Case Report

Appendiceal mucocele: benign and malignant

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Summary

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Appendicular mucocele is a rare lesion. It is a descriptive term denoting an obstructive dilatation of the appendicular lumen by mucinous secretions. Mucinous cystadenoma and cystadenocarcinoma account for 60 - 70% of all mucoceles. Less common causes are retention cyst, mucosal hyperplasia, carcinoid, appendicolith, endometriosis, adhesions and volvulus. The clinical presentation is usually non-specific with 50% of cases being an incidental finding at surgery.

We present two rare cases of mucinous cystadenoma and cystadenocarcinoma respectively, the 1st case, a female patient was presented by right lower quadrant pain and right iliac fossa tenderness, the diagnosis was made by computed tomography (CT) scanning, she underwent right hemicolectomy and histopathology result was mucinous cystadenoma. The diagnosis of the 2nd case, a male patient was incidental finding in CT scanning, physical examination revealed firm, non tender mass at suprapubic and right iliac fossa regions, he underwent right hemicolectomy with en-block resection of the sigmoid colon and histopathology result was mucinous adenocarcinoma.

Benign and malignant appendiceal mucocele may present as an incidental surgical or radiological finding. Right hemicolectomy for cases in which the pathological diagnosis has not been established are recommend.

Introduction

Appendiceal mucocele is a rare, but well-recognized entity that can mimic several common clinical syndromes or present as an incidental surgical or radiological finding. It has 0.2% to 0.4% prevalence among appendectomies (1,2). The term mucocele is widely used in diagnosing both benign and malignant lesions, but specific criteria are being proposed for definitive diagnosis and surgical management of appendiceal mucocele (3). While some neoplasms with malignant potential may be treated definitively by resection, other seemingly benign lesions must be treated conservatively due to complications that ensue from...
peritoneal and cecal inoculation and the possibility of progression to malignancy\cite{4,5}. Mucocele can result from mucosal hyperplasia, mucinous cystadenoma, mucinous cystadenocarcinoma or simple mucocele due to obstruction of the appendix base by faecolith. Signs and symptoms occur in fewer than 50% of cases and are generally associated with malignancy\cite{3}. These include pain in the right lower abdominal quadrant, an abdominal mass, weight loss, nausea, vomiting, change in bowel habits, anemia, and hematochezia. Depending on the location of the appendix, other signs may be observed, such as hematuria. The most dreaded complication of benign or malignant mucocele is pseudomyxoma peritonei, which is difficult to treat surgically or medically. It has an uncertain prognosis, with a 5-year survival rate between 53\% and 75\%\cite{5,6}.

More than half of appendiceal mucoceles are mucinous cystadenomas, most of which can be treated by appendectomy alone, with careful exploratory laparotomy for mucinous peritoneal adhesions typical of pseudomyxoma\cite{7}. Wide resection of the appendix, however, is currently the standard for conservative surgical management of unspecified appendiceal mucocele.

**Reports of Cases**

**Case 1**

A sixty-year-old woman presented with a constant, dull right lower quadrant pain of 8 days duration, her pain did not radiate and was not affected by any activity. She reported no changes in her appetite or bowel habits and any hematochezia or melena. The patient had diabetes mellitus for last 2 years controlled by oral hypoglycemic agents, but she had no history of surgery. On physical examination, she was not febrile and hemodynamically stable. The abdominal examination was normal except for tenderness and guarding over the right iliac fossa. Laboratory analysis was unremarkable. Computed tomography scanning revealed a 13.4 x 6.1 x 5.6cm cystic lesion with thin enhancing wall and internal densities seen in the right iliac fossa consistent with mucocele of the appendix. The CT scan also showed, portal vein thrombosis with varicose veins at porta hepatitis (Fig 1).

**Fig 1: Axial CT section through ascending colon. A large tumor is demonstrated (arrow).**

The patient underwent laparotomy and right hemocelectomy. Large appendicular mass adherent to overlying small bowels was noted. Exploration of the peritoneum did not show evidence of malignancy. Pathological examination of the surgical specimen revealed a cecum and ascending colon 22cm long with 14cm of terminal ileum. The mucosal surface was grossly normal. The appendix was markedly dilated and measured 17 x 7 x 5cm and cut surface showed lumen distended by thick gelatinous mucus (Fig 2).

**Fig 2: Shows mucinous contents upon opening of cyst**
The tumor was diagnosed on histological section as mucinous cystadenoma. The patient postoperative course was uneventful, and she was discharged home on postoperative day 7 on warfarin for life as the cause of portal vein thrombosis was discovered to be due to protein C deficiency.

Case 2
A 58-year-old man was on regular follow-up for renal stone, computed tomography for kidney, ureter and bladder. CT KUB scanning accidentally found a soft tissue mass involving the right iliac fossa region indenting the medial border of the cecum. He gave history of fresh blood in the stool for 4 days and mild weight loss. The patient had a history of left varicocelectomy and extracorporeal shock wave lithotripsy (ESWL) for right renal stone. On physical examination, he was afebrile and hemodynamically stable. The abdominal examination revealed left inguinal crease incision, divarication of the recti muscle and 14 x 10cm lobulated, firm, non tender mass, at the suprapubic and right iliac fossa region. Laboratory analysis was unremarkable. Computed tomography scanning revealed a 10 x 12cm cystic mass lesion in the right iliac fossa adherent to bowel loops and specks of calcification within noted consistent with mucocele of the appendix or gastro intestinal tumor e.g. GIST (Fig 3).

Pathological examination of the surgical specimen revealed cecum and ascending colon measures 35cm long. The cecum shows mucinous mass measuring 14cm in maximum diameter stuck to and penetrating sigmoid. The appendix was not identified grossly. The tumor was diagnosed on histological section as mucinous adenocarcinoma infiltrating the wall and extending into the serosa. The seven lymph nodes were examined and the surgical cut edges were free of neoplasm. The features were those of mucinous adenocarcinoma of the appendix. The patient was discharged home on post operative day 7. He was readmitted after 10 days with sepsis. Laparotomy and drainage of intraperitoneal collections were performed. The patient was discharged home in good condition on warfarin because he developed DVT, and is currently undergoing chemotherapy.
Case Report
Appendiceal mucocele Abubaker AM Elhaj

Discussion
Some consider the term mucocele to encompass a large group of conditions involving the appendix, pancreas, or ovaries with diverse morphological features and pathogenesis. These conditions share the common feature of obstructive process or hyperplasia of mucinous epithelium, or both leading to gross mucinous accumulation. Others consider mucocele to be a strictly neoplastic process that can spread to lymph nodes, extend into surrounding tissue, or seed the peritoneum(7). The latter description encompasses most mucocele-related diagnoses, and thus, right hemicolecetomy is an appropriate first step in managing suspicious mucinous collections of the appendix. Histopathological findings can confirm whether further tests are needed for workup of malignancy. If a patient has a mucinous cystadenoma of the appendix, as was the case with our first patient, then right hemicolecetomy is curative (except in cases that are complicated by pseudomyxoma peritonei).

In a retrospective study of 135 patients with appendiceal mucocele, 55% were women; other reports have shown a distinct male predominance ratio of 4:1(3,8). Indications for removal of appendiceal mucocele are evolving as diagnostic procedures that lead to surgery for a wide variety of concomitant conditions improve. Forty percent of patients in the 135-patient study went into the operating room specifically for treatment of symptoms or to confirm a diagnosis, while the remaining 60% had their appendiceal mucocele removed on incidental finding. Although CT scanning is usually accurate in imaging a fluid-filled appendix, the appendix was often missed on CT scans for workup of coexisting conditions. This may explain the high incidence of surgical diagnosis for other conditions. Still, in the workup for a patient's symptoms of right lower quadrant abdominal pain or a palpable abdominal mass, CT scanning has high sensitivity and specificity for detecting an abnormal appendix. Also CT scanning has the advantage of allowing precise observation of the relationship between the lesion and adjacent organs and any other abnormalities associated with the mucocele.

Ultrasonography and endoscopy are becoming the standards for confirming CT findings before taking the patient to the operating room. Endoscopic biopsy and pathological determination can further guide the operative procedure if hemicolecetomy is wanted to be avoided. In suspected cases of appendiceal mucocele, fine needle aspiration should be avoided to preserve integrity of the appendix and prevent tumor inoculation(1,6-11).

Although complications from appendiceal mucocele are minimal, there is evidence that complications are associated with concomitant neoplasms, these occurred in about one-third of patients in the retrospective study and undoubtedly contributed to the high number of incidental findings of mucocele. While the incidence of ovarian and uterine neoplasms might be explained in part by the high number of gynaecologic procedures reported, the association between appendiceal mucocele and colonic neoplasms is well-established(3,12,13). This additional evidence makes a strong case for the use of surveillance colonoscopy and removal of polyps in any patient with an appendiceal mucocele. Most investigators agree that the adenoma-adenocarcinoma sequence is similar to the colonic polyp-adenocarcinoma sequence(14).

No cystadenomas smaller than 2cm have been reported, which suggests that all mucoceles larger than 2cm should be removed to eliminate the chance of progression to malignancy. Furthermore, all patients, whether they have benign or malignant appendiceal mucocele, should be evaluated for pseudomyxoma peritonei. Although the condition is more common in malignant appendiceal mucocele (95% occurrence rate as opposed to 13% in patients with
nonmalignant appendiceal mucocele), the grave consequences of pseudomyxoma peritonei and the somewhat better prognosis for patients whose condition is diagnosed and treated early should challenge the view that simple surgical resection and careful exploration for every diagnosed appendiceal mucocele is sufficient (5).

Appendiceal mucocele presents a challenge to the surgeon who does not appreciate the effect of pathological diagnosis on the operative procedure. Appendectomy alone should be performed only on mucinous lesions that are determined to be non-neoplastic after biopsy. If appendectomy is performed, precautions should be taken to minimize the risk of seeding the peritoneal cavity with mucinous tumour during manipulation.

Some suggested that if distention of the appendiceal lumen is observed laparoscopically, suggesting a mucocele as a possible diagnosis, conversion to open appendectomy may be the most prudent surgical judgment because of the increased risk of rupture and subsequent pseudomyxoma peritonei (15). However, appendectomy alone? Whether open or laparoscopic? It does not guarantee the removal of all neoplastic tissue, including extensions into surrounding tissues and lymph nodes (2).

Mucocele of the appendix is uncommon and the greatest percentage is of the benign variety. Its incidence is estimated at 0.224% (16). The incidence of malignant mucocele is one out of 4,300 surgically removed appendices (17). The association of malignant mucocele with diffuse thromboembolic disease is rather interesting in the experience of the authors, a hitherto unreported condition. It is well recognized that a large percentage of cases of carcinoma of the pancreas is associated with thromboembolic disease. It is estimated that approximately 50% of such cases have an associated thromboembolic condition. They may occur anywhere in the body and characteristically wax and wane, giving rise to a migrating type of phlebitis, (Trousseau sign). Thromboplastin-like substances are supposed to be released and are thought to play a causal role in the genesis of this phenomenon. A direct causal relationship between malignant mucocele of the appendix and thromboembolic disease has not been completely established; however, it is known that mucus producing carcinomas, in particular pancreatic carcinomas, are associated with an increase of the antithrombin titer (18-21). As in our 2nd case, who developed deep vein thrombosis of the both lower limbs despite of all prophylactic measures were done (he received clexane 40mg daily, thromboembolic detergent stockings and early mobilization, but he didn't checked for thrombogenic diseases).

In conclusion, preoperative diagnosis of appendicular mucocele is very important for the selection of an adequate surgical method to prevent peritoneal dissemination, to prevent intraoperative and postoperative complication, and repeated surgery. Ultrasonography, computed tomography (CT), colonoscopy and intra-operative frozen sections are used for diagnostics. We recommend right hemicolecotomy for cases in which the pathological diagnosis has not been established and for any cases suspicious for adenocarcinoma or complicated by concomitant adenocarcinomatous disease or pseudomyxoma peritonei.

**References**
