CASE REPORTS

Duodenal mucosa-associated lymphoid tissue lymphoma treated by eradication of Helicobacter pylori: report of 2 cases including EUS findings

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Although the GI tract is the most common site of low-grade B-cell lymphoma of mucosa-associated lymphoid tissue (MALT),1,2 duodenal MALT lymphoma is rare.3-10 The eradication of Helicobacter pylori is now considered the treatment of choice for gastric MALT lymphoma, yet therapeutic strategies for duodenal MALT lymphoma are still in dispute.3-10 This is a report of 2 cases of duodenal MALT lymphoma, both treated by H pylori eradication. The use of EUS to predict efficacy of eradication is discussed. Informed consent was obtained from each patient for EUS and H pylori eradication therapy.

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Case 1
A 65-year-old woman was admitted because of abdominal discomfort. Examination was negative for hepatosplenomegaly and lymphadenopathy. Laboratory data and imaging studies, including bone marrow aspiration, chest and abdominal CT, and gallium scintigraphy, were normal. Upper endoscopy disclosed irregular granular mucosa with mucosal defects in the anterior wall of the duodenal bulb (Fig. 1A). Although no abnormalities were evident in the stomach at endoscopy, the presence of H pylori infection was confirmed by histology, culture, and a rapid urease test. Histologic examination of the biopsy from the duodenal lesion disclosed diffuse infiltration by centrocyte-like cells with lymphoepithelial lesions in the lamina propria (Fig. 1B). Immunostaining of the tumor cells with the B-cell marker CD20 was positive.

EUS with a 20 MHz catheter US probe (GF-UM3R, Olympus Optical Co., Ltd., Tokyo, Japan) demonstrated focal thickening of the second sonographic layer of the duodenal wall, but the third layer seemed to be well preserved (Fig. 1C). These findings suggested that the lymphoma was restricted to the mucosa or to the superficial portion of the submucosa.14 Periduodenal lymph node enlargement was not observed. Based on these findings, a diagnosis was made of primary duodenal MALT lymphoma, stage EI1 according to the Ann-Arbor system as modified by the German MALT Lymphoma Study Group.12,13

The patient was treated with a 2-week course of orally administered rabeprazole (20 mg/day), amoxicillin (1500 mg/day), clarithromycin (600 mg/day), and metronidazole (500 mg/day) to eradicate the infection. Endoscopy 6 weeks after the conclusion of the treatment regimen showed that the duodenal lesion had improved (Fig. 1D). Biopsies disclosed mildly inflamed duodenal mucosa without evidence of residual lymphoma cells. Gastric biopsy specimens were obtained and histology, culture, and a rapid urease test were negative for H pylori.

Case 2
A 48-year-old asymptomatic woman was referred for a general health evaluation. Examination and laboratory data, including abdominal CT and gallium scintigraphy, disclosed no abnormalities. Upper endoscopy revealed a normal-appearing stomach, but there were granular areas with ulceration in the duodenal bulb (Fig. 2A). Biopsy specimens from the lesion revealed dense infiltration by CD20-positive centrocyte-like cells and lymphoepithelial lesions.

EUS demonstrated a hypoechoic mass in the second and third sonographic layers of the duodenal wall (Fig. 2B), suggesting that the tumor had massively invaded the deep portion of the submucosa.14 However, there was no periduodenal lymphadenopathy. Based on these findings, a diagnosis was made of primary duodenal MALT lymphoma, stage EI1.12,13 Gastric biopsy specimens were obtained and histology, culture, and a rapid urease test were positive for H pylori infection.

The patient was treated with a 2-week course of orally administered rabeprazole (20 mg/day), amoxicillin (1500 mg/day), and clarithromycin (600 mg/day). Eight weeks after completion of the treatment regimen a 13C breath test was negative. Although follow-up endoscopy at this time demonstrated eradication of H pylori (negative histology, culture, and rapid urease test) granular areas were still present in the duodenal bulb. A large number of remnant lymphoma cells were identified in duodenal biopsy specimens. EUS revealed residual hypoechoic tumor within the second and the third sonographic layers (Fig. 2C). An alternative therapy, oral monochemotherapy with cyclophosphamide, was recommended to the patient.9

DISCUSSION

Primary malignant lymphoma of the duodenum is rare, accounting for only 2% to 4% of primary GI lymphomas15,16 and 5% to 16% of small intestinal lymphomas.17,18 Thirty-three reported cases of duo-
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denal lymphoma were identified in English language publications.3-10,19-38 Among these cases, only 12 were specified as duodenal MALT lymphoma.3-10 Endoscopic features of duodenal MALT lymphoma include multiple erosions, ulcerations, nodular or granular mucosa, and a polypoid appearance, or a combination of these findings.3-10 The endoscopic features in the 2 cases reported here are similar to those reported previously.

The EUS findings of duodenal MALT lymphoma are not well documented. Case 1 reported here had a focal thickening of the second sonographic layer with a preservation of the third layer (Fig. 1C), whereas in Case 2 a hypoechoic mass was located within the second and third layers. These EUS findings are similar to those of gastric MALT lymphoma.14 To date, EUS findings of duodenal MALT lymphoma have been presented in only a single report.9 Lepicard et al.9 found by EUS that a MALT lymphoma in the second portion of the duodenum had invaded the muscularis propria and had involved multiple periduodenal lymph nodes.

Surgical resection or chemotherapy have been recommended for the majority of patients with duodenal MALT lymphoma.4,5,7-9 However, treatment by H pylori eradication has been attempted in 3 cases.3,6,10 The duodenal lesions disappeared in 2 cases,3,10 whereas the lesion in the other case

Figure 1. Case 1. A, Chromoendoscopic view (0.2% indigo carmine solution) showing irregular granular mucosa with mucosal defects on anterior wall of duodenal bulb. B, Photomicrograph of duodenal biopsy specimen showing diffuse infiltration of centrocyte-like cells with lymphoepithelial lesions (H&E, orig. mag. ×400). C, EUS image of focal thickening of second sonographic layer of duodenal wall with preservation of third layer (arrows). D, Endoscopic view 6 weeks after conclusion of eradication therapy showing regression of duodenal lesions.
remained unchanged after eradication of *H pylori*. Complete remission was achieved after *H pylori* eradication in Case 1 reported here, but in Case 2 lymphoma cells were still present after eradication. Thus, the efficacy of the eradication therapy for duodenal MALT lymphoma remains controversial.

EUS staging is useful in gastric MALT lymphoma for predicting the response to eradication therapy. In a previous study, complete remission was obtained in 93% of cases of gastric MALT lymphoma restricted to the mucosa, whereas the remission rate was only 20% in cases with deep submucosal invasion. As in gastric MALT lymphoma, complete remission was achieved in Case 1 in which the lymphoma was found at EUS to be restricted to the mucosa, but in Case 2 with deep submucosal invasion, residual lymphoma was present after *H pylori* eradication. Therefore, assessment of deep submucosal invasion by EUS may be valuable for predicting the efficacy of *H pylori* eradication even in duodenal MALT lymphoma. However, this is speculative and further studies are necessary to confirm these observations.

REFERENCES


