecting a mucocele of the appendix as a cause of right lower abdominal pain and highlights that proper preoperative diagnosis and surgical planning followed by early surgical management can help contain the disease in a benign state.

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