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Laparoscopic treatment of mucinous adenocarcinoma of jejunum associated with celiac disease. Case report

R. VECCHIO[1], S. MARCHESE[1], P. GANGEMI[2], G. ALONGI[1], F. FERLA[1], C. SPATARO[1], E. INTAGLIATA[1]

SUMMARY: **Laparoscopic treatment of mucinous adenocarcinoma of jejunum associated with celiac disease. Case report.**

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Mucinous adenocarcinoma of the small bowel is very rare, and only few cases have been described in the literature. Association of this tumor with celiac disease has never been published. The authors report a unique case of jejunal mucinous adenocarcinoma in which a concomitant celiac disease has been histologically recognized. The difficult diagnosis, the role of laparoscopic surgery and the relationship between small bowel tumors and celiac disease are discussed.

A 49-year-old man presented with recurrent melena, nausea, vomiting and anemia. A stenosis of the jejunum was documented by means of CT scan and video capsule enteroscopy. A laparoscopy was scheduled. A tumor, found in the first jejunal loop, was removed by laparoscopic surgery. Histopathology revealed a rare mucinous adenocarcinoma associated with epithelial changes secondary to celiac disease.

Although small bowel tumors are rare entity, in patients with celiac disease complaining of symptoms related to altered intestinal transit or occult bleeding, an appropriate work-up should be planned for diagnosis. Mucinous type intestinal adenocarcinoma, even if never published before, could be observed. Laparoscopic surgery is often essential for the diagnosis and treatment.

RIASSUNTO: **Trattamento laparoscopico di adenocarcinoma del digiuno associato a malattia celiaca. Case report.**

R. VECCHIO, S. MARCHESE, P. GANGEMI, G. ALONGI, F. FERLA, C. SPATARO, E. INTAGLIATA

L'adenocarcinoma mucinoso dell'intestino tenue è molto raro e soltanto pochi casi sono stati descritti in letteratura. L'associazione di questo tumore con la malattia celiaca non è mai stata descritta. Gli autori riportano un singolare caso di adenocarcinoma mucinoso digiunale nel quale una concomitante malattia celiaca è stata diagnosticata istologicamente. Sono discussi la diagnosi difficile, il ruolo della chirurgia laparoscopica e la relazione tra i tumori dell'intestino tenue e la malattia celiaca.

Un uomo di 49 anni presentava melena ricorrente, nausea, vomito e anemia. Una stenosi del digiuno veniva documentata da una indagine TC e da una enteroscopia videocapsulare. Un intervento laparoscopico veniva programmato. Un tumore individuato nella prima ansa digiunale veniva rimosso. Lo studio anatomopatologico rivelava un raro adenocarcinoma mucinoso associato a cambiamenti epiteliali secondari a malattia celiaca.

Sebbene i tumori del piccolo intestino siano rari, uno studio diagnostico appropriato è necessario in pazienti celiaci che riferiscono sintomi di alterato transito o sanguinamento occulto. La chirurgia laparoscopica è spesso essenziale per la diagnosi e per il trattamento.

KEY WORDS: Mucinous adenocarcinoma - Jejunum - Celiac disease - Laparoscopy.
Adenocarcinoma mucinoso - Digiuno - Malattia celiaca - Laparoscopia.

Introduction

Celiac disease (CD) is an autoimmune disorder, resulting from the interaction between gluten, immunogenetic and environmental factors, which today is often diagnosed also in adults (1). Several studies have suggested an increased risk of gastrointestinal malignancies, particularly cancers of small bowel, in patients with CD compared to general population (1-3).

Adenocarcinoma of the small bowel has been occasionally described in patients with CD and only few case reports have been published (4-6). In our knowledge mucinous adenocarcinoma of the small bowel associated with CD has never been described.

The Authors report a unique case of mucinous adenocarcinoma associated with CD localized in the first jeju-

University of Catania,Catania, Italy
"Policlinico Vittorio Emanuele" Hospital, Catania, Italy
[1] Department of Surgery, Laparoscopic Surgery Unit
[2] Department of Pathology

nal loop and point out diagnostic difficulties, and the role of laparoscopic surgery in its management.

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 ($pT_3N_1M_x$ – 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.

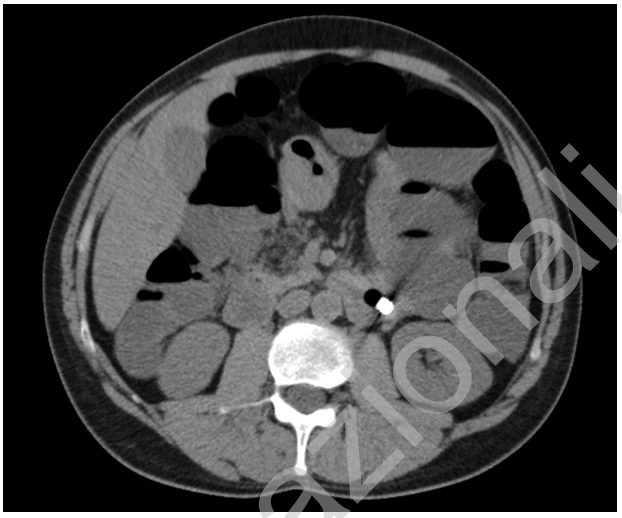

Fig. 1 - Small bowel CT scan. Jejunal occlusion with entrapped enteroscopic capsule.

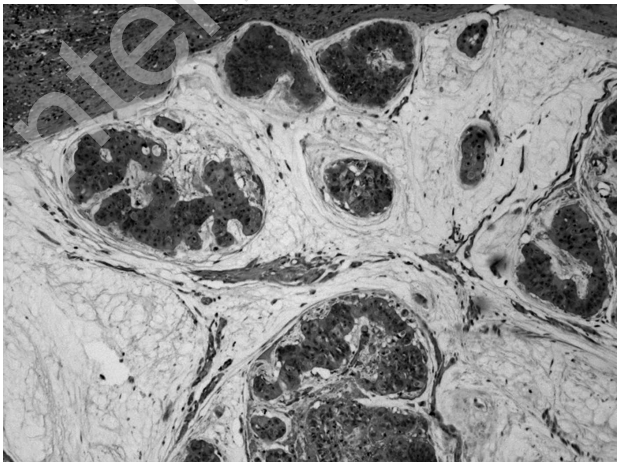

Fig. 2 - Mucinous adenocarcinoma: histological pattern (E&E, original magnification, x50).

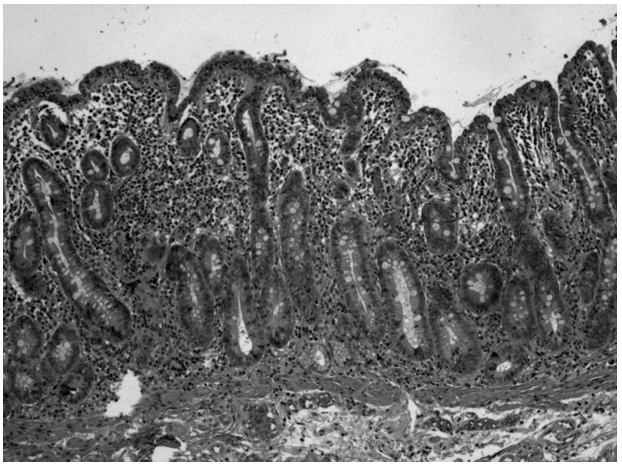

Fig. 3 - Histology. Epithelial changes secondary to celiac disease (E&E, original magnification, x50).

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 case 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 hystologically 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 malignancies. 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.

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