

Title: Distal Ileum Adenocarcinoma Masquerading As Acute Appendicitis

Chan Wei Chin¹, Firdaus Hayati²

1. Department of Surgery, Queen Elizabeth, Ministry of Health Malaysia, Kota Kinabalu, Sabah, Malaysia
2. Department of Surgery, Faculty of Medicine and Health Sciences, University Malaysia Sabah, Kota Kinabalu, Sabah, Malaysia

Introduction

Small bowel cancer is a rare malignancy. It only comprises less than 5% of all gastrointestinal malignancies. The lifetime risk of developing small bowel cancer has been reported to be as low as 0.3%. Amongst the histology subtypes of small bowel cancer, in descending order, it comprises neuroendocrine tumour (40%), adenocarcinoma (40%), sarcoma, lymphoma, gastrointestinal stromal tumour and secondaries.

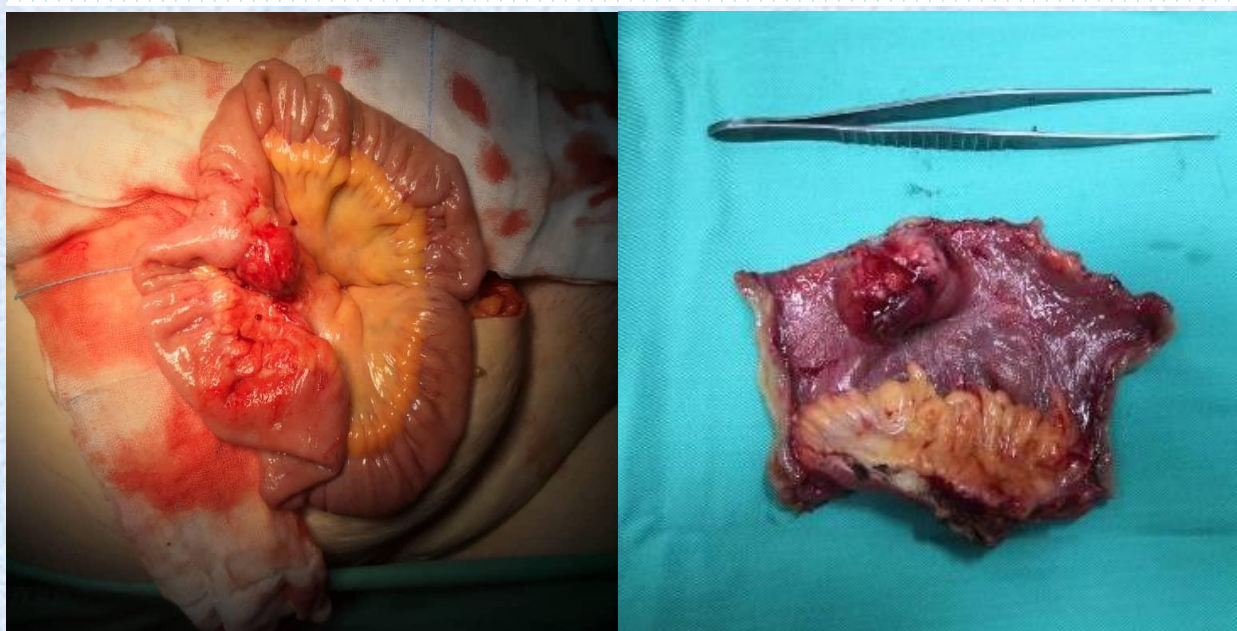
Here we present a case of a 50-year-old lady with signs and symptoms suggestive of acute appendicitis, with suspicion of small bowel adenocarcinoma being made intraoperatively and histopathological examination proven as small bowel adenocarcinoma.

Case Report

A 50-year-old lady presented to our emergency department with acute onset abdominal pain for 2 days over the right lower quadrant, pricking type, continuous, with a pain score of 8. She had fever and nausea for 1 day, after the onset of abdominal pain. Otherwise, there is no altered bowel habit, constitutional symptoms, history of TB contact, personal history of malignancy, or family history of malignancy. On examination, her abdomen was tender over the right iliac fossa; McBurney's and Rovsing's signs are positive, with a vague mass felt over the right iliac fossa. Blood investigations show leukocytosis and hypochromic microcytic anaemia.

Emergency diagnostic laparoscopy was performed due to persistent pain over the right iliac fossa. Intraoperative findings revealed an inflamed mass at the distal ileum, 40 to 50 cm from the ileocecal junction. Otherwise, the caecum, appendix, visualised colon, other parts of the small bowel, gallbladder, stomach and gynecologic organs are normal. It was converted into lower midline laparotomy due to limited working space. Noted a 3x4cm mass at distal ileum, 40 to 50 cm from ileocecal junction, able to separate from the mesentery. The mass arised from the antimesenteric border. The decision was made for segmental resection of the small bowel with 5cm proximal and 5cm distal margins, followed by primary end-to-end anastomosis. Histopathological examination proven it to be small bowel adenocarcinoma. The patient is 2 months post-operation and on oncology follow-up for adjuvant chemotherapy.

Figures



Intraoperative pictures demonstrating the small bowel adenocarcinoma

Discussion

The small bowel comprises up to 75% of the whole length of the gastrointestinal tract. Despite this, small bowel cancer is much rarer than other gastrointestinal tumours. Pathogenesis of small bowel adenocarcinoma is still in study due to the rarity of the disease. As with other gastrointestinal cancers, smoking and alcohol consumption are predisposing factors of small bowel adenocarcinoma. A study has also shown that Crohn's disease and celiac disease both have a significantly higher risk of developing small bowel adenocarcinoma.

The clinical presentation of small bowel adenocarcinoma is not specific. These include abdominal pain, weight loss, nausea, and vomiting. A few may present with emergencies such as bowel obstruction, perforation, and bleeding. Patients should be evaluated with blood and radiological investigations. Full blood count, and tumour markers including carcinoembryonic antigen (CEA), and carbohydrate antigen CA19-9 should be taken. Oesophagogastroduodenoscopy (OGDS) and endoscopic ultrasound (EUS) can be done in patients with suspected duodenal adenocarcinoma.

Treatment of the disease depends on its staging. Surgical treatment is indicated for localised and locoregional small bowel adenocarcinoma. Oncological resection with an adequate margin with en-bloc lymphadenectomy is recommended. Adjuvant medical therapy for small bowel adenocarcinoma, on the other hand, still needs to be developed due to its rarity and limited number available for study. MSI/MMR status is an essential piece of information to consider. There is an ongoing BALLAD trial evaluating the potential effect of adjuvant therapy with either 5-FU/leucovorin or 5-FU/leucovorin and oxaliplatin with observation in patients with local and locoregional small bowel adenocarcinoma.

On the other hand, management of metastatic small bowel adenocarcinoma differs. Patients with limited metastasis to visceral organs may be suitable for metastasectomy. Patients with resectable peritoneal carcinomatosis may benefit from cytoreductive surgery; however, this requires multidisciplinary effort with an experienced team at a well-established centre. Systemic therapy for metastatic small bowel adenocarcinoma is still under trial. FOLFOX or CAPEOX, FOLFOXIRI, and Bevacizumab are recommended first-line therapy. Pembrolizumab or Nivolumab as subsequent line therapy.

It is reported that small bowel adenocarcinomas have a poor prognosis, with 5-year overall survival rates of 30-79% for localised disease, and 3-19% for metastatic disease. [9]

Conclusion

We report this case to highlight the importance of considering small bowel adenocarcinoma as a cause of abdominal pain to avoid delay in diagnosis and for earlier initiation of treatment. In discordance between clinical and radiological findings, having a high suspicion index is important. With this, we aim to highlight the importance of high clinical suspicion during the management of patients during daily practice and to stimulate interest in further studies for systemic therapy and post-treatment surveillance of this rare disease.