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Acute Peritonitis Revealing a Gastrointestinal T-Cell Lymphoma Associated with Celiac Disease

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Abstract **Case Report**

This case report describes a 52-year-old man who had undiagnosed and poorly managed celiac disease, and presented with acute peritonitis. Further examination revealed a rare complication of celiac disease, which was a T-cell lymphoma of the gastrointestinal tract. Imaging is important in confirming the diagnosis and chemotherapy is the main treatment option.

Keywords: T cell lymphoma-peritonitis- Celiac Disease- surgery- bowel.

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INTRODUCTION

In 1937, Farely and Mackie were the first to describe celiac disease with intestinal T-cell lymphoma during autopsies of patients [1]. T-cell lymphoma associated with enteropathy is a rare complication of celiac disease, which develops on T lymphocytes in the gastrointestinal tract. It is a lymphoma of poor prognosis. We report the case of a poorly managed man with celiac disease admitted in emergency for acute peritonitis revealing a T-cell lymphoma.

CASE

This is a case report of a 52-year-old man with undiagnosed and poorly managed celiac disease who presented with acute peritonitis revealing a rare complication of celiac disease: a T-cell lymphoma of the gastrointestinal tract. The patient had been experiencing abdominal pain, nausea, and vomiting for days. Upon examination, he had diffuse abdominal guarding, and a computed tomography (CT) scan revealed a large amount of fluid and pneumoperitoneum. Laboratory tests showed leukocytosis with neutrophilic predominance and high levels of C-reactive protein. In surgery, an extensive purulent fluid was found along with false membranes, a perforation at the 15th cm of the duodenojejunal junction, stenosis at 115 cm, and multiple lymph nodes. The pathological examination of the surgical specimen confirmed a T-cell lymphoma associated with celiac disease stage 3c of modified Marsh (CD30 positive, CD56 negative). The patient was referred for chemotherapy but died after the first cycle due to hemorrhagic shock.

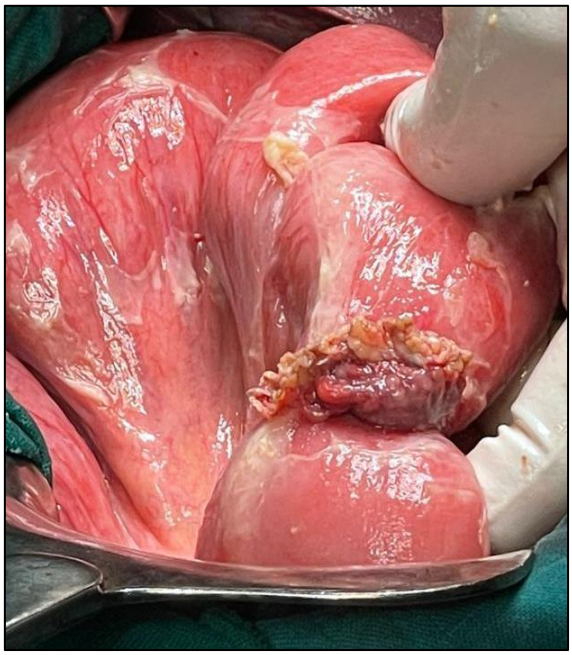

Figure 1: A bowel stenosis

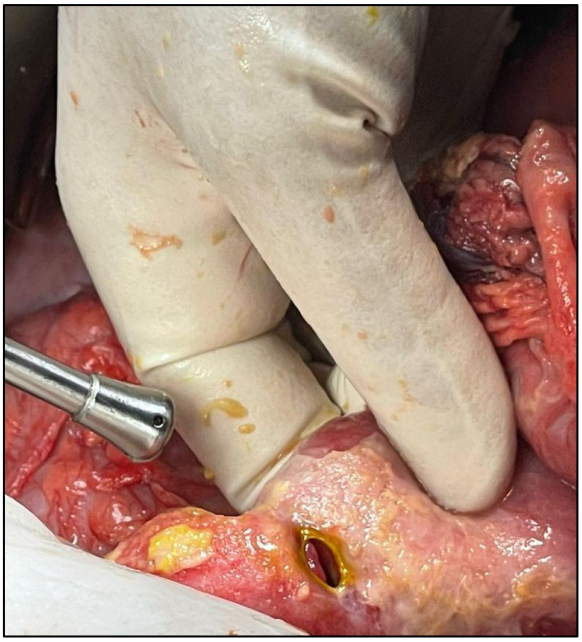

Figure 2: Intestinal perforation

Figure 1: Surgical piece of the tumor

DISCUSSION

Celiac disease is an autoimmune disease triggered by gluten consumption, affecting 0.2-2% of genetically predisposed individuals, leading to chronic inflammation of the small intestine [2]. T-cell lymphoma associated with celiac disease is relatively rare, representing less than 5% of lymphomas associated with celiac disease, with an unknown mechanism of occurrence. The risk is higher when the patient is poorly or not managed, or if the diagnosis is delayed [3].

The average age of diagnosis is 60 years with a male predominance [4]. The clinical presentation includes abdominal pain, anorexia, diarrhea, rarely a tumor syndrome, but in more than 40% of cases, it is a surgical emergency [5]. Imaging plays a crucial role in confirming the diagnosis, with abdominal CT as the preferred modality for characterizing lesions, evaluating lymph node involvement and potential metastases [5]. Acute peritonitis allowed in this case the discovery of T-cell lymphoma associated with celiac disease, with 90% of cases involving the small intestine but may also be multifocal. The WHO has classified T-cell lymphoma into two types: Type 1 lymphoma expresses CD3 in 80-90% of cases, and type 2 lymphoma expresses CD56. The pathological examination of ower surgical specimen confirmed a CD3-expressing digestive T-cell lymphoma associated with celiac disease.

Chemotherapy is the mainstay of treatment, but T-cell lymphoma is generally resistant to chemotherapy with a high rate of relapse [6, 7]. Surgery is indicated only for treating urgent complications such as stenosis, perforation, or pre-perforative resection [8]. Despite all therapies, EATL remains a lymphoma with a poor prognosis due to treatment resistance, complications such as perforation, bleeding, sepsis, and local relapse, with a 5-year cumulative survival rate of 20%, depending on the patient's ability to tolerate the treatments [9].

CONCLUSION

T-lymphoma associated enteropathy is a rare and serious complication of celiac disease; it can be revealed by acute peritonitis. Its treatment, which is not yet standardized, combines early surgery with intensive chemotherapy. The prognosis of EATL remains poor. Its prevention requires an early diagnosis of CD and its consequent management management with a gluten-free diet.

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