Crohn’s disease of the appendix with enterocutaneous fistula post-appendicectomy: An approach to management

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Abstract

Context: Isolated involvement of the appendix in Crohn’s disease is reported to be 0.2% to 1.8%, and is usually associated with ileocaecal Crohn’s disease in 25% of ileal and 50% of caecal disease. While appendicitis in a patient who was previously diagnosed to have ileocaecal Crohn’s may be managed with appendicectomy and ileocaecal resection, appendicectomy alone when performed for appendicitis in a patient with unsuspected ileocaecal Crohn's disease could lead to postoperative complications including enterocutaneous fistula. Case Report: A young female patient who underwent appendicectomy elsewhere for acute appendicitis presented to us with a persistent enterocutaneous fistula of 6 weeks duration. She had complained of general ill health and occasional altered bowel habits for 6 months prior to the acute appendicitis presentation. Our investigations, including a CT scan, suggested the possibility of ileocaecal Crohn’s disease. She underwent excision of the enterocutaneous fistula and ileocaecal resection, and histopathology of the resected specimen confirmed Crohn’s disease. In the postoperative period she received mesasalazine. When last seen 2 years later during her regular follow-up, she was found to be in good health. Conclusion: The possibility of ileocaecal Crohn’s disease should be considered in patients presenting with unexplained postoperative enterocutaneous fistula following appendicectomy. A high index of clinical suspicion is required to make a prompt diagnosis and institute appropriate further treatment in form of ileocaecal resection.

Keywords: Crohn’s disease, appendicitis, ileocaecal resections, enterocutaneous fistula.

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Introduction

Crohn’s disease is a chronic inflammatory condition that may involve the entire GI tract. Histological features include granuloma, lymphoid aggregates, fissures and ulcers extending into muscularis propria, and transmural inflammation [1, 2, 3]. Crohn’s disease has been reported to be the cause of acute appendicitis in 0.2% to 1.8% of all the appendicectomies in some series [1, 3]. Operations described in the literature for Crohn’s disease of the appendix include appendicectomy with caecectomy and ileocaecal resection [2, 3, 4]. However, patients who undergo appendicectomy alone are at the risk of complications including enterocutaneous fistula in 34% to 58% of the cases [3, 4]. These patients may require further surgical intervention in the form of ileocaecal resection.

Case Report

A 24-year-old unmarried female patient presented to us with a persistent fistula in the right iliac fossa following an appendicectomy done in another hospital 6 weeks previously. On further questioning, she informed us that she had presented to this hospital with a 2-week history of abdominal pain, confined to the right lower abdomen. She complained of general ill health and occasional altered bowel habits for 6 months prior to this acute episode.
An enquiry with the doctors who had initially seen her revealed that at the time of presentation for acute appendicitis she was quite unwell with fever, tachycardia, and dehydration. Her abdomen was tender all over with guarding, more pronounced over the right lower half. Her white cell count was 24 x 10^9/L. An X-ray of her abdomen showed a few fluid levels and an ultrasound scan revealed free fluid in the abdomen with an associated right iliac fossa mass.

A diagnosis of pelvic peritonitis secondary to acute appendicitis was made by the physicians, and she was taken up for surgery after adequate resuscitation. The abdomen was approached through a right McBurney's incision. There was pus in the abdomen with an appendicular mass, and the appendix appeared oedematous, thickened, and congested. An appendicectomy was carried out followed by a thorough irrigation of the abdominal cavity. The abdomen was closed after placing a drain, which was removed on the 3rd post-operative day. In spite of antibiotic administration, she developed a wound infection and persistent pyrexia. She was managed conservatively with antibiotics and anti-inflammatory medication and eventually improved, except for a persistent purulent discharge from the wound. She then decided to seek our opinion and was admitted under our care for further investigation and management.

The appendicular histopathology was retrieved from the previous hospital and revealed transmural inflammation with granulomas suggestive of Crohn’s disease. A computerized tomography (CT) scan of the abdomen carried out in our hospital showed pericaecal collection with an inflammatory mass in front of the caecum (Fig. 1).

The abdomen was surgically explored through the previous incision after excising the fistula leading into the caecum. An inflammatory mass associated with the caecum was noted. The appendicular stump had not healed, and was draining into a cavity which was communicating with the wound, indicating a complex enterocutaneous fistula. A limited right hemicolectomy was carried out and continuity established with a primary anastomosis of macroscopically-appearing healthy bowel of the ascending colon and terminal ileum. A specimen of the resected caecum revealed the cobblestone appearance of the mucosa, strongly suggesting the possibility of underlying caecal Crohn’s disease (Fig. 2). The histology was reported as inflammatory bowel disease (IBD) consistent with Crohn’s disease (Fig. 3). The patient’s post-operative recovery was uneventful. She was referred to a gastroenterologist and was being treated with mesalazine and a regular annual colonoscopy. When last seen at two years post-surgery she continued to remain in good health.

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Discussion
Crohn’s disease typically affects the ileum, colon, and perianal region. Crohn’s disease may involve the appendix by extension from the terminal ileum or the caecum and present as an acute or sub-acute appendicitis [1-5]. Crohn’s disease limited to the appendix usually affects young adults in their 20s and 30s, although it is not limited to this age group and can occur at any age[3,6,7]. The clinical presentation is variable, with acute right iliac fossa pain suggestive of acute appendicitis in about 85% of patients [2, 3, 4], and chronic pain and a palpable mass in the right iliac fossa in about 25% of patients [3, 4]. When the preoperative diagnosis is acute appendicitis, Crohn’s disease should be suspected when an atypical or protracted clinical course is present in a patient prior to the appendicitis, particularly in areas where Crohn’s disease is prevalent [1, 2, 3]. Bowel obstruction can be another acute presentation of Crohn’s disease of the appendix [3].
Crohn’s disease is diagnosed by a combination of clinical manifestations and radiological findings including barium studies, showing string strictures, fissures, and fistula in the ileum and caecum [5, 6, 7, 8]. Recently, gray scale sonography and color Doppler-flow features have been used in diagnosing Crohn’s disease [9]. When the disease is located atypically in the appendix, histological evaluation is required since during surgery a conclusive diagnosis is difficult [1, 3, 6]. Macroscopically, the appendix will be seen to be enlarged, swollen and adherent to the surrounding structures, these findings being secondary to chronic inflammation. Histologically, the disease is characterized by transmural inflammation with thickening of the appendiceal wall, epitheloid granulomas, lymphoid aggregates, and mucosal ulceration [3, 5, 6, 7]. Other histological features are multinucleated giant Langherhan’s cells, crypt abscess, neural hyperplasia and lymphangiectasia [6].

Differential diagnosis includes the presence of foreign bodies and diverticulitis of the appendix, which also give rise to chronic granulomatous inflammation with induration and fibrosis of the appendiceal wall, and granulomatous disease of unknown aetiology such as appendiceal sarcoidosis, which is rare and often accompanies systemic disease [1, 3, 4, 8]. Differential diagnosis should also include infectious diseases such as tuberculosis, actinomycosis and Yersenia infection [1, 3, 10, 11]. A rare fungal infection such as histoplasmosis or blastomycosis, or a parasitic infestation such as Schistosoma or Enterobius vermicularis infestation can elicit an appendiceal granulomatous reaction [3, 6]. The diagnostic difficulty arises when a patient undergoes an appendicectomy for suspected acute appendicitis and the surgeon unexpectedly encounters ileocaecal pathology whose nature is difficult to ascertain in an emergency situation. In view of this, a definitive treatment in the form of ileocaecal resection for conditions like Crohn’s disease may be difficult to carry out. While a frozen section may be helpful to differentiate some of these conditions it may not always be feasible in an emergency situation.

If Crohn’s disease of the appendix is suspected and is limited to the appendix then appendicectomy alone is a routine surgical procedure with very low intraoperative or postoperative mortality and a low rate of fistula formation [1, 2, 3]. The postoperative enterocutaneous fistula incidence rate in Crohn’s disease restricted to appendix alone has been reported to be 3.5%, whereas in patients with Crohn’s disease of the ileocaecal segment, as in our patient, the postoperative enterocutaneous rate rises to 34% to 58% [2, 3, 4]. Weston et al reported that the majority of the patients whose first presentation of Crohn’s disease simulates appendicitis and who undergo appendicectomy alone leaving the ileocaecal segment in place to be treated medically, returned postoperatively within 3 years with symptoms. A significant percentage (38%) of these patients returned within one year and most of them (77%) were frequently ill with their disease in the interim period [4]. Patients who had an ileocolic resection at the time of the initial operation did not require early reoperation, did not develop short bowel syndrome, and did not have significant postoperative complications [4]. In the postoperative period these patients receive medical therapy to keep the disease quiescent such as salazopyrine, 5-ASA, prednisilone, and azothioprine [2, 4]. Crohn’s disease-related fistula have also been treated with infliximab with some success [12]. Despite improvements in medical therapy, between 70% to 90% of the patients with Crohn’s disease will eventually require surgical intervention, and approximately half of these will require additional operations [13]. Because of the high risk of reoperations and a relatively young age at the time of first operation, a minimally invasive approach has recently been used to carry out the ileocolic resections [13]. A recent meta-analysis of laparoscopic ileocolic resection has revealed a shorter hospital stay compared to open resection, while the morbidity rates were equal and conversion rates were acceptable [13].

The average interval between surgery and recurrence after appendicectomy for Crohn’s disease of the appendix is 4 years, after which some claim the recurrence rate to be almost nil. However, others have reported a recurrence rate of 10% over an 8-year follow-up and is much higher in patients with Crohn’s disease of the appendix associated with ileocaecal Crohn’s disease [1]. Most authors recommend periodic surveillance visits with radiological and endoscopic examination of the small intestinal and colon for a period of at least 3 years to promptly detect recurrence [10]. However, others claim that patients with Crohn’s disease should be followed for 10 years, and if disease free by then are considered cured [11].

**Conclusion**

Crohn’s disease of the appendix is usually associated with Crohn’s disease of the ileum and caecum. Isolated Crohn’s disease of the appendix is rare. Appendicectomy will suffice in those who present subacutely, are diagnosed preoperatively by clinical signs, and radiographic evidence shows that the disease is restricted to the appendix alone. However, in the presence of ileocaecal involvement, resection of the ileocaecal segment may be required to prevent postoperative complications of enterocutaneous fistula. While patients who develop enterocutaneous fistula post-appendicectomy in an unsuspected Crohn’s disease case have been treated with infliximab with some success, the majority of the patients required ileocaecal resection. Moreover, these patients still require long term surveillance to promptly detect recurrence.

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