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Helicobacter pylori–Negative MALT Lymphoma Presenting as a Massive Recurrent Gastrointestinal Hemorrhage

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Abstract

Primary gastric lymphoma is rare, representing 5% of all primary gastric neoplasms. The presenting complaints of gastric mucosa-associated lymphoid tissue (MALT) lymphoma are usually nonspecific. However, life-threatening gastrointestinal bleeding from the stomach is unusual and sparsely reported. While studies reveal an indolent course, we present a case that presented with massive and recurrent hematemesis leading to hypovolemic shock secondary to endoscopically confirmed MALT lymphoma, which was treated with radiotherapy to achieve remission. She had no autoimmune diseases and tested negative for *Helicobacter pylori*. Our case emphasizes the importance of early diagnosis and timely intensive radiotherapy of a localized but aggressive gastric MALT lymphoma.

Keywords

MALT lymphoma, gastrointestinal bleed, MALToma

Introduction

Non-Hodgkin's lymphoma (NHL) represents about 4% of all cancers in the United States, and its incidence has been on the rise since the 1970s.[1] Primary gastric lymphoma is rare, representing 4% to 20% of all NHLs and roughly 5% of all primary gastric neoplasms.[1] Histological subtypes of primary gastric lymphoma are broadly classified into a high-grade diffuse large B-cell lymphoma and a low-grade B-cell lymphoma of mucosa-associated lymphoid tissue (MALT).[2]

While the stomach is the most common site of localization of MALT lymphoma, other involved sites include salivary glands, skin, lungs, thyroid, other gastrointestinal (GI) sites, and liver.[3,4] The clinical presentation for gastric MALT lymphoma is nonspecific and includes symptoms such as dyspepsia, epigastric pain, nausea, and anemia due to chronic/occult GI bleeding.[2,4] There additionally exists a strong relationship between *Helicobacter pylori* and gastric lymphomas.[5] Massive GI bleeding is unusual and sparsely reported in the literature.

We present a case of an elderly woman who presented with massive and recurrent hematemesis leading to hypovolemic shock secondary to biopsy confirmed MALT lymphoma, which was treated with radiotherapy to achieve remission.

Case Presentation

We present the case of a 68-year-old woman who presented with weakness and syncope after large volume hematemesis. She was not on any prior antiplatelet or anticoagulant therapy. Physical examination revealed tachycardia to 127 bpm (beats per minute), hypotension to 76/45 mm Hg, and conjunctival pallor. Her hemoglobin and hematocrit on admission were 8.5 g/dL and 25.6%, respectively. She was managed with IV (intravenous) fluids, packed red blood cell transfusion, and Pantoprazole and Octreotide infusions. An upper endoscopy (esophagogastroduodenoscopy) within 24 hours revealed erythematous, friable, spontaneously oozing mucosa with neovascularization in gastric fundus, which was treated with bipolarcautery as depicted in Figures 1 and 2. Biopsy was not obtained due to recent bleeding. She was discharged on oral Pantoprazole.

Two days later, she was readmitted due to severe recurrent hematemesis with similar findings on repeat esophagogastroduodenoscopy with the addition of an adherent clot over the area of previously oozing and friable mucosa, as depicted in Figures 3 and 4. Bleeding was controlled with argon plasma coagulation and biopsies were obtained from the gastric fundus. Due to the recurrent bleeding, an angiogram was

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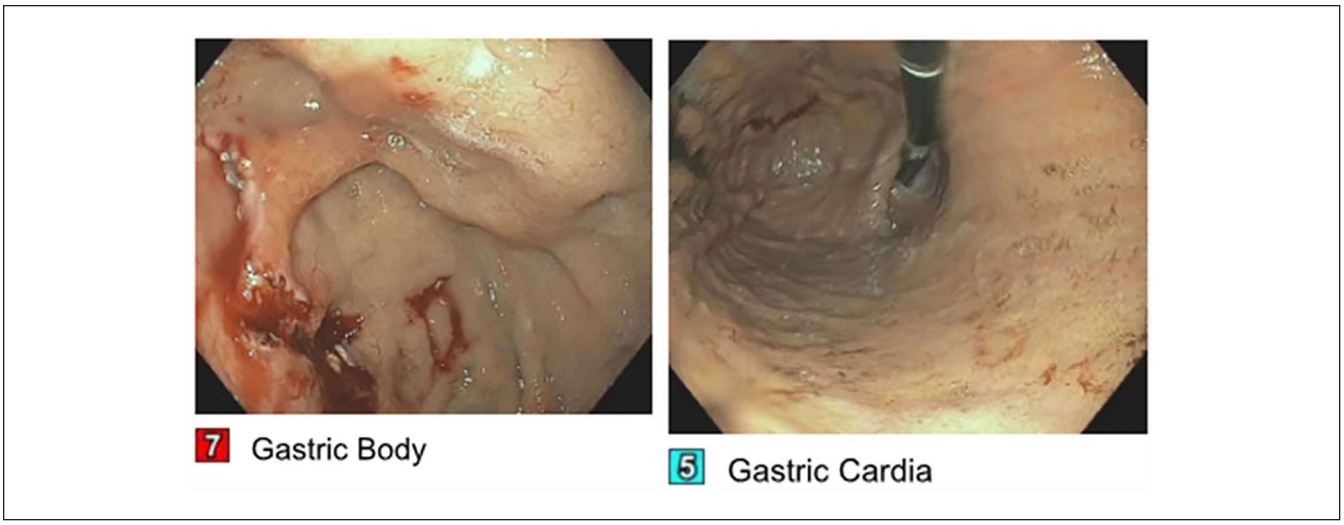
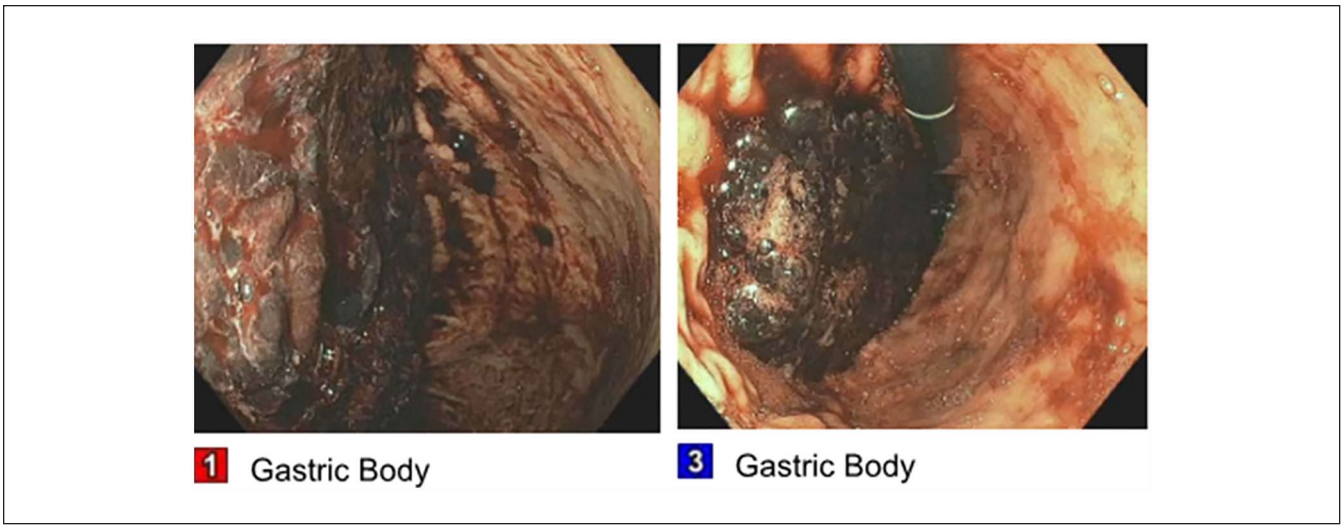


Figures 1 and 2. Erythematous, friable, and spontaneously oozing mucosa. This was the initial endoscopy on presentation.



Figures 3 and 4. An adherent clot over the area of previously oozing and friable mucosa. Biopsies were taken during this time. Bleeding was controlled with argon plasma coagulation (APC).

performed. Active bleeding could not be seen. Prophylactically, the left gastric artery was embolized. The biopsies revealed low-grade B cell MALT lymphoma with negative *H pylori*. She was treated with 20 fractions of radiotherapy with the intention to cure. She did not experience further episodes of bleeding and has remained in remission since.

Discussion

Although studies have shown an indolent nature for GI MALT lymphomas,[6] our case illustrates a life-threatening presentation of a rare low-grade B-cell MALT lymphoma as a massive and recurrent hematemesis. In the study mentioned above, Thieblemont et al[6] analyzed 108 patients to find the overall survival rate of 87% and a 10-year recurrence-free rate of 76%, and Song et al[7] studied 122 patients to report 90.3% overall survival rate and 76.2% hinting toward the indolent nature of this disease.

Song et al[7] also reported GI bleeding in 19 (15.6%) patients but did not comment on the hemodynamic instability, which appears to be sparsely reported. Obleagă et al[8] and Bestari et al[9] reported a case each of upper GI hemorrhage with hypovolemic shock as the first sign of MALT lymphoma which was similar to our case.

Over time, there have been large-scale epidemiological studies unequivocally pointing toward the association between chronic *H pylori* infection and development of MALT lymphoma with cytotoxin-associated gene A implicated in a sustained proliferation of B lymphocytes.[5] In 75% of these cases, eradication of *H pylori* is associated with

achieving remission of MALT lymphoma.[10] Interestingly, our case tested negative for acute or chronic *H pylori* infection.

Similarly, there has also been a 1000-fold increase in the development of marginal zone lymphomas (most common being MALT) when associated with autoimmune processes such as Sjogren syndrome, systemic lupus erythematosus[11]; however, our case lacked any such risk factors.

Dreyling et al[12] recommend that *H pylori*-negative, localized, and recurrent disease when treated with radiotherapy bears favorable outcomes, as seen in our case. Chemotherapy and/or immunotherapy is given when patients fail to respond to radiotherapy or develop metastasis. Long-term endoscopic and laboratory follow-up is needed as gastric MALT lymphoma is associated with a significant risk of gastric adenocarcinoma[13] and increased incidence of gastric cancer and residual NHL. Research in the field remains ongoing with agents such as vorinostat, idelalisib, everolimus, and ibrutinib under clinical investigation.[14]

The management of non-variceal bleeding too has undergone tremendous changes in the recent past. While we adopted the time-tested argon plasma coagulation for management in our case, the advent of modalities such as the Hemospray, the "Endo Clot," and the Blood Stopper appear to have had a paramount impact.[15]

While most gastric malignancies present with GI bleeding, we wanted to highlight the fact that hemodynamically unstable bleeds may not necessarily be varices. The case further emphasizes the importance of early diagnosis and timely intensive radiotherapy of a localized but aggressive gastric MALT lymphoma, which induced remission and cessation of GI bleeding.

Authors' Note

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Declaration of Conflicting Interests

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Ethics Approval

Our institution does not require ethical approval for reporting individual cases nor case series.

Informed Consent

Written informed consent was obtained from the patient(s) for their anonymized patient information to be published in this article.

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