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Abdominal Pain in a 17-Year-Old Girl With Pancreatic Insufficient Cystic Fibrosis

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Case Presentation

A 17-year-old Caucasian girl with known cystic fibrosis (CF; F508del/621 +1G > T), associated pancreatic insufficiency, and congenital hypothyroidism presented by telephone to the University of Minnesota Cystic Fibrosis Clinic with 1 week of low-grade fevers, fatigue, and 1 month of intermittent abdominal pain described as diffuse and crampy in nature. Her abdominal symptoms were thought to be secondary to a prior diagnosis of constipation and were relieved with as-needed polyethylene glycol. She also reported having an upper respiratory infection 1 week prior to when the fevers began; however, her respiratory symptoms had completely resolved without treatment. Given the duration of symptoms, she was instructed to be evaluated the next day at a local primary care clinic. At that time, she did not have an identifiable source of infection, she had no respiratory symptoms, and a monospot was negative; however, she had an elevated white blood cell (WBC) count. The primary care physician was concerned for an early CF pulmonary exacerbation so she was prescribed a 2-week course of oral ciprofloxacin.

The following day, the patient called the CF clinic with a fever to 102°F, decreased appetite, and sharp upper abdominal pain that caused her to electively stop her high-frequency chest wall oscillatory therapy treatments. Secondary to these complaints, she was seen in the CF clinic where her abdominal pain was described as sharp and localized to the lower quadrants. She reported normal stools for the past few days, nausea, one episode of emesis, and a new productive cough. On presentation, she was tachycardic (heart rate 114) and tachypneic (respiratory rate 32) with a temperature of 100.1°F, blood pressure of 111/74, and an oxygen saturation of 94% on room air. Physical exam was significant for pallor and bilateral lower quadrant abdominal pain with guarding.

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Hospital Course

She was admitted for further evaluation and management of an acute abdomen. Laboratory studies on admission were the following: hemoglobin 10.8 (normal range = 11.7-15.7 g/dL), WBC 20 000 (80% neutrophils), erythrocyte sedimentation rate 72 (0-20 mm/h), C-reactive protein 188 (0-8 mg/L), negative β -hCG, negative chlamydia/gonorrhea polymerase chain reaction, and urine analysis/urine culture with no sign of infection. A chest X-ray showed underlying chronic changes of CF with no new or acute process. An abdominal X-ray demonstrated moderate stool with prominent small bowel loops and probable fecalization in the terminal ileum, suggestive of distal intestinal obstruction syndrome (DIOS). An abdominal and pelvic ultrasound revealed a large multilocular left ovarian cyst, $17.7 \times 8.5 \times 18.8$ cm, with no indication of torsion. The appendix was not visualized on ultrasound, so an abdominal and pelvic computed tomography (CT) with oral and intravenous contrast was obtained. The CT confirmed the presence of the ovarian cyst (Figure 1). In addition, a 5-cm inflammatory mass suggestive of an appendiceal abscess was identified (Figure 2).

The patient was taken to the operating room for a diagnostic laparoscopy with likely laparotomy. An appendiceal abscess and large ovarian cyst were both identified. The appendiceal abscess was resected laparoscopically, but due to concern for rupturing the large ovarian cyst, an open left oophorectomy was performed. Pathological examination identified a mucinous cystadenoma of the left ovary with no evidence of malignancy. She was treated with intravenous levofloxacin and metronidazole for 1 week as an inpatient, and oral levofloxacin and metronidazole on discharge to complete a 2-week course. Blood cultures remained negative and she has since made a complete recovery.

Final Diagnosis

(1) Appendiceal abscess, (2) mucinous cystadenoma of the left ovary, and (3) cystic fibrosis

Discussion

Cystic fibrosis is an autosomal recessive disorder known for its associated lung disease that results in early mortality; however, gastrointestinal (GI) complications can also be frequent and debilitating.¹ CF is caused by a mutation of the CF transmembrane conductance regulator (CFTR) gene, which codes for a protein responsible for chloride transport across epithelial membranes. In addition to its presence on the epithelial surface of the lungs, the CFTR protein is present in the GI tract, pancreatic ducts, and biliary system. When chloride transport is impaired, salt and water transport is disrupted, which results in thickened secretions in the affected organs. These thick secretions can cause a variety of GI complications including pancreatic insufficiency, CF-related diabetes, chronic pancreatitis, constipation, intussusception, focal biliary cirrhosis, cholangiectasis, and DIOS.¹

Constipation is a common problem in people with CF, affecting over 40% of the patient population, and occasionally it can progress to DIOS and an acute abdomen. DIOS occurs in 16% to 21% of the CF patient population.¹ It is hypothesized that DIOS is caused by altered bile absorption that leads to high fecal fat, which increases stool viscosity and results in obstruction typically found in the right lower quadrant (RLQ). Clinically, it presents with

acute, colicky periumbilical or RLQ abdominal pain that can mimic appendicitis.¹ The literature illustrates how a history of constipation and DIOS can cloud the diagnostic picture in the setting of abdominal pain in a CF patient. Shields et al describe a series of 9 CF patients with appendicitis, and 5 of the 9 patients were initially thought to have DIOS. This resulted in a significant delay in diagnosis and treatment.² Chen et al report a case of a 21-year-old Taiwanese male with CF who was found to have an intestinal obstruction and appendicitis, requiring open disimpaction and an appendectomy.³ Our patient had a history of intermittent abdominal pain relieved with polyethylene glycol 1 month prior to the acute presentation, making constipation with possible obstruction high on the initial differential diagnosis as a cause of her abdominal symptoms. Her initial imaging findings also raised concern for DIOS, which may have delayed the true diagnosis of appendicitis.

A second confounding factor that may have delayed our patient's diagnosis is the lower incidence of appendicitis in the CF population than in the general population. Appendicitis is commonly found on the list of differential diagnoses for a child or teenager with abdominal pain; however, the incidence of appendicitis in the CF population may be as low as 1% to 2%,⁴ compared with 7% in the general population.² The proposed mechanism by which CF patients have a lower incidence of appendicitis is that it may be related to protective eosinophilic secretions in the appendix that distend the appendiceal lumen, making it less prone to occlusion and acute inflammation.⁴ While it is necessary to include CF-related causes of abdominal pain in our patient's differential diagnosis, it is also critical to consider the more common causes of illness in our adolescent female presenting with abdominal pain, fever, and an elevated WBC count.

In addition to the history of constipation in our patient and the need for consideration of CF-related illnesses, the ciprofloxacin prescribed to our patient for a possible CF pulmonary exacerbation did not relieve our patient's symptoms and may have played an additional role in delaying the true diagnosis of appendicitis. Antibiotics are commonly prescribed to many CF patients for prophylaxis or treatment of pulmonary illnesses²; however, antibiotic use early in the course of illness or for other reasons has been proven to cause a therapeutic delay greater than 48 hours in pediatric patients with appendicitis.⁵ It also increases the difficulty of obtaining accurate diagnostic imaging. Patients who have received antibiotics require more CT scans than ultrasounds and an increased number of imaging studies overall, resulting in increased radiation exposure.⁵

The delayed diagnosis of appendicitis is concerning as it can result in appendiceal perforation and abscess formation; however, even in the general population, diagnosis remains a challenge.³ While appendicitis does have some key physical exam and laboratory findings that can increase clinical suspicion, many of these findings have significant overlap with other abdominal conditions or may vary depending on whether the patient presents either early on or at a late stage of illness.⁶ The Alvarado score is a points system that can help determine risk of appendicitis based on symptoms and findings. A 2011 meta-analysis calculated 99% sensitivity with a score of 5 or below.⁶ Our patient had anorexia, nausea and vomiting, elevated temperature, leukocytosis, left shift of WBC count, tenderness in RLQ, which gives her a total of 8 points and puts her at high risk of appendicitis (Figure 3). It is important to note that some of our patient's abdominal discomfort could be attributed to the

ovarian mucinous cystadenoma. There is no known relationship between mucinous cystadenoma of the ovary and CF, as was confirmed by a literature search on the topic. Our patient's mucinous cystadenoma is attributed to a coincidental finding.

In conclusion, this case highlights the importance of keeping a broad differential diagnosis for patients, even those with a known chronic disease. It is also crucial to revisit the differential diagnosis if therapies are not successful at improving symptoms. The cause of our patient's delayed diagnosis was likely multifactorial, including her history of constipation relieved by polyethylene glycol, use of ciprofloxacin, imaging findings concerning for DIOS, and presence of an ovarian mucinous cystadenoma. Described using one of our favorite clinical idioms, even if you have seen many zebras recently, those hoofbeats are still more likely to be horses.

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Figure 1.
Computed tomography scan of the abdomen and pelvis revealed a large cystic mass in the pelvis.

Figure 2.
Computed tomography scan of the abdomen and pelvis revealed a 5-cm inflammatory mass in the right central abdomen (arrow); its location suggests an appendiceal abscess.

Figure 3.
Alvarado score for the diagnosis of appendicitis.⁶