Waldenström’s macroglobulinemia is a rare hematological neoplasm that infrequently involves the gastrointestinal tract.

We report a case of a 53-year-old female patient, who was admitted for severe iron-deficiency anemia without evidence of blood loss. Colonic evaluation was normal and esophagogastroduodenoscopy revealed a brown-colored second portion of duodenum with the classic pale appearance of the tip of the folds (Fig. 1). Intestinal biopsies revealed distorted microvilli and isolated acellular, homogeneous eosinophilic deposits, positive for periodic acid-Schiff stain (PAS+), in the lymphatic vessels (Fig. 2). Parasitic causes, celiac disease, and amyloidosis were ruled out. On the basis of the above findings, a bone marrow biopsy was done, which was consistent with large B-cell diffuse lymphoma. Hyper-gammaglobulinemia with an IgM monoclonal band and elevated lactate dehydrogenase (LDH) were also present. The patient was diagnosed as having lymphoplasmacytoid lymphoma/Waldenström’s macroglobulinemia with lymphatic transformation to large B-cell diffuse lymphoma in the bone marrow. Thoracic, abdominal, and pelvic computed tomography (CT) scans and positron emission tomography (PET) revealed no abnormal findings. Six cycles of chemotherapy and intrathecal prophylaxis were successfully tolerated. Complete remission was supported by lack of abnormal findings on esophagogastroduodenoscopy (Fig. 3) and a duodenal biopsy (Fig. 4), and in the bone marrow.

Waldenström’s macroglobulinemia is more prevalent in males and in white and elderly people. Clinical features are related to tumor infiltration, hyperviscosity, cryoglobulinemia, and amyloidosis, which are due to the specific properties of monoclonal IgM. Affected organs include the nervous system, bones, kidneys, lungs, liver, spleen, and skin. The small intestine is the most commonly affected segment of the gastrointestinal tract [1–3]; only few cases of gastric compromise have been described [4]. Diarrhea, intestinal pseudo-obstruction, protein-losing enteropathy, and overt or occult bleeding [5] are features indicative of gastrointestinal involvement.

In summary, we present a case with unusual gastrointestinal involvement of a rare hematological malignancy and a lack of specific gastrointestinal symptoms, which was diagnosed following suspicious duodenal biopsy findings and improved markedly after treatment.

Competing interests: None

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Fig. 4  Histological appearance of the duodenum after treatment, showing disappearance of eosinophilic deposits.

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Corresponding author

M. C. Bolino
Gastroenterologia Diagnostica y Terapeutica – Endoscopy
Beruti 2347
PB A’ C1117AAA
CABA
Buenos Aires
Argentina
Fax: +1-905-5223454
carolinabolino@gedyt.com.ar