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Visible intestinal peristalsis and chronic diarrhea due to a rare lymphoproliferative disease

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Authors´ contribution: GRAM: performed the autopsy, analyzed the histological data and wrote the manuscript. RVP: assisted the patient and wrote the manuscript. MLB, FMR: performed and analyzed the immunofluorescence assay and reviewed the manuscript. ACFS: assisted the patient and reviewed the manuscript. CGM: reviewed the manuscript. JV: analyzed the histological data and wrote the manuscript. MCS: assisted the patient and reviewed the manuscript.

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Keywords: Chronic diarrhea. Lymphoproliferative disorder. Lymphoma.

Dear Editor:

We present the case of a 45-year-old man with watery diarrhea for 2 years, leading to marked weight loss (52kg). On admission, the patient presented with pallor, dehydration and cachexia. Abdominal examination revealed increased bowel sounds, painful and visible intestinal peristalsis, suggesting intestinal obstruction. Upper digestive endoscopy showed mosaic duodenitis (Fig.1A). Duodenal histology showed villous atrophy and an intense infiltrate of small lymphocytes in the lamina propria (Fig. 1B). Immunohistochemical staining was positive for CD3, CD4 (Fig. 1C) and CD8 (Fig. 1D). Colonoscopy showed mild ileal atrophy and biopsies revealed dense ileal and colonic lymphoid infiltrates. Abdominal tomography and magnetic resonance enterography showed no strictures or intestinal occlusion. There was no response to a gluten-free diet and nutritional support. Due to suspicion of autoimmune enteropathy, treatment with methylprednisolone was performed, with significant improvement of diarrhea. However, abdominal pain remained, even after high doses of analgesics, antispasmodics, and opioids. Finally, the patient developed pulmonary infection, septic shock and died 3 months after admission.

Autopsy findings: The abdominal cavity presented various adherences between the intestinal loops (Fig. 1E). Examination of the gastrointestinal mucosa displayed no exophytic or ulcerated lesions, but occasional “mosaic-like” areas were seen, especially on the gastric surface (Fig. 1F). The immunohistochemical phenotype evidenced, again, a T-cell infiltrate and diffuse positivity for both CD4 and CD8. Further analysis using an immunofluorescence assay confirmed double expressor CD4+/CD8+ lymphoid cells (Fig. 1G, 1H, and 1I). No expression of CD30 or CD56. The diagnosis of CD4+/CD8+ indolent T-cell lymphoma of the GI tract (iTCL-GI) was, then, made.

Discussion

The iTCL-GI is a neoplastic disorder recognized in the latest 2022 WHO Classification for Hematolymphoid Tumors [1]. It is characterized by a clonal proliferation of small T-lymphocytes along the mucosa and submucosa of the GI tract [2]. The natural history of iTCL-GI is a chronic and relapsing course, with a poor response to chemotherapy [3]. It can eventually be mistaken with reactive conditions, such as celiac disease and autoimmune enteropathy, which also may present an increased number of lymphocytes in the lamina propria. Our patient had both chronic diarrhea and signs of intestinal occlusion, and the diagnosis was only achieved at the autopsy. Of note, we also described an aberrant immunophenotype of the lymphoid cells (CD4+/CD8+), which has only a few similar prior reports [4,5].

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Figure

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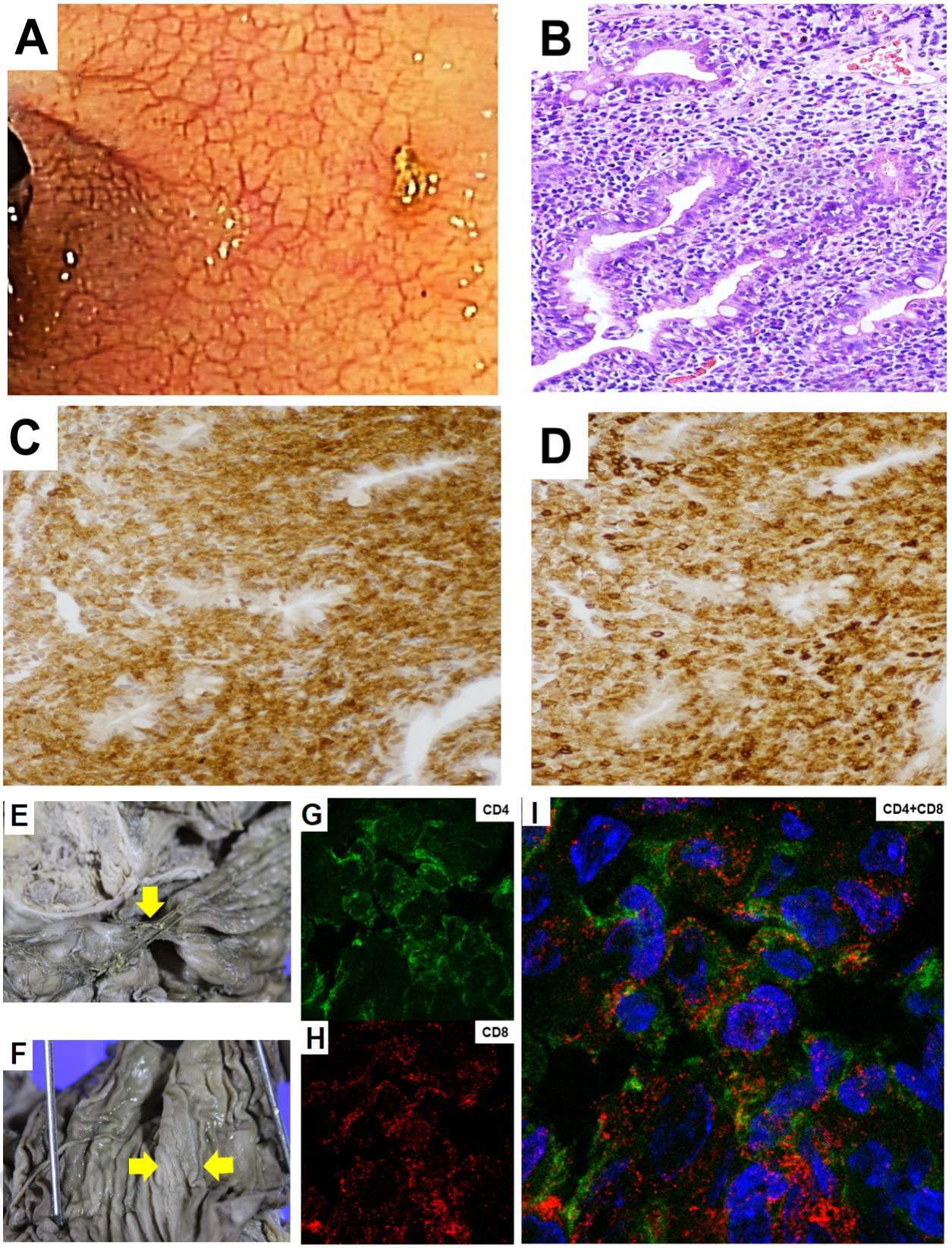

Figure 1. Endoscopic, histopathological findings in the duodenum and autopsy. Upper endoscopy **(A)** showed a mosaic-patterned duodenitis. Microscopic examination evidenced a dense lymphoid infiltrate in the lamina propria, (100x magnification) **(B)**. Immunohistochemistry showed a diffuse lymphoid positivity for CD4, (200x magnification) **(C)** and CD8, (200x magnification) **(D)**. One of the many adherences among intestinal loops is highlighted by the yellow arrow **(E)**. A mosaic-like pattern in the gastric mucosa is delimited by the yellow arrows) **(F)**. After consultation with an hematopathologist, an indirect immunofluorescence was performed in the tissue samples and confirmed that the lymphocytes stained for CD4 **(G)** and CD8 **(H)**, with coexpression of both antigens in the same cells **(I)**.