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Single Case

Early, Isolated Duodenal Mucosa-Associated Lymphoid Tissue Lymphoma Presenting without Symptoms or Grossly Apparent Endoscopic Lesions and Diagnosed by Random Duodenal Biopsies

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Keywords
Duodenum · Lymphoma · Mucosa-associated lymphoid tissue lymphoma · *Helicobacter pylori* · Esophagogastroduodenoscopy

Abstract
Clinical data regarding mucosa-associated lymphoid tissue lymphoma (MALToma) solely involving the duodenum are sparse because of the relative rarity of the disease. A comprehensive literature review revealed only 17 cases reported until 2004, and only a moderate number of cases have been reported since. MALToma can be asymptomatic in its very early stages but frequently produces localized or nonspecific symptoms, including early satiety, abdominal pain, vomiting, and involuntary weight loss in later stages. While gastric

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MALToma is strongly associated with gastric *Helicobacter pylori* infection, duodenal MALToma is often unassociated with *H. pylori* infection. A 74-year-old female presented with only dysphagia (without symptoms referable to a duodenal lesion), without systemic 'B' symptoms, and with no evident duodenal lesions at esophagogastroduodenoscopy; however, she was diagnosed with duodenal MALToma by pathologic examination of random duodenal biopsies performed to exclude celiac disease. An important clinical feature of this case is that duodenal MALToma was diagnosed by pathologic analysis of duodenal biopsies despite (1) no endoscopically apparent duodenal lesions; (2) duodenal involvement without gastric involvement; (3) lack of symptoms attributable to duodenal MALToma, and (4) absence of evident *H. pylori* infection. This work shows that early duodenal MALToma can be difficult to diagnose because of absent symptoms, absence of gastric involvement, absence of endoscopic abnormalities, and absence of *H. pylori* infection; it may require random duodenal biopsies for diagnosis.

Introduction

Mucosa-associated lymphoid tissue lymphoma (MALToma) is the third most common non-Hodgkin's lymphoma, representing about 8% of all non-Hodgkin's lymphomas [1, 2]. MALTomas occur predominantly in the stomach [3]. Much data have accumulated about the pathogenesis, clinical presentation, pathology, and treatment of gastric MALTomas because these are relatively common. The normal stomach contains scant lymphoid tissue, but gastric infection with *Helicobacter pylori* leads to accumulation of CD4+ lymphocytes and B lymphocytes in the gastric lamina propria, followed by B-lymphocyte proliferation and the formation of lymphoid follicles [4]. Continued activation, replication, and proliferation of these lymphocytes can lead to MALToma and transformed lymphocytes [4]. Indeed, gastric MALToma is highly associated with *H. pylori* infection, with up to 90% of patients with gastric MALToma having serologic markers of *H. pylori* infection [4].

The close association between *H. pylori* infection and MALToma is strikingly demonstrated by complete histologic, long-term remission in 50–80% of patients with localized, early, gastric MALTomas after *H. pylori* eradication, using combination proton pump inhibitor and antibiotic therapy [5–7]. Patients initially treated with *H. pylori* eradication therapy require a follow-up to confirm *H. pylori* eradication as well as retreatment with another *H. pylori* eradication regimen if the infection was not entirely eradicated [8]. Patients achieving *H. pylori* eradication should then undergo periodic surveillance esophagogastroduodenoscopy (EGD) until complete histologic response is achieved and thereafter undergo ongoing surveillance EGD to confirm persistence of both complete histologic response and *H. pylori* eradication [8]. However, advanced MALTomas, associated with chromosomal t(11;18) translocation, are unlikely to remit with anti-*H. pylori* therapy and thus generally require local radiotherapy, chemotherapy, or surgery [9].

Contrariwise, data regarding the clinical presentation, natural history, pathophysiology, and therapy of duodenal MALTomas are scant because duodenal MALTomas are relatively rare [10, 11]. Duodenal MALTomas appear to have a different pathophysiology and therapy than gastric MALTomas. For example, duodenal MALTomas are relatively rarely associated with *H. pylori* infection and, therefore, are not usually treated with a combination of antibiotic and proton pump inhibitor therapy to eradicate *H. pylori* [12]. We present a patient with localized duodenal MALToma who presented *without* (1) attributable symptoms, (2) endoscopically apparent duodenal lesions, and (3) evident *H. pylori* infection. This work illus-

trates the clinically important finding that early duodenal MALTomas may present without symptoms and may require random duodenal biopsies for diagnosis; we then reviewed the literature on duodenal MALTomas to contrast the biology and natural history of duodenal MALTomas with that of gastric MALTomas and describe what is known or unknown about duodenal MALTomas.

Methods

A comprehensive, computerized, literature review was performed using PubMed with the following MeSH (medical subject headings) or key words: 'gastrointestinal MALToma'; 'small bowel MALToma'; 'intestinal MALToma'; 'gastric MALToma', or 'endoscopy' or 'esophagogastroduodenoscopy' and 'MALToma'. This case received approval/exemption by the Institutional Review Board of William Beaumont Hospital at Royal Oak on March 4, 2016.

Case Report

A 74-year-old woman, with a past medical history of end-stage renal disease, mild chronic obstructive pulmonary disease, left ventricular hypertrophy, mild chronic iron deficiency anemia, chronic gastroesophageal reflux disease treated with proton pump inhibitors, and no known autoimmune disorders presented with dysphagia for solids without abdominal pain or other gastrointestinal (GI) symptoms, and without systemic 'B' symptoms of pyrexia, night sweats, or weight loss. She had a 10-pack-year history of smoking cigarettes, but had quit smoking 10 years earlier. She drank alcohol only socially and did not use any illicit drugs. Physical examination revealed a blood pressure of 145/78 mm Hg, a heart rate of 98 beats/min, and a temperature of 36.5°C. The abdominal examination was unremarkable, including findings of a nontender abdomen, normoactive bowel sounds, and no hepatosplenomegaly. Rectal examination revealed no fecal occult blood. Laboratory analysis showed a hemoglobin level of 10.3 g/dl, a mean corpuscular volume of 87 fl, a serum iron level of 25 μg/dl, and a total iron binding capacity of 223 μg/dl (iron saturation 11%, compatible with iron deficiency anemia). The serum levels of sodium were 136 mmol/l, potassium 3.7 mmol/l, chloride 98 mmol/l, and bicarbonate 26 mmol/l. The serum creatinine level was 2.71 mg/dl, and blood urea nitrogen was 27 mg/dl. Serum tests of liver function were all within normal limits, except that the alkaline phosphatase level was 263 U/l. The albumin level was low at 2.7 g/dl. The lactate dehydrogenase level was 206 U/l (normal range 100–238). A barium esophagogram revealed no abnormalities. EGD revealed no gross duodenal lesions and some digested food obscuring about 20% of the descending duodenal mucosa despite her having fasted for 8 h before EGD (fig. 1). There were no gastric or esophageal lesions. Histologic analysis of esophageal biopsies, performed because of dysphagia, revealed no lesions. Histologic analysis of endoscopic biopsies of the descending duodenum, performed to exclude celiac disease in a patient with suspected iron deficiency anemia, revealed a prominent submucosal lymphocytic infiltrate with scattered plasma cells, consistent with low-grade B-cell non-Hodgkin's lymphoma and otherwise normal mucosa (fig. 2a, b). Immunohistochemistry predominantly revealed B cells, with a strong expression of CD43 and a coexpression of BCL2, findings highly compatible with extranodular marginal zone MALToma (fig. 2c, d). Microscopic examination of gastroduodenal biopsies using a modified Giemsa stain (Diff Quick stain; Electron Microscopy Sciences, Hatfield, Pa., USA) was negative

for *H. pylori*, as was immunohistochemistry using antibodies to *H. pylori*. The serum IgG antibody level to *H. pylori* was 0.41 U/ml (levels of 0.00–0.89 U/ml compatible with no infection). Serum tissue transglutaminase IgA antibody levels were 7.1 U/ml (<20 U/ml negative for celiac disease), with a normal serum IgA level and no monoclonal gammopathy. An abdominal PET scan revealed small, minimally fludeoxyglucose-avid, mesenteric and periaortic lymph nodes. The patient agreed to undergo treatment for duodenal MALToma in the future.

Discussion

MALToma is histologically characterized by diffuse infiltration of neoplastic, centrocyte-like cells distributed in marginal zones around reactive, secondary, lymphoid follicles in the lamina propria [2, 13]. GI MALTomas usually occur in the stomach, sometimes the jejunum ileum, and rarely the duodenum. For example, in a study of 307 cases of primary GI non-Hodgkin's lymphoma, 244 cases (80%) had gastric lymphoma and 63 cases (20%) intestinal lymphoma [14], whereas in another study of 150 GI lymphomas, 105 patients had gastric lymphoma, 27 patients had jejunoileal lymphoma, and only 1 patient had primary duodenal lymphoma [15]. The literature review of duodenal MALTomas identified only 17 cases reported before 2004 [16], and only moderately more cases have been reported since [11, 17–34].

Gastric MALTomas commonly present with GI symptoms of epigastric pain, nausea and vomiting, and gross upper GI bleeding, or with constitutional symptoms of loss of appetite, involuntary weight loss, and night sweats [35]. Symptoms from duodenal lymphomas depend upon lesion location, size, and degree of luminal obstruction [36]. Common clinical presentations include abdominal pain, GI bleeding, and anemia. Najem et al. [37] divided duodenal lymphomas into the following 4 groups based on growth characteristics and symptomatology: (1) obstructive lesions associated with early satiety, vomiting, and postprandial pain; (2) ulcerating lesions associated with GI bleeding, manifesting as hematemesis, melena, or anemia; (3) penetrating lesions associated with typical symptoms of peptic ulcers, and (4) periampullary lesions associated with jaundice. Weight loss may occur in any group. Acute pancreatitis or gastric outlet obstruction are rare presentations [11, 38]. However, duodenal MALTomas are occasionally asymptomatic, or present with nonspecific abdominal pain, especially when diagnosed as early and small lesions [11].

In a review of 38 publications incorporating 2,000 patients with gastric MALTomas, patients almost always presented with endoscopic abnormalities, including gastric erosions, nodularity, enlarged folds, ulceration, polyps, mass, petechiae, or hyperemic mucosa; gastric MALToma without endoscopic abnormalities was rare [39, 40]. Duodenal MALTomas appear similarly at EGD [41]. A comprehensive literature review revealed that duodenal MALTomas rarely present with no endoscopically apparent lesions [16–18, 20–23, 25–27, 30, 31, 34, 42–48].

The current case report illustrates that duodenal MALTomas may be unassociated with *H. pylori* infection. *H. pylori* is strongly associated with gastric MALTomas and is believed to play a role in the pathogenesis of MALTomas by stimulating lymphocyte activation, proliferation, and transformation from chronic inflammation induced by *H. pylori* infection, but its role in duodenal MALTomas is unclear. Duodenal MALTomas are typically independent of *H. pylori* infection [40] and generally persist despite treatment and eradication of *H. pylori* [16]. However, duodenal MALTomas may occasionally regress after *H. pylori* eradication [41].

Due to its rarity, no consensus exists regarding duodenal MALToma therapy [29]. Surgery is often recommended to eradicate and achieve a nearly 100% cure for localized duodenal lymphoma/MALToma, as demonstrated in a study which included 15 patients with duodenal lymphomas/MALTomas [49]. Some authorities argue that surgery may be too aggressive and advocate for chemotherapy [29]. In a study of 4 patients treated with cyclophosphamide 100 mg/day for 18 months without surgery, 2 patients had complete remission for 2 years and 1 patient had complete remission for 65 months [50]. Rituximab has been used for gastric (but not duodenal) MALToma: 20 of 26 patients with gastric non-Hodgkin's lymphoma achieved an objective response with rituximab [51]. In the currently reported case, MALToma therapy was postponed, in accord with the patient's preferences, because the lesion was asymptomatic, localized, and detected very early.

In this case report, the presenting symptom of dysphagia that prompted EGD is not attributable to the duodenal MALToma. The major finding of the current work is that very early duodenal MALTomas can present with no symptoms, produce no grossly evident duodenal lesions at EGD, and may require biopsies of normal appearing duodenal tissue for diagnosis. The lack of mucosal lesions at EGD with early disease may reflect the predominant involvement of the lamina propria or deeper tissue without superficial mucosal involvement.

Study limitations primarily include that this report is retrospective and involves only 1 case. Secondly, the entire descending duodenum was not visualized at EGD because of residual partly digested food, and, therefore, a small duodenal lesion hidden behind partly digested food could not be entirely excluded. However, the great majority of the descending duodenum was visualized at EGD, and pathologic examination of an endoscopic biopsy from normal appearing duodenal mucosa revealed MALToma. Thirdly, we cannot completely exclude that the reported iron deficiency anemia arose from GI bleeding from the MALToma. However, the patient had no fecal occult blood and no (ulcerating) lesions detected at EGD. Furthermore, it is unlikely that the iron deficiency anemia resulted from malabsorption of ingested iron because the patient had early duodenal MALToma, as indicated by no evident endoscopic lesions, as well as indolent MALToma, as indicated by the absence of all of the following 5 aggressive factors: evident disseminated disease [52], peripheral blood involvement [53], presence of 'B' symptoms, an elevated lactate dehydrogenase [54], and a monoclonal gammopathy [55]. Fourthly, the clinical impact of the current report of clinically occult duodenal MALToma is limited by a lack of consensus regarding therapy for asymptomatic, early duodenal MALToma.

Statement of Ethics

The authors report no ethical problem.

Disclosure Statement

The authors report no conflicts of interest. This paper does not discuss any confidential pharmaceutical industry data reviewed by Dr. Cappell as a consultant for the United States Food and Drug Administration (FDA) Advisory Committee on Gastrointestinal Drugs. Dr. Cappell is a speaker for Movantik, a drug jointly manufactured by AstraZeneca and Daiichi Sankyo. This work does not discuss any drug or medical device manufactured or marketed by AstraZeneca or Daiichi Sankyo.

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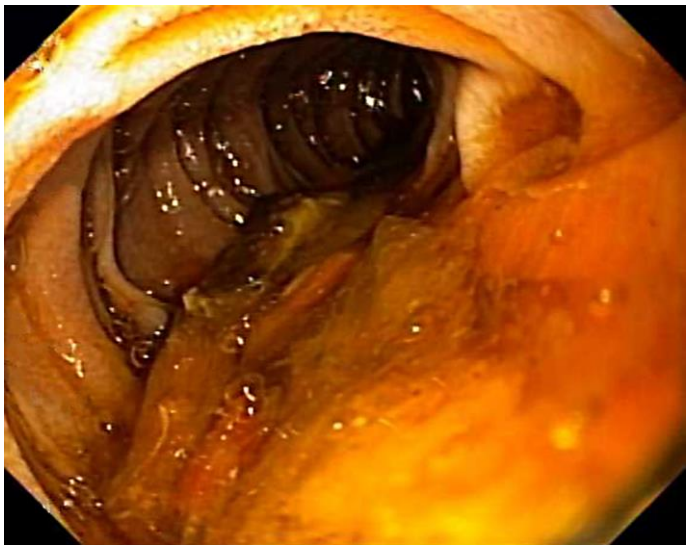

Fig. 1. Endoscopic appearance of the descending duodenum in a 74-year-old female showing retained, partially digested food in a small part of the descending duodenum, and no evident endoscopic abnormalities in the remaining 80% of the descending duodenum. Pathologic examination of endoscopic biopsies of normal appearing mucosa of the descending duodenum revealed MALToma.

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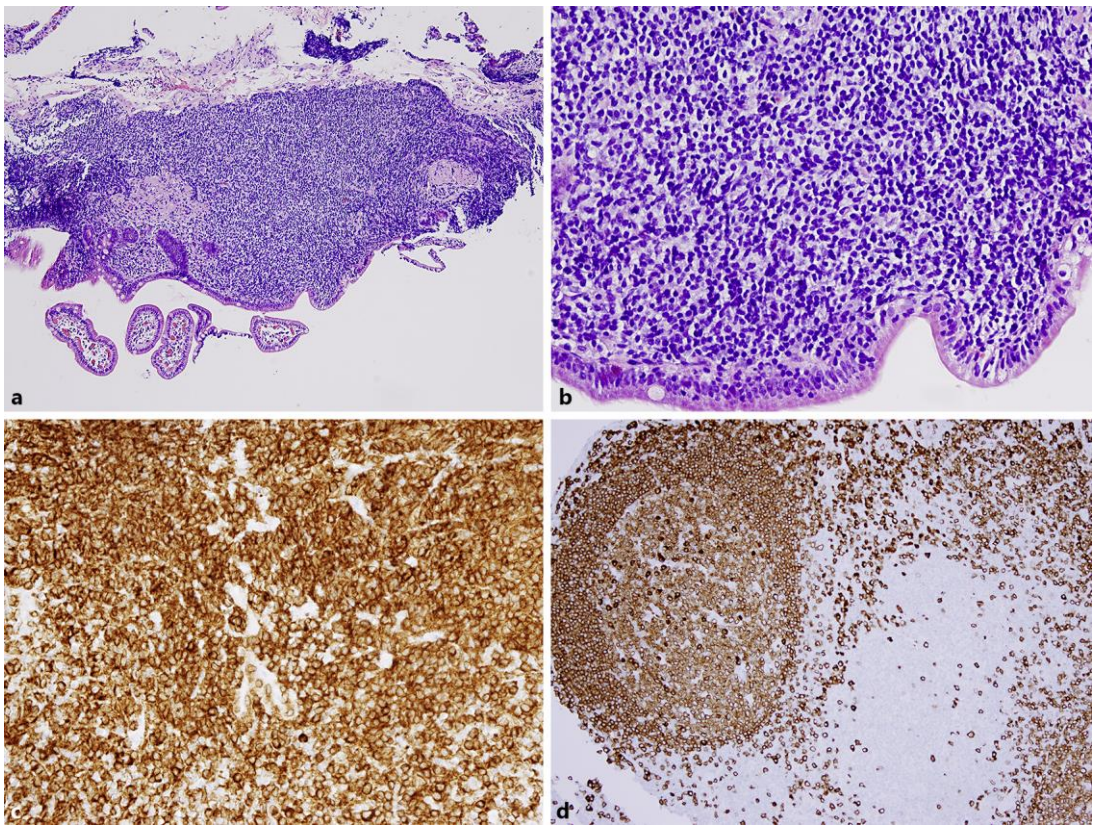

Fig. 2. Histopathologic examination of endoscopic biopsies of the descending duodenum in a 74-year-old woman with no evident endoscopic duodenal lesions. **a** Low-power photomicrograph from a duodenal biopsy showing a dense infiltrate of small mature lymphocytes extensively involving the submucosa, with otherwise unremarkable duodenal mucosa (H&E, ×10). **b** High-power photomicrograph from a duodenal biopsy showing a monotonous proliferation of small mature lymphocytes, with some evident monocytoid differentiation (H&E, ×40). **c** Immunohistochemistry for the B-cell marker CD79a shows strong positivity in most cells of the lymphocytic infiltrate, highly consistent with the diagnosis of a B-cell lymphoma (×40). **d** Normal tonsil, used as an external control, demonstrates positive staining with CD79a on the left in a lymphoid follicle and absent staining with CD79a on the right in an adjacent area containing T cells.