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CASE REPORT

Extranodal Marginal Zone B Cell (MALT) Lymphoma of the Esophagus

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Introduction

Approximately, one-quarter of all non-Hodgkin's lymphoma cases manifest extranodally, half of which are located in the gastrointestinal tract. The majority of the cases are mature B cell neoplasms which arise in the stomach and are highly associated with *H. pylori* infection [1]. In contrast, primary esophageal lymphoma is exceedingly rare, with only 18 cases reported to date in the world literature, nearly all of which are from Eastern Asia [2, 3]. Moreover, primary esophageal lymphoma does not have a clear association with *H. pylori*. Due to the rare occurrence of esophageal mucosa-associated lymphoid tissue (MALT) lymphoma, there is a paucity of data regarding clinical findings, endoscopic features, treatment regimen, and overall prognosis.

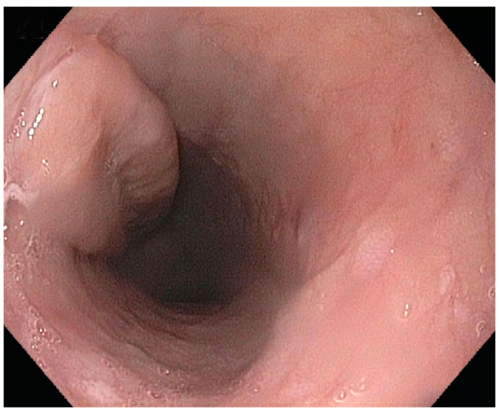

Fig. 1 Endoscopic findings of a 2 cm long, raised, longitudinally-indentated, soft, subepithelial lesion located in the mid-esophagus.

Case Report

A 72-year-old non-Hispanic White woman with well-controlled gastroesophageal reflux underwent esophagogastroduodenoscopy (EGD) for new-onset anemia. This revealed a 2 cm long, raised, longitudinally indentated, soft,

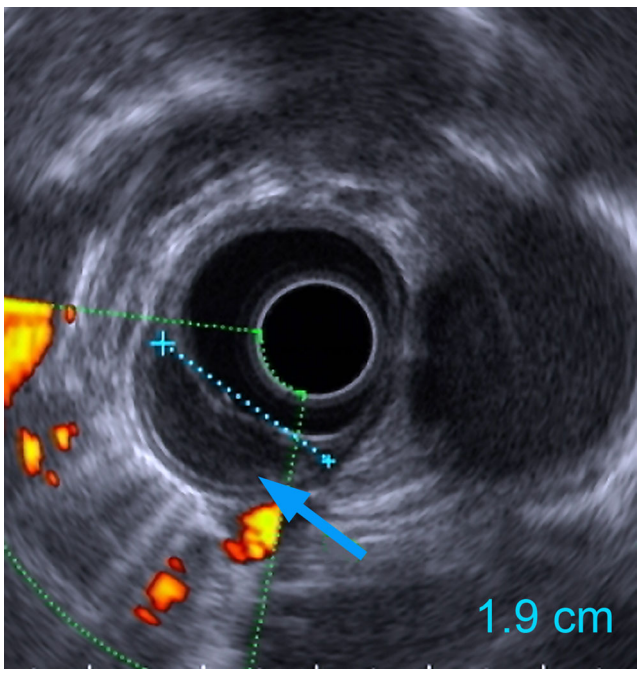

Fig. 2 Endoscopic ultrasound demonstrating a hypoechoic, well-marginated, deep mucosal lesion of unclear etiology

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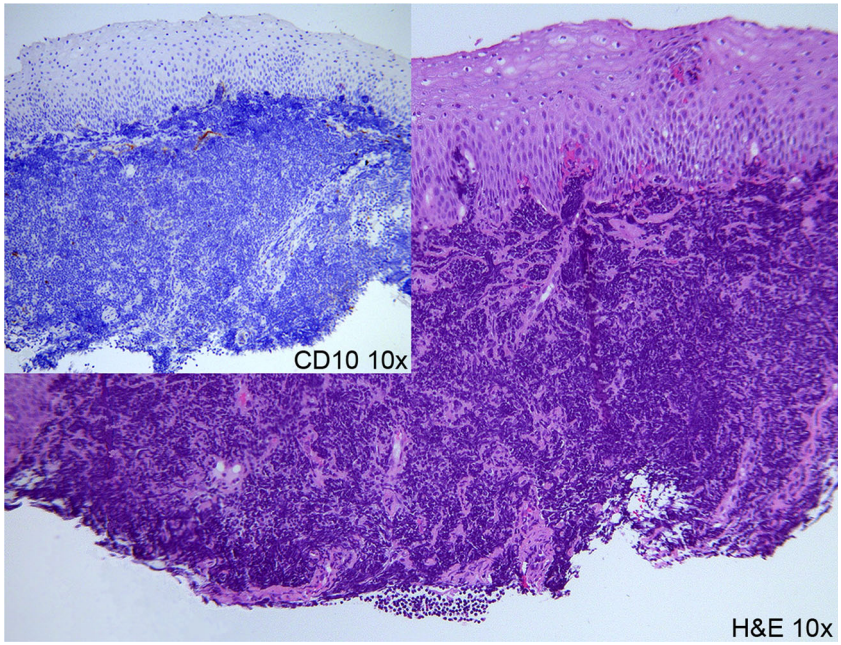

Fig. 3 Endoscopic esophageal mucosal biopsies showing (A) diffuse, dense lymphocytic infiltration on hematoxylin and eosin stain (magnification × 10) and (B) the absence of CD10 expression

subepithelial lesion (Fig. 1) located 25 cm from the incisors. Endoscopic ultrasound (EUS) demonstrated a well-marginated, hypoechoic, deep mucosal lesion (Fig. 2); the esophageal wall layer pattern proximal and distal to the lesion was normal, and there was no lymphadenopathy. Given the benign appearance, negative (reactive) mucosal biopsies and absence of symptoms, repeat EGD-EUS in 1 year was recommended.

One year later, the lesion at 25 cm appeared unchanged endoscopically; however, an additional 1-cm lesion was seen at 35 cm which exhibited analogous endoscopic and EUS features. Biopsies unexpectedly demonstrated a dense, atypical infiltrate with numerous small lymphocytes with round-ovoid nuclei, condensed chromatin, and scant cytoplasm. Immunohistochemistry revealed abundant CD20+ and BCL2+ B lymphocytes negative for BCL6 and CD5/10 (Fig. 3). The morphologic and immunohistochemical features were consistent with low grade, extranodal marginal zone B cell (i.e., mucosa-associated lymphoid tissue, MALT) lymphoma. Notably, *H. pylori* testing was negative, and PET/CT revealed only localized disease (Fig. 4). The patient was seen in consultation by hematology and initiated on chemotherapy with weekly rituximab infusions. At 6 months, the patient was asymptomatic, with PET/CT demonstrating disease remission. Repeat EGD at 1 year showed a resolution of the distal esophageal lesion and a significant improvement of the lesion at 25 cm.

Discussion

MALT lymphoma is a low-grade B cell lymphoma which is found at mucosal sites where marginal zone B cells are

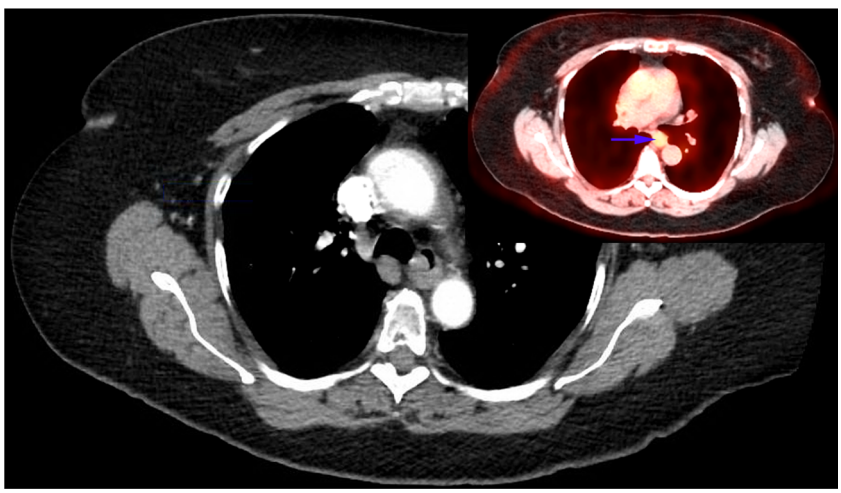

Fig. 4 Contrast-enhanced axial computed tomogram of the chest demonstrating the raised, soft tissue esophageal lesion and (B) positron emission tomography revealing only regional focal

present. MALT lymphoma has been reported in a variety of organs such as the thyroid [4], thymus [5], colon [6, 7], lung [8], skin [9], and salivary glands [10]. However, the gastrointestinal tract is the most common site for MALT lymphomas with most cases reported in the stomach [11]. Approximately, 90% of MALT lymphomas reported in the stomach have been associated with *H. pylori* infection [12]. Primary esophageal MALT lymphoma is extremely rare, and unlike gastric MALT lymphomas, it has not been associated with coexisting *H. pylori* infection.

Here, we report a rare case of esophageal MALT lymphoma which is presented as a submucosal lesion in the esophagus. Based on our case, as well as a review of the literature, it appears that a linear, cranio-caudally oriented, central indentation/ridge in a subepithelial lesion may be a cardinal feature of esophageal MALT lymphoma [12]. This unique finding may assist gastrointestinal endoscopists in recognizing the rare case of esophageal MALT lymphoma and thereby, expedite definitive diagnosis and potentially curative therapy. The overall prognosis of esophageal MALT lymphoma is good due to its slow disease progression and various treatment strategies such as chemotherapy, radiotherapy, and resection have all shown to be effective.

Author Contributions AK assisted with the manuscript preparation and editing. JHT drafted the manuscript and organized images. AMM provided histopathology data and images and edited the manuscript. KP edited the manuscript and provided the endoscopic and radiographic images and is the article guarantor.

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest.

Informed Consent Informed consent was obtained for publication of the case details.

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