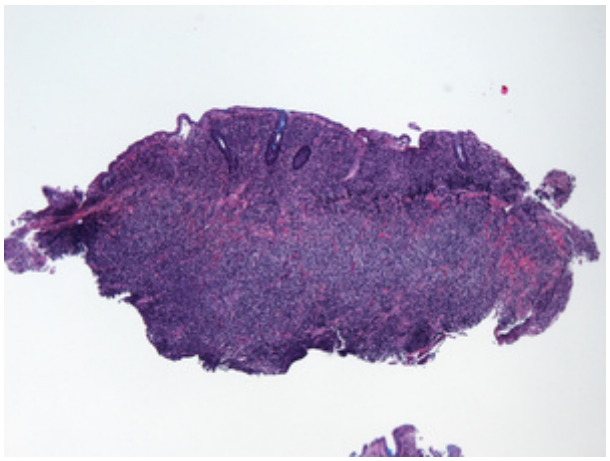
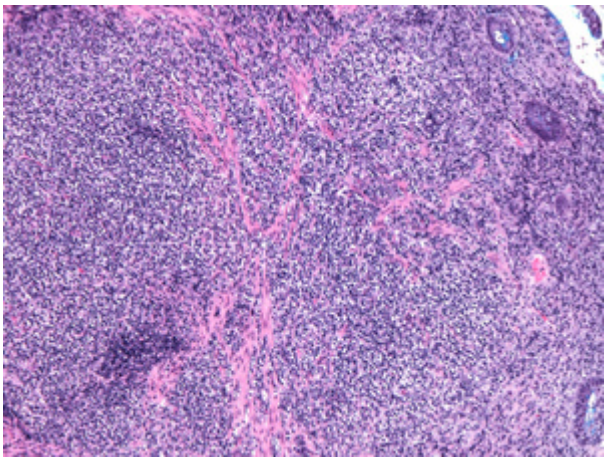
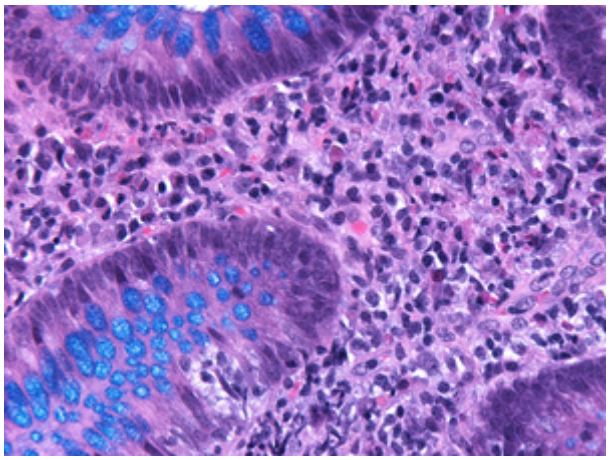
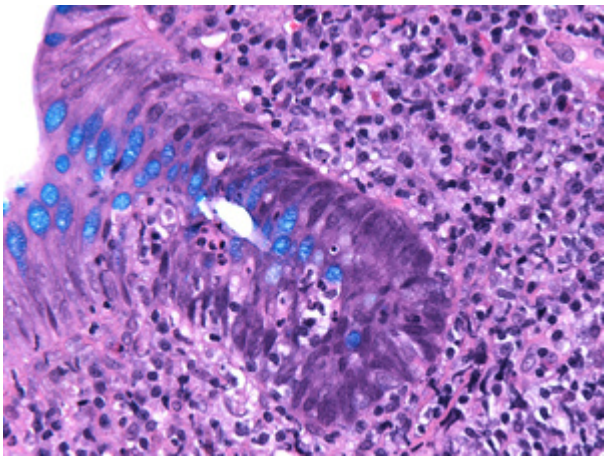
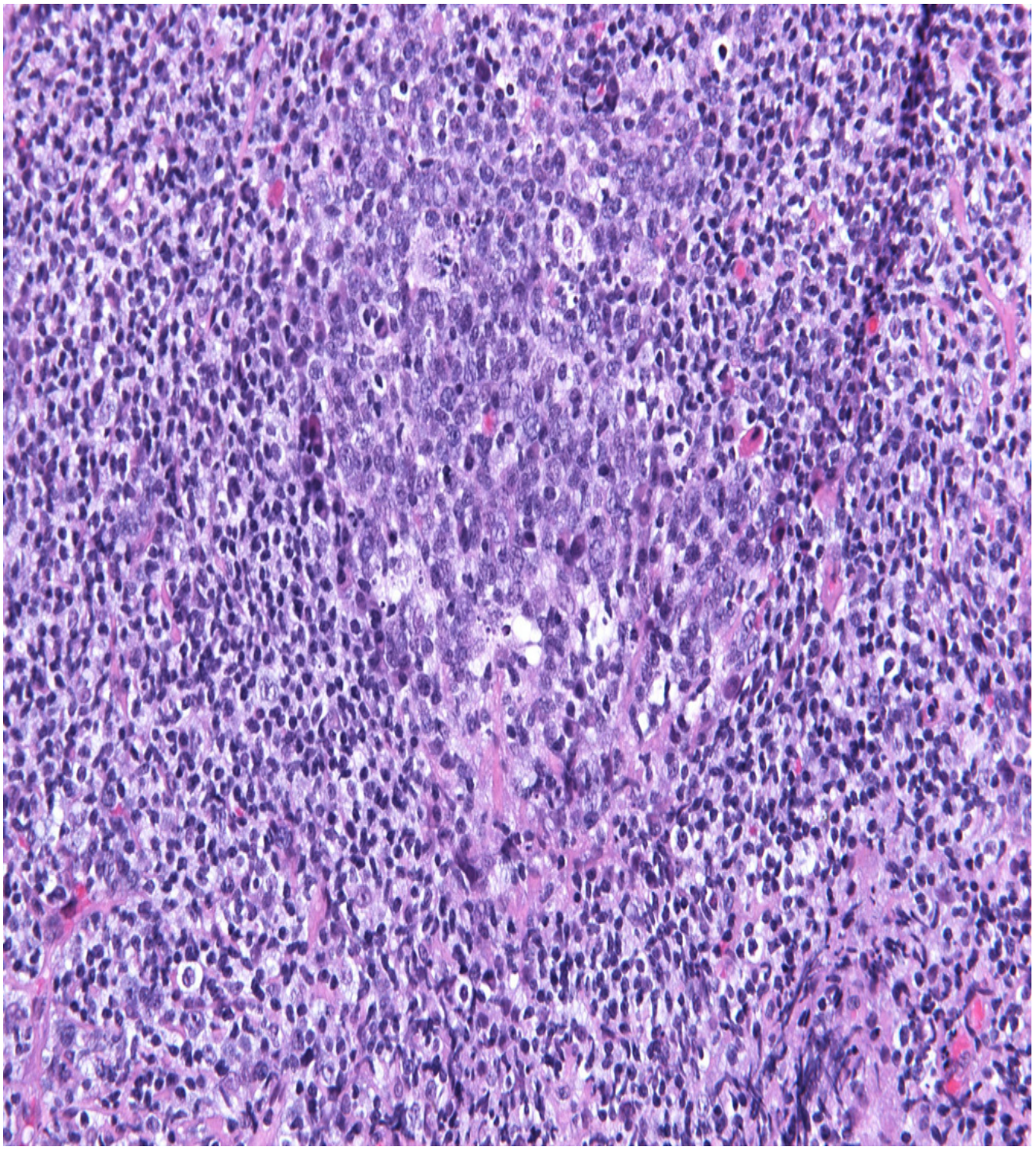
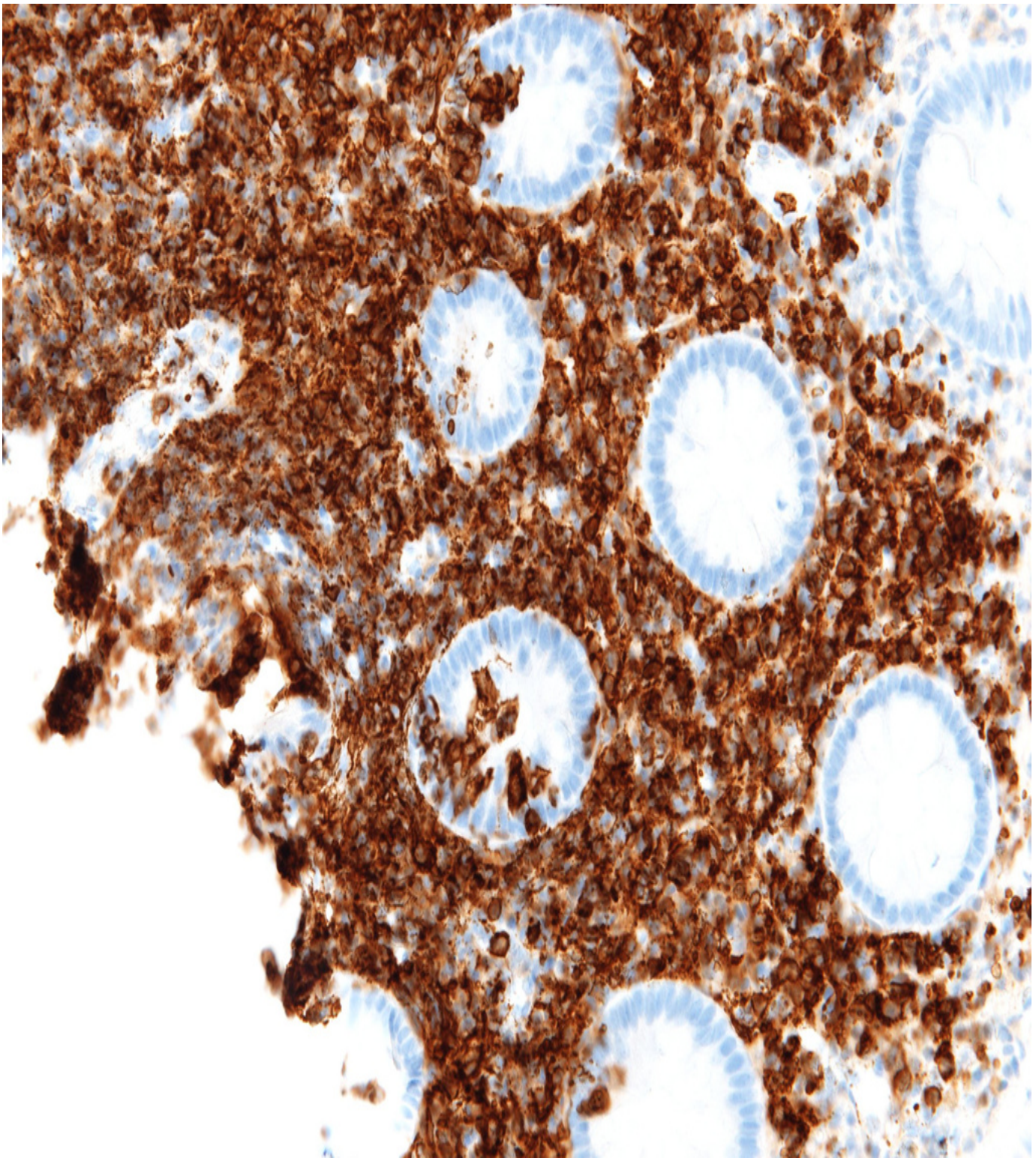
January 2015

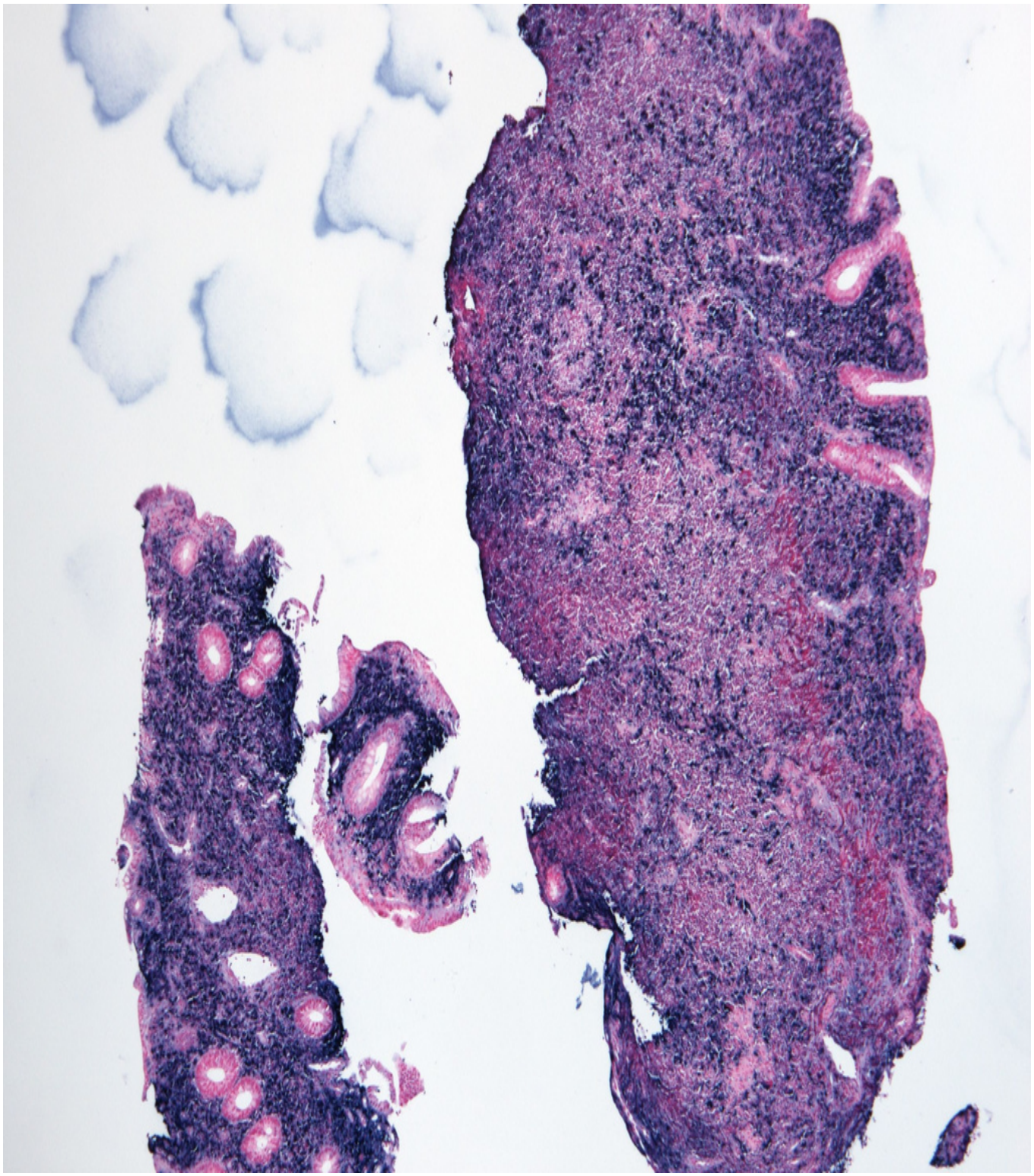
Colonoscopy in a 62-year-old male with a three month history of diarrhoea, abdominal pain and hematochezia. Clinical suspicion of inflammatory bowel disease (IBD). Elevated erythematous mucosa and aftous ulcerations in lower left colon.

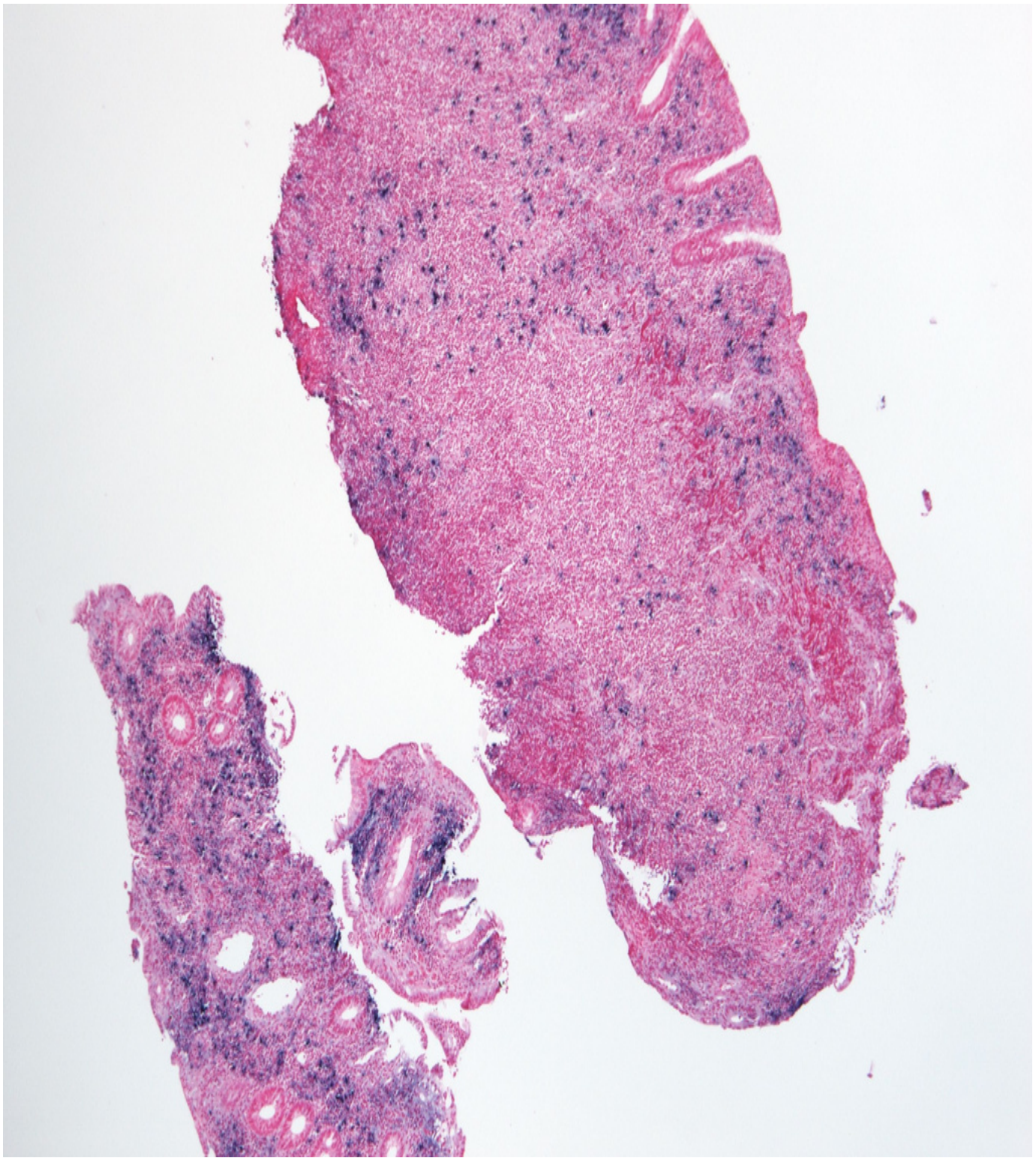
What is your diagnosis?









Diagnosis:

Intestinal MALT lymphoma (Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue).

Comment:

Mucosa with decreased crypt density shows intense lymphatic infiltration involving lamina propria, muscularis mucosae and submucosa (Panels A-B). Partial epithelial destruction is present, with lymphoepithelial lesions and focal cryptitis (Panels C-D). Small/medium sized atypical lymphatic cells are admixed with eosinophils and reactive germinal centers (Panel E). Atypical lymphatic cells are immunohistochemically CD20+ (Panel F), bcl-

2+ and CD43+, and negative for CD3, CD5, CD10, CD21, CD23, bcl-6 and cyclin D1. Plasmacytosis, pronounced in superficial lamina propria, shows kappa light chain restriction (Panel G kappa; Panel H lambda).

Intestinal lymphomas account for 0.3% of large intestinal malignancies and 3% of primary GIT lymphomas. MALT lymphomas of colorectum are very rare.

Diagnosis is based on morphology, immunophenotype, genetic and clinical features. Histologically, lymphoma cells infiltrate around reactive B-cell follicles (Panel E), in marginal zone distribution and eventually colonise follicles. Marginal zone cells resemble centrocytes, have relatively abundant pale cytoplasm (monocytoid appearance) and immunohistochemical characteristics of post-germinal centre B-cells. Lymphoepithelial lesions and plasmacytic differentiation are frequently observed. Transformation to diffuse large B-cell lymphoma may occur.

Differential diagnosis includes reactive lymphoid infiltration, other small B-cell lymphomas, and IBD, a clinical and histological mimic. Both entities may coexist or precede one another.

MALT lymphoma is considered an indolent disease, with distinct tendency of intraorgan and interorgan dissemination in late stages. Recurrences may involve many extranodal sites. Although primary intestinal MALT lymphomas exist, dissemination of (gastric) MALT lymphoma is diagnosed more frequently, and was also present in our case (not shown). Therefore active search for primary gastric lymphoma is always required during staging procedure. Successful regression of colorectal MALT lymphoma was described in some HP positive patients after HP eradication.

For further reading:

- Chen M, Semrad TJ, Wang J. Diagnosis of primary gastrointestinal lymphomas and mimics. Oncology - Theory & Practice. ISBN: 978-1-922227-80-5
- Holubar SD, Dozois EJ, Loftus EV Jr, et al. Primary intestinal lymphoma in patients with inflammatory bowel disease: a descriptive series from the prebiologic therapy era. Inflamm Bowel Dis. 2011;17:1557–1563.
- Isaacson PG, Chott A, Nakamura S, Muller-Hermelink HK, Harris NL, Swerdlow SH. Extranodal marginal zone lymphoma of mucossa-associated lymphoid tissue (MALT lymphoma). In: Swerdlow SH, Campo E, Harris NL, Jaffe ES, Pileri SA, Stein H, Thiele J, Wardiman JW, eds. WHO classification of tumours of haematopoietic and lymphoid tissue. 4th edn. Lyon: IARC, 2008; 214-9.
- Matsuo S, Mizuta Y, Hayashi T et al. Mucosa-associated lymphoid tissue lymphoma of the transverse colon: A case report. World J Gastroenterol. 2006; 12: 5573-6.
- Quayle FJ, Lowney JK. Colorectal lymphoma. Clin Colon Rectal Surg. 2006; 19: 49–53.
- Raderer M, Streubel B, Woehrer S et al. High relapse rate in patients with MALT lymphoma warrants lifelong follow-up.Clin Cancer Res. 2005; 11; 3349–3352.
- Shaheen S, Guddati AK. Secondary mucosa-associated lymphoid tissue (MALT) lymphoma of the colon. Med Oncol. 2013; 30: 502.
- Streubel B, Seitz G, Stolte M, Birner P, Chott A, Raderer M. MALT lymphoma associated genetic aberrations occur at different frequencies in primary and secondary intestinal MALT lymphomas. Gut. 2006; 55: 1581-5.

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