

## Objectives:

- To recognize that Mucosa Associated Lymphoid Tissue (MALT) lymphoma may occur outside the stomach and in *Helicobacter pylori*-negative patients.
- To highlight the importance of optimal high-quality long-term follow-up via colonoscopies with biopsies for chronic UC patients.

## Introduction:

- MALT lymphomas are extra-nodal marginal zone B-cell lymphomas, mostly found in the stomach associated with *H. pylori* infections and rarely presenting in inflammatory bowel disease<sup>1</sup>.
- Rectal MALT lymphoma is very uncommon and often presents as painful defecation, rectal bleeding, or rectal pressure/prolapse.
- Here, we present a rare case of an asymptomatic female with ulcerative colitis (UC) with benign-appearing rectal polyps during a routine colonoscopy, found to be MALT lymphoma on biopsy.

## Case Presentation:

- The patient is a currently asymptomatic 57-year-old Female with UC presenting for a routine screening colonoscopy.**
- Past medical history includes UC and pre-diabetes. UC is left sided, with a flare frequency of 1-2 per year, most recent flare was 4 months prior (after the COVID-19 vaccine). She is a never-smoker and former occasional alcohol user. Current medications include oral Olsalazine 500 mg twice daily, low dose prednisone, and mesalamine suppositories as needed.
- She is well appearing with a BMI of 29.49. Vitals are within normal limits. Physical exam is unremarkable: abdomen is flat, non-distended, non-tender in all 4 quadrants, no masses or hernias appreciated, no rebound or guarding.

## Initial Diagnostic studies:

- Screening colonoscopy** revealed two 7 mm sessile, non-bleeding rectal polyps (Figure 2), removed with a cold biopsy forceps, with an area of surrounding congested, erythematous, friable, and ulcerated mucosa in the rectosigmoid colon (Figure 1). Biopsies of the abnormal mucosa were taken with a cold forceps for histology.
- Pathology of routine samples:** chronic nonspecific colitis of the cecum, ascending colon, descending colon, sigmoid colon, and rectum, consistent with IBD. The transverse colon sample was normal colonic mucosa.
- Hematopathology of the rectal polyps showed marked lymphoplasmacytic infiltrate and extra-nodal marginal zone lymphoma of MALT.**
- Referred immuno-stains for CMV and Congo red stain were negative. **Specifically, staining for spirochetes (including *H. pylori*) was also negative.**

## Follow-up diagnostic studies and plan:

- Pertinent Lab findings:** ESR 40, Immunofixation panel showing slightly elevated IgG (1650), no M-spike on SPEP, elevated free Kappa light chains 3.13, normal free lambda light chains, normal free kappa/lambda light chain ratio
- CT Abdomen and Pelvis w/ IV contrast:** featureless chronic colitis from the splenic flexure to the rectum, fibrofatty proliferation, no active inflammation, no mass-like bowel wall thickening + nonspecific rounded <1cm retroperitoneal lymphadenopathy, stable on follow-up, presumed reactive
- Follow-up oncology recommendations included watchful waiting and close CT and colonoscopy surveillance.**

Figure 1. Inflamed rectosigmoid mucosa

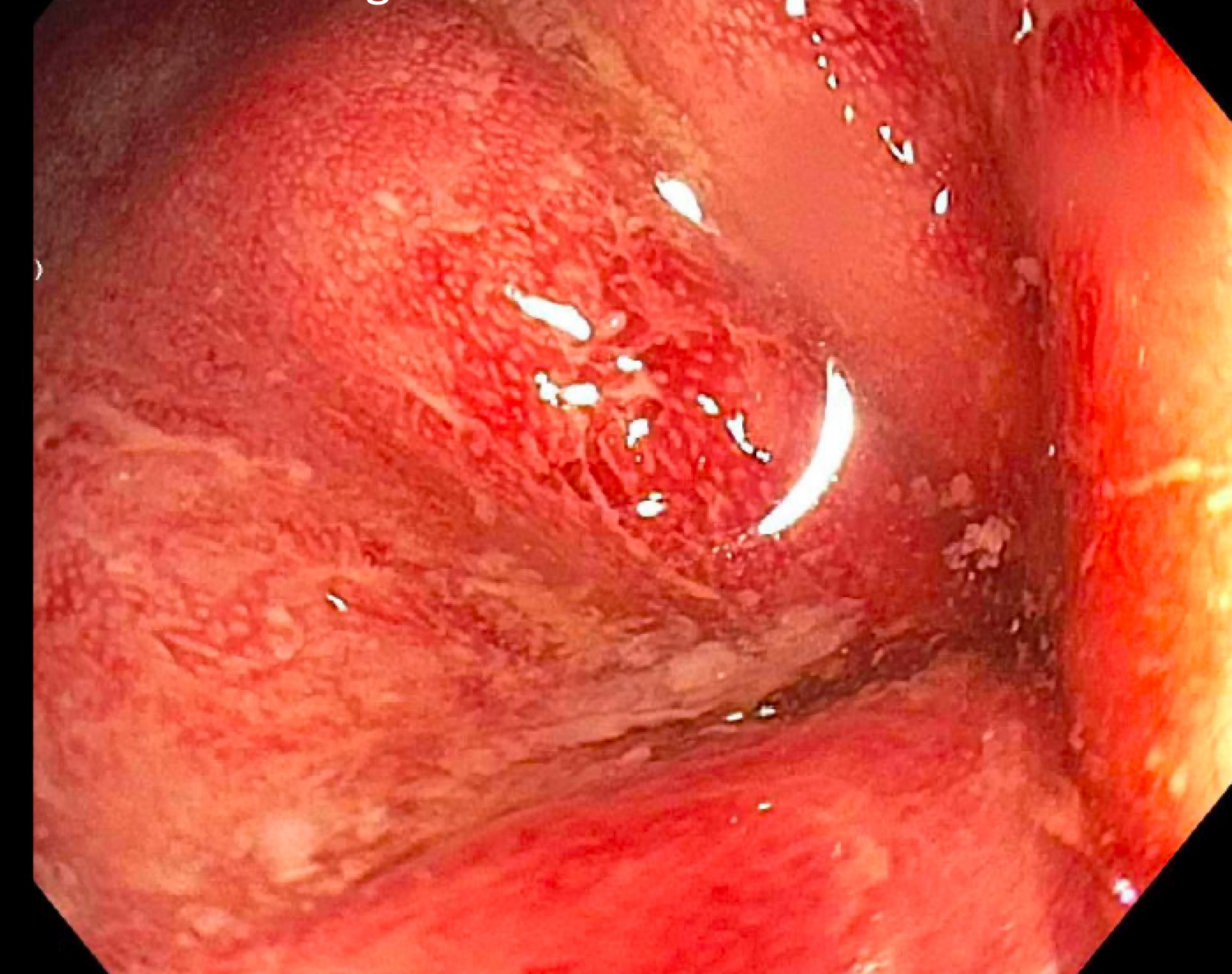


Figure 2. Two 7 mm sessile rectal polyps

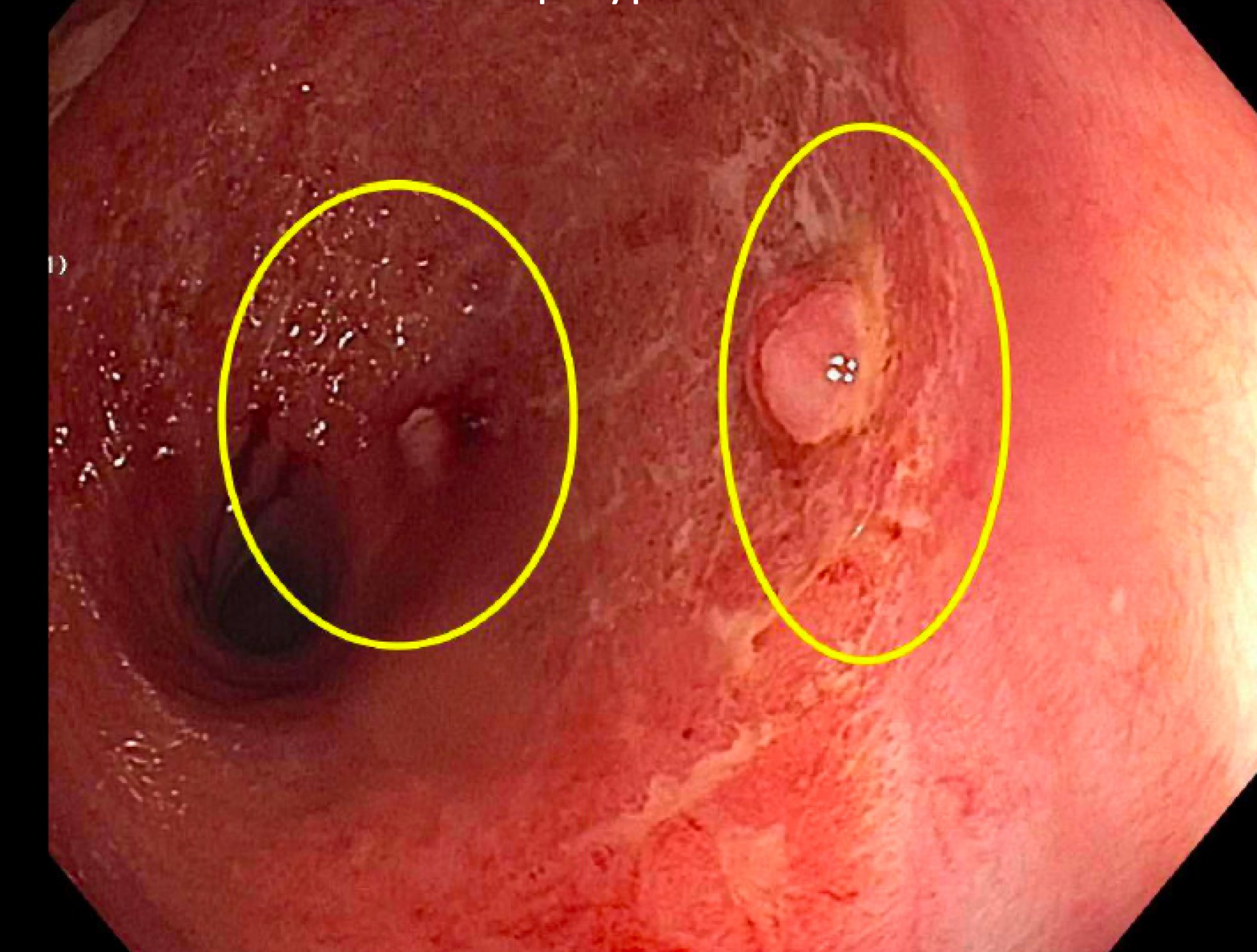
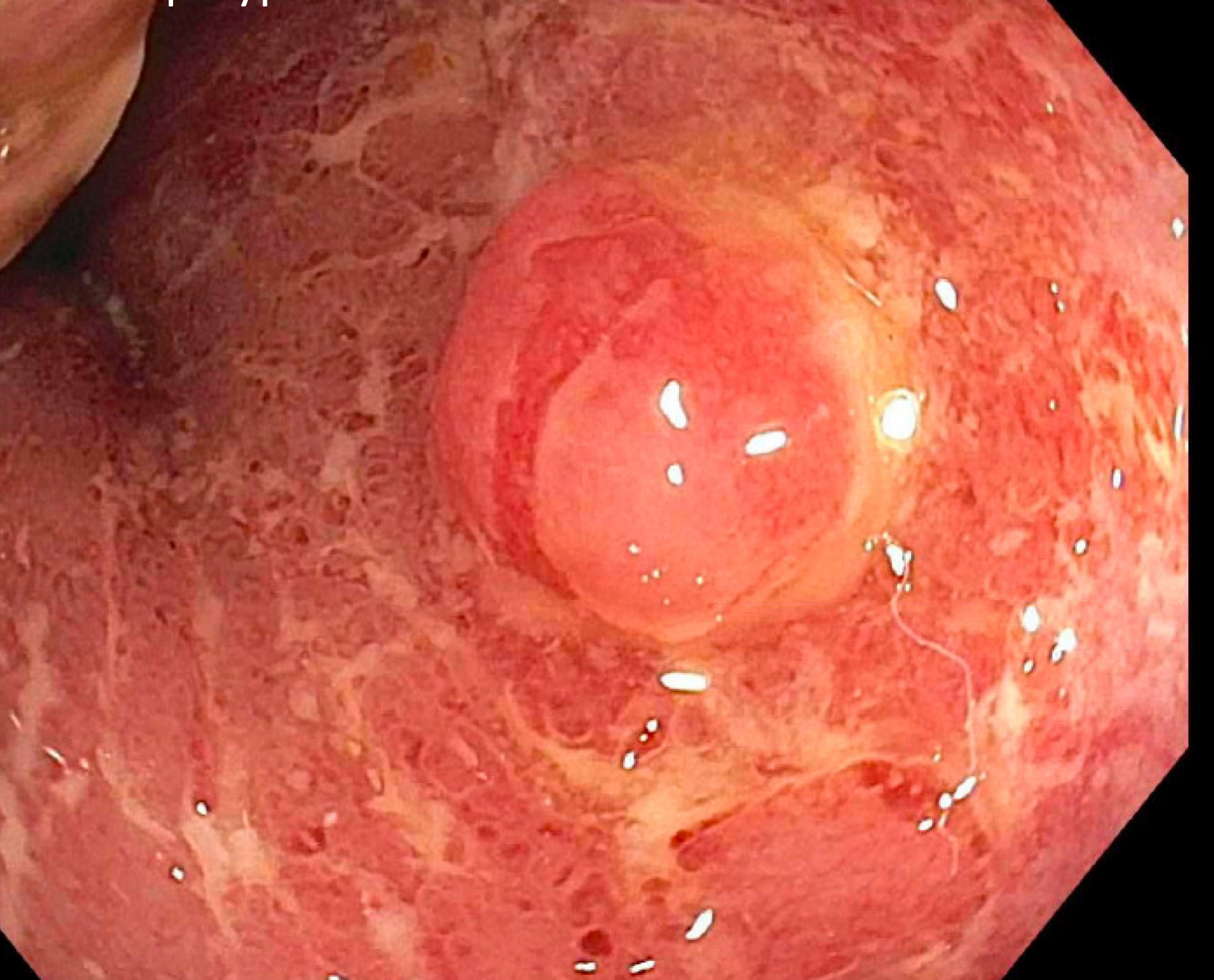


Figure 3. Rectal polyp



## Discussion:

**Rectal MALT lymphoma is a rare entity with unclear management options.**

- The limited treatment modalities for rectal MALT lymphoma in UC would include watchful waiting, surgical resection, endoscopic mucosal resection, radiation, and/or chemotherapy (rituximab, cyclophosphamide, vincristine, and prednisone or R-CHOP)<sup>2</sup>.
- H. pylori* infections, though strongly linked with *gastric* MALT lymphoma, have not been shown to be correlated with *rectal* MALT lymphoma<sup>4</sup>. There have been cases, however, demonstrating regression of the disease in response to *H. pylori* eradication therapy.<sup>3</sup>
- Given that patients with UC have chronic UC-associated colonic inflammation, lymphoma is often difficult to detect during colonoscopy. It is frequently masqueraded by ulcerations and pseudo-polyps<sup>5</sup>.
- More definitive treatments such as surgical resection could be warranted given that the recurrence of MALT lymphoma is difficult to distinguish from the existing chronic inflammation of the colonic mucosa.

## Conclusion:

Long term follow-up data is sparse and definitive management of rectal MALT lymphoma in UC patients remains a clinical conundrum. Thus, these patients require reliable high-quality long-term close follow-up utilizing a multidisciplinary approach.

## References:

- Adachi K, Ohtsuka H, Kozai Y. Primary Rectal Mucosa-Associated Lymphoid Tissue Lymphoma. Clin Gastroenterol Hepatol. 2016 May;14(5):e52-3. doi: 10.1016/j.cgh.2015.08.021. Epub 2015 Aug 22. PMID: 26305065.
- Zhou, TT., Wang, XL. & Liu, W. Primary Rectal Mucosa-Associated Lymphoid Tissue Lymphoma. J Gastrointest Surg 25, 2997–2998 (2021)
- Nakase H, Okazaki K, Ohana M, Ikeda K, Uchida K, Uose S, Itoh T, Iwano M, Watanabe N, Yazumi S, Kawanami C, Inoue F, Chiba T. The possible involvement of micro-organisms other than *Helicobacter pylori* in the development of rectal MALT lymphoma in *H. pylori*-negative patients. Endoscopy. 2002 Apr;34(4):343-6. doi: 10.1055/s-2002-23643. PMID: 11932795.
- Terada T. Extranodal marginal zone B-cell lymphoma of Mucosa-Associated Lymphoid Tissue (MALT lymphoma) in ulcerative colitis. Saudi J Gastroenterol. 2014 Sep-Oct;20(5):319-22. doi: 10.4103/1319-3767.141696. PMID: 25253369; PMCID: PMC4196349.
- Politis DS, Katsanos KH, Tsianos EV, Christodoulou DK. Pseudopolyps in inflammatory bowel diseases: Have we learned enough? World J Gastroenterol. 2017 Mar 7;23(9):1541-1551. doi: 10.3748/wjg.v23.i9.1541. PMID: 28321155; PMCID: PMC5340806.