

Introduction

- Follicular lymphoma (FL) accounts for approximately 4% of gastrointestinal (GI) lymphomas.¹
- Of the follicular lymphomas found in the GI tract, duodenal-type follicular lymphoma (D-FL) make up 63-89% of cases.^{2,3}
- D-FL is often incidentally discovered, lacks invasion into deeper layers of the GI tract, and indolent in nature.⁴
- The Lugano staging classification is recommended for staging and prognosis. D-FL 5-year overall survival and progression free survival rate is 100% and >90%, respectively.^{3,5}
- Due to these unique characteristics, D-FL is recognized as a distinct variant of follicular lymphoma in the World Health Organization (WHO) classifications.^{5,6}
- Given the excellent prognosis and rare occurrence of progression to high stage FL or transformation to diffuse large B cell lymphoma, a “watch and wait” therapeutic approach is often taken.^{4,7,8,9}

Case Description

- A 39-year-old female with no pertinent past medical history presented with refractory dyspepsia. She denied any fevers, chills, night sweats, fatigue, weight loss, nausea, vomiting, diarrhea, melena, or hematochezia.
- Her endoscopy was notable for gastritis, duodenal atrophy, and three 5-mm pale nodules in the second part of the duodenum (Figures 1A-C).
- Initial biopsy of these polyps showed atypical lymphoid follicular aggregates. Outside pathologists’ subsequent review noted atypical lymphoid cells positive for CD10, BCL-2, BCL-6, and CD20 markers (Figures 2A, 2B), consistent with duodenal-type follicular lymphoma.
- Additional workup:
 - LDH, SPEP with immunofixation, HIV/HBV/HCV serologies were negative.
 - Full body CT and PET-CT showed no evidence of metabolically active lymphoma elsewhere in the body.
 - Antegrade small bowel enteroscopy to the mid-jejunum and colonoscopy were unremarkable.
- Per the Lugano staging system for gastrointestinal lymphomas, the patient was diagnosed with stage I (confined to the gastrointestinal tract) D-FL, and a “watch and wait” approach was recommended by Oncology.

Endoscopy

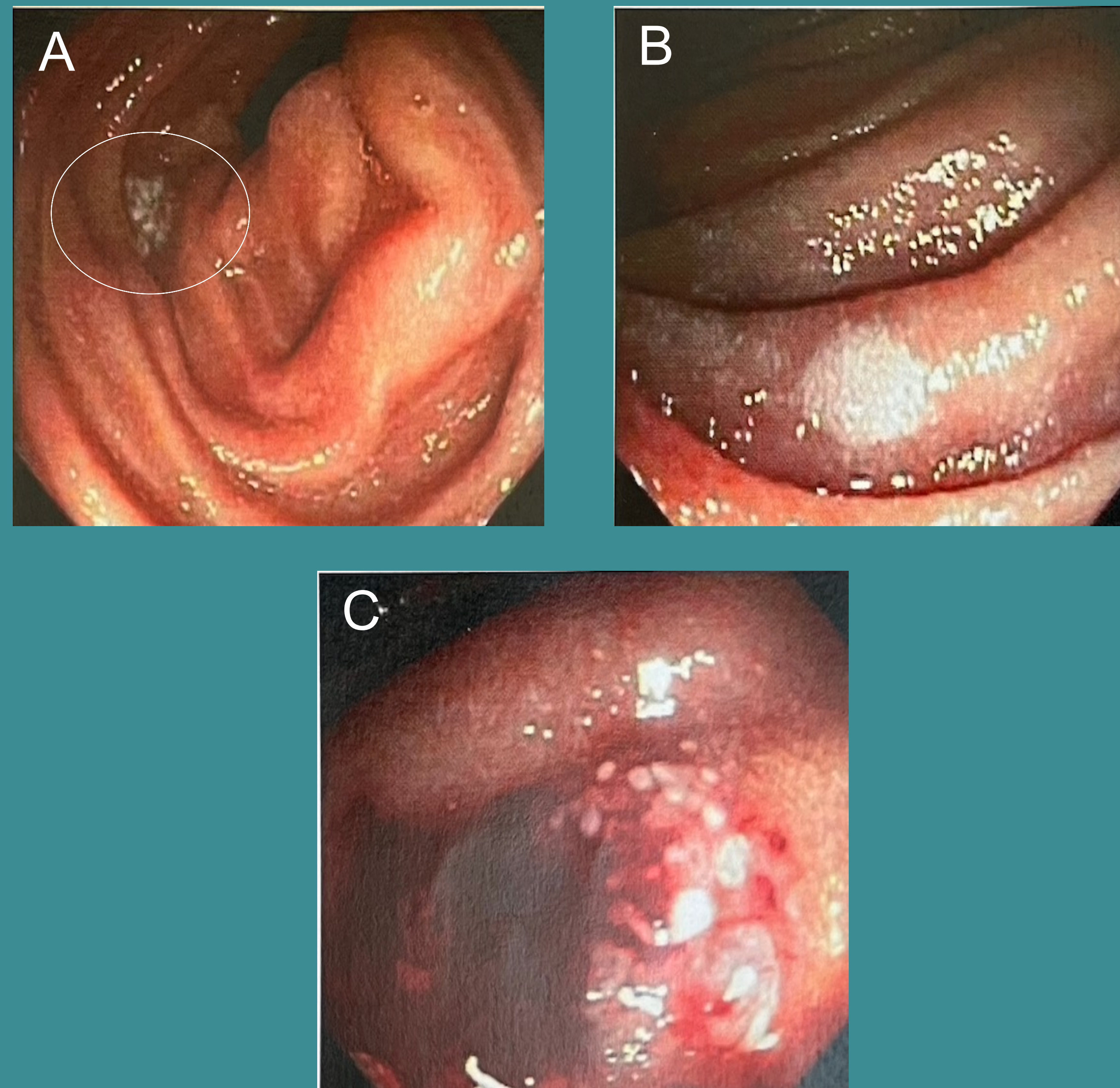


Figure 1A-1C: Endoscopic images of the duodenal second portion. Three distinct 5-mm pale nodules were seen and biopsies. First, second, and third lesions are seen in figure 1A, 1B, and 1C, respectively.

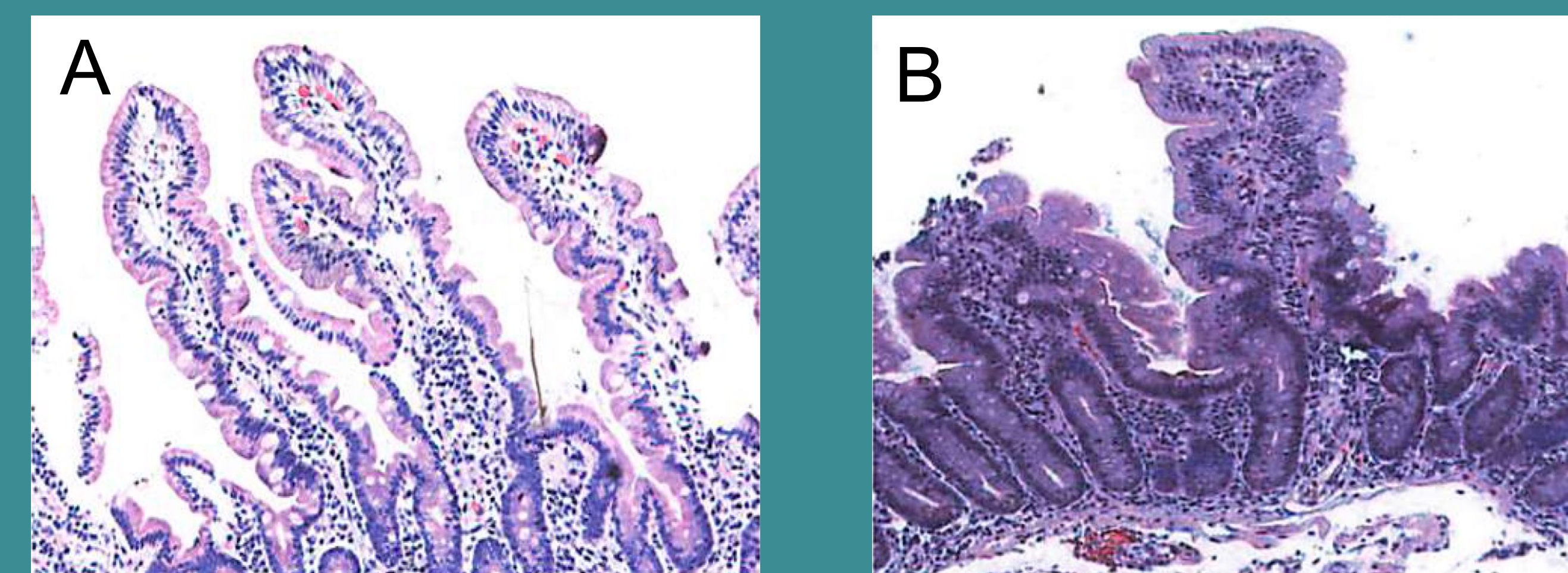


Figure 2A,2B: Duodenal biopsy include fragments with an atypical lymphoid infiltrate, comprised of a majority of small cells, without significant mitotic activity or cellular atypia. Immunohistochemical stains for CD10, BCL-2, BCL-6, and CD20 are positive in atypical follicles.

Discussion

- As in our case, most patients do not have symptoms attributable to D-FL and are incidentally diagnosed on EGD.
- Endoscopic Findings:
 - D-FL most often presents with multiple low-stage, white nodules in the second portion of the duodenum.³⁻⁴
 - Additional lesions are found in the remaining small bowel in 85% of D-FL cases, warranting thorough small bowel assessment with enteroscopy or capsule endoscopy.³
 - Gastric and/or colorectal involvement is exceedingly rare.^{3,4}
- Treatment options include “watch and wait”, chemotherapy, radiation, chemotherapy plus radiation, or surgical resection. However, there is no consensus on first-line treatment.
- Though radiation therapy and/or chemotherapy leads to higher rates of clinical remission, it does not result in a clinically significant reduction in progressive disease.^{4,7}
 - Radiation therapy is limited by the difficulty in identifying further involvement in the rest of the small bowel to effectively radiate all lesions.⁹
- Based on many retrospective case series, it appears “watch and wait” is a reasonable approach for asymptomatic D-FL without nodal or systemic spread.^{2,4,7-9}
- There is no evidence regarding the optimal surveillance interval for patients under the “watch and wait” treatment strategy.⁸
- Future studies with longer duration of follow up and variable surveillance strategies are needed to better understand the natural course of D-FL.

References

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