Computed Tomographic presentation of obstructive jejunal adenocarcinoma associated with celiac disease and incomplete intestinal malrotation

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A B S T R A C T

INTRODUCTION: Small bowel adenocarcinoma is a rare entity most frequently observed with celiac disease. This is the first case report on the association of celiac disease, small bowel adenocarcinoma and intestinal malrotation.

CASE REPORT: A 40-year-old male patient diagnosed with celiac disease since the age of 5 years complained of epigastric pain and vomiting for three days. Computed tomography (CT) showed a significant gastroduodenal dilatation with thickened intestinal wall proximal to the duodenojejunal flexure. The lumen contained a food bezoar in the center. The duodenojejunal angle was abnormally on the right side of the abdomen and the superior mesenteric vein was anterior to the superior mesenteric artery. Endoscopy after aspiration found a hemi-circumferential and irregular mass which bled at the contact of fibroscope. Biopsies showed an adenocarcinoma and small bowel resection was performed.

DISCUSSION: Celiac disease is associated with a high risk of small bowel cancer. The association of incomplete intestinal malrotation, duodenojejunal flexure tumor and celiac disease made the surgery challenging.

CONCLUSION: Patients with celiac disease should be carefully monitored and endoscopic or radiologic investigations should be carried out in patients with any doubtful symptoms.

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1. Introduction

Small bowel adenocarcinoma is a rare but it is most commonly seen in patients with celiac disease. The jejunum is less frequently involved than the duodenum. Celiac disease is considered as a risk factor for small bowel adenocarcinoma and lymphoma [1,2]. Many authors have reported the association of celiac disease with small bowel adenocarcinoma but this is the first time these two conditions are associated with incomplete intestinal malrotation.

2. Case presentation

A 40-year-old man had a history of celiac disease since the age of 5 years. The diagnosis was established after a positive serological test of IgA-tTG antibodies and jejunal biopsy which showed marsh II lesions. He had a regular follow up with good adherence to gluten-free diet. Six months prior to this acute episode, he developed cerebral vein thrombosis and was treated with antithrombotics.

Three days before this hospital admission, he experienced epigastric pain, vomiting and an absolute constipation. The abdomen was not severely distended on examination. The biochemistry results were all normal. There was no pancreatic enzymes elevation. The infectious indicators, Prothrombin Ratio and the International Normalized Ratio were normal. Computed Tomography with oral and intravenous contrast was performed. The stomach and duodenum were shown to be dilated. The level of the obstruction was at the duodenojejunal junction. There was a 6 cm long irregular wall thickening of the jejunum which was enhanced with iodinsed contrast injection. The bowel lumen was completely obstructed by a bezoar (Fig. 1). The Treitz angle was at the right side of the abdomen and the superior mesenteric vein was anterior to the superior mesenteric artery (Fig. 2). The caecum was not in the right iliac fossa but at a higher level in the right abdomen. These radiological features were consistent with an incomplete intestinal malrotation.
malrotation. There were no other bowel anomalies. After nasogastric aspiration, an upper gastrointestinal endoscopy was performed which confirmed the CT findings. Histological study of the biopsied specimens showed an infiltrative cancerous proliferation.

Operation was performed by a surgeon who was informed about the patient’s anatomical condition. During surgery, there was a tumor of the Treitz (Fig. 3). Resection of the small bowel carrying tumor was done with an end-to-end anastomosis. Five mesenteric lymph nodes were removed.

The final histopathological diagnosis showed a well differentiated adenocarcinoma of the jejunum with 30% of mucinous component (Fig. 4). The operation was R0 resection and all the 5 lymph nodes were negative for malignancy.

No chemotherapy was administered following the bowel cancer resection. The postoperative course was uneventful. The patient was well on follow-up at 6 months after surgery.

3. Discussion

Only a few studies have reported on upper intestinal obstruction caused by a jejunal adenocarcinoma [3]. To the best of our knowledge, this is the first report on a small bowel adenocarcinoma (SBA) complicating celiac disease and associated with incomplete intestinal malrotation.

SBAs are rare although the incidence is increasing. They represent only 5% of digestive tumors [1]. The predominant location of adenocarcinoma is in the duodenum and proximal jejunum, with
decreasing incidence distally. Celiac disease is an immune-based reaction to dietary gluten that induces epithelial changes. It has a high potential risk to develop adenocarcinoma as well as lymphoma of small bowel [4]. The clinical presentation and diagnosis of SBA are usually delayed. Bowel obstruction is most often seen with jejunal and ileal locations.

Malrotation of midgut is defined as an anomaly of rotation and fixation of the midgut. Stinger classified several types of malrotation according to the embryologic state of development [5]. Intestinal malrotation with complications is often seen in infants and children. Most adult cases are discovered incidentally [6].

Multi detector Computed Tomography Enteroclysis (MDCTE) should be performed to rule out carcinoma in patients with celiac disease who have symptoms despite adherence to a strict gluten-free diet [7]. Typical feature of SBA on MDCTE is a concentric lumen narrowing with irregular edges, but the tumor can also appear as polypoid lesions with well-defined surface and margins [8]. Extraluminal infiltration may appear as fat stranding on CT. MRI, when available offers an excellent soft tissue contrast with T2 weighted fast spin-echo sequences. True fast imaging with steady-state precession and gadolinium enhanced sequences help to depict intraluminal tumors of the small bowel. Lymphoma is also more common with celiac disease but it usually appears as a coarse segmental wall thickening with ulceration and necrosis and these features are associated with lymphoidenopathies. A video capsule endoscopy is a good alternative in patients with established celiac disease with alarming symptoms [9].

With the expanse of cross sectional imaging, adult intestinal malrotation is increasingly diagnosed. In our case, radiological features of duodenojejunal angle on the right side of the spine, and the ileocecal junction below the liver are consistent with a Stage II of the midgut rotation with 180° counterclockwise rotation.

Complete resection (R0) of the primary tumor with mesenteric loco-regional lymph nodes resection is mandatory. This resection is decided on by the location and histology of the tumor [10]. In the context of invasion, neoadjuvant therapy should be considered, and resection reconsidered after 2–3 months of chemotherapy. No standard adjuvant regimen has been defined in the literature due to the lack of randomised controlled trials [1]. Disease burden in lymph nodes and microscopic vascular invasion on histopathology indicate poor prognosis [10].

The risk of volvulus or internal hernia is extremely low in adult patients who do not have complete malrotation. In our case, the surgeon chose not to perform prophylactic Ladd’s procedure because of the low risk of midgut volvulus.

Conflicts of interest

All authors declare there is no conflict of interest.

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Ethical approval

This case report does not require any ethical approval. All the figures are anonymized.

Consent

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Authors contribution

We certify that we have participated sufficiently in the intellectual content, conception and design of this work or the analysis and interpretation of the data, as well as the writing of the manuscript, to take public responsibility for it and have agreed to have our name listed as a contributor.

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