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Chronic bowel obstruction secondary to MALT lymphoma☆

Oclusión intestinal crónica secundaria a linfoma tipo MALT

In healthy adults, mucosa-associated lymphoid tissue (MALT) accounts for 80% of all immune cells of the body. MALT has 3 functions: to protect mucous membranes against pathogens; to prevent the uptake of antigens in food, commensal microorganisms, and airborne matter; and to prevent a pathologic immune response to external antigens if they cross the mucosal barrier.[1,2]

MALT lymphoma is a marginal zone-type indolent B cell non-Hodgkin lymphoma (NHL) There are 3 main types of marginal zone lymphomas (MZLs): splenic MZL, extranodal MZL of MALT, and nodal MZL. MALT lymphoma belongs to the extranodal MZL group. Biopsies of MALT lymphoma show perivascular and parafollicular infiltration by atypical monocytoid lymphocytes with folded nuclear edges that are positive for CD19, CD20, and CD79a, but negative for CD5, mainly, as well as CD10.[2]

This disease is associated with White advanced-age (>60 years) populations and patients present with lymphocytosis, with or without cytopenia and splenomegaly. They usually present with lymphadenopathy and can have fever, night sweats, and weight loss greater than 10% of their previous total body weight (known as B symptoms).[3]

A 66-year-old woman sought medical attention, due to colicky pain that increased during food intake. She presented with intake intolerance of 6-month progression and immediate postprandial pain, with nausea and occasional vomiting. After vomiting she had pain that was accompanied by the sensation of a hard abdominal mass. She showed no signs of bowel obstruction. A relevant fact of her medical history was that she had a second-degree relative with Hodgkin's lymphoma. Physical examination revealed a soft depressible abdomen, increased peristalsis upon superficial palpation, and increased volume in the left hypochondrium. The rest of the physical examination was normal.

The patient had previously undergone colonoscopy for gastrointestinal symptoms and was diagnosed with lymphocytic colitis. Laboratory test results reported carcinoembryonic antigen 1.45 ng/mL and CA 19−9 2.5 U/mL.

Preoperative complete blood count and blood chemistry analyses, as well as coagulation tests, were within normal limits, with adequate controls.

The following laboratory work-up results stood out: glucose 92 mg/dl, blood urea nitrogen 7.9 mg/dl, creatinine 0.5 mg/dl, blood urea nitrogen/creatinine ratio 15.8, total cholesterol 136 mg/dl, and triglycerides 67 mg/dl.

Contrast and non-contrast computed tomography (CT) of the abdomen and pelvis, with axial views and multiplanar reformation, were carried out (Fig. 1).

The CT scans identified small bowel segments distended with air and neutral fluid. There was homogeneously enhanced concentric wall thickening (with no stratification pattern) up to 21 mm thick at the level of the distal ileum, which conditioned a narrowing of approximately 70% of the lumen, causing retrograde dilation of the ileum. Air and residual material were observed in the colon.

The thickening of the distal ileum walls ended with a retrograde obstructive defect and suspicious data of hepatic and para-aortic retroperitoneal lymph node metastatic activity, suggesting a carcinoid tumor. Exploratory laparotomy revealed a small bowel tumor at 210 cm from the angle of Treitz and 160 cm from the ileocecal valve that obstructed 80% of the intestinal lumen, causing wall thickening up to 10 cm. Intestinal resection with a 10 cm proximal and distal extension of healthy tissue was performed, along with an end-to-end intestinal anastomosis. Liver examination identified smooth edges and no superficial or deep tumors were palpated. Given the absence of bulges or macroscopic alterations, retroperitoneal exploration was not performed.

The diagnosis was extranodal marginal zone lymphoma, clinical stage II Be, low-risk MALT-IPI. The results of the complementary immunohistochemical tests were:

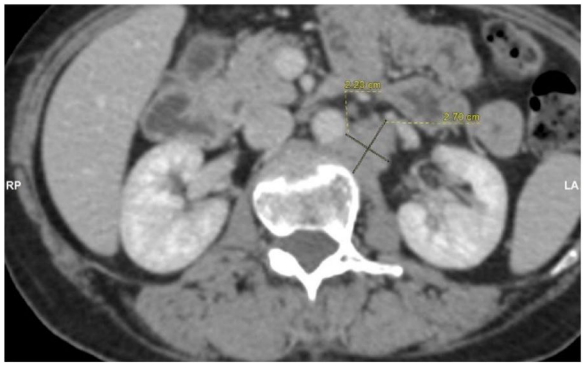

Figure 1 Computed axial tomography scan showing a left-sided 2.40 × 2.70 cm mass in the ileum.

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Figure 2 Positive immunohistochemical staining (CD20) in the atypical lymphoid infiltrate. The color brown shows CD20 positivity.

CD79: positive in 100% of the neoplastic cells; CD20: positive in 100% of the neoplastic cells (Fig. 2); CD43: positive in 80% of the neoplastic cells; CD5: positive in 20% of the neoplastic cells; CD10: negative; BCL-2: focally positive; Cyclin D1: negative; Kappa: negative; Lambda: negative; and CD23: negative.

The patient was released on hospitalization day 5, with adequate oral intake of food. At follow-up at the hematology service, 6 cycles of rituximab-bendamustine were started. She had complete response, with positron-emission tomography (PET) follow-up. Fifteen months after the surgery there were no signs of tumor activity.

MALT lymphoma is a non-Hodgkin extranodal marginal zone lymphoma that most commonly presents in the stomach, but other sites include the skin, salivary glands, lung, small bowel, thyroid, etc.[2,3]

Patients with gastric lymphoma typically present with nonspecific symptoms, frequently with the most common gastric conditions, such as peptic ulcer disease, gastric adenocarcinoma, and non-ulcerous dyspepsia.[4]

Like the rest of the lymphomas, diagnosis is based on lesion histology, complete blood count, and biochemical analyses. Endoscopic examination is necessary in cases of gastrointestinal or pulmonary lymphoma. Disease stage is determined through magnetic resonance imaging (MRI) and CT scanning. Bone marrow biopsy is also performed.[5]

The differential diagnosis includes B cell lymphoma, diffuse large B cell lymphoma, all types of non-Hodgkin lymphoma, and *Helicobacter pylori* infection.[5]

Ethical considerations

The authors declare that this case report contains no personal information that could identify the patient, thus informed consent was not required, but nevertheless, it was obtained for the publication of this work. This case report meets the current bioethics research regulations. Given that the patient's health was not compromised, authorization by the institution's ethics committee was not required.

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Author contributions

Concept, P.I.D.-H., C.A.C.-F., and J.G.L.-V.; Research, P.I.D.-H., C.A.C.-F., J.G.L.-V., U.V.-R., R.M.M.-L., and C.A.C.-F.; Writing/reparation of the original draft, P.I.D.-H., C.A.C.-F., J.G.L.-V., U.V.-R., R.M.M.-L., and C.A.C.-F.; Writing/review and editing, P.I.D.-H., C.A.C.-F., J.G.L.-V., U.V.-R., R.M.M.-L., and C.A.C.-F.; Supervision, C.A.C.-F.

All the authors have read and approved the present version of the manuscript to be published.

Conflict of interest

The authors declare that there is no conflict of interest.

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Intestinal perforation secondary to systemic mastocytosis: Report of an exceptional case☆

Perforación intestinal secundaria a mastocitosis sistémica: reporte de un caso excepcional

Mastocytosis is a rare disease characterized by the anomalous proliferation and accumulation of mast cells in one or more organs. Activating mutations of the c-KIT receptor tyrosine kinase gene is the postulated pathogenesis; the D816 V mutation has been detected in more than 90% of cases. Mast cell accumulation can be produced at the level of the skin (cutaneous mastocytosis) or other organs (systemic mastocytosis [SM]), including gastrointestinal involvement.[1]

A 73-year-old man, with an unremarkable past medical history, was referred from the cardiology service for anemic syndrome study. The outstanding finding in the physical examination was splenomegaly. A computed tomography (CT) scan revealed an enlarged 23 cm spleen and abdominal and mediastinal lymphadenopathy. Bone marrow biopsy was performed, and immunohistochemistry (IHC) testing identified the presence of abnormal mast cells that were positive for CD68, CD117, and tryptase, with a positive cKIT mutation (D816 V). The patient was diagnosed with SM associated with monoclonal hemopathy involving the skin, spleen, lymph nodes, bone, and bone marrow. Because there was no clinical progression, periodic follow-up with no treatment was decided upon.

Five years later, the patient was admitted to the gastroenterology service due to microcytic anemia (hemoglobin 9.6 g/dl), melena of 15-day progression, and chronic diarrhea. Microbiologic studies were negative. Endoscopic studies were carried out, in which gastroscopy revealed millimetric pseudovascular lesions in the second part of the duodenum that were friable when touched. Suspected to be the cause of the gastrointestinal bleeding, the lesions were photocoagulated with argon plasma (Fig. 1A and B); colonoscopy findings were unremarkable. Random biopsies of the digestive tract were taken during the two studies and showed dense cellular accumulations, with foci of more than 15 anomalous mast cells. IHC revealed cKIT and tryptase positivity, CD30+, and CD2−, consistent with gastrointestinal involvement of the SM.

Six months later, the patient arrived at the emergency room for the sudden onset of abdominal pain. Abdominal CT scanning identified chronic thrombosis of the suprahepatic veins and inferior vena cava, development of collateral veins, chronic liver disease, and ascites, all suggesting chronic Budd–Chiari syndrome (Fig. 2A and B). The patient's condition worsened within a few hours, presenting with fever and hemodynamic instability. Antibiotic and vasoactive amines were started. He developed septic shock, and because there was no clear diagnosis from the imaging study, emergency laparotomy was performed, identifying fecaloid peritonitis and complete circumferential rupture, 10 cm from the angle of Treitz, of an intestinal segment opening into the cavity. Given the situation of generalized peritonitis and refractory septic shock in a patient with chronic, aggressive, and incurable disease, only surgical damage control was carried out (resection of the affected intestinal segment and primary closure of the perforation). The patient died a

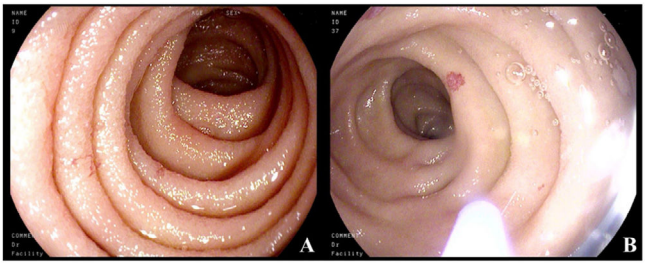

Figure 1 (A) Normal mucosa of the second part of the duodenum, with millimetric erythematous lesions. (B) Catheter for applying argon plasma. The normal mucosa is interspersed with millimetric, rounded, pseudovascular, erythematous lesions with spontaneous bleeding from the passage of the endoscope.

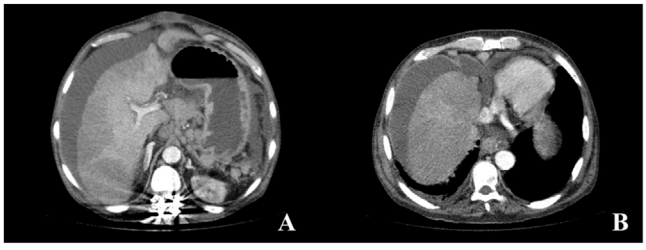

Figure 2 Abdominal CT images with intravenous contrast material in the portal phase, axial MIP reconstruction. (A and B) Signs of chronic liver disease, chronic thrombosis of the suprahepatic veins and inferior vena cava, development of collateral perigastric and perihepatic veins, as well as ascites in all the compartments.

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