Introduction

Celiac disease (CD) is an autoimmune disorder, resulting from the interaction between gluten, immuno-
Laparoscopic treatment of mucinous adenocarcinoma of jejunum associated with celiac disease. Case report

Case report

A 49-year-old male was admitted to Authors’ Department for recurrent melena. At the presentation the patient showed severe anemia with a hemoglobin level of 7.00 g/dl. The clinical history revealed a two years complaints consisting in nausea and recurrent vomiting. An upper endoscopy and a colonoscopy, both performed before hospital admission, were negative. Capsule enteroscopy, accomplished at another hospital two months earlier, was inconclusive.

After admission, a small bowel CT scan reported a circumferential involvement with stenosis and occlusion at the level of the first jejunal loop where the enteroscopic capsule was entrapped (Fig. 1). A laparoscopy was scheduled for diagnostic purpose and, in case, for treatment of the jejunal stenosis.

With the patient placed in supine position an Hasson trocar was inserted in the umbilical area and other two 5 mm trocars were laterally inserted in the midclavicular lines. Laparoscopic exploration of the abdominal cavity showed a mass localized in the jejunum at the level of the Treitz’s ligament. Resection of the first jejunal loop was thereby accomplished and a latero-lateral duodeno-jejunal anastomosis was performed by means of stapling device.

The histological study of the surgical specimen revealed a poorly differentiated jejunal mucinous adenocarcinoma with large mucin lakes that contained malignant epithelium (Fig. 2) and metastatic mesenteric lymph nodes (pT N M G3). In association with the tumor the pathologist also described an altered small intestinal architecture with partial villous atrophy, hyperplasia of glandular crypts and significant increased number of intraepithelial lymphocytes (Fig. 3). The histological pattern was compatible with the diagnosis of celiac disease which was confirmed by serological immunological tests.

The postoperative phase was unremarkable. Gluten-free diet was recommended and chemotherapy was planned.

Discussion

Despite the small intestine represents the 90% of the surface area and the 75% of the length of the entire gastrointestinal tract, malignancies represent merely 2% of the cancers of the whole digestive system and they are very uncommon in the general population (1,7).

An association between small bowel’s tumors and celiac disease has been reported for the first time in 1958 (8). More recent studies pointed out that celiac disease highly increases the risk of small intestinal malignancies (2-3). According to different statistics small intestinal cancer is up to 82 times more common in patients suffering from celiac disease than in general population (2-3).

Immunologic disorders with increased permeability to oncogenic factors and malabsorption of protective vitamins A and E may be responsible of the increased risk of cancer in CD patients (1,9), but no conclusive studies have been published on this matter. Gluten-free diet seems to decrease the incidence of malignancies in CD (10) but its preventive role in small bowel cancer is controversial.
The diagnosis of jejuno-ileal tumors is always difficult. Clinical history is usually nonspecific and consequently surgery is often delayed (1-11). Moreover, instrumental examinations often fail in adding useful information to the diagnosis (11), and only surgery, with the removal of the tumor, often allows the definitive diagnosis.

Signs and symptoms are usually vague and poorly defined (11). Despite recent advances in radiology and endoscopy, diagnostic evaluation of small bowel is still challenging for a definitive diagnosis. CT scan and/or video capsule enteroscopy may reveal a disease in the small bowel, but, like in our case, they are not conclusive about the etiology. This can result in a delayed diagnosis (11).

In Authors’ opinion a small bowel tumor has always to be suspected when long lasting gastrointestinal nonspecific symptoms are referred by patients, especially if an underlying CD is present. A laparoscopic exploration, in case of doubtful or suspicious radiological or endoscopic imaging, should be considered essential for the diagnosis and the treatment.

Even if most patients with malignancies of the small bowel present with a long standing history of malabsorption, in certain cases a jejunal adenocarcinoma can be the first presentation of an underlying CD (9). In our case, gluten-sensitive enteropathy was diagnosed after the surgical specimen was histologically analyzed.

Mucinous adenocarcinoma is an extremely rare tumor of small bowel and only three cases have been formerly described in the literature (4-6). Our report of a mucinous adenocarcinoma of the small bowel in a patient with an underlying CD is unique, since in our knowledge no previous reports have been published.

Conclusions

Our case may confirm the association of CD with small bowel cancer which can be mucinous in type, with a poor prognosis. The association with CD suggests an oncological screening in patients with malabsorption syndrome. Anyway, long standing symptoms of malabsorption with suspected alteration of the transit or occult bleeding should induce to a work-up in order to exclude malignancy. Mucinous type intestinal adenocarcinoma, even if never published before, could be observed. Laparoscopic surgery is essential in the management of small intestinal tumors associated with CD.

Disclosure statement

No competing financial interests exist.

References