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CASE REPORT | SMALL BOWEL

Monomorphic Epitheliotropic Intestinal T-Cell Lymphoma of the Duodenum

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ABSTRACT

Monomorphic epitheliotropic intestinal T-cell lymphomas (MEITLs) are rare neoplasms that carry a poor prognosis. MEITLs originating in the duodenum are uncommon. There are only 3 published case reports of primary duodenal MEITLs. They are typically found in the jejunum or ileum because these parts of the small bowel have more lymphoid tissue. We present a 41-year-old man with weight loss and abdominal pain for 2 months. Imaging showed a heterogeneous duodenal mass, and subsequent endoscopy demonstrated a fungating, ulcerative mass with stigmata of recent bleeding in the duodenal sweep. Pathology from the biopsy revealed an MEITL.

KEYWORDS: lymphoma; MEITL; duodenum; T-cell lymphoma; EATL

INTRODUCTION

Monomorphic epitheliotropic intestinal T-cell lymphomas (MEITLs), formerly known as type 2 enteropathy-associated T-cell lymphoma, are rare peripheral T-cell lymphomas that arise in the small intestine.[1,2] MEITLs occur spontaneously and are distinct from Type I enteropathy associated T-cell lymphomas (EATLs), which are malignancies strongly associated with celiac disease.[3] MEITLs are usually diagnosed later in the disease course when patients experience symptoms from complications, such as a perforation or a bowel obstruction.[4–6] Before that point, patients may endorse vague gastrointestinal (GI) symptoms that are often dismissed.[6]

Primary GI tract lymphomas can occur in all parts of the GI tract and are differentiated using histomorphology.[1,4] Within the small bowel, MEITLs are primarily found in the jejunum and ileum; these segments are rich in lymphoid tissue and are more likely to give rise to lymphomas.[3,7] MEITLs originating in the duodenum, consequently, are rare. In fact, there are only 3 previously reported cases of primary duodenal MEITLs.[1,5,8] We present a case of a previously healthy man found to have MEITL of his duodenum.

CASE REPORT

A 41-year-old man presented to the emergency department with 2 months of decreased oral intake and abdominal pain. Initially, he described the pain as a cramping sensation associated with postprandial bloating that was worse with greasy or spicy foods. In the 24 hours before admission, the pain became a sharp, stabbing sensation localized to his epigastrium. He reported a 10 lb weight loss and intermittent night sweats over these 2 months. He denied nausea, vomiting, fevers, or changes in bowel movements.

He was born in Puerto Rico and moved to New York City a decade earlier. He denied any significant alcohol or tobacco use. He also denied any history of gastrointestinal cancers in the family. On initial physical examination, the patient was diaphoretic and tachycardic. His abdomen was distended and diffusely tender, with more prominent pain in the right upper quadrant. There was no rebound tenderness or involuntary guarding. There were no palpable masses, lymphadenopathy, or hepatosplenomegaly. Laboratory test results were notable for a leukocytosis to 19,000 cells/μL, hemoglobin of 13.2 g/dL, and 540,000 platelets/μL. A large, heterogeneous duodenal mass with surrounding fat stranding and fluid was seen on abdominal/pelvic computed tomography

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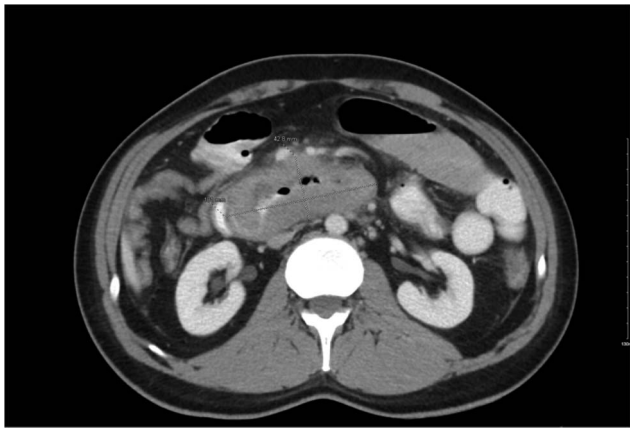

Figure 1. Abdominal and pelvic computed tomography shows a large, heterogeneous duodenal mass (10.1 cm × 4.3 cm) with surrounding fat stranding and fluid.

(Figure 1). It also showed bowel distension proximal and distal to the mass with a possible transition point in the mid-abdomen that was concerning for a possible closed-loop obstruction.

The patient underwent nasogastric tube decompression with successful relief of obstruction. Subsequent esophagogastroduodenoscopy showed a large, fungating, ulcerative mass with stigmata of recent bleeding in the duodenal sweep that was biopsied (Figure 2). Esophagogastroduodenoscopy also revealed erythematous mucosa in the gastric body and duodenitis. The second and third portions of the duodenum were normal.

Pathology of the duodenal mass demonstrated MEITL (Figure 3). Lymphoma cells were positive for CD3, CD7, CD8, and CD56 with Ki-67 = 80%. They were negative for granzyme B and Epstein-Barr encoding region. The patient was then started on chemotherapy with a plan for 4 rounds of CHOP (doxorubicin, cyclophosphamide, etoposide, and vincristine).

Five days after the first round of chemotherapy, he presented again to the emergency department for severe abdominal pain where he was found to have a small obstruction. The hospitalization was complicated by neutropenic fever and sepsis, requiring an intensive care unit admission. His condition

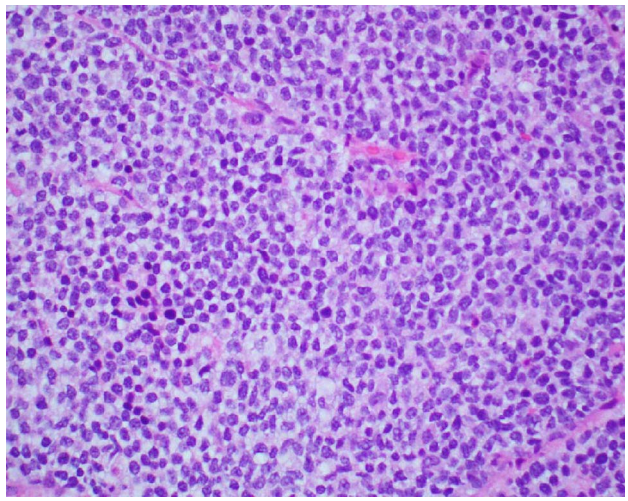

Figure 3. Pathology slide from esophagogastroduodenoscopy biopsies demonstrating monomorphic epitheliotropic T-cell lymphoma.

improved on intravenous antibiotics and was discharged home to resume chemotherapy.

DISCUSSION

In general, lymphomas of the gastrointestinal tract are rare. A Japanese population-based study found that nearly 80% of all gastrointestinal lymphomas originate in the stomach and large intestine; only 6% involve the duodenum.[9] A similar study in the United States showed that follicular lymphomas make up the majority of all duodenal lymphomas (61.9%), followed by diffuse large B-cell lymphomas and mucosa-associated lymphoid tissue lymphomas. Peripheral T-cell lymphomas are the least common subtype, comprising 2.6% of all duodenal lymphomas.[10]

Previously, all enteropathy-associated T-cell lymphomas were considered the same entity. In 2008, the World Health Organization classification system started to bifurcate Type I EATLs from Type II EATLs based on epidemiological and histological differences.[11] In 2017, the World Health Organization further clarified its guidelines by proposing that Type II EATLs be renamed as MEITLs, per the disease's unique pathology

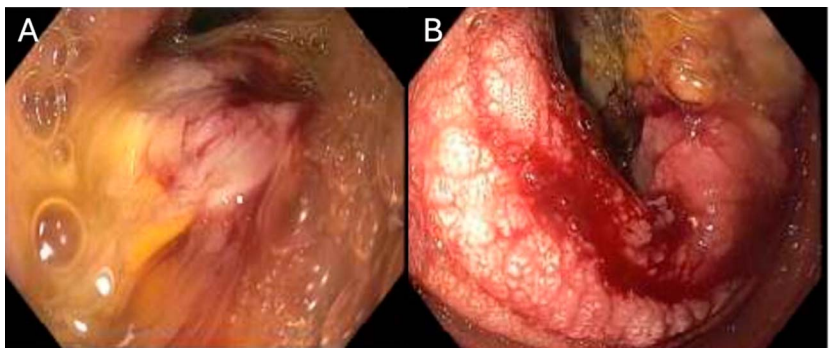

Figure 2. Endoscopy images showing a large, ulcerated mass in the (A) duodenal bulb and (B) second portion of the duodenum.

findings.[12] Histologically, MEITLs present as small- to medium-sized monomorphic cells with darkly stained, round nuclei that tend to be $CD3^+$, $CD4^-$, $CD8^+$, $CD56^+$, and TIA-1^+.[2] Meanwhile, Type I EATL cells are pleomorphic, medium to large in size with prominent nucleoli that are $CD3^+$, $CD4^-$, $CD5^-$, $CD8^{+/-}$, $CD43^+$, TIA-1^+, and $CD103^+$.[8] These lymphocytes are usually embedded in a background of inflammatory cells, which is unlike the MEITL histology.[13]

MEITLs carry a poor prognosis as they are aggressive malignancies diagnosed in the late stages. A retrospective analysis of 42 individuals demonstrated a mean survival time of 2.4–27.2 months.[7] Of these, 50% of the primary sites were in the jejunum, 45% were in the ileum, and none in the duodenum.[7] In previously published cases of duodenal MEITLs, each patient died within 2 years of diagnosis, despite treatment.[1,5,8] Given the paucity of cases of duodenal MEITLs, it is unclear whetner the location of the malignancy affects disease course or outcomes.

Research on evidence-based treatment protocols for MEITLs is scarce. Most accepted therapeutic regimens have been formulated from Type I EATLs and adapted for MEITLs.[1] There are no standardized therapies for MEITLs, but a typical regimen consists of surgery, chemotherapy, and/or autologous stem cell transplantation (ASCT).[14,15] An anthracycline-based regimen, such as CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisone), is associated with longer survival times and has been the mainstay of therapy.[2,3] A retrospective study of 54 patients with EATL found improved remission and 5-year survival rates in patients who underwent chemotherapy +/− surgery compared with surgery alone.[16] Protocols have gradually added a consolidation chemotherapy phase with ASCT after the initial anthracycline-based induction. In a study where CHOP was followed by a consolidation regimen and/or ASCT, a 65% complete remission rate and a 60% 5-year survival rate were achieved.[17] For our patient, he is currently undergoing CHOP with plans for bone marrow transplant after chemotherapy concludes.

DISCLOSURES

Author contributions: S. Babbar and J. Cerezo: substantially contributed to the design of the article and interpreting the relevant literature; drafted the article and revised it critically for important intellectual content; and approved the final manuscript. R. Williams: revised the article critically for important intellectual content; approved the final manuscript. S. Babbar is the article guarantor.

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Informed consent was obtained for this case report.

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