Isolated Appendiceal Crohn’s Disease

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ABSTRACT

Since its first description in 1953, granulomatous appendicitis is still considered a rare occurrence. Herein we present a 45-year-old Caucasian female who was evaluated for right lower quadrant abdominal pain. She was found to have inflammation of the appendix on computed tomography (CT) scan and mild elevation of inflammatory markers with leukocytosis. She was treated with antibiotics with resolution of her symptoms; however, her abdominal pain recurred. She underwent colonoscopy, which revealed an endoscopically unremarkable terminal ileum and cecum with no definite histologic evidence of Crohn’s disease (CD). The patient subsequently underwent laparoscopic appendectomy and was found to have histologic evidence of CD in the appendix. A capsule endoscopy following the operation did not show evidence of upper GI tract involvement. The patient remained symptom-free postoperatively. Isolated CD of the appendix is a rare occurrence. Appendectomy is often sufficient for patients who are diagnosed with radiographic and histologic evidence that show the disease is restricted to the appendix alone.

INTRODUCTION

Since its first description in 1953,¹ granulomatous appendicitis is still considered a rare occurrence, with incidence ranging from 0.2-0.62% of all appendectomies.² ³ Although there are various etiologies of appendiceal granulomas, isolated Crohn’s disease (CD) of the appendix has become more recognized with approximately 229 reported cases to date.¹ ⁷ ⁹ ₁₂ ₁₄ ₂₃

CASE PRESENTATION

A 45-year-old Caucasian female with no significant past medical history presented with abdominal pain. Twelve days prior to presentation, the patient developed constant, cramping abdominal pain localized to the right lower quadrant. She denied other associated symptoms including fever, nausea, vomiting, constipation, diarrhea, melena, or hematochezia. The pain began the day after she completed a 5-day course of azithromycin for a sinus infection. She denied nonsteroidal anti-inflammatory drug (NSAID) use. She endorsed drinking one glass of alcohol

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per month and denied tobacco or other drug use. The patient also acknowledged recent travel to Mexico two months before the onset of symptoms but denied any illnesses at that time.

The patient initially thought her pain was ovarian in origin and presented to her gynecologist. A pelvic ultrasound was performed and based on the results she was advised to go to the emergency department (ED) for further evaluation. A CT scan with contrast was performed in the ED and revealed a soft tissue density measuring 5.7 x 3 cm at the base of the cecum with surrounding inflammation of the adjacent fat. Patient was admitted for possible appendicitis or other gastrointestinal issue. A second CT scan with contrast obtained two days later showed extensive thickening of the bowel in the right lower quadrant with no defined appendix. Laboratory investigation performed during hospitalization was unremarkable with exception of elevated inflammatory markers and mild leukocytosis (C-reactive protein [CRP] 5.46 mg/dL, erythrocyte sedimentation rate [ESR] 36 mm/hr, white blood cell count [WBC] 12.3 K/UL). Serologic markers of inflammatory bowel disease were negative. The patient was treated with IV antibiotics for five days and discharged on a 21-day course of oral levofoxacin twice daily and metronidazole three times daily. A repeat CT scan performed two weeks after her hospitalization showed a dilated appendix with improved cecal wall inflammation. At this time, her inflammation improved and her symptoms had resolved. A colonoscopy with possible scheduled appendectomy was recommended. The patient presented to clinic, and a colonoscopy was scheduled for the following month. However, the patient’s symptoms recurred a week after her clinic visit (one month after initial hospitalization), and a second course of oral antibiotics was prescribed (10 days of ciprofloxacin twice daily and metronidazole three times daily) with subsequent resolution of symptoms. Outpatient colonoscopy was performed two weeks later.

The terminal ileum was examined up to 10 cm from the ileocecal valve and appeared grossly normal. The rest of the colon including the cecum appeared normal. The rectum had a slightly granular appearance. Biopsies were obtained and showed mild excess of intraepithelial lymphocytes and eosinophils in the terminal ileum but no definitive features of inflammatory bowel disease. Biopsies from the ascending, transverse, descending, and sigmoid colon were all normal. Rectal biopsies showed focal active proctocolitis with minimal chronicity consistent with acute self-limited proctocolitis from infection versus bowel preparation effects.

Given radiographic evidence of persistently dilated appendix and chronic return of symptoms, appendectomy was determined to be the best course of action. The patient was taken for an uncomplicated laparoscopic appendectomy, which was notable for a thickened and enlarged appendix. Pathologic review of the specimen was performed and the findings were consistent

![Figure 1. Low power global view of the appendix. Transmural lymphoid aggregates including rosary sign (arrows), stricture, fibrosis, smooth muscle proliferation, and mild architectural disarray (Hematoxylin & eosin [H&E] stain, 20x magnification).](image-url)
with Crohn’s disease of the appendix. Histology revealed chronic active appendicitis characterized by architectural disarray, crypt abscess, cryptitis, basal lymphoplasmacytosis, and excess Paneth cells. Frequent non-necrotizing sarcoid-type microgranulomas and small granulomas were also present as well as transmural lymphoid aggregates, smooth muscle proliferation, and fibrous obliteration of the appendiceal lumen (Figure 1 and 2). Notably, the specimen margins were free of ulcer. Further staining was negative for fungi, acid-fast organisms, and cytomegalovirus.

Postoperatively, the patient recovered without complication. Capsule endoscopy performed one month after surgery showed no evidence of CD in the small bowel or cecum. The patient remained symptom-free and reported no further episodes of abdominal pain during her last follow-up visit three months after surgery. Magnetic resonance imaging (MRI) of the abdomen and pelvis performed at that time was normal and showed no further areas of bowel inflammation.

**DISCUSSION**

Review of current literature reveals that CD limited to the appendix usually affects young adults with a mean age of 21-29 years old. However, our case demonstrates that this phenomenon is not limited to this age group, and indeed, the oldest reported patient with appendiceal CD was 66. There is also a male predominance with a ratio between 1.3:1 and 2:1. Clinical presentation of our patient was similar to other reports,

![Image](image-url)
which included protracted and indolent abdominal pain localized to the right lower quadrant. These symptoms combined with radiographic evidence of bowel inflammation lend way to a wide differential including, but not limited to, appendicitis, diverticulitis, inflammatory bowel disease, and bowel ischemia. It is prudent to prescribe antibiotics that provide broad enteric and anaerobic coverage as well as perform a colonoscopy if there is no risk of perforation. Negative biopsies, such as in our case, can help narrow the differential.

It appears that with isolated appendiceal disease, an appendectomy is a safe and effective means of treatment. One series reports 65% of patients had abdominal symptoms for more than five days before surgery with a maximum of seven weeks prior to surgery. Our patient underwent surgery about nine weeks after initial presentation. Post-appendectomy, recurrence rate varies from 0-10% with an average interval between surgery and recurrence of four years.

Due to the benign long-term course of isolated CD of the appendix, there is still some debate whether it can be a true representation of CD, which is a disease characterized by high rates of recurrence and fistulization. It has been suggested to be categorized as a completely distinct process altogether known as idiopathic (primary) granulomatous appendicitis. However, characteristic histologic differences favor the notion that CD of the appendix and idiopathic granulomatous appendicitis are two separate entities. Dudley and Dean first described these differences and noted an increased number of granulomas per tissue section in idiopathic granulomatous appendicitis compared to appendiceal CD where the granulomas were more dispersed. A study from Stangl et al. identifies the presence of lymphoid aggregates at the border of the muscularis propria and subserosa as an important distinguishing factor of CD in the appendix. Obliteration of the appendiceal lumen was also significantly greater in CD patients (31% vs. 12% in controls). These histologic findings are very consistent with findings from our case and serve as further evidence that we have identified a true experience of CD isolated to the appendix. Furthermore, our case demonstrates that an appendectomy is sufficient treatment and can be effective even in the delayed setting as late as nine weeks after initial presentation of symptoms.

**LEARNING POINTS**

- Crohn’s disease isolated to the appendix is a rare phenomenon and appears to have a more benign course compared to typical CD with decreased fistula rates and recurrence rates postoperatively.
- With isolated CD of the appendix, an appendectomy is a safe and effective means of treatment and can be performed in the delayed setting.
- Additional studies examining underlying histology and correlation with long-term follow up are warranted to further delineate idiopathic granulomatous appendicitis from true isolated appendiceal CD.

**REFERENCES**


