Unusual association of diseases/symptoms

Appendico-colic fistula complicating appendicitis in cystic fibrosis

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Summary

The case of a boy with cystic fibrosis who presented with an unusual complication of appendicitis is reported. Delayed presentation, with complications of appendicitis such as perforation and abscess formation, is not uncommon in cystic fibrosis; however, this case represents the first report of an isolated appendico-colic fistula following appendicitis in association with cystic fibrosis.

BACKGROUND

Cystic fibrosis is the most common autosomal recessive condition and can present with a myriad of potential causes for abdominal pain.

CASE PRESENTATION

A 13-year-old boy with cystic fibrosis presented with an 18-month history of intermittent, mild central abdominal and right-lower quadrant pain. There were no other associ-

Figure 1 Gastrograffin enema demonstrating a right iliac fossa soft tissue mass displacing the terminal ileum and an appendico-colic fistula (white arrow) between the base of the appendix (dashed arrow) and the sigmoid colon (dotted arrow).

ated gastrointestinal symptoms. Medications included prophylactic antibiotics and pancreatic enzyme supplementation. On examination the patient was afebrile with a heart rate of 80 bpm. Abdominal examination revealed a mass in the right iliac fossa, which was mildly tender but with no evidence of rebound tenderness or guarding.

INVESTIGATIONS

Haematological and biochemical investigations were normal. Abdominal ultrasound demonstrated a small ill-defined fluid collection in the right iliac fossa. Gastrograffin enema identified a soft tissue mass, which was displacing the ileum, together with a fistula between the sigmoid colon and the caecum (figure 1).

TREATMENT

Following optimisation of respiratory function with antibiotic treatment and intensive physiotherapy the patient underwent a laparotomy. Dense adhesions were identified involving the ileum, caecum and sigmoid colon. A fistula was evident running from the stump of a retrocaecal appendix to the sigmoid colon. The sigmoid colon adjacent to the fistula was oedematous and inflamed as was the terminal ileum and caecum. The fistula was excised and a sigmoid colectomy performed together with a right hemicolectomy.

OUTCOME AND FOLLOW-UP

The patient made a good but slow recovery and was discharged on the 11th postoperative day. Histological examination of the resection specimen indicated that the fistula was lined by chronically inflamed colonic mucosa suggesting that the primary pathology had been acute appendicitis.

DISCUSSION

Cystic fibrosis is the most common lethal autosomal condition: a defect in chloride channels in exocrine glands resulting in abnormal secretions. While the primary manifestation of cystic fibrosis is usually related to respiratory pathology, the gastrointestinal tract is a common site of cystic fibrosis-related complications. Pancreatic insufficiency¹ and pancreatitis² are well-known associations. Hepatobiliary complications range from fatty liver infiltrates, liver nodularity and biliary cirrhosis,¹ cholelithiasis³ and intrahepatic microlithiasis.⁵ Oesophageal reflux and Barrett's oesophagus are documented.⁶ Distal intestinal obstructive syndrome is not uncommon⁷ and has recently been reviewed.⁸ Ileocecal and colonic stenoses⁹ (with wall thickening) and fibrosing colonopathy¹⁰ may be related to replacement pancreatic enzyme treatment. Intussusception^{11 12} and rectal prolapse are other known associations.¹³

Acute appendicitis develops in up to 1.5% of cystic fibrosis patients—a rate that is significantly lower than the 7% seen in the general population.¹⁴ However, a high percentage of these patients present late with appendiceal abscesses as the disease process is frequently masked by antibiotic treatment prescribed for respiratory infections. For this reason, and since they generally have a poor respiratory status, their morbidity and mortality following appendicectomy is significantly higher.¹⁴

A detailed review of the literature revealed only five cases^{15–19} of fistulation between appendix and colon (in English language case reports) all in the presence of diverticular disease. A sixth case describes the first case of appendico-sigmoid fistulation in the absence of diverticular disease.²⁰ We describe the second documented occurrence of isolated fistulation between the appendix and sigmoid colon—the first in the presence of cystic fibrosis and the increased risk of appendicular complications.

Learning points

- Cystic fibrosis can present with a myriad of abdominal problems.
- While rates of appendicitis are low, they present late with appendiceal abscesses.
- Appendicocolic fistulation has been reported in the presence of diverticulitis; only one previous case has been documented in the absence of diverticulitis.

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Competing interests None.

Patient consent Obtained.

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