Duodenal adenocarcinoma might be the cause of intractable nausea and vomiting in patient with coeliac disease

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Abstract

Coeliac disease (CD) is an autoimmune disorder which leads to chronic inflammation of the gut. Untreated CD is associated with upper gastrointestinal malignancies. Small-bowel lymphoma and adenocarcinoma are recognized complications of untreated coeliac disease (CD). We report the case of a 43-year-old male suffering from CD who was treated with a gluten-free diet one year, presenting with complaints of intractable nausea and vomiting. After several studies, he underwent push enteroscopy, which identified one large mass lesion in the third part of duodenum. However, histopathological examination showed adenocarcinoma. Subsequently, a duodenal segment resection was performed. After surgery, the patient recovered well and left our hospital in good condition. Clinicians should take into account that adenocarcinoma is rare but associated with CD particularly in CD patients with worrying symptoms such as nausea and vomiting unresponsiveness to treatment and these patients should be screened for long term complications like malignancy.

Keywords: Coeliac disease, Adenocarcinoma, Malignancy.

Introduction

Coeliac disease (CD) is a unique autoimmune disorder which results from the interaction between gluten and immune, genetic and environmental factors (1). Clinical manifestations of CD differ greatly among patients, varying from vague symptoms such as fatigue or malaise to a classical malabsorption syndrome including diarrhoea and steatorrhoea accompanied by abdominal pain or discomfort (2). The small intestine constitutes 90% of the mucosal surface area of the gastrointestinal tract, but is the site of <5% of all gastrointestinal malignancies (3).

Malignant tumors of the small intestine are rare all over the world (4), with a global incidence of less than 1 per 100 000 population (5). Cancers of the small intestine or small bowel cancer (SBC) account for only 0.42% of total cancer cases and 2.3% of cancers of digestive system in the United States (6); while in Canada, 0.37% and 1.78% respectively (7). Mortality of the cancer is even lower, accounting for only 0.2% of the total cancer deaths in the United States and in Canada (6,7).

Several studies have suggested increased mortality from gastrointestinal malignancies in...
patients with CD compared to the general population (8-10). We report a case of duodenal carcinoma in a patient suffering for CD.

Case report

A 43-year-old man with intractable nausea and vomiting 2-3 hours after eating and anemia had been worked up. In primary evaluation laboratory data showed: Hemoglobin=11.2g/dl, MCV=72.8fl, Serum Iron=23mcg/dl, Ferritin=10.3ng/ml, Stool exam: Occult blood (-), Anti-Tissue-Transglutaminase IgA=26.5(+), Esophagogastroduodenoscopy (EGD): Mild antral gastritis, Mild atrophic folds + scalloping in the second part of duodenum (D2) (Figure 1).

Figure 1. Mild atrophic folds and scalloping in D2

Biopsy of D2 revealed mild to moderate villous atrophy with increased numbers of intraepithelial lymphocytes (up to 40-50) accompanied by changes consistent with CD Marsh type 3a and colonoscopy survey showed Grade1 hemorrhoids.

In his initial referred to clinic in other center the physicians suggested that coeliac disease causes to these symptoms so they prescribed gluten free diet. After that nausea and vomiting subsided initially but recurred again. In order to study central nervous system etiology of vomiting, brain MRI was done and it was normal. Barium transit showed loop separation and segmentation with possibility of edema and mesenteric thickening (Figure 2).

Figure 2. Barium transit showing loop separation and segmentation with possibility of edema and mesenteric thickening

In abdominopelvic CT scan, we did not have any evidence of organ disorder, lymphadenopathy, ascites and other significant pathologic change (Figure 3).

Figure 3. Abdominopelvic CT scan with IV and oral contrast did not show any evidence of organ disorder or significant pathologic change
Due to several admissions because of intractable nausea and vomiting, EGD with duodenal biopsy was done again and pathologic examination revealed few lymphoid aggregates in antrum and no evidence of CD in D2.

We decided to perform push enteroscopy to find etiology of recurrent nausea and vomiting, so we detected one large ulcerated mass with partially obstruction in D3 (Figure 4).

Pathologic examination revealed tumoral tissue composed of irregular villi and gland-like structures covered by highly atypical cells having large pleomorphic nuclei (Adenocarcinoma) (Figure 5). The patient was undergone Surgery (Duodenal segmental resection and duodenojejunal side to side anastomosis) successfully.

Discussion

Approximately 30%-40% of the cancers observed in the small intestine are adenocarcinomas, a percentage much lower than the proportion in the colon where the overwhelming majority is adenocarcinomas. Most of the tumors located in the duodenum and the duodenal-jejunal junction are adenocarcinomas (11,12).

Small-bowel lymphoma and adenocarcinoma are recognized complications of coeliac disease (13,14). The association between carcinoma of the small bowel and CD was first reported in 1958 (15). Since then, the association between CD and small bowel carcinoma is persistent although only based on a small number of patients of which the greater part is in the form of case reports.

Next to the rarity of these lesions, these patients are presenting usually with vague and poorly defined signs and symptoms, often delaying a correct diagnosis. Furthermore, conventional radiographic studies of the upper and lower intestinal tract often appear normal (16).

Despite the fact that a lifelong gluten free diet (GFD) leads to noticeable clinical and histological improvement in about 70% of patients (17), studies about the protective effect of GFD on malignancy are scarce. Most studies suggest that a GFD leads to a decreased incidence of lymphoma, cancer of the mouth, pharynx and esophagus and a decreased mortality in CD (18–21).

The present patient is a reality that CD is a serious disease with serious complications such as duodenal adenocarcinoma. Despite the association between CD and small bowel malignancy, currently
there are no recommendations for screening for this type of malignancy. Our patient was suffering from recurrent nausea and vomiting and several different treatments, eg, gluten free diet did not improve them significantly so we performed push enteroscopy for evaluation of the initial parts of small intestine. We recommend that clinicians should take into small bowel adenocarcinoma is rare but associated with CD particularly in CD patients with worrying symptoms such as nausea and vomiting unresponsiveness to treatment especially in combination with weight loss. In these patients evaluation of small intestine with push enteroscopy or single or double balloon enteroscopy with biopsy should be performed.

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