

## GIPS COM

### Case History

A 44-year-old female presented with a 1 month history of mild right lower quadrant pain, which slowly evolved into diffuse lower abdominal discomfort. On physical exam, the abdomen was soft, non-tender, and non-distended, and there was no rebound or guarding. Abdominal CT revealed an enlarged/dilated appendix. Appendectomy was performed.



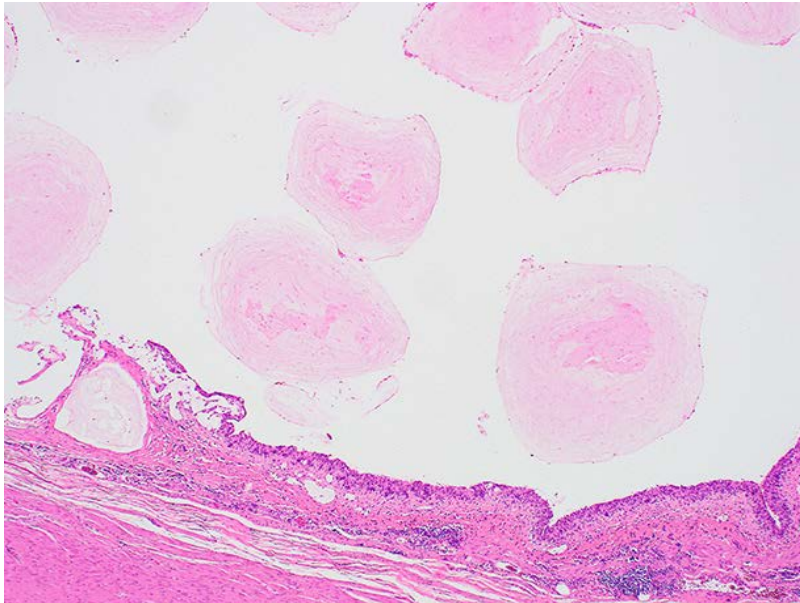
**Figure 1A.**



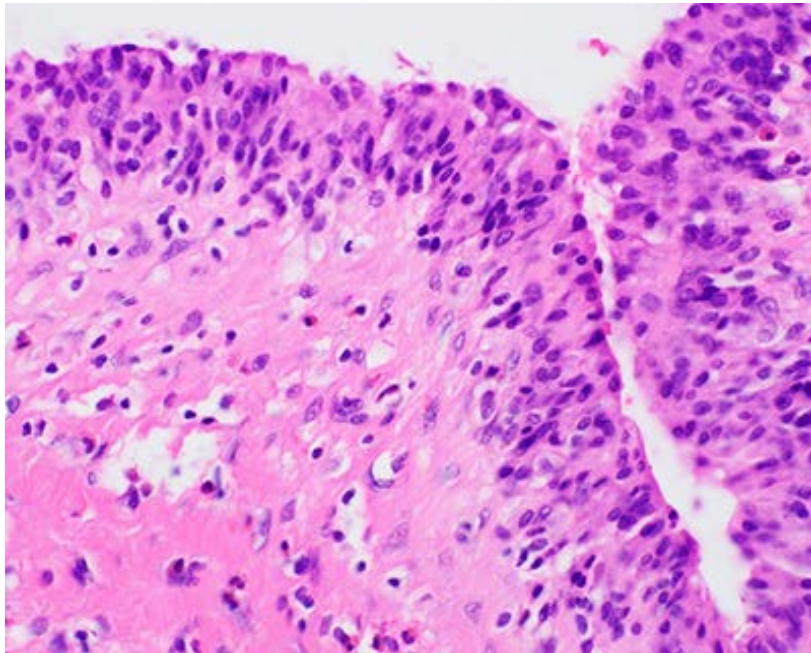
**Figure 1B**



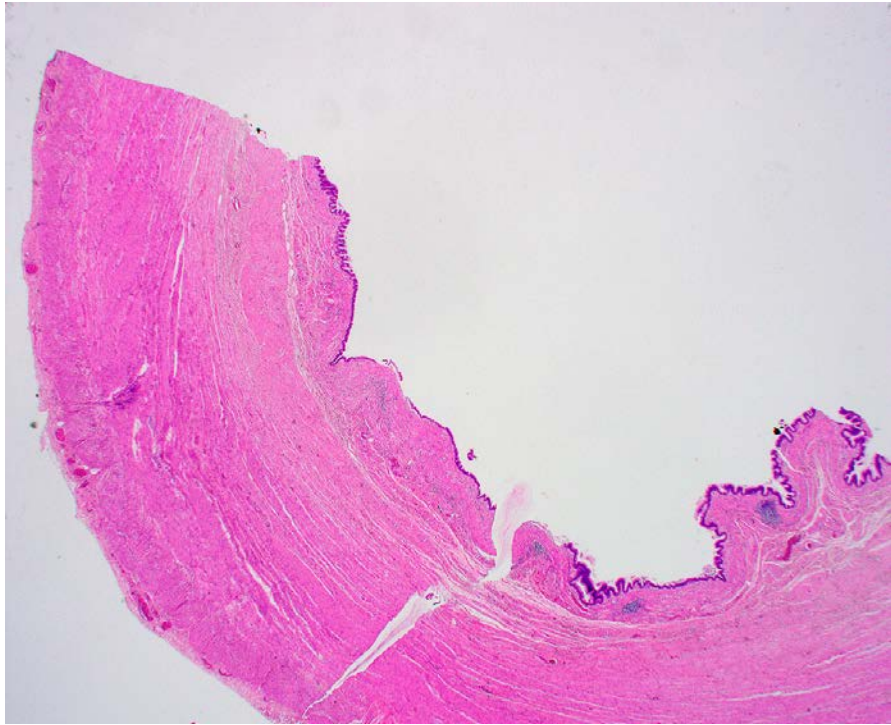
**Figure 1C**



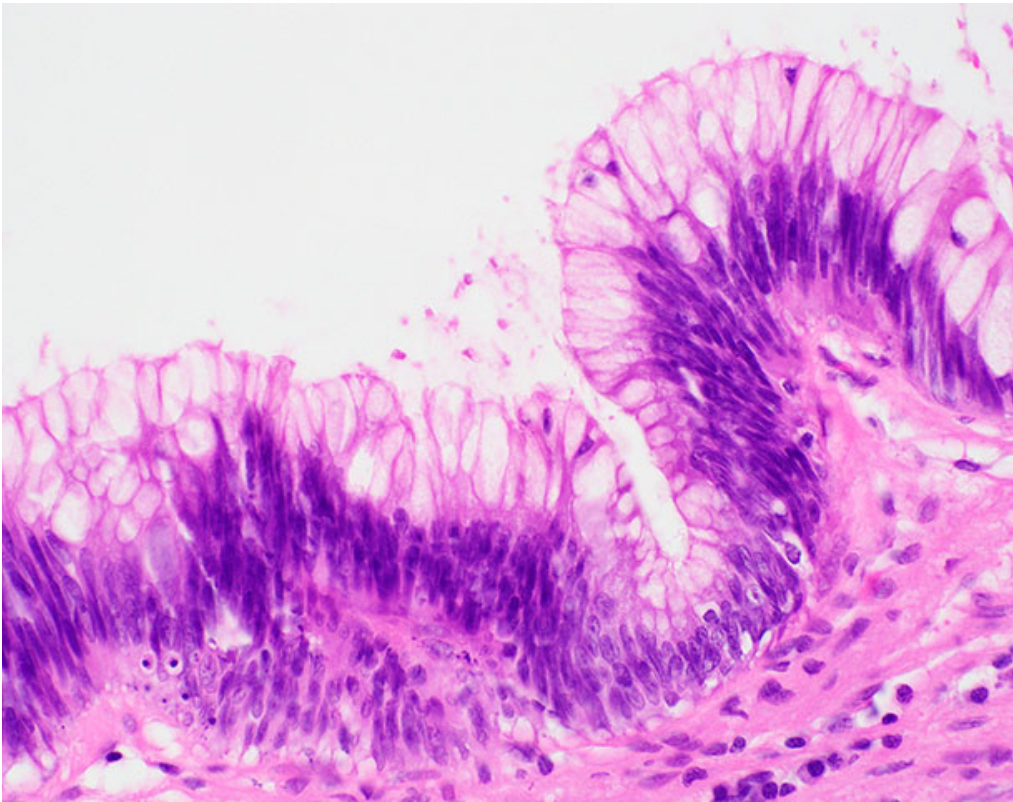
**Figure 2A; H&E Original magnification 40x**



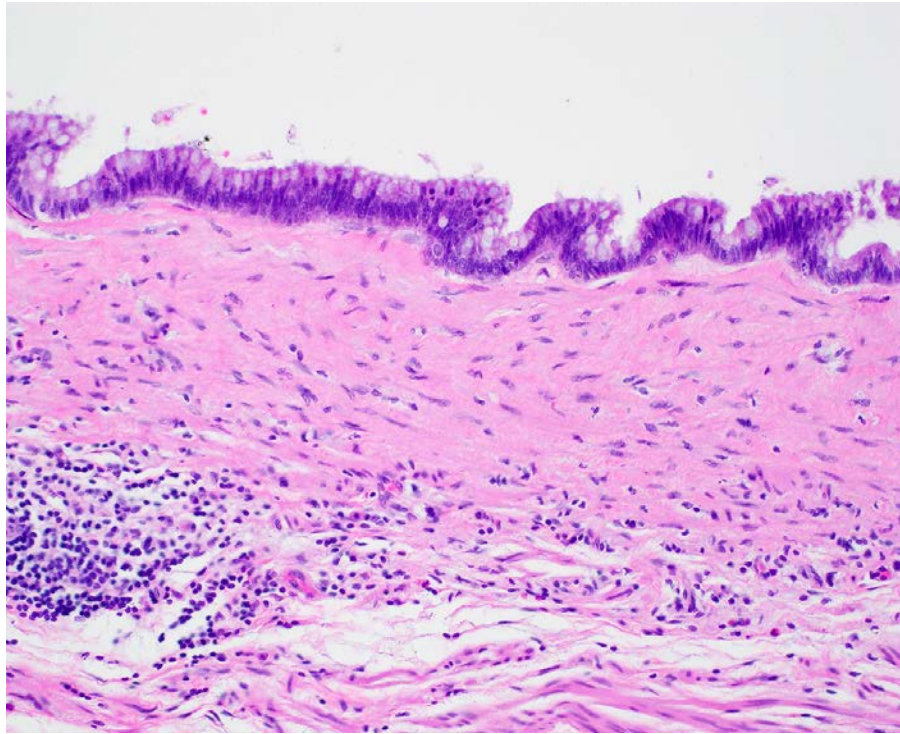
**Figure 2B; H&E original magnification 400x**



**Figure 2C; H&E Original magnification 12.5x**



**Figure 2D; Original magnification 400x**



**Figure 2E; H&E original magnification 200x**

**What is the most likely diagnosis?**

- A. Ovarian serous borderline tumor
- B. Desiccant ingestion r
- C. Myxoglobulosis and low grade appendiceal mucinous neoplasm (LAMN)
- D. Appendiceal helminthiasis
- E. Florid papillary mesothelial proliferation

### **Answer: (C) Myxoglobulosis and LAMN**

Gross inspection revealed a dilated appendix (Figure 1A) with a surface remarkable for soft opalescent beads on the serosa and within the specimen container. The lumen was filled with the opalescent beads (Figures 1B and 1C). The beads on the serosal surface were not attached, and by report, were from procedural disruption after the specimen was removed from the patient. The appendix was removed with a rim of cecum at the proximal margin, and the orifice appeared free of any mechanical obstruction. Microscopic examination revealed eosinophilic laminations containing a few necrotic cells (Fig 2A), present in the lumen and embedded in the mucosa. Portions of the mucosa were replaced by a prominent pseudosynovial reaction (Figure 2B). The remaining appendiceal lumen was lined by a monotonous epithelium with low grade dysplasia, an undulating growth pattern and areas of fibrosis obliterating the muscularis mucosae consistent with LAMN (Figure 2C, D, and E).

### **Discussion**

First described by Latham in 1897, myxoglobulosis is a rare variant of “mucocele” characterized by mucinous globules distributed throughout the lumen, which are similar in appearance to fish eggs<sup>1,2</sup>. “Mucocele” is a clinical term used to describe dilatation of the appendix due to mucin, and it does not denote a specific etiology. Literature reports “mucocele” in 0.2%-0.3% of appendectomy specimens, of which 0.35% to 0.8% contain myxoglobulosis<sup>3,4</sup>. Most cases were discovered incidentally on autopsy or laparotomy<sup>4</sup>; but some patients present with signs and symptoms of acute appendicitis<sup>3,5</sup>. Patients often present in their 6<sup>th</sup> to 7<sup>th</sup> decades and there is a slight female predominance<sup>5</sup>. Gross examination reveals an enlarged, dilated appendix containing translucent to opalescent globules located within the lumen<sup>6</sup>. Occasionally, these globules may be calcified<sup>4</sup>. The pathogenesis of myxoglobulosis remains unknown. Most literature states a requirement for several factors: (1) partial or complete obstruction of the proximal appendix (inversion, fecalith, epithelial hyperplasia, inflammation, or carcinoid tumor)<sup>3,6</sup>, and (2) there must be mucin production distal to the obstruction<sup>4,6</sup>. The mechanism of mucin globule formation is unknown, although authors have proposed a crystallization-like phenomenon, with nucleation of mucin around sloughed granulation tissue or necrotic epithelial cells<sup>2,7,8</sup>. Unfortunately, the majority of literature regarding myxoglobulosis harkens from the “mucocele” era, during which LAMN and HAMN were not distinguished from dilated appendices lacking an epithelial neoplasm. Given the reported “requirement” for obstruction of the appendix for myxoglobulosis, one would expect most cases were associated with non-neoplastic, true “mucoceles”. Recent reports of myxoglobulosis have reported abnormalities of the epithelium, with one report interpreting it as hyperplastic/adenomatous change<sup>6</sup>, and another interpreting it as appendiceal mucinous neoplasm<sup>9</sup>.

Differential diagnoses to consider for the epithelium include reactive non-neoplastic epithelium, or an appendiceal serrated polyp flattened by the presence of myxoglobulosis. Appendiceal serrated polyps are characterized by serrated epithelium with preserved lamina propria and preservation of the architecture of the appendix. Epithelium with reactive change should demonstrate varied cell types, while this case is comprised of a monotonous epithelium with features characteristic for LAMN. In our case, there was no apparent obstruction of the appendiceal orifice, and the nuclear and cytoplasmic atypia, undulating growth, and patchy obliteration of the muscularis mucosae justify a diagnosis of LAMN.

Appendiceal helminthiasis (D) does not typically fill the lumen with eggs. While there is limited information on the pathology of silica desiccant ingestion (B), there would not be any necrotic cells in the center of the globules. Ovarian serous borderline tumor (A) and florid papillary mesothelial proliferation (E) would be attached to the surface, and do not fill the lumen of the appendix. Microscopic examination revealed the presence of LAMN in the appendix.

## Figure Legends

Figure 1. Gross examination of appendectomy specimen. External examination (A) of the unfixed specimen revealed a soft, dilated appendix with opalescent soft beads on the surface of the specimen and in the specimen container. The beads were not attached to the surface of the appendix. Post-fixation, the specimen was bivalved (B, C) to reveal a lumen that was packed with variably sized beads (0.1 to 0.2 cm).

Figure 2. Microscopic examination revealed mucinous globules in the lumen (A). The lumen was lined by prominent pseudosynovial reaction (B) and LAMN (C, D, E).

## References

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