



CASE REPORT

Asymptomatic ileal adenocarcinoma in the setting of undiagnosed Crohn's disease

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INTRODUCTION

The incidence of inflammatory bowel disease (IBD) is increasing with rates of around 6/100 000 for Crohn's disease with a marked rise in the age group between twenty to forty years^[1]. Small bowel carcinomas are uncommon representing less than 5% of all gastrointestinal malignancies^[2-7]. Prognosis is unfavorable with 1 and 2 year survival of 30%-60%, depending on the stage of the cancer^[8-12]. Crohn's disease has been associated with an elevated risk for the development of small bowel adenocarcinoma^[9,13-20] and chronic inflammation is implicated in this neoplastic progression^[21]. Although most adenocarcinomas arise in the duodenum, those associated with Crohn's disease generally occur in the ileum 20 years or more after the onset of Crohn's disease^[9,11,12]. In most cases, detection of small bowel carcinoma associated with Crohn's disease is at operation for other reasons or due to obstructive symptoms^[22-25].

Neither the risk factors nor screening for early diagnosis of small bowel adenocarcinoma in patients with Crohn's disease have been established. Here we report on an asymptomatic patient who on routine screening colonoscopy was diagnosed with adenocarcinoma of the terminal ileum which eventually led to the establishment of his Crohn's disease.

CASE REPORT

A 53-year old man presented for evaluation of a small bowel mass found on a screening colonoscopy. He denied any episodes of abdominal pain or diarrhea. He noted that his father had been diagnosed with Crohn's disease at the time of his death. He subsequently underwent a computed tomography (CT) examination

Abstract

A 53-year old previously healthy male underwent a screening colonoscopy for detection of a potential colorectal neoplasm. The terminal ileum was intubated and a mass was noted. Examination of the colon was normal. The biopsy of the ileal mass was consistent with an adenocarcinoma arising from the terminal ileum. His father who had never been previously ill from gastrointestinal disease died of natural causes, but was found to have Crohn's disease postmortem. The patient underwent exploratory laparotomy and a right hemicolectomy with a 30 cm section of terminal ileum in continuity. Findings were consistent with ileal adenocarcinoma in the setting of Crohn's disease. The patient made an uneventful recovery. The pathology was stage 1 adenocarcinoma. This is a unique case in that on a screening colonoscopy, a favorable ileal adenocarcinoma was discovered in the setting of asymptomatic, undiagnosed ileal Crohn's disease in a patient whose father had Crohn's disease diagnosed postmortem.

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Key words: Ileal adenocarcinoma; Crohn's disease; Colonoscopy



Figure 1 CT scan of the abdomen showing thickened and spiculated ileal wall.

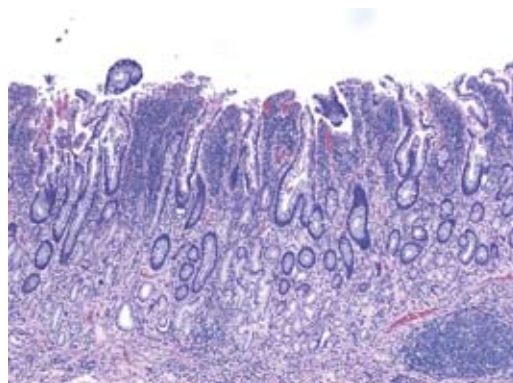


Figure 3 Crohn's changes in the terminal ileum (HE, x 400).

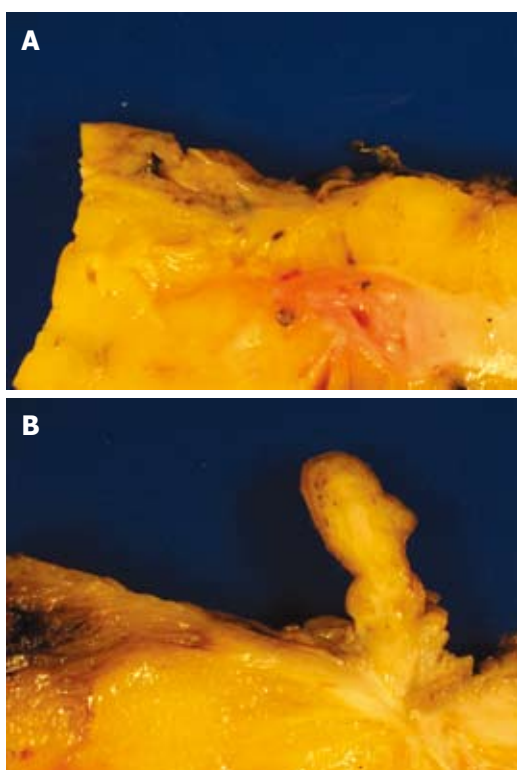


Figure 2 A: Stricture adjacent to the ileocecal valve; B: 3 cm polyp, sections of which revealed a white spiculated area extending into the underlying fat.

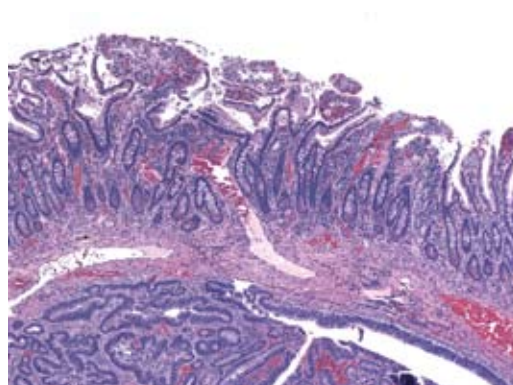


Figure 4 Adenocarcinoma invading the muscularis propria (HE, x 400).

of his abdomen and pelvis with both oral and intravenous contrast which revealed a thickened and spiculated wall of the terminal ileum with fistulization from the terminal ileum to the cecum (Figure 1). Given these findings, he was taken to the operating room for exploration.

On exploration, he was found to have a dense inflammatory mass in the right lower quadrant with multiple adhesions to the abdominal wall and bladder. Further exploration revealed a mass in the terminal ileum with dense adhesions involving the terminal ileum and cecum. Given these findings, a right hemicolectomy with a concomitant terminal ileal resection was performed. The surgical specimen consisted of a 20 cm segment of the cecum and a 53 cm segment of the terminal ileum with pericolic fat averaging 9 cm in diameter.

Macroscopic examination revealed Crohn's disease involving the distal most segment of the terminal ileum with a stricture adjacent to the ileocecal valve (Figure 2A). Adjacent to the stricture was a 3 cm polyp, sections of which revealed a white spiculated area that extended into the underlying fat (Figure 2B).

Histopathological examination in the area of the stricture revealed areas of chronic inflammatory change with marked architectural distortion and extensive pseudo-pyloric metaplasia in the mucosa. Extensive transmural lymphoid aggregates, muscular hypertrophy, neural hyperplasia, and submucosal fibrosis, compatible with Crohn's disease were also noted (Figure 3). In many areas, the mucosa was sitting directly in contact with the muscularis propria without any intervening submucosa. In the background of this inflammation, areas of low and high grade dysplasia were identified. Scattered glands were seen infiltrating into the markedly hypertrophic and disorganized muscularis mucosa. Isolated tumor glands were also seen in the submucosa and as discussed above, in some areas, there were hardly any intervening submucosa with the tumor glands infiltrating the muscularis propria (Figure 4). The size of the tumor was difficult to estimate, as grossly no obvious mass was identified. However, from microscopic sections, it was estimated that the intramucosal carcinoma extended over a 2.0 cm area. The staging of this tumor was also difficult due to complex architecture and histological changes in the bowel wall. However, due to the presence

of occasional tumor glands in the muscularis propria, it was staged as pT2. Twenty-nine lymph nodes were harvested and all were negative for carcinoma. The patient recovered well from surgery.

DISCUSSION

A thorough search of the worldwide literature has revealed that the case presented here is the first ever of a patient with ileal adenocarcinoma as the first manifestation of Crohn's disease in an otherwise asymptomatic patient. There are, however, other reports of ileal adenocarcinoma as the first manifestation of Crohn's disease in patients with vague abdominal complaints^[26,27].

In 1956, Ginzburg *et al* first described carcinoma as a rare complication of small bowel Crohn's disease^[28]. To date, several cases of small bowel carcinoma in Crohn's disease have been reported^[29]. The most common small bowel carcinoma is adenocarcinoma, and risk factors include long-standing disease, surgically bypassed loops, male sex, onset of disease before the age of 30 years, and associated chronic active disease with strictures and fistulas^[11,30-32].

Munkholm *et al* reported an incidence of 0.54% of small bowel cancer in Crohn's disease compared to an expected rate of 0.04% ($P = 0.0001$)^[30]. In a later study, the same group reported a more than 60-fold increased risk of small bowel adenocarcinoma independent of age and sex in patients with Crohn's disease^[33]. The lifetime prevalence of small bowel adenocarcinoma in patients with Crohn's disease is 1%-3%^[34]. The combination of genetic susceptibility, mechanical irritation and surgery has been suggested as possible etiologies for this elevated risk in Crohn's disease. Small bowel carcinomas are also mostly localized to strictures^[22,35,36]. Immunosuppression has also been implicated^[32].

These occult carcinomas pose a challenge to conventional diagnostic investigations such as upper or lower gastrointestinal endoscopy and small bowel series. CT has now emerged as the imaging modality of choice^[37-40]. Magnetic resonance imaging (MRI)^[41], double-contrast enteroclysis^[42], and video wireless capsule endoscopy^[43,44] have also been promising.

Survival rates for small bowel malignancies are much worse than for large bowel cancers with a mean survival of 6 mo as compared to 65 mo for the latter^[9]. Mortality rates range from 30%-60% depending on the stage of the carcinoma^[8-12]. Poor prognostic factors include positive resection margins, extramural venous spread, lymph node metastases, poor tumor differentiation, depth of tumor, and a history of Crohn's disease^[45].

Only small studies have evaluated adjuvant therapy for small bowel adenocarcinoma^[46,47]. These studies are not in patients with Crohn's disease and randomized controlled trials are definitely needed.

We report a patient with ileal adenocarcinoma as the first manifestation of Crohn's disease. The patient had no symptoms of Crohn's disease and had only a family history of Crohn's disease. This case report, as well as others discussed above, highlight the need for a higher

index of suspicion in screening for Crohn's disease and initiating a regular follow-up with ileocolonoscopy.

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