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Travis Magdaleno MD

Lehigh Valley Health Network, travis.magdaleno@lvhn.org

Anastasia Shnitser MD

Lehigh Valley Health Network, anastasia.shnitser@lvhn.org

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Acquired Small Bowel Mucosa-Associated Lymphoid Tissue Lymphoma in the Absence of *Helicobacter pylori*

¹Travis Magdaleno, DO and ²Anastasia Shnitser, MD

¹Department of Medicine, ²Department of Gastroenterology, Lehigh Valley Health Network, Allentown, Pennsylvania

INTRODUCTION

Comprising the most extensive component of the human lymphatic system, mucosa-associated lymphoid tissue (MALT) is a protective system of extra-nodal lymphoid cells distributed throughout the body.¹ Malignancies that develop within these tissues are regarded as extra-nodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma) and represent about 8% of all non-Hodgkin lymphomas.^{2,3} Most MALT lymphomas develop within the stomach and have been associated with chronic inflammation from infectious or autoimmune etiologies.³ Fewer than 30% are noted to arise from within the small intestine.³ *Helicobacter pylori* (*H. pylori*) has been a well described risk factor for the development of gastric MALT lymphomas, however its role in duodenal disease is unclear as only a limited number of reports have been described.⁴ We present a unique case in which a MALT lymphoma, discovered within the duodenum, developed from a duodenal ulcer in the absence of autoimmune or infectious causes.

CASE REPORT

A 56-year-old male with a medical history of hypertension presented to his primary care doctor for evaluation of progressive fatigue and melena over the previous few weeks. Laboratory data revealed a microcytic anemia with a hemoglobin of 6.6 g/dL. Initial upper endoscopy revealed a large white based non-bleeding ulcer with heaped edges within the second portion of the duodenum (Figure 1). Duodenal biopsies revealed active chronic duodenitis with evidence of gastric metaplasia. Antral biopsies were performed and *H.pylori* staining was negative. Computed tomography of the abdomen and pelvis revealed wall-thickening of the junction of the second and third portions of the duodenum. He was started on a proton pump inhibitor. Follow up endoscopy 3 months later revealed persistence of the duodenal lesion with a new nodularity component (Figure 2). Repeat biopsy was consistent with a MALT lymphoma (Figure 3). Follow up positron emission tomography (PET) scan and bone marrow biopsy revealed stage 1E disease without evidence of lymph node involvement or metastasis. He completed 4 cycles of rituximab therapy with follow up PET scan revealing partial tumor regression.

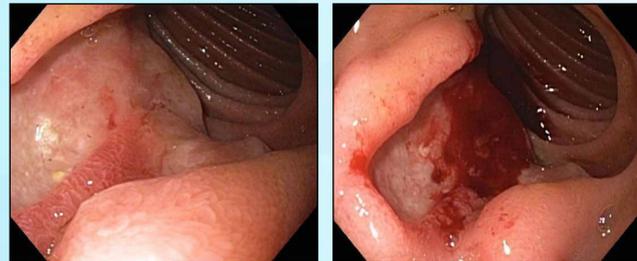


Figure 1: Initial EGD revealing a large white based ulcer with heaped edges within the second portion of the duodenum.

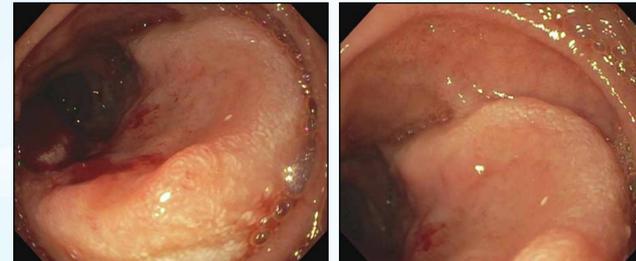


Figure 2: Repeat Endoscopy 3months later revealing development of duodenal lesion with nodularity component.

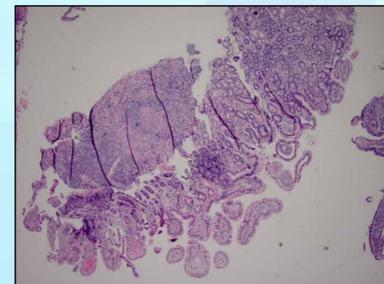


Figure 3: Endoscopic biopsy of duodenal mass revealing dense infiltration of glandular tissue by lymphoid neoplastic cells.

DISCUSSION

- Primary duodenal MALT lymphoma is exceedingly rare with a small number of case reports 1995⁴
- Symptoms of gastrointestinal MALT Lymphoma are dependent on location of growth however may include:
 - Indigestion, abdominal pain
 - Night sweats, weight loss, low grade fevers
 - Iron deficiency anemia, melena
 - Constipation and even bowel obstruction in severe cases
- Diagnosis is made endoscopically with biopsies revealing:
 - Massive lymphomatous infiltration of CD20+ neoplastic cells within lymphoid follicles⁵
- The role of autoimmune diseases and *H. pylori* has been clearly established in gastric disease however is unclear in duodenal manifestations.
 - Few studies have reported regression following treatment in *H. pylori* positive cases⁶
 - Few studies have reported remission with combination chemotherapy including CVP (cyclophosphamide, vincristine, and prednisolone) and rituximab^{3,8}
- In our case report, we have presented the progression of a duodenal ulceration to a MALT lymphoma
 - Duodenal ulcerations in itself may represent a significant risk factor for MALT lymphoma development
- Clinical experience and judgment is crucial in its diagnosis. Although duodenal ulceration does not currently require follow up endoscopy, unusual or atypical appearing lesions should always be investigated with biopsy and followed clinically for resolution.

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