

CLINICAL NOTE

## Cavitating mesenteric lymph node syndrome: a rare complication of celiac disease

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### ABSTRACT

Among the many complications of celiac disease, mesenteric lymph node syndrome cavitated is considered one of the rarest, there is few case series published in the literature. The etiology and pathophysiology are unknown but because of its high mortality rate, estimated to be around 50%, it should recognize at an early stage in order to institute appropriate therapy as soon as possible.

**Key words:** Celiac disease. Mesenteric lymph nodes. Gluten-free diet. Hyposplenism. Lymphoma.

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### INTRODUCTION

The cavitated mesenteric lymph node syndrome is a complication of celiac disease, very rare and also potentially serious as its presence increases mortality rate by almost a 50% (1). Their characteristics were described for the first time, in 1969, for Hemet (2) and its relationship with celiac disease was established by Gleeson in 1973 (3). Since then, they have been very few cases reported in the world literature, remaining its etiology still unclear. It has been suggested that a significant alteration in the permeability of the intestinal barrier may favor massive and chron-

ic exposure of mesenteric lymph nodes to specific antigens (including bacterial antigens), resulting in the deposition of immune complexes in the vessels of the lymph nodes and, therefore, causing ischemic damage in these nodes (4).

### CASE REPORT

Male 65 years old, diagnosed with celiac disease of 10 years of evolution. From the time of diagnosis, the patient remained with high degree of anti-tissue transglutaminase antibodies (ATGT) and repeated crisis of watery diarrhea, diagnosed of "dietetic default". In the past 6 months, these crises were becoming more frequent, even after following a strict gluten-free diet accompanied with a loss of 20 kg, without accompanying constitutional syndrome or fever. He joined our department for study. Among the analytical data highlighted a blood count with mild thrombocytosis (602,000 platelets), but otherwise normal formula, and in the biochemical analysis, an albumin of 2.1 g/dl, a pre-albumin of 8.82 g/dL with cholesterol (HDL 21 mg/dL and LDL 55 mg/dL), a ferritin of 1054 ng/mL, transferrin 80 mg/dL, vitamin B12 12 pg/mL, folic acid and 5 pg/dL. The tumor markers were negative. The ATGT was 76 UI (+++). The serum protein: IgG 2480 mg/dL, IgA 189 mg/dL, IgM 26.8 mg/dL. CT findings showed a dilated portal vein, 2 cm, with a liver of normal size and morphology. Spleen, bowel loops and mesentery was normal. Gastroscopy was performed which showed multiple gastric ulcers, and duodenal biopsies were taken was diagnosed with a marked atrophy of the villi with increased intraepithelial lymphocytes, compatible with celiac disease. The colonoscopy was normal.

Given these findings and the lack of clinical improvement was decided to perform video capsule endoscopy (VCE), which was observed at 3 hours of exploration, multiple ulcerations and areas of stenosis in the jejunum medium, suggestive of refractory celiac disease, although not

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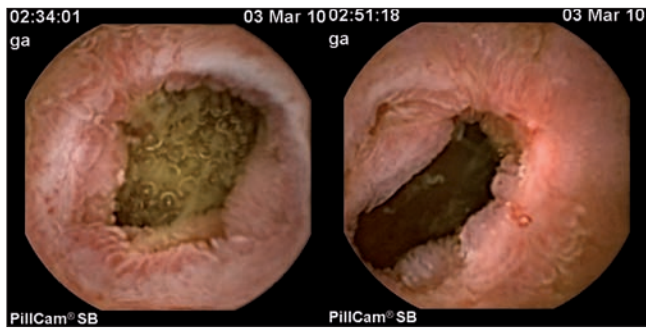


Fig. 1. Two hours and 30 minutes after the start of the scan are displayed at the jejunum, atrophic and irregular mucosa with areas of fibrin, diffuse erythema and ulceration confers a degree of stenosis. These findings suggest as a main cause celiac disease or refractory lymphoma.

ruling lymphoma (Fig. 1). Exploratory laparoscopy was performed to biopsy of the jejunum and possible lymphadenopathy, uncovering a, 1.5 cm in diameter in the mesentery, which was removed.

On histology, the lymph node showed very peculiar macroscopic alterations, mainly by the presence of a central cavity void of content and irregular walls, it was deep inside a creamy whitish material (Fig. 2). In the microscopic examination, this material was consistent with a fibrinous necrotic tissue including degenerate cellular debris, small lipid vacuoles, and polynuclear xanthomatous macrophages. In some isolated focus of this material could be observed suggestive images (not conclusive) of bacteria in short bacillary morphology. In the outer zone, the node kept a narrow rim of normal lymphoid tissue-looking but not very reactive (atrophic) (Fig. 3) and a moderate infiltrate plasmocytorous. Ziehl techniques and PAS were negative. It ruled out the presence of malignancy (lymphoma).

Bowel biopsy showed an erosion of the mucosa that had been replaced by a nonspecific inflammatory tissue character. In a separate piece mucosa showed a better preserved but with lymphocytic inflammatory infiltration and moderate partial atrophy of the villi, compatible with celiac disease (Fig. 4).

All these findings were consistent with “celiac disease and cavitated mesenteric lymph node syndrome.

Corticosteroid treatment was initiated at doses 1 mg/kg and after 5 days of intravenous therapy, the patient was discharged with clear clinical improvement. The patient received maintenance oral corticosteroid treatment and in the review month, remained asymptomatic and ATGT antibodies levels declined.

## DISCUSSION

The cavitating mesenteric lymph node syndrome (CMLNS) is a very rare complication of celiac disease, the lack of recognition and proper treatment can determine a



Fig. 2. Macroscopic appearance of the lymph node that largely has been replaced by a soft white tissue consistency and creamy, which includes a central cavity void of content and boundary anfractuous.

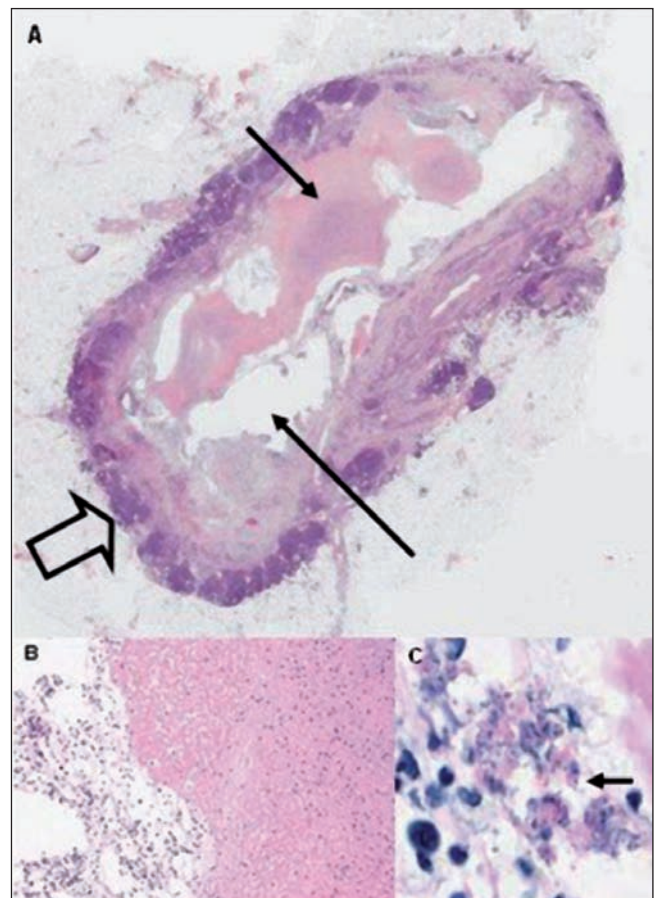


Fig. 3. A. Lymph node showing a central cavitation (long arrow) much necrotic material surrounded by a fibrinous appearance (short arrow). In the periphery there is a border node tissue hypotrophic (arrow) B. Detail of necrotic material including cytoplasm vacuolated macrophages and polynuclear C. As a focal and immersed in the central necrotic area could see some bacterial forms short bacillary appearance (arrow).

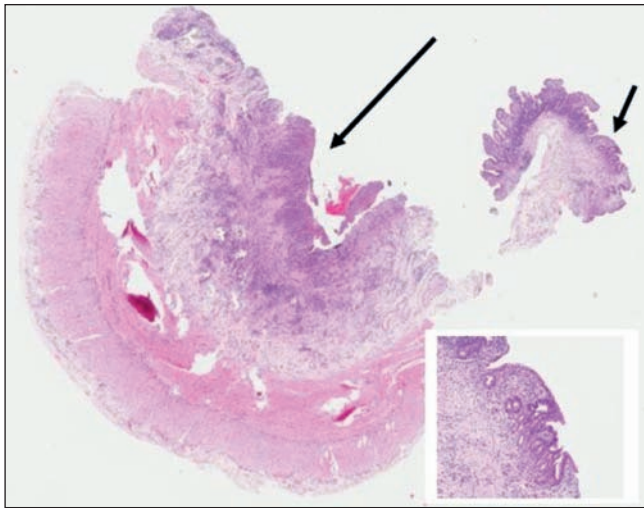


Fig. 4. Image of the jejunum wall where there is an ulceration of the mucosa that is replaced by inflammatory tissue (large arrow) and adjacent, a small piece of mucosa most respected but moderate partial villous atrophy and chronic inflammation (small arrow). These alterations are best seen in the box below.

fatal course of the disease. In its pathogenesis is suggested that a significant alteration of the permeability of the intestinal barrier could facilitate the massive and chronic exposure to certain antigens (including bacterial antigens), resulting, by deposition of immune complexes in the node vascular endothelium, ischemic damage and a consequent necrosis of the node.

In this syndrome, apart from the typical findings of atrophy and inflammation of the intestinal mucosa as described in celiac disease, are other signs, such as splenic atrophy and hypofunction, curiously not present in our case. However, the absence of these signs does not exclude the diagnosis (5). While hyposplenism has been documented

in other diseases of the digestive tract, cavitation of mesenteric lymph nodes has only been described in connection with celiac disease, and not in other diseases such as, for example, inflammatory bowel disease.

Clinically, one should suspect the existence of this complication in cases of celiac disease with a poor response to withdrawal of gluten from the diet. The use of radiological techniques such as ultrasonography and abdominal CT could detect pathological mesenteric lymph nodes, which will make it to an early diagnosis. We believe that these tests should be performed more frequently in cases of celiac disease with poor response to gluten withdrawal. However, laparotomy is usually necessary to obtain adenopathy overlooking the pathological confirmation of the picture. Histologically, there is an empty shell cavity immersed in a necrotic-lipid material, in turn surrounded by a remnant of atrophic lymph node tissue.

In addition to the typical dietary restrictions for celiac disease, treatment is based on intensive use of intravenous steroids. Still, the mortality rate approaches 50%, mainly due to electrolyte disorders, infectious complications and cachexia.

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