Small Bowel Mucinous Adenocarcinoma Complicating Crohn’s Disease: A Case Report

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ABSTRACT

The incidence of Crohn’s disease (CD) is increasing over the last three decades. But due to high prevalence of tuberculosis, there is still a delay in diagnosing CD. As a result, CD usually presents with complications like strictures, internal fistulas or intra-abdominal abscess. Small bowel adenocarcinoma (SBA) is the rarest among the many complications of CD. All over the world, less than 200 cases of SBA associated with CD have been reported. Both diseases presenting simultaneously has never been reported. We present a case of mucinous adenocarcinoma of terminal ileum complicating CD, which presented simultaneously, and was managed successfully by laparoscopic right hemicolectomy with adjuvant chemotherapy.

Key words: Ileal adenocarcinoma, Crohn’s disease, laparoscopic right hemicolecotomy.

INTRODUCTION

Crohn’s disease (CD) is a recognized risk factor for small bowel adenocarcinoma (SBA). Many meta-analytic studies have shown 28.4 times increased risk for SBA in patients with CD when compared to general population{1}. The increasing availability of imaging studies, colonoscopy-ileoscopy, small bowel enteroscopy, serological markers like anti-Saccharomyces cerevisiae antibody (ASCA) for CD and molecular diagnostic techniques to differentiate CD from tuberculosis in tissue, have added significantly to the ability of clinicians to diagnose CD and SBA. CD usually presents with subacute intestinal obstruction, diarrhea, weight loss and obscure gastrointestinal bleeding. Most common complications associated with CD are stricture and fistula formation. Other complications seen are abscess and perforation{2}. We report a case of CD with SBA which were diagnosed simultaneously, and review the published literature.

Case history

A 36-year-old woman presented to hospital with recurrent episodes of colicky abdominal pain and occasional vomiting for six months. She was able to tolerate liquid diet only. She lost 4 kilograms of body...
weight over two months. There was no history of fever, diarrhea or bleeding per rectum. She never had similar complaints before this. Her family history was insignificant.

Her abdominal examination revealed mild right iliac fossa tenderness without any palpable lump. Routine blood investigations and sonogram of abdomen were normal. CT scan with contrast showed diffuse enhancement with dilatation of small bowel with a stricture segment in the distal ileum (Figure 1). A provisional diagnosis of CD with stricture was made. Antegrade and retrograde enteroscopy showed a partially obstructing polyoidal mass lesion with ulceration in the terminal ileum. The pathology revealed tubulovillous adenoma with focal high grade dysplasia. IgA antibody to Saccharomyces cerevisiae (ASCA IgA) was negative. Anti-neutrophil cytoplasmic antibodies (c-ANCA and p-ANCA) were both negative. C-reactive protein (CRP) level was normal.

Diagnostic laparoscopy revealed a terminal ileal growth with multiple enlarged mesenteric lymph nodes causing intestinal obstruction. An ileo-ileal fistula was seen between loops of terminal ileum at the site of growth. Laparoscopic right hemicolectomy with ileotransverse anastomosis was done (Figure 2). Biopsy of the specimen revealed mucinous adenocarcinoma of the terminal ileum in a background of multiple sessile tubulovillous adenomas with high grade dysplasia (Figure 3). Both resection margins and all the removed regional lymph nodes were negative for cancer. Non-neoplastic small bowel mucosa showed features suggestive of Crohn’s disease. Post operative period was uneventful. She was started on adjuvant chemotherapy.

DISCUSSION

Three decades ago, CD was a rare disease and the diagnosis was often confused with tuberculosis due to the latter’s high prevalence. Many population-based studies have demonstrated that the incidence and prevalence of CD all over the world have increased since the mid-1970s. The disease seems to be most common in northern Europe and North America, but is probably increasing also in Asia and Africa(3).

SBA complicating CD is rare. The first report of adenocarcinoma of small intestine complicating CD was by Ginzburg et al in 1956(4). Fever than 200 cases of SBA complicating CD have been reported to date all over the world(4).

The pathogenesis of SBA arising in CD is chronic inflammation (inflammation-dysplasia-cancer sequence)(5). Extended duration of CD, distal jejunal and ileal location, male sex, small bowel bypass loops, stric-
Figure 3. H&E photomicrographs of (a) mucinous adenocarcinoma of the terminal ileum, at 10x magnification and (b) ileal mucosa showing architectural distortion of crypts, typical of Crohn’s disease, at 60x magnification.

...chronic fistulous disease and younger age at diagnosis are likely risk factors for developing CD associated SBA\(^1\). The mean duration of CD prior to diagnosis of SBA is 19 years (range 0-48 years)\(^1\). In our case, both CD and SBA were diagnosed simultaneously, with the patient having non-specific symptoms for six months.

The highest incidence of CD associated SBA is in the distal jejunum and ileum. The diagnosis is usually delayed due to non-specific symptoms. As a result, infiltration with extension into adjacent tissues and bowel obstruction are common at the time of diagnosis. Resection of the tumor with adequate margins is the primary modality of treatment. Prognosis of SBA is poor, with the mortality at 1 and 2 years ranging from 30-60% depending on the stage of the cancer\(^6\). Other bad prognostic factors are positive surgical margins, poor differentiation, increasing depth of tumor invasion, positive lymph nodes and extramural venous spread\(^7\).

**CONCLUSION**

Although SBA is rare in CD, it is still important for clinicians to be aware of this disease. It can present insidiously as well, at times even before the diagnosis of CD is made, as in our case. Stricture biopsies should be taken whenever strictureplasty is performed to exclude malignancy.

**REFERENCES**