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Case Report

Free Perforation of the Colon: Emergent Presentation of an Advanced T-Cell Intestinal Lymphoma

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Abstract

A 35-yr-old female with presented emergently with acute and severe abdominal pain and vomiting for only a few hours duration. Investigations led to recognition of a perforated viscous due to a colon perforation. Extensive intra-abdominal involvement of the small intestine, colon and mesenteric lymph nodes was present at exploratory laparotomy with a peripheral T-cell lymphoma. Rapid deterioration led to her death. This case represents a very dramatic presentation of an ominous disease that is usually indolent in its development and presentation, sometimes associated with and complicating celiac disease and/or dermatitis herpetiformis.

Keywords: Sprue-like Intestinal Disease; Celiac Disease; Malignant Lymphoma; Colonic Perforation

Prior studies have shown that different sites in the gastrointestinal tract may be the site of malignant lymphoma [1-3]. In addition, different forms of lymphoma may complicate underlying celiac disease, an immune-mediated small intestinal mucosal disease related to gluten ingestion [4]. Finally, lymphoma in celiac disease may occur in different extra-intestinal sites, including the liver and spleen, even without intestinal involvement [5-7].

Lymphoma often presents as a complication of established celiac disease, with an obstructing intestinal lesion or with ulceration. Rarely, feeding a high gluten diet in lymphoma may precipitate underlying celiac disease [8,9]. A more dramatic acute abdominal presentation of lymphoma may has also be reported with perforation leading to peritonitis [10]. Likely, most perforations result from a pre-existent malignant intestinal ulcer. Our previously published experience emphasized that the small intestine was the usual site of intestinal perforation complicating an underlying lymphoma, most often jejunal in site and frequently associated with unrecognized celiac disease [10]. The present case was unusual and deserved emphasis owing to its site of perforation in the colon.

Case Report

A 35-yr old otherwise well female presented to our hospital emergency centre with an 8 to 10 hour history of continued abdominal pain and vomiting. There were no other prior symptoms with this event or family history of gastrointestinal disease. At age 25, she had a pruritic skin rash on the extensor surfaces of her extremities thought to be possible dermatitis herpetiformis. This resolved spontaneously, however, without treatment and did

not recur. No further investigations were done at that time. No pharmacologic or dietary treatment for her skin rash was provided.

Physical examination in the emergency centre demonstrated generalized peritonitis, pyrexia of 40C and tachycardia. Blood studies revealed a white cell count of 21,000 and plain abdominal radiographs showed free air below the diaphragm. A diagnosis of a perforated viscus was made and she was transferred immediately to the operating room. Within the abdominal cavity, there were copious amounts of foul murky fluid. A small perforation in the proximal transverse colon near the hepatic flexure was located. In addition, an extended segment (38 cm) of abnormally thickened jejunum was resected and treated with a side-to-side anastomosis. A section of mesentery along with enlarged lymph nodes were also removed. A right hemi-colectomy with an ileostomy was completed, followed by saline wash-outs and insertion of drains in right and left para-colic gutters. She was transferred to intensive care, but her clinical course was complicated by progressively deteriorating renal function and respiratory failure.

Pathological exam revealed a non-Hodgkin's type of lymphoma with involvement of the lymph nodes in the greater omentum and resected mesentery. Both small and large bowel showed ulceration with focal lympho-epithelial lesions along with flattened sprue-like small intestinal mucosal changes and focal areas of heavy lymphocytic infiltrates extending through the muscular propria into the serosa. Areas of increased mitoses and a starry sky appearance was also noted. Immunophenotyping studies of her small and large bowel indicated an extensive peripheral T-cell lymphoma.

Discussion

This patient presented emergently with a perforated colonic ulcer due to extensive intestinal and lymph node involvement with a peripheral T-cell lymphoma. Most striking was the lack of reported symptoms antedating her brief illness in spite of extensive abdominal involvement with this malignancy. Previous reports, particularly in established celiac disease, have emphasized the development of small intestinal lymphoma, even in those without overt initial pathological features of classical celiac disease, i.e., so-called latent celiac disease [8,9]. This report further emphasizes that free perforation may occur with lymphoma involvement of the colon, not just in small intestinal disease, particularly in patients with celiac disease and/or dermatitis herpetiformis.

A very similar rare acute presentation of gastric perforation due to a T-cell lymphoma was also recently described [11] emphasizing that free perforation may occur anywhere within the gastrointestinal tract, including the stomach in this setting. It is of particular interest that in this latter gastric perforation case, celiac disease with poor diet adherence was noted [11]. In our patient, despite some apparent histopathologic features of underlying sprue-like intestinal enteropathy [12], celiac disease could only be historically suspected because of the prior history of a closely associated skin disorder, dermatitis herpetiformis [13,14]. Formal studies of her small intestine after treatment with a gluten-free diet were obviously not possible owing to her acute presentation and death.

Celiac disease may present with colonic lymphoma [15,16] and prior clinical reports have described an indolent clinical course [17]. Perforation of the cecum or sigmoid colon [18] have been described with emphasis on a high mortality rate and need for early diagnosis [18]. B-cell type lymphoma has also been described in some series [19]. Synchronous perforation of lymphoma in the small bowel and colon without celiac disease have also been noted [20]. Interestingly, in a survey of 68 intestinal T-cell lymphoma cases from China, misdiagnosis of Crohn's disease, intestinal tuberculosis and cancer were reported [21]. In this series, more than half of all cases also involved the colon [21]. A case from Korea [22] emphasized another dramatically rapid clinical course with a fatal outcome following colonic perforation in an enteropathy-associated-T-cell lymphoma. In contrast, another study of 6 immunocompetent Chinese men [23] described a prolonged and indolent history of abdominal pain and diarrhea for 4 to 13 years. In these patients, longitudinal immunopheno-typing suggested that disease progression may be linked to a gain of novel gene mutations during the clinical course [23].

Presentation with a perforated viscous represents one end of the clinical spectrum of lymphoma, often in an otherwise clinically indolent and widespread intestinal disease. This presentation is not only dramatic and may actually lead to an initial lymphoma diagnosis but represents in an immediate and ominous prognosis because of complicating septic peritonitis.

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