A Case of Isolated Colonic MALT Lymphoma and Synchronous Celiac Disease

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Introduction

- Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma) is uncommon, accounting for 5% of all non-Hodgkin lymphomas
- The most common site is the stomach with isolated colonic involvement comprising only 2.5% of all MALT lymphomas and less than 0.5% of all colon cancers
- Up to 50% of cases are asymptomatic with a variable endoscopic appearance (polypoidal, ulcerative or mass-like) and there is a lack of clear guidance on management
- While gastric MALT lymphomas are strongly linked to active Helicobacter pylori (H. pylori) infection they have also been associated with autoimmune diseases such as celiac disease (CD); a link that is uncertain with the colonic variety

Case Description

- A 50-year-old asymptomatic male with no significant past medical history underwent a screening colonoscopy which was notable for multiple irregular, small, sessile polyps
- Histopathological and immunohistochemical analysis confirmed multifocal MALT lymphoma
- Staging positron emission tomography (PET) showed a thickened duodenum and jejunum with FDG uptake
- Enteroscopy was endoscopically normal, but small bowel biopsies of both the duodenum and jejunum returned consistent with CD
- Follow up CD serologic testing was positive for IgA anti-tissue transglutaminase antibodies (anti-TTg IgA)

Figure 1. Colonoscopy shows a 4-6 mm sized sessile polyp arising in the cecum

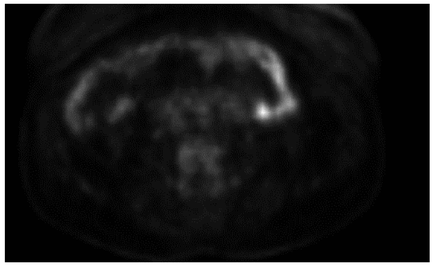

Figure 2. Staging FDG PET/CT with diffuse small bowel FDG uptake

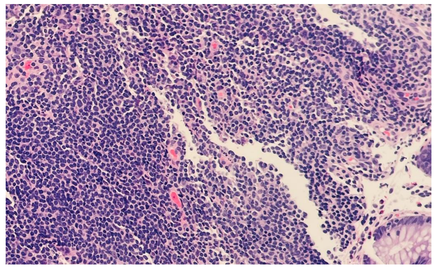

Figure 3. Colonic biopsy specimen shows a diffuse and dense infiltration of small-sized lymphocytes and lymphoepithelial lesions (H&E staining, ×20)

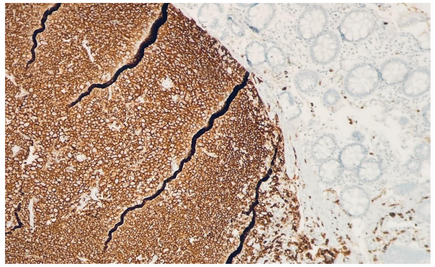

Figure 4. Immunohistochemical staining shows markedly increased infiltrative small-sized lymphocytes with positivity for B-cell marker CD20 (×20)

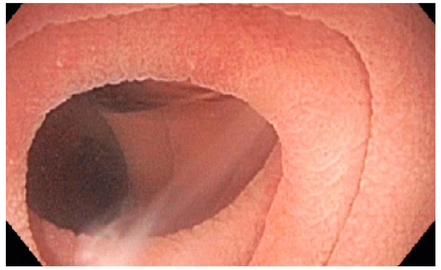

Figure 5. Endoscopic water-immersion technique demonstrating absence of villi and scalloping in the jejunum

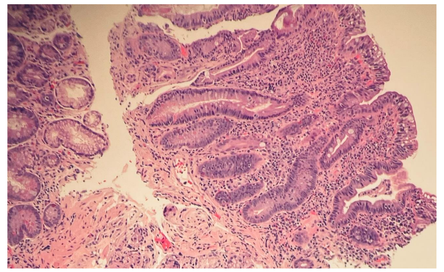

Figure 6. Duodenal biopsy specimen shows villous blunting and increased intraepithelial lymphocytes (H&E staining, ×10)

Case Description Continued

- Due to the low stage of lymphoma and excellent prognosis, patient opted for clinical observation
- After initiating a strict gluten free diet (GFD), patient's previously unreported abdominal bloating markedly improved and anti-TTg IgA normalized
- Several months later, repeat PET/CT showed resolution of the small bowel FDG avidity and no evidence of metastatic disease or adenopathy
- Our patient has now been on a GFD for four years
- Close follow up with annual bidirectional endoscopy and whole-body PET/CT imaging have shown a lack of disease progression

Discussion

- While a causal relationship between enteropathy-associated T-cell lymphoma and CD is well established, data supporting an increased risk of MALT lymphoma is less robust, with one study suggesting an estimated odds ratio of 3.5
- The overall risk of NHL lymphoma in CD is higher when there is active inflammation in the small bowel and lower when CD is quiescent
- While current treatment of colonic MALT lymphoma includes surveillance for low stage disease with the possibility of chemotherapy or resection, the prognosis is excellent regardless of therapeutic modality
- There is insufficient data at this time to recommend treatment of synchronous CD as an adjunct, but this would theoretically be of benefit and warrants further investigation

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