

# Mucinous cystadenoma presenting as appendiceal mucocele

Nirav Kishor Desai, Vladimir Rubinshteyn, Mena Ayoub, Asaf Gave

## ABSTRACT

**Introduction:** Appendiceal mucocele is a complex entity consisting of several different pathologies with a spectrum of clinical presentation. **Case Report:** We present a case of a 52-year-old male presented with a two-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. **Conclusion:** We review the pathology as well as the different pathological conditions that may lead to an appendiceal mucocele.

**Keywords:** Appendix, Appendiceal mucocele, Cystadenocarcinoma, Cystadenoma, Intussusception, Mcburney's point, Mucooid, Ki-67

### How to cite this article

Desai NK, Rubinshteyn V, Ayoub M, Gave A. Mucinous cystadenoma presenting as appendiceal mucocele. Int J Case Rep Images 2016;7(3):181–184.

Article ID: Z01201603CR10618ND

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doi:10.5348/ijcri-201631-CR-10618

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Received: 05 November 2015

Accepted: 23 December 2015

Published: 01 March 2016

## 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) scan. 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, presented with two 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 2.3 cm at the base and 1.5 cm towards the tip. No periappendiceal 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.

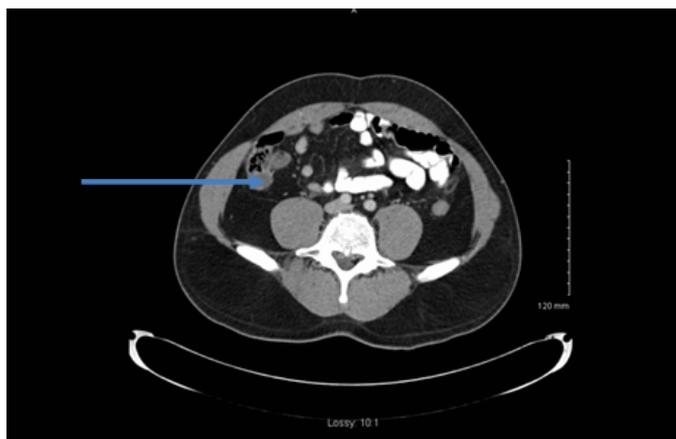


Figure 1: Base of the appendix.

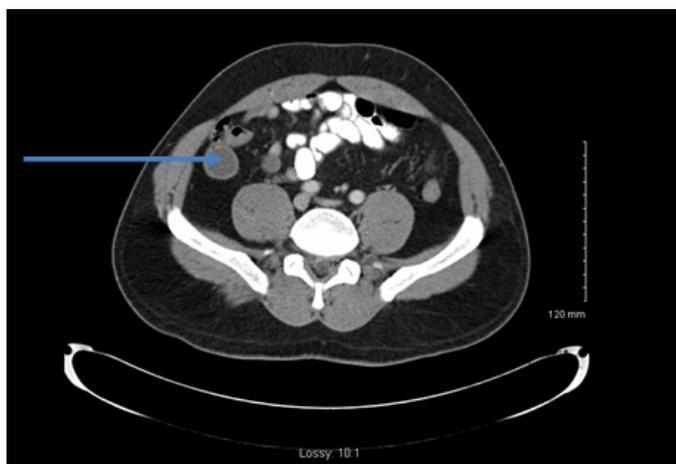


Figure 2: Body of the Appendix.

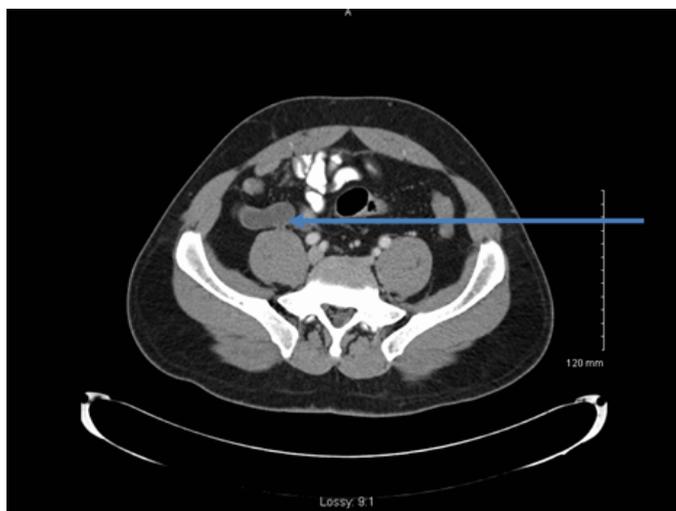


Figure 3: Distal tip of the Appendix.

### 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 9.5 cm in length and 2.1 cm in diameter with no perforation noted. The appendix was markedly dilated with an unremarkable tip. The wall measured 2 mm thick and the mucocele consisted of thick pale white mucoid material. A gross picture of our specimen along with cross-section 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 7.0 cm in length with negative 2.0 cm 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-



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.

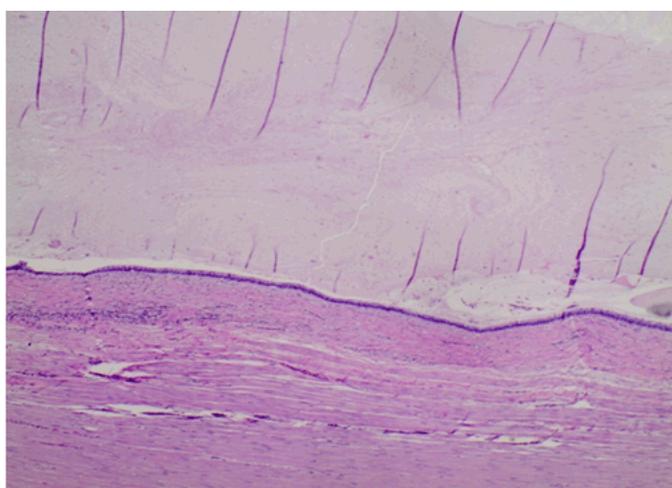


Figure 6: Low-power histological view of a appendiceal mucinous cystadenoma.

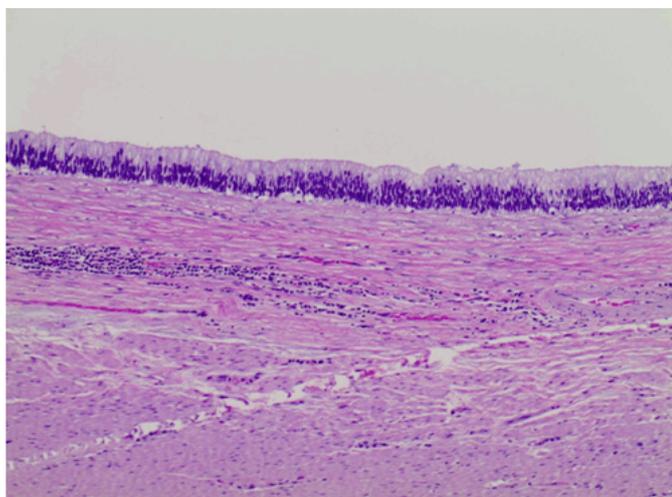


Figure 7: High-power histological view of a appendiceal mucinous cystadenoma.

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 straight-forward 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 cystadenocarcinoma has a five-year survival that ranges from 6–100% based on tumor stage [8].

## 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 7 cm mucinous cystadenoma in a patient presenting with abdominal pain.

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## Author Contributions

Nirav Kishor Desai – 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

Vladimir Rubinshteyn – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Mena Ayoub – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Asaf Gave – 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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