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A case of mucinous cystadenoma of appendix: Management and review

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ABSTRACT

Introduction: Though mucinous cystadenoma of the appendix represents the majority among all primary tumors, tumors of the appendix are still rare. They most commonly present as a mucocele of the appendix during abdominal imaging studies. Case Report: We report a case of a 52 year-old female who was found to have an enlarged and prolapsed base of the appendix into the cecum during a routine colonoscopic examination for colorectal cancer screening. A computed axial tomography of the abdomen showed a mucocele of the appendix. The patient underwent a laparoscopic hemicolectomy. Histopathological examination revealed a mucinous cystadenoma of the appendix. As these carry a high risk of presence of other coexisting colorectal and ovarian malignancies, a detailed intra-operative exploration of the gastrointestinal tract, ovaries and peritoneum was conducted. A periodic follow-up colonoscopic examination, and examination of the pelvic organs was followed as per the recommendations in such cases. Conclusion: Our case highlights the importance of diagnosing mucoceles of the appendix, which are rare and often pose a diagnostic challenge. Moreover, their association with coexisting malignancies further warrants the need for a thorough investigation and a broader approach towards the management.

Keywords: Appendiceal tumors, Appendix, Mucinous cystadenoma, Mucocele

How to cite this article

Mohan S, Shayaan M, Griech-McCleery CA, Roy S. A case of mucinous cystadenoma of appendix: Management and review. J Case Rep Images Med 2017;3:25–30.

Article ID: 100036Z09SM2017

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doi:10.5348/Z09-2017-36-CR-8

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Received: 07 June 2017
Accepted: 26 June 2017
Published: 11 July 2017

INTRODUCTION

Mucinous cystadenomas of the appendix are rare and are often discovered unexpectedly [1–3]. They mostly present in the form of a mucocele. Mucocoele of the appendix denotes an obstructive dilatation of the appendiceal lumen due to abnormal accumulation of mucus resulting in a cystic dilatation of the lumen, which may be related to a variety of pathological conditions [2, 3]. It is a daunting task to offer only a major surgical option to an asymptomatic patient with seemingly benign tumor of the appendix. However, the unpredictable

course of the mucocele; driven by the accumulating mucus, the pressure exerted with distortion of the surrounding tissue, and difficulty in obtaining an accurate histopathological diagnosis makes the diagnosis difficult [3–6]. The latter may allow a neoplastic epithelium of the mucocele to remain unrecognized unless a detailed histological examination of the appendix is undertaken. Additionally, there are reports of coexisting colorectal and ovarian malignancies in association with about a fifth of all mucoceles [4–8]. Hence, the management plan should not only address the removal of the mucocele, but it should also include a detailed examination of the gastrointestinal tract and utero-pelvic organs to look for other neoplastic lesions that may coexist [2–8].

CASE REPORT

A 52-year-old female underwent an annual well visit examination. Her long-term medical problem of gastro-esophageal reflux disorder was under good control with oral omeprazole 40 mg daily. She suffered from occasional episodes of abdominal bloating sensation that was associated with mild heartburn for more than five years. She had no abdominal pain, nausea, vomiting, fever, constipation, diarrhea, hematemesis, malena, or hematochezia. Additional medical history was significant for bipolar disorder that was controlled with quetiapine.

Physical examination of the patient was unremarkable. Her diagnostic and routine blood tests that included a complete blood count, comprehensive metabolic panel, and lipid profile, were all within normal range. Her family history was significant for colon polyps hence she was advised to undergo a colonoscopy for colon cancer screening. Her colonoscopic examination revealed an enlarged and prolapsed base of the appendix into the cecal lumen, and a few descending colonic diverticuli (Figure 1). She was referred to undergo a contrast-enhanced computed tomography (CT) scan of the abdomen and pelvis. It showed approximately 3.0x1.2 cm mass within the proximal aspect of the appendix, without evidence of surrounding inflammation. The mass had a uniform hypodense appearance, suggestive of a mucocele. Rest of the gastrointestinal tract, solid organs, and pelvic structures were normal (Figure 2).

The patient was referred to a colorectal surgeon who performed a diagnostic laparoscopy that showed a 3-cm smooth mass at the base of the appendix. Based upon the tumor size of greater than 2 cm and tumor protrusion into the cecum, a right ileocolic resection (hemicolectomy) and lymph node dissection was performed. Histopathological analysis of the mass showed abundant accumulation of mucinous material in the lumen, areas of mucus membrane showing columnar epithelium, and severely flattened epithelium that could be either secondary to exerted pressure of the mucinous material in the lumen, or due to metaplasia (Figure 3). The proximal, distal and radial margins of resection were negative for tumor.

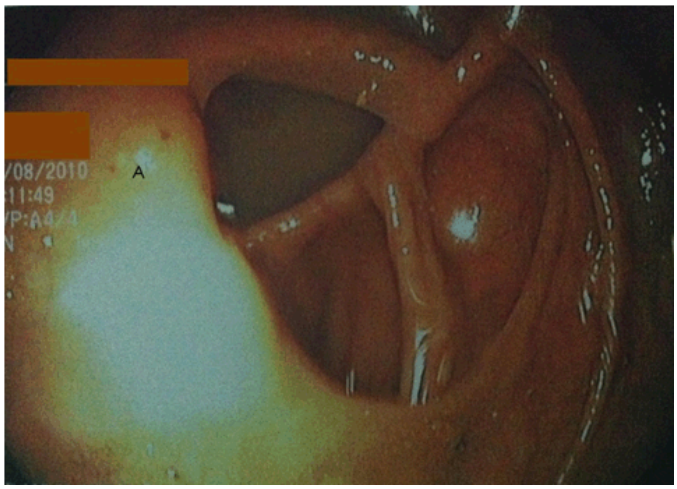

Figure 1: Colonoscopic view of cecum showing ileocecal valve.

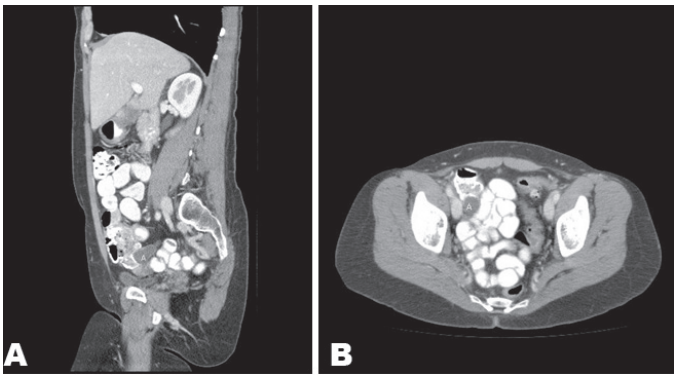

Figure 2: (A, B) Computed tomography scan of the abdomen and pelvis showing a mass within the proximal aspect of the appendix.

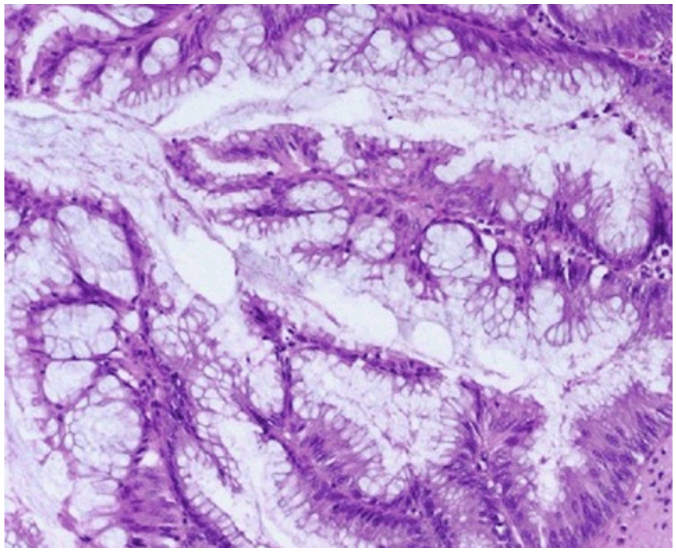

Figure 3: Histopathologic appearance of mucinous cystadenoma of the appendix showing epithelial lining that is made up of intestinal type epithelium mostly crowded columnar cells with basal, elongated, hyperchromatic nuclei, and large amount of apical mucin.

Twenty-two lymph nodes were all negative for metastatic tumor. The patient recovered uneventfully.

DISCUSSION

Mucoceles constitute about 0.2–0.3% of all appendectomies [1]. It can be caused by a variety of neoplastic and non-neoplastic pathological conditions of the appendix; such as mucinous cystadenoma (63%), mucosal hyperplasia (25%), mucinous cystadenocarcinoma (11%), and rest as retention cyst [2]. Based on retrospective case series, appendiceal mucoceles are second only to carcinoid tumors [3]. Mucocele of non-neoplastic origin is often caused by a fecalith obstructing the base of the appendix, resulting into accumulation of mucus and subsequent dilation[1, 4, 5].

Mucinous cystadenomas have high frequency in women compared to men with a ratio of 4:1, and it tends to affect patients over 50 years of age [6]. A rare variant of appendiceal mucocele that is known as myxoglobulosis, has also been reported. Their incidence varies from 0.35–8% of all mucoceles [7, 8]. The diagnostic hallmark variety is the collection of intra-luminal globules containing mucous, hence attributed as a caviar appendix. Most of the benign mucoceles, including mucinous adenomas, are asymptomatic. These are mostly detected incidentally during ultrasonography, computed tomography and other radiographic examinations of gastrointestinal tract, or during a laparotomy [5]. Symptomatic manifestation may result from inflammation, like acute appendicitis; or they can present with a palpable mass such as intussusception, gastrointestinal bleeding, ureteral obstruction or hematuria, and increasing abdominal girth due to the malignant tumor extending into the peritoneal cavity, or rupture of a malignant mucocele resulting in to pseudomyxoma peritonei [1, 9, 10].

Making a clinical diagnosis of appendiceal mucocele is difficult. Therefore, preoperative diagnosis is mostly relied on imaging studies [11]. Computed tomography scan of abdomen is an ideal test because it is more sensitive than ultrasonography for the detection of mural calcifications within the mucinous neoplasm [12]. The detection of mural curvilinear calcifications is highly suggestive of the diagnosis while contrast-enhancing nodules suggest cystadenocarcinoma [9]. Magnetic resonance imaging features of appendiceal cystadenoma recapitulate the CT findings of a cystic mass with low signal intensity on T1-weighted images and high signal intensity on T2-weighted images. Colonoscopy, on the other hand, shows extrinsic compression or mass protrusion of the appendiceal orifice as seen in this case. It has also been reported that several diseases like hydrosalpinx, ovarian cysts or renal cysts mimic appendiceal mucocele in their ultrasonography and CT scan, further enhancing their uncharacteristic clinical symptoms [1].

There are reports of coexistence of mucinous appendiceal tumors and colonic neoplasms. In one series, 21.4% patients with appendiceal mucocele had coexistent colon carcinomas [13]. Hence, a colonoscopy should be performed before surgical treatment as well as during follow-up. Although, an accurate histopathological interpretation of the appendiceal mucocele is an important determinant of the management, it is a challenging task with presence of tenacious mucous, distortion of surrounding normal tissue and subtle changes in columnar epithelium [14].

Apparently benign looking mucin-producing mucoceles may grow rapidly or create excessive mucin production. Abdominal organs may malfunction due to pressure exerted by a sudden and progressive accumulation of mucin in the abdominal cavity that may result into a fatal outcome. These cases warrant surgical intervention [14–17]. Post-surgical data shows that the survival outcomes from the benign, or low-grade mucin-producing tumors (like, mucinous cystadenoma) are much better than outcomes for high-grade mucin-producing tumors (like, mucinous cystadenocarcinoma. Our patient's mucinous cystadenoma was located near the base of the appendix, protruding into the cecum, and it was more than 2 cm in dimension; hence a right hemicolectomy (ileocolic resection) was performed. The abdominal cavity was explored and 22 lymph nodes were resected. None of the lymph nodes showed malignant transformation.

The choice of surgical approach has evolved from laparotomy to a laparoscopic approach. Some authors were cautious about laparoscopic removal of appendiceal mucocoeles, due to potential risk of peritoneal seeding and subsequent pseudomyxoma peritonei. A case report from González Moreno et al. [15] showed diffuse peritoneal carcinomatosis detected in a patient after nine months of laparoscopic resection of an appendiceal cystadenocarcinoma. However, a case series study by Rangarajan et al. [16] showed that eight patients who underwent laparoscopic removal of appendiceal mucocoeles remained asymptomatic during a two-year follow-up. More favorable long-term outcomes of laparoscopic approach have made it a procedure of choice for experienced colorectal surgeons. It is minimally invasive as demonstrated by minimal postoperative pain and quick recovery [17, 18]. Simple appendectomy is indicated for the appendiceal mucocele, retention cyst, benign tumor of appendix, tumor of 1–2 cm size, and localized pseudomyxoma peritonei [3, 18]. A right hemicolectomy is indicated when the mucocele involves the cecum or any other adjacent organ(s), or if its size exceeds 2 cm, or if there is evidence of lymphatic involvement, or if it is a proven malignancy [3, 18]. A detailed intra-operative exploration of the gastrointestinal tract, ovaries and peritoneum, and a follow-up colonoscopy and pelvic examination are required due to its high association with other colon and ovarian malignancies, as discussed earlier. Patients with a simple or benign neoplastic mucocele have an excellent postoperative prognosis, with five-year survival rates of 91–100%, even in cases

of extension of mucus into the extra-appendiceal spaces. However, the clinical course of diffuse pseudomyxoma peritonei is insidious and unrelenting. Treatment consists of aggressive surgical debulking of all apparent mucinous tissue and, in women, bilateral oophorectomy, or total abdominal hysterectomy. This approach, compared with appendectomy alone, leads to a significant improvement in the survival rate and a decrease in recurrence rate [12]. Due to a significantly high association of coexistent colorectal and pelvic malignancies our patient was enrolled into higher-risk colorectal cancer screening, and high-risk gynecological malignancy screening programs. At 6th month and at 12th month follow-up, our patient remained symptom free.

CONCLUSION

To summarize, mucinous cystadenomas are a rare subtype of appendiceal mucoceles, characterized by distended, mucous filled appendix. They may be found incidentally on abdominal computed tomography scan or colonoscopy, and pathological examination of the excised appendix remains the mainstay of diagnosis. Several studies in the past have reported an association between appendiceal mucoceles and tumors of ovary, endometrium, breast, gastrointestinal tract and kidney, warranting closer surveillance and monitoring for this patient population.

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Author Contributions

Sachin Mohan – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Mehwish Shayaan – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Satyajeet Roy – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Cynthia A. Griech-McCleery – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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Article citation: Mohan S, Shayaan M, Griech-McCleery CA, Roy S. A case of mucinous cystadenoma of appendix: Management and review. J Case Rep Images Med 2017;3:25–30.

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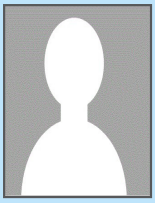

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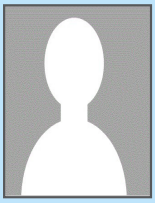

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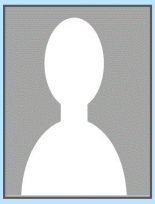

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