Early View Article: Online published version of an accepted article before publication in the final form.

Journal Name: International Journal of Case Reports and Images (IJCRI)

Type of Article: Case Report

Title: Mucinous cystadenoma presenting as appendiceal mucocele

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doi: To be assigned

Early view version published: December 23, 2015

How to cite the article: Desai N, Rubinshteyn V, Ayoub M, Gave A. Mucinous cystadenoma presenting as appendiceal mucocele. International Journal of Case Reports and Images (IJCRI). Forthcoming 2015.

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Short Running Title: Appendiceal Mucocele

Guarantor of Submission: The corresponding author is the guarantor of submission.
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ABSTRACT
Appendiceal mucocele is a complex entity consisting of several different pathologies with a spectrum of clinical presentation. We present a case of a 52-year-old male that presented with a 2-day history of abdominal pain. He was taken to the operating room for an open appendectomy and found to have a large mucocele of the appendix. We review his pathology as well as the different pathological conditions that may lead to an appendiceal mucocele.

Keywords: Appendix, appendiceal mucocele, intussusception, mucoid, Ki-67, p53, cystadenoma, cystadenocarcinoma, Mcburney’s,
INTRODUCTION
An appendiceal mucocele is a well-described entity that may manifest with a variety of clinical symptoms which range from abdominal pain to intussusception. [1-3] This abnormality is also often found incidentally on imaging or in the operating room for different indications. The majority of literature describing this entity comes from isolated case reports on its presentation and treatment. The largest study in the literature identifies 135 patients with this histological diagnosis with the majority having been found at time of surgery for a synchronous lesion. We describe a patient that presented to our institution with abdominal pain with an appendiceal mucocele found on computed tomography(CT)imaging. We report our technique used for performing a successful open procedure along with the pathological results.

CASE REPORT
Our patient is a 52-year-old male, who presented with 2 days of right-sided abdominal pain associated with nausea. His past medical history included hypertension and hyperlipidemia, and surgical history was non-contributory. On physical examination, he was hemodynamically stable and his abdomen was soft with tenderness in the right lower quadrant associated with voluntary guarding. His white blood cell count was 7.7 with 85% neutrophils seen on differential. His other laboratory values were unremarkable. At our institution, workup in the emergency department included a CT scan of his abdomen and pelvis with oral and intravenous contrast. The appendix was noted to be dilated with fluid measuring 2.3cm at the base and 1.5cm towards the tip. No peri-appendiceal fat stranding was seen. Representative cuts may be seen of the base, body and distal tip in Figures 1-3, respectively. Incidentally, a pancreas divisum was discovered; otherwise the scan was unremarkable. Based on the patient’s clinical presentation and imaging, he was scheduled for an open appendectomy.

Surgical Technique
The patient was taken to the operating room, where he was first prepped and draped. Intravenous cefoxitin was delivered within one hour prior to skin incision. A
horizontal incision was made measuring 4-5cm over Mcburney’s point. The subcutaneous tissues and anterior sheath were divided with an electrocautery and the abdominal muscles were separated bluntly so that the posterior sheath could be opened using Metzenbaum scissors. The appendix was grasped and delivered out of the abdominal cavity and was found to be grossly dilated. Care was taken not to injure the appendix or cause rupture. An Endo GIA stapler was used to divide the appendix at the base, and the mesentery was divided and ligated using a silk suture ligature. We closed the posterior and anterior sheath separately with a running #1 Vicryl. The wound was irrigated and the skin subsequently closed with Monocryl.

Pathology
Our specimen was placed in formalin and sent for permanent examination by our Pathology department. On gross examination, the appendix was 9.5cm in length and 2.1cm in diameter with no perforation noted. The appendix was markedly dilated with an unremarkable tip. The wall measured 2mm thick and the mucocele consisted of thick pale white mucoid material. A gross picture of our specimen along with cross-sections can be seen in Figures 4 and 5, respectively. Immunostaining for p53 tumor suppressor and stathmin oncogene 18 were focally positive. Furthermore, immunostain for Ki-67 protein revealed increased proliferative activity. On histological diagnosis, the tumor was a mucinous cystadenoma measuring 7.0cm in length with negative 2.0cm resection margins. A representative portion of H&E stained slides of our specimen can be seen in low and high-power view in Figures 6 and 7, respectively.

DISCUSSION
Appendiceal mucocele is a broad inclusive entity that comprises all lesions that lead to a distended, mucus-filled appendix. The entity itself is rare and in one series found in approximately 0.3% of appendectomy specimens [4]. Its presence has been shown to have a slight female predominance and is typically found in the 5th or 6th decade of life [4]. Histologically, these lesions have been subdivided into four separate groups: mucosal hyperplasia, simple or retention cysts with degenerative epithelial changes, mucinous cystadenomas, and mucinous cystadenocarcinomas. Non-neoplastic lesions have a higher prevalence with one study reporting
prevalence rates of 52, 20, 18, and 10 percent in these categories, respectively [5-6]. To add, an association has been reported between the tumors mentioned and other tumors of the gastrointestinal tract, ovary, endometrium, breast, and kidney.

Management of an appendiceal mucocele typically mandates resection since lesions that may appear benign on imaging studies could be found malignant on histological analysis. Laparoscopy has been proven successful in the literature for isolated cases. Except for the most straightforward cases, open resection is encouraged to minimize the possibility for rupture and subsequent peritoneal contamination[7].

Appendectomy is deemed adequate in the first three subgroups as well as for cystadenocarcinoma without mesenteric, adjacent organ or peritoneal involvement. Involvement of any of these surrounding structures or the peritoneum necessitates a right hemicolectomy or laparotomy and debulking if peritoneal disease is seen. The prognosis of these lesions is excellent for the first three subgroups while mucinous cystadenocarcinomahas a 5-year survival that ranges from 6-100% based on tumor stage.

**CONCLUSION**

Appendiceal mucocele is a broad, complex entity with histological characterization showing four distinct subgroups. We emphasize that careful operative planning is necessary as pathological diagnosis is needed to definitively exclude malignancy. We successfully performed an open appendectomy with negative margins for a large 7cm mucinous cystadenoma in a patient presenting with abdominal pain.

**REFERENCES**


FIGURE LEGENDS

Figure 1: Base of the Appendix

Figure 2: Body of the Appendix

Figure 3: Distal tip of the Appendix

Figure 4: Dilated appendiceal base and body. The tip is unremarkable

Figure 5: Cut sections of the appendix which consisted of thick pale white mucoid jelly-like material

Figure 6: Low-power histological view of a appendiceal mucinous cystadenoma

Figure 7: High-power histological view of a appendiceal mucinous cystadenoma
FIGURES

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Figure 5: Cut sections of the appendix which consisted of thick pale white mucoid jelly-like material

Figure 6: Low-power histological view of a appendiceal mucinous cystadenoma
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