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Case Report

Lymphoma of the Small Intestine: Case Report

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Abstract

Primary malignant tumors of the small intestine are rare and correspond to less than 2% of all tumors of the alimentary canal. The gastrointestinal tract is the most common location of extranodal lymphomas, affecting the small intestine in only 9% of the cases. Its manifestation through free perforation is uncommon. The objective of this report is the description of a patient bearer of a lymphoma of the perforated small intestine, with discussion of clinical presentation, diagnosis, complications and treatment.

T.G.B. 66 years of age, experienced symptoms of colic-type abdominal pain of six months, with worsening in the last 30 days due to uncontrollable vomiting. Weight loss during the period was approximately 10 kg. Alterations of intestinal habit were denied. Patient was bearer of celiac disease with symptoms compensated by diet. After image exams, patient was submitted to segmentary resection of the small intestine for lymphoma, with associated obstruction and good intestinal recovery. Adjuvant chemotherapy made with good evolution. After one year, patient entered in the emergency service with acute abdominal perforation. Patient was submitted to a new laparotomy that demonstrated a recurrence of the lymphoma in the proximal jejunum, with free perforation in the cavity. A segmentary enterectomy was made with good evolution. Bone marrow transplant was proposed but not done. Rescue chemotherapy was made with death after 6 months. Obstruction and perforation secondary of small intestine lymphomas are rare. The survival after resections for perforation is poor. Most patients that recover from the surgery have less than eight months of survival in agreement with the literature.

Keywords: Lymphoma;Intestinal Neoplasms;Intestinal Perforation;Celiac Disease

Introduction

Primary malignant tumors of the small intestine are rare and they correspond to less than 2% of all tumors of the alimentary canal.[1] The gastrointestinal tract is the location most common of extranodal, lymphomas.[2] The stomach is affected in 75% of the cases and the small intestine in only 9%. [2] Lymphomas constitute 15 to 20% of the neoplasms of the small intestine. [3] They are classified as primary (predominance of intestinal compromise, with regional lymphadenopathy) or secondary (superficial lymphadenopathy or mediastinal, with involvement of the spleen and liver).[4] They appear insidiously, with unspecific symptoms of abdominal colic, nausea, vomiting, indisposition and fatigue. Its diagnosis is difficult because the complementary examinations (intestinal transport and tomography) present low sensibility and specificity.[4] Some cases can appear as acute obstructive symptoms, intussusception or perforation (complicated forms); however, these complications are uncommon.

The objective of this report is the description of a patient bearer of lymphoma of the small intestine with discussion of clinical presentation, diagnosis, complications and treatment.

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Case Report

T.G.B., 66 years old, Jewish, married, white, born and raised in Curitiba (PR), experienced symptoms of colic-type abdominal pain in the flank and left hypochondrium of 6 months of evolution, with worsening in the last 30 days due to uncontrollable vomiting. Weight loss approached 10 kg in the period. Patient denied diarrhea or alterations of intestinal habit and presented lack of appetite in the final days. Patient was a bearer of celiac disease with symptoms compensated by diet. Patient also presented hypothyroidism compensated by clinical treatment. Patient denied previous surgeries. There was negative familial history for neoplasms. Patient denied heavy alcohol or tobacco use. Upper digestive endoscopy was made in another service, with active gastric ulcer diagnosis. Patient did not present improvement with pantoprazole given orally and after the onset of uncontrollable vomiting, was directed to the Coloproctology Service of Hospital Universitário Cajuru (HUC).

The physical exam presented excellent general state, normal skin color, stable vital signs, with abdomen plane, flaccid, with light pain and palpation of the left hemi-abdomen, without palpable masses or signs of peritoneal irritation. Segmentary general exam was without alterations; normal laboratory exams. High digestive endoscopy was repeated with report of scarred gastric ulcer, in spite of the permanence of symptoms. Intestinal transport demonstrated luminal constriction in the jejunum, with irregularity in the mucous membrane and upstream dilatation (Figure 1). With the hypothesis of high intestinal occlusion, an abdominal tomography was made, which demonstrated the occlusion level in the jejunum, of a small mass of 3 cm in diameter (Figure 2).

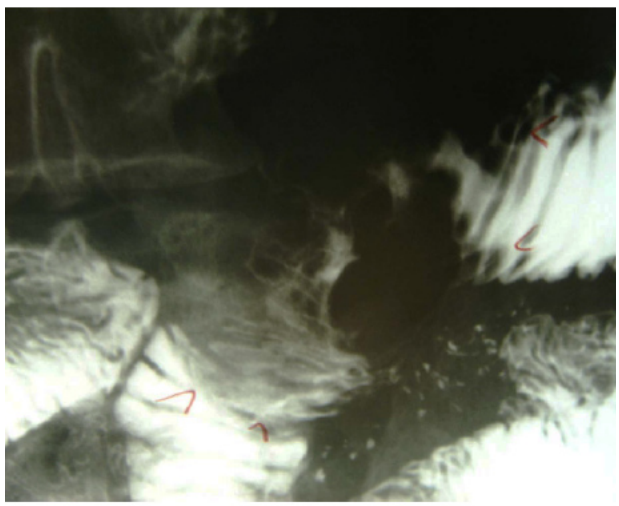

Figure 1 - Intestinal transport: luminal constriction in the jejunum, with irregularity in the mucous membrane and upstream dilatation.

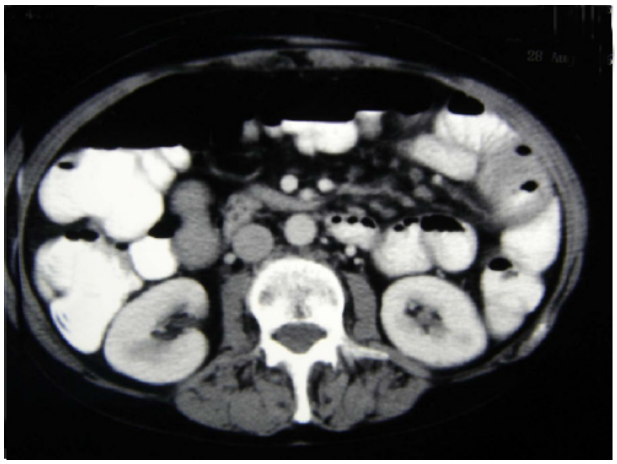

Figure 2 - Abdominal tomography: occlusion level in the jejunum with a small lesion of 3 cm in diameter.

Patient was submitted to exploratory laparotomy with evidence of tumorous lesion of the jejunum, and intestinal subocclusion (Figures 3 and 4). Resection of the affected segment was made with oncologic margins and manual end-to-end anastomosis. There

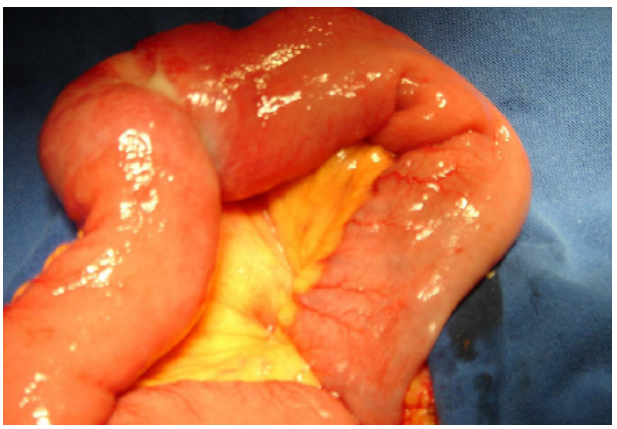

Figure 3 - Macroscopic aspect of a jejunal tumorous lesion with mesentary retraction (first resection).

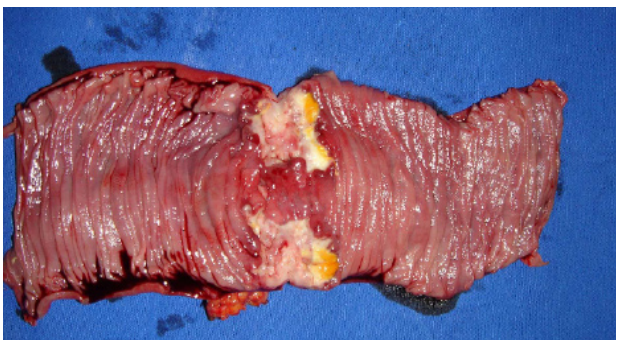

Figure 4 – Surgically resected specimen: ulcerated lesion of the jejunum (first resection)

were no other synchronous lesions in the small bowel, and the liver aspect was of normal consistency. Patient had excellent postoperative evolution with hospital discharge in 3 days. A fever of 38 degrees developed in the 12 days that followed the operation, with laboratory exams and normal image. Patient experienced symptom improvement. The histological report demonstrated the treatment of jejunal lymphoma. Immunohistochemical analysis proved the diagnosis of T-cell lymphoma. Patient submitted to chemotherapy with cyclophosfamide, vincristine, adriblastine and prednisone with good evolution.

Patient remained asymptomatic for a period of 1 year. Returned to HUC Emergency with symptoms of intense abdominal pain, with radiological investigation evidenced free air in the peritoneal cavity, confirming the condition of acute perforated abdomen. Submitted to a new exploratory laparotomy, which demonstrated lymphoma recurrence in the proximal jejunum with free perforation in the cavity (Figures 5 and 6). Segmentary enterectomy was made with end-to-end anastomosis, with good evolution. The proposal of a bone marrow transplant was made, which was not carried out. Rescue chemotherapy was initiated, with death after 6 months.

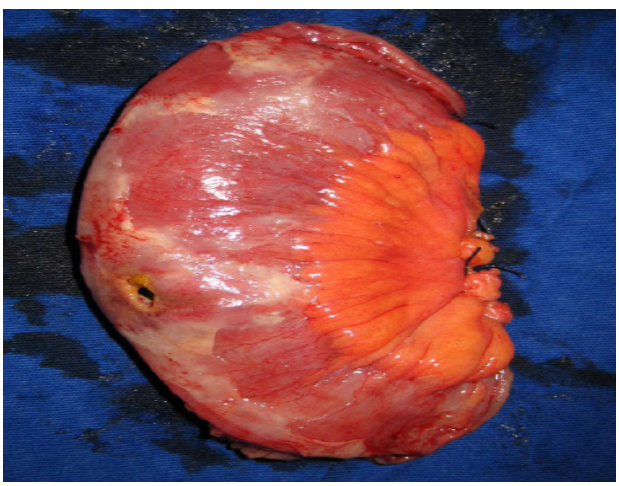

Figure 5 – Lymphoma recurrence in the proximal jejunum with free perforation (second resection).

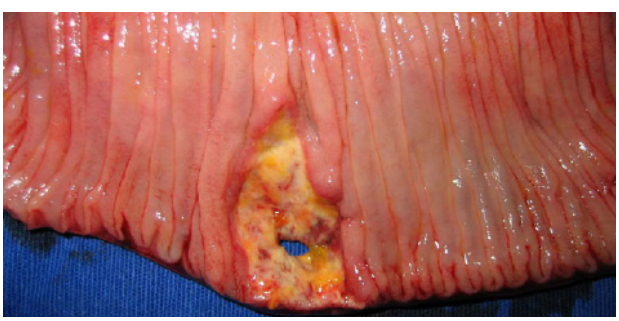

Figure 6 - Lymphoma recurrence in the proximal jejunum with perforation (second operation).

Discussion

Lymphomas of the small intestine can appear as primary tumors or as components of a systemic disease.[2] They are classified as Western type or Mediterranean type. These are usually associated to immunoproliferative disease of the small intestine or alpha-chain disease. [5] They generally manifest as infiltrative and diffuse lesions, most commonly in the intestine proximal. The Western type habitually comes as lesions in mass, ulcerated, protruded or infiltrative and they are most common in the segment distal of the small intestine. [5] However, they still can be histologically classified as low or high grade and can originate from B cells or T cells. Other described classification is of the primary and secondary lymphomas, in agreement with the occurrence of affected nodal at distance. They affect the masculine sex twice as much and are more frequent in individuals of white race. The presentation is bimodal, with a peak below 15 years of age and another above 50 years of age. [4]

Studies show there to be malignant complications in bearers of celiac disease. Around 4.4% of these patients develop lymphomas. [6] When this occurs, the presentation is usually around 50 years of age, with clinical profile compatible with relapse of the symptoms in patients with previous history of celiac disease.[6] Histological evidence of villose atrophy is frequent in the jejunal mucous membrane not affected by the tumor, unless a diet exempt of gluten is being followed.[7,8] In the related case, the previous diagnosis of celiac disease had been made more than 20 years previous and the patient presented control of the symptoms with diet.

The symptoms are unspecific and can include systemic manifestations, as fever and night sweats. Most of the bearers of Western-type or primary lymphoma undergo surgery for occlusive complications or for the discovery of intestinal masses. The free perforation from these tumors is an uncommon finding. [9] The related patient presented the two complications (obstruction and perforation) in an interval of one year, which shows the aggressiveness of this tumor type.

The pathogenesis of the perforation through the tumor is uncertain. Possible mechanisms include substitution of the mucous membrane for tumorous cells, followed by necrosis and ischemia for tumor embolization, increasing the pressure for obstruction and tumorous necrosis by chemotherapy. [9] Systemic

steroids can also induce tumorous necrosis and intestinal perforation. [10]

The role of adjuvant therapy is quite debated. Chemotherapy is indicated for patients with nodal involvement or after complete resection of high grade tumors.This seems to increase the disease free survival and overall survival, mainly when associated with surgery.[4] Adjuvant radiation can diminish the local occurrence of the disease but does not alter survival. This related patient underwent chemotherapy with apparent control of the disease and did not undergo radiotherapy.

Postoperative survival for perforation is poor. Most of the patients that suffer this complication have advanced disease, frequently associated to nutritional, endocrine, hematological and immune disturbances. Many bearers that recover from the surgery have survival less than eight months. [9] In the related case, the poor prognostic demonstrated the low survival in the cases with associated complications.

We conclude that the diagnostic hypothesis of lymphoma of the small intestine should be remembered in cases of celiac disease with long evolution, mainly with atypical symptoms and at the recent beginning. Complications as obstruction and intestinal perforation, secondary to small intestine lymphomas, are rare. The resection of the affected intestinal segment associated to chemotherapy seems to be the best treatment form.The prognostic and survival of these patients are limited.

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