Unusual presentation of more common disease/injury

Enterocutaneous fistula as early presentation of Crohn's disease in an adult woman

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Correspondence to Dr Javariah I Asghar, asgh0002@umn.edu**Summary**

Crohn's disease is a chronic transmural inflammatory process that primarily affects the gastrointestinal tract, with several extra-intestinal manifestations. Though patients usually present with vague abdominal pain, initial presentation can be at an advanced stage or with extra-intestinal pathology. The authors report the case of a 59-year-old woman, who presented with a tubo-ovarian abscess that resulted in a protracted non-healing enterocutaneous fistula, which was eventually diagnosed as Crohn's disease approximately 2 years after initial presentation. Relevant literature is reviewed, as well as time-points where an earlier diagnosis could have been made thereby underscoring the importance of considering Crohn's Disease in a non-healing enterocutaneous fistula in an otherwise asymptomatic patient.

BACKGROUND

Crohn's disease (CD) is a chronic, transmural, inflammatory bowel process characterised by pathology in the distal ileum, though it can occur anywhere along the gastrointestinal tract.¹ Patients usually present with abdominal pain in the third decade of life, with or without fever, weight loss, fatigue and protracted diarrhoea. However, atypical and rare presentations of intestinal and extra-intestinal CD have been described in the literature.²⁻⁵ The disease incidence in North America is 3.1 to 14.6 cases per 100 000 persons per year, with 40% of cases presenting with ileocolic disease, 30% limited to the small bowel, and 30% with disease in the colon or anorectum. The transmural nature of inflammation predisposes bowel to the formation of abscesses or fistulae. Diagnosis is usually made based on clinical history, presentation and biopsy samples obtained via colonoscopy.

Our case describes a woman in her sixth decade of life, without any antecedent history of typical abdominal symptoms, who presented with a pelvic mass that after protracted surgical treatment, lead to a diagnosis of CD. We discuss clinical events, which may have warranted further investigation and the need for including CD in the differential diagnosis of non-healing enterocutaneous fistulae.

CASE PRESENTATION

Our patient is a previously healthy 59-year-old woman, who was admitted to a community hospital in August 2009 with abdominal pain of short duration and diagnosed with a tubo-ovarian mass. She had been normotensive and afebrile without changes in bowel habits. She did not have any significant personal or family history of gastrointestinal or gynaecologic pathology. She was staged appropriately and underwent a total abdominal hysterectomy and bilateral salpingo-oophorectomy. Intraoperatively, she was found to have endometriosis and a hepatic haemangioma. The patient did well postoperatively and was subsequently discharged home. Final pathology of the tubo-ovarian mass revealed an abscess.

Two months later, she presented to our emergency department with a 4-day history of abdominal pain, nausea, emesis and an inability to eat. On examination, she had a temperature of 36.1°C but without tachycardia or hypotension. Her abdomen was distended and tender to palpation with diminished bowel sounds.

INVESTIGATIONS

Haematologic and biochemical investigations were significant for a leukocytosis of $15 \times 10^9/\text{dl}$ and an international normalised ratio of 1.88. Imaging studies, including CT scans revealed a possible bowel leak with multiple air-fluid levels and free fluid in the pelvis (figure 1a,b).

TREATMENT

She underwent an exploratory laparotomy that revealed stool-covered bowel and intraperitoneal fluid. After multiple attempts at localising the leak, including intraoperative flexible sigmoidoscopy, a small epithelialised channel was discovered 20 cm proximal to the ileocecal valve. The fistula was resected and an ileostomy created. Histopathology of the resected bowel revealed ulceration, mural fibrosis and inflammation. The patient did well postoperatively though required supplemental parenteral and subsequent enteral nutrition due to significant malnutrition. She was eventually discharged home.

The ileostomy was reversed in March 2010 without complications. However, the patient developed a chronic, non-healing wound at the site that required several intraoperative debridements over several months. Imaging studies, including CT scans with sinus tract visualisation and small bowel follow-through, did not indicate a fistula or intraabdominal abscess. During this time, she had no other symptom of disease except significant malnutrition. In March 2011, she underwent an exploratory laparotomy for wound exploration and to better evaluate the right lower quadrant. Intraoperatively, she was found to have densely adherent bowel to the abdominal wall and two

Figure 1 (a) CT abdomen indicating right upper quadrant fluid collection (red arrow). (b) CT abdomen and pelvis indicating dense fluid collection and associated inflammation in the pelvis (red arrow).

enterocutaneous fistulae. These were taken down, along with an ileocectomy including the previous ileal anastomosis. Final pathology revealed inflammation consistent with CD.

OUTCOME AND FOLLOW-UP

She was diagnosed with silent ileal CD. Her subsequent postoperative course was uncomplicated with negative magnetic resonance enterography and Crohn's serologies. Given the high likelihood of recurrence in fistulising CD, she is being closely monitored but has not experienced a recurrence yet and is doing well.

DISCUSSION

Our patient had no family history of inflammatory bowel disease (IBD) or any history of prior symptoms of abdominal pain, diarrhoea or gastrointestinal haemorrhage. Her initial presentation of a tubo-ovarian abscess may have been the result of an internal enteric fistula. Following the second laparotomy, the patient's prolonged state of severe malnutrition, for which she required supplemental enteral and occasional parenteral nutrition, may have been a partial manifestation of the terminal ileal mucosal inflammation and partly due to the significantly higher metabolic requirements to heal a chronic wound, but was attributed to the metabolic demands of postoperative healing. The treatment with ileostomy, several courses of antibiotics to treat cellulitis and a chronic wound, and the resection of only a small portion of the terminal ileum that could not have resulted in malnutrition, should have prompted investigation into a chronic inflammatory process, such as CD, but may likely have distracted from it, as these are standard treatments in the pretext of IBD treatment. Histopathology, following the patient's second laparotomy was suggestive of CD but was insufficient for a definitive diagnosis. Given the unusual and rare presentation of CD described in the literature, a higher index of suspicion may have helped make an earlier diagnosis. At the time of the second laparotomy, specific antibody testing, such as the anti*Saccharomyces cerevisiae* antibody, may have been informative as it is 40% sensitive and 100% specific for CD.

Mounting evidence exists regarding the efficacy of biologics, such as antitumour necrosis factor α antibodies, in inducing CD remission and decreasing recurrence rates in patients with moderate to severe exacerbation, fistulating pathology or steroid-refractory disease.⁶ However, these studies do not distinguish between small bowel or peri-anal fistulas. In fact, most studies have grouped various types of fistulas together while in practice, therapy is tailored for the patient's presentation and varies by type of fistula. A growing cohort of studies has evaluated the use of biologics in inducing remission or maintaining quiescence of CD after surgical resection.^{7 8} The results remain controversial: though these studies have reported high rates of success, the number of patients treated in each study is small and most have grouped strictures and fistulas together in the treatment cohorts. Biologics, in this patient population, have been recommended for patients with known CD who are at high risk of recurrence postoperatively. Given the side effects associated with biologics, we discussed treatment options with our patient and chose to observe her clinically and endoscopically postresection.

In conclusion, we believe a higher index of suspicion for IBD is warranted when treating cases of chronic, non-healing abdominal wounds following gastrointestinal or pelvic surgery. Findings of transmural intestinal ulceration and fibrosis on histopathology, severe chronic malnutrition of unclear aetiology and fistulation at the site of non-healing wounds should further raise suspicion for Crohn's aetiology.

Learning points

- Higher index of suspicion for IBD is warranted when treating cases of chronic, non-healing abdominal wounds following gastrointestinal or pelvic surgery.
- Severe chronic malnutrition of unclear aetiology and fistulation at the site of non-healing wounds should raise suspicion for CD.
- Transmural intestinal ulceration and fibrosis on histopathology should prompt investigation of CD as possible aetiology even in the absence of other symptoms.

Competing interests None.

Patient consent Obtained.

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