

SMALL BOWEL CARCINOMA ASSOCIATED WITH CROHN'S DISEASE: CLINICAL REVIEW AND CASE REPORT

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ABSTRACT

Crohn's disease is a chronic relapsing and remitting inflammation of the bowel involving all its layers. A small bowel adenocarcinoma in Crohn's disease is a rare entity. The literature about this disease is reviewed. A case report of a 41-year male patient illustrates the significance of the diagnostic and therapeutic procedures.

Key words: Crohn's disease, small bowel adenocarcinoma, diagnosis, treatment, case report

Crohn's disease is a chronic relapsing and remitting inflammation of the bowel involving all its layers (13). A small bowel adenocarcinoma in Crohn's disease was first described by Ginsburg *et al.* in 1956 (6). Since then an increasing number of cases have been identified (7). It is a rare complication of Crohn's disease that typically involves the ileum. The data, however, are limited and based on case reports, retrospective studies, and reviews of the literature (4). The symptoms are often nonspecific and the diagnosis is usually delayed. As a result, infiltration and possible extension into adjacent tissues are common findings (4). The patients often complain of abdominal pain, weight loss, anorexia, bleeding and perforations. In general, localized adenocarcinomas of the small bowel are best managed with wide segmental surgical resection.

The role of adjuvant chemotherapy for small intestine adenocarcinomas remains undefined. Scanty data suggest that they are sensitive to 5-FU- and irinotecan-based chemotherapy (5,10,11). The adjuvant chemotherapy used is similar to that for colorectal cancer. The prognosis of Crohn's

adenocarcinoma of the small bowel remains disturbingly poor. Negative prognostic factors for survival include positive surgical margins for cancer, lymph node metastases, extramural venous spread and poor tumour differentiation (1). Regional node metastases are more common in jejunal or ileal adenocarcinomas (2,9).

Despite advances in imaging and therapeutic techniques, one- and two-year mortality rates range between 30% and 60%, depending on the stage of disease (8,12). According to the Bulgarian national cancer registry in 2009 (3), the number of new small intestine cancer cases is 44 (17 females and 27 males), without specifying the presence of Crohn's disease.

Table 1 shows cancer incidence rates per 100000 inhabitants in Bulgaria and abroad.

Table 1. Cancer incidence rates per 100000 inhabitants

Gender	Bulgaria	World standardized rate
males	0,7	0,4
females	0,4	0,2

The information about Crohn's disease associated with adenocarcinoma is poorly represented in the Bulgarian medical literature.

Table 2 demonstrates the clinical characteristics in 157 previously reported cases of small bowel carcinoma in Crohn's disease (4).

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Table 2. Clinical characteristics in 157 previously reported cases of small bowel carcinoma in Crohn's disease (4)

total number of patients	157
duration of Crohn's disease before diagnosis of cancer (range and mean)	0-48 (19,0)
age at cancer diagnosis (range and mean)	21- 86 (43,3)
Tumour localization	
jejunum	24
ileum	115
unavailable	18
stricture plasty	4
primary anastomosis site	3
other	
perforation at presentation	1

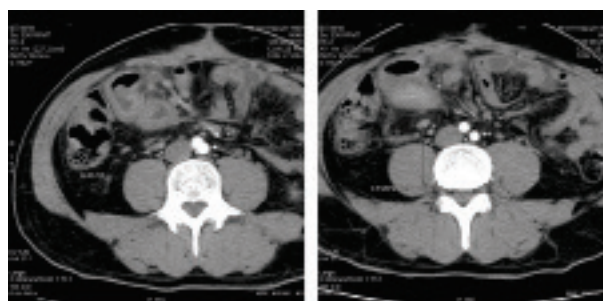
That's why patients with documented Crohn's disease in whom symptoms referable to the upper gastrointestinal tract develop should undergo diagnostic examination including barium and CT scanning as the results from endoscopy and biopsy can be vague or misleading.

Case report

This case is a proof that a small bowel carcinoma associated with Crohn' disease is rare. A 41-year-old male presented to the Department of Internal Medicine in November 2010 with a history of referred intermittent episodes of abdominal pain and diarrhea for the last few years. He was admitted with a working diagnosis of 'Crohn's disease' and corticosteroids were administered. During the last week the abdominal pain became more severe. Physical examination showed tenderness in the epigastrium and the right lower quadrant on the palpation with positive peritoneal irritation. Blood tests didn't show any signs of inflammation.

Abdominal x-rays showed air-fluid levels. Abdominal ultrasound performed for reassessment of the disease displayed a small bowel structure in the ileocecal region and dilated small bowel loops (Fig. 1).

Abdominal CT scan revealed 8-mm ileum wall thickness, fluid-filled loops in the small intestine, a small amount of visible air in those loops and enlarged mesenteric and retroperitoneal lymph nodes (Fig. 2).

*Fig. 1. Abdominal ultrasound images**Fig. 2. Abdominal CT images*

After acute aggravation of patient's status, he underwent urgent surgery.

During laparotomy, purulent exudate in the pelvis (300 mL) as well as extremely dilated small bowel loops with adhesion and high-grade inflammatory alteration were identified. Besides there were a 2-cm perforation of the ileum, retroperitoneal appendix with thick wall, multiple lymph nodes, subdiaphragmatic collection of purulent exudate (100 mL) and pancreatic edema with hemorrhagic collection.

The following operations were performed: laparotomy, debridement, resection of the terminal ileum (100 cm³), right hemicolectomy with continuity restored by side-to-side ileotransversal anastomosis, cholecystectomy, abdominal lavage and drainage.

The histological examinations revealed sings of Crohn's disease with fissuring and formation of fistulae, 'skip lesions, a ring-cell carcinoma infiltrating the muscularis propria and subserosa of the ileum, tumour emboli into lymph and venous vessels of the subserosa, and tumour emboli in the appendix. One out of 20 lymph nodes removed was positive for a metastasis of a poorly differentiated adenocarcinoma. The surgical margins were negative for cancer.

The patient recovered well from surgery and underwent 4 cycles of FOLFOX chemotherapy.

In September 2011, during the follow-up, the patient complained of abdominal distention, pain in the left lower quadrant and constipation. There was a physical evidence of *large* amounts of *ascitic* fluid. The abdominal ultrasound showed a conglomeration of bowel loops, dilated common bile duct at the level of papilla Vateri, bilateral compression of the ureters at the level of the pelvic floor and ascites. After the evacuation of the ascitic fluid, patient's general status became stable and he was discharged with normal bowel function. It was accepted to be a cytologically proven ascites of cancer origin.

In October 2011, the patient was hospitalized again in the Department of Surgery with severe abdominal pain and vomiting. The abdominal x-rays showed air-fluid levels in the small bowel. With symptoms and *signs of acute abdomen*, he underwent emergency surgery. The exploratory laparotomy identified a conglomeration of small bowel loops without clear demarcation line between them, thick descending and sigmoid colon, and carcinosis of the parietal peritoneum mainly expressed in the upper part of the abdomen. Enterostomy was performed for the most dilated small bowel loop for upper gastrointestinal tract decompression. There were no other possible options for its reconstruction. After surgery, the patient was administered parenteral nutrition, painkillers and additional symptomatic therapy. In spite of restored motility and functional enterostoma, he died one year after the first operation because of multiple organ failure.

It can be concluded that Crohn's disease associated with small bowel adenocarcinoma leads to progressive multiple organ failure due to bowel stricture, fistulae and perforation, on the one hand, and due to carcinoma intoxication, on the other hand.

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