CASE REPORT

Acute appendicitis mimicking intestinal obstruction in a patient with cystic fibrosis

Chun-Han Chen\textsuperscript{a}, Cheng-Chih Chang\textsuperscript{a}, Bor-Yau Yang\textsuperscript{b}, Paul Y. Lin\textsuperscript{c}, Chia-Siu Wang\textsuperscript{a,d,}\textsuperscript{*}

\textsuperscript{a} Department of General Surgery, Chang-Gung Memorial Hospital, Chia-yi, Taiwan
\textsuperscript{b} Department of Radiology, Chang-Gung Memorial Hospital, Chia-yi, Taiwan
\textsuperscript{c} Pathology, Chang-Gung Memorial Hospital, Chia-yi, Taiwan
\textsuperscript{d} Chang Gung University, Kweishan, Taoyuan, Taiwan

Received 11 August 2009; received in revised form 24 November 2009; accepted 15 December 2009

KEYWORDS
abdominal pain; acute appendicitis; cystic fibrosis; distal intestinal obstruction syndrome

Cystic fibrosis (CF) is an inherited disease of the secretory glands caused by mutations of the cystic fibrosis transmembrane regulator (CFTR) gene. The clinical manifestations of CF are repetitive lung infections, biliary cirrhosis, pancreatic abnormalities, and gastrointestinal disorders. We report a 21-year-old Taiwanese man with CF who had abdominal pain for 2 days. The diagnosis of CF had been confirmed by peripheral blood analysis of the CFTR gene 5 years before admission. He presented to the emergency department with nausea, vomiting, abdominal distension, and crampy abdominal pain, which is atypical for acute appendicitis. The physical examination and a series of studies revealed intestinal obstruction, but acute appendicitis could not be ruled out. After conservative treatment, together with empiric antibiotics, the refractory abdominal pain and leukocytosis with a left-shift warranted surgical intervention. A diagnostic laparoscopy revealed a swollen, hyperemic appendix, a severely distended small intestine, and serous ascites. The laparoscopic procedure was converted to a laparotomy for open disimpaction and appendectomy. He was discharged on the eighth postoperative day. The histologic examination of the appendix was consistent with early appendicitis. In conclusion, acute abdominal pain in adult CF patients is often associated with intestinal obstruction syndrome. The presentation of concurrent appendicitis may be indolent and lead not only to diagnostic difficulties, but also a number of therapeutic choices.

Copyright © 2012, Elsevier Taiwan LLC & Formosan Medical Association. All rights reserved.

* Corresponding author. 6 West Section, Chia-Pu Road, Putz City, Chia-yi, Taiwan.
E-mail address: wangcs@adm.cgmh.org.tw (C.-S. Wang).

0929-6646/$ - see front matter Copyright © 2012, Elsevier Taiwan LLC & Formosan Medical Association. All rights reserved.
http://dx.doi.org/10.1016/j.jfma.2012.07.011
Introduction

Cystic fibrosis (CF) is a congenital disorder attributed to mutations of the cystic fibrosis transmembrane regulator (CFTR) gene, which was cloned in 1989.1–3 The clinical manifestations of CF are repetitive pulmonary infections, biliary cirrhosis, pancreatic abnormalities, and gastrointestinal complications. Cystic fibrosis is rare in Taiwan, but the true prevalence rate is not available. A dramatic improvement in survival has occurred in the past 3 decades. The median survival of patients with CF in the United States is predicted to exceed 40 years for a patient born in the 1990s, whereas it was only 16 years for those patients born in the 1970s.4 As a result, adult patients with CF will increase in number and proportion in the future. Previous knowledge about the gastrointestinal diseases in patients with CF was mainly derived from pediatric patients. Since abdominal pain is not an uncommon complaint in patients with CF, care providers should be aware of the differential diagnosis and management in the adult population. We present an unusual case involving a patient with CF who had acute abdominal pain in adulthood and a successful surgical outcome.

Case report

A 21-year-old Taiwanese man had periodic episodes of pneumonia and pneumothoraces since childhood. He had undergone several operations involving video-assisted bullae ligation and pleurodesis in the lungs bilaterally. Sweat chloride tests were remarkably high (>100 meq/l), but it was not until 5 years previously that a diagnosis of CF was confirmed by peripheral blood analysis of CFTR gene (heterozygous c.1898 + 5G > T and heterozygous p. I1023R). In reviewing his past history, chronic constipation was a persistent problem and he experienced crampy abdominal pain before defecation. There was no history of abdominal surgery or pancreatitis, and he never took pancreatic enzyme replacement therapy. Two days before he presented to our emergency department, he had the onset of intermittent, crampy, abdominal pain associated with a poor appetite, nausea, and vomiting. The symptoms began in the periumbilical area and shifted to the lower abdomen (right side predominant) before he came to the hospital. On physical examination, he had diminished bowel sounds, abdominal distension, bilateral lower abdomen tenderness, and equivocal rebound pain. There was no palpable mass in the right lower quadrant of the abdomen. A series of laboratory check-ups showed leukocytosis (WBC, 13.5 x 10⁹/L with 70% mature granulocytes and 22% lymphocytes) and an elevated C-reactive protein level (13.7 mg/L). Loops of distended small bowel with visible air in the cecum were demonstrated on plain film. Computed tomography (CT) revealed low-density intraluminal masses containing air bubbles, exhibiting a mottled appearance in the terminal ileum and cecum with ascites (Fig. 1A). A swollen appendix (1.0 cm in diameter) with minimal periappendiceal inflammation was also demonstrated (Fig. 1B). A barium enema excluded a colonic stricture, but the appendix was not opacified. Filling defects within the cecum and terminal ileum were also detected. Empiric antibiotics (cefazolin, gentamicin, and metronidazole) were prescribed. After repeated sodium phosphate enemas, there was only a small amount of watery defecation without obvious passage of flatus on the day of admission. Surgery was recommended due to the worsening abdominal pain and a leukocytosis with a left shift (WBC, 13.4 x 10⁹/L with 82% granulocytes and 2% late metamyelocytes) on the day after admission.

At laparoscopy, a distended small bowel with serous ascites and a hyperemic, distended appendix were found (Fig. 2). The laparoscopy was converted to a laparotomy. A thickened mucoid impaction was palpated in the terminal ileum and cecum, measuring 30 cm in total length, with collapse of the transverse and descending colon. The impaction was manually crushed and advanced into the transverse colon. Inspissated mucosal content was expressed from the lumen of the appendix specimen after the appendectomy. Later the same day, extubation was done successfully in the intensive care unit, and the patient was transferred to the ward the next morning. The postoperative course was uneventful. The cultures of the ascites fluid yield no bacterial growth. The histologic

Figure 1  (A) Contrast enhanced abdominal computer tomography (CT) revealed low-density intraluminal masses containing air bubbles and exhibiting a mottled appearance (arrows) in terminal ileum and cecum, where ascites could also be found in pericecal area. (B) Another CT image showed a swollen appendix with minimal periappendiceal inflammation (arrows).
The medical therapy of DIOS included pancreas enzyme replacement, laxatives, mucolytic agent (N-acetylcysteine), and osmotically active hypertonic agent (gastrografin).8 Conservative treatment of DIOS is usually effective,9 but clinical deterioration and signs of bowel ischemia warrant operation. However, surgery could be hazardous in patients with CF due to poor nutrient status and underlying lung disease. Surgical strategies, including open disimpaction, appendicostomy tube irrigation, enterostomy (for evacuation of impaction and irrigation) with or without a T-tube, and bowel resection, have been proposed.8 The choices of different procedures were basically made at operation to achieve bowel patency. Unfortunately, with a very limited case number, there were no available data concerning the recurrence rate following surgical treatment in literature.

In a CF patient with DIOS, refractory abdominal pain should merit a thorough investigation with higher index of suspicion. While Crohn’s disease was not a likely diagnosis according to the history presented, volvulus, intussusception, and colon carcinoma could be excluded by CT scan. However, the differential diagnosis among DIOS, appendicitis and fibrosing colonopathy might not be straightforward. Fibrosing colonopathy, a form of colonic stricture complicating CF first reported on 1994,10 has clinical presentations, such as abdominal pain and features of intestinal obstruction, may mimic DIOS.5 Barium enema provides reliable clues on the diagnosis of fibrosing colonopathy; the characteristic radiological findings include: colonic stricture, shortening of ascending colon, and abnormal haustra.11 The treatment of choice is surgical resection and the diagnosis is confirmed by histological examinations. To our knowledge, there was no optimal diagnostic criterion of fibrosing colonopathy on CT scan. Nevertheless, barium enema should be performed with extreme care in those with acute abdominal pain.

The incidence of acute appendicitis in patients with CF has been reported to be 1% to 2%, which is lower than the 7% occurring in the general population.12 The reason for the low incidence of appendicitis in CF patients was not clear. A protective effect of mucous secretion within appendix had been proposed that it might help to keep the lumen from total occlusion and therefore decreasing the risk of acute inflammation.12 Prolonged use of antibiotics for the pulmonary infections in CF patients might also account for the low incidence of appendicitis. Appendicitis among CF patients is usually associated with delayed diagnosis, substantial perforation, and abscess formation.13 Delayed diagnosis was related to extended surgery in a Toronto study; specifically, three of four patients with delayed diagnosis of appendicitis underwent laparotomy, while five other patients with an early diagnosis were cured by appendectomy.14 Nevertheless, the early diagnosis of appendicitis remains a diagnostic challenge. There is no single clinical score that has been accepted as an ideal diagnostic tool for acute appendicitis in general population, but a high positive predictive rate of 94% in adult males has been reported using the Alvarado score.15 The Alvarado score has 8 variables, including migratory abdominal pain, anorexia, nausea (vomiting), pain on pressure in the right lower quadrant, rebound tenderness, fever, leukocytosis, and neutrophilia, in which right lower quadrant pain and leukocytosis were each assigned 2 points, with a total of 10 points. In the case presented herein, nausea/vomiting, anorexia, rebound pain, and leukocytosis scored 5 points, which would have predicted a low probability of acute appendicitis and encouraged further imaging studies.16 The common diagnostic criteria of acute appendicitis on CT scan include increased appendiceal diameter, appendicolithiasis, wall thickening, and signs of periappendiceal inflammation. In CF patients, however, an image diagnosis might lack accuracy, as the outer diameter of the appendix is not a reliable criterion.17 A swollen appendix with ascites, as existed in the case herein, should not be mistaken as a ruptured appendicitis. The barium enema has a limited role on the diagnosis of acute appendicitis since it is low in accuracy and might result in severe complications including perforation.18 A mass effect on the cecum has been reported in patients with CF on barium enema; however, it is not a reliable sign of appendicitis.19 We recommend a diagnostic laparoscopy to minimize the risk of an unnecessary laparotomy in CF patients with suspected appendicitis, since laparoscopic appendectomy has diagnostic and therapeutic advantages in the general population.20

Therapeutic strategies for CF patients with intestinal obstruction and concurrent appendicitis have never been
well elucidated in the literature. Only limited information is available from a few case reports and retrospective series.\textsuperscript{14,21–24} Surgical mortality has been reported involving a 19-year-old woman with DIOS, who had symptoms initially thought to be appendicitis, after a subsequent laparotomy for postappendectomy ileus.\textsuperscript{22} On the other hand, a case with persistent medical treatment of DIOS for nearly 2 months has been related to appendiceal abscess formation and eventual resection of the cecum and terminal ileum.\textsuperscript{23} Furthermore, appendectomy without disimpaction may result in prolonged postoperative ileus.\textsuperscript{24,25} A wide variation in presentation of appendicitis in CF patients leads not only to diagnostic difficulties, but also numerous therapeutic options. Some authors have advocated incidental appendectomy in CF patients undergoing other abdominal procedures.\textsuperscript{26} When laparoscopy reveals evidence of DIOS and appendicitis, conversion to open surgery combining appendectomy and disimpaction is a safe and effective option.

In conclusion, acute abdominal pain in CF patients is often related to DIOS, and it could mimic (or mask) symptoms of appendicitis. Although the incidence of appendicitis is low in patients with CF, there are no simple ways to exclude the diagnosis. Laparoscopy provides valuable information on the differential diagnosis and could potentially change the definitive treatment. Surgical intervention is justified in those with DIOS refractory to medical treatment or in those who have deteriorated clinically.

References