

A Presentation of a Follicular Lymphoma Incidentally Found in the Duodenum

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ABSTRACT

Duodenal lymphomas are considered a rare etiology in the GI tract and are found incidentally in around one out of every 5000 esophago-gastroduodenoscopies. These tumors are distinct from nodal follicular lymphomas but share some morphologic, immunochemical, and histological features. Further staging is performed via CT scan, PET scan for classification of the tumor burden of the disease. We present

a case report of a healthy 63 year old male who was found to have an indolent variant of the follicular lymphoma classified as stage 1E follicular lymphoma. This disease process is generally benign and usually confers a good overall prognosis. Follicular duodenal lymphomas were further classified in 2016 by the World health organization; as these tumors have increased in incidence due to advancing techniques in endoscopies and determining whether or not treatment options are needed for these tumors which have excellent outcomes. Our case highlights that a watchful wait approach can be a good option for patients who do not present with concerns of systemic disease.

Key words: Antiviral infection; Aloe-emodin; Epigenetic butyrate; case reports; *Kampo* medicine; Aloe vera juice ingestion

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INTRODUCTION

Duodenal lymphomas are considered to be a clinical variant of a follicular lymphoma that is confined to the small intestine. Most cases of primary follicular lymphoma involve the second portion of the duodenum and are incidentally found on endoscopy. Although, the risk factors of primary gastrointestinal follicular lymphoma are yet to be understood; they are generally benign tumors which remain remarkably indolent and usually do not develop into tumorous growth, rarely disseminates, or transform into high grade lesions^[1-4].

Patients are generally asymptomatic on presentation but may involve having certain chief complaints of abdominal pain, intestinal obstruction or diarrhea. The median age of discovery is noted to be around 56 years and previous case reports have noted to be equal prevalence in both males and females^[5]. On Endoscopic evaluation, the duodenal lymphoma may present as multiple white nodules or polyps measuring < 10-20 mm, while larger masses have been docu-

mented^[1]. The prevalence of duodenal lymphomas are considered to be higher in regions such as Japan, where there is a greater amount of surveillance due to increased amounts of peptic ulcer disease and gastric cancer^[6].

CASE PRESENTATION

We present a 63 yo caucasian male with PMHx significant for GERD, colon polyps on previous colonoscopy who presented to an outpatient office for evaluation of GERD and follow-up for concerns of possible polyps seen on a previous colonoscopy. Patient underwent a colonoscopy about 7 years ago with unknown findings. He was recommended to follow with a repeat colonoscopy 5 years later but did not do so. He reported having a family history of colo-rectal cancer that was diagnosed in his father in the mid-late 60s. In addition, the patient reported a history of GERD-like symptoms for the past ten years with intolerance to spicy/acidic foods a few times a week. He stated he did take omeprazole over the counter 20mg daily. Patient did not report any concerns of alarm symptoms of weight loss, odynophagia, dysphagia, nausea, vomiting, diarrhea, melena, hematochezia, melena or hematemesis.

On the physical exam, the patient's abdominal exam was benign with no concerns of tenderness or guarding noted. No additional blood work was performed on admission and the patient was scheduled for an EGD and colonoscopy for further evaluation of his ongo-

ing GERD flare up and to monitor for esophageal dysplasia. During endoscopic evaluation, a nodularity was noted in the second part of the duodenum that was biopsied with cold forceps and biopsies were performed in the antrum to rule out H. Pylori (Figure 3A). EGD findings were consistent with esophagitis, gastritis and an irregular Z line at the E/G junction. While his colonoscopy did reveal three polyps which were biopsied and only 2 were considered precancerous. He was diagnosed with Barrett's esophagus without dysplasia and was discharged on pantoprazole 40mg daily.

Pathology results from the nodularity in the second part of the duodenum was found to show small intestinal mucosa with few atypical lymphoid follicles and lymphoid aggregates within the mucosa and lamina propria (Figure 1A and 1B). The lymphoid follicles were composed of mainly small to medium sized lymphocytes with infrequent large cells. Immunohistological stains were performed and were found to be positive for CD20, CD10, and BCL2 (Figure 2A).

Morphological findings were consistent with follicular type B cell lymphoma. Fish testing for BCL2/IGH was performed and a BCL2/IGH gene rearrangement was detected. Further imaging studies to classify staging included a PET SCAN, which was found to have symmetric distribution involving the tonsils with no foci seen in the chest abdomen or pelvis. Patient was followed by an outpatient ENT who described the uptake of the PET scan as a physiologic response likely due to his history of chronic cryptic tonsillitis. Bone marrow biopsy was relatively benign and did not show any further signs of

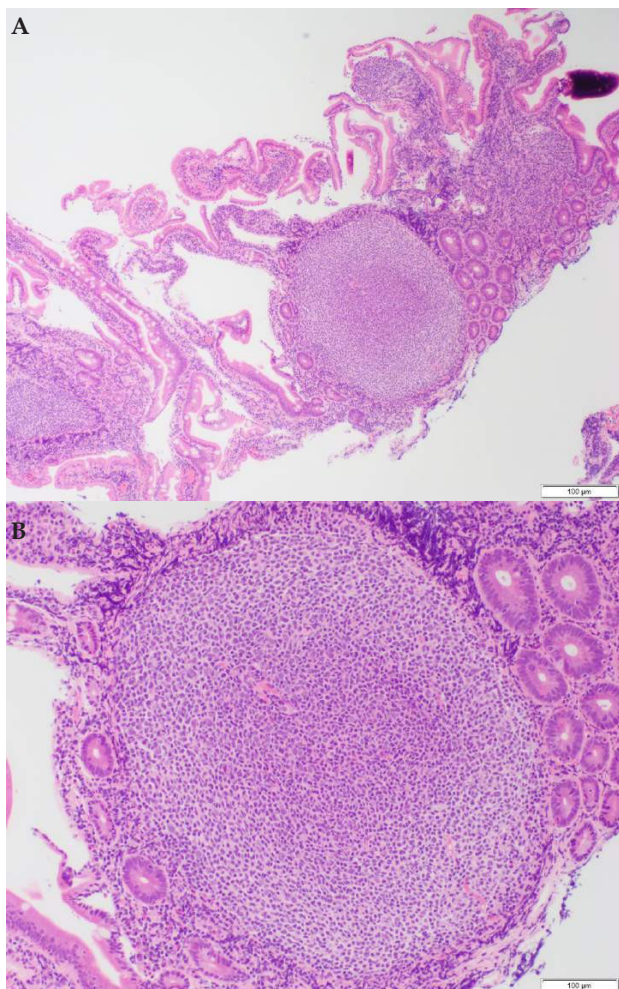


Figure 1 A Follicular lymphoma stained with hematoxylin and eosin (low power); B: Follicular lymphoma stained with hematoxylin and eosin (high power).

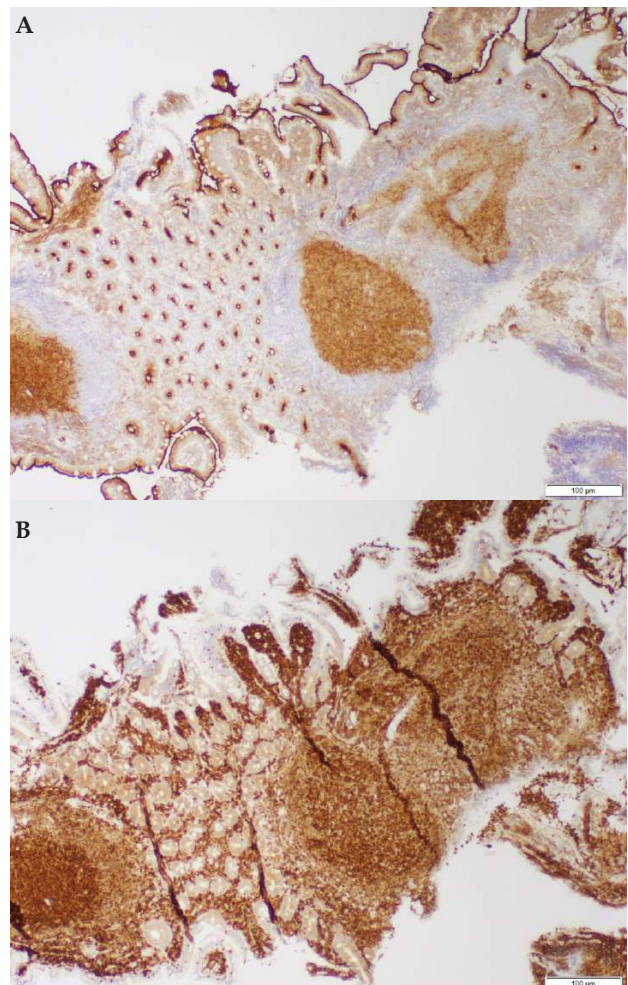


Figure 2 A Follicular lymphoma stained with antibody with CD10; B: Follicular lymphoma stained with antibody to Bcl-2.



Figure 3 EGD with nodularity seen at the second part of the duodenum.

leukemia/lymphoma.

Upon further discussion with his oncologist, the patient pursued active surveillance of his disease. Patient was classified as having stage 1E local regional follicular lymphoma and was rescheduled for an endoscopy in about 6 months with no need for chemotherapy or radiation at this time.

DISCUSSION

The gastrointestinal tract accounts for over 40% of extranodal lymphomas. Follicular lymphomas consist of only 1 to 3% of all primary GI lymphomas despite being the most common nodal lymphoma in the western population^[1]. These lymphomas reassemble nodal follicular lymphomas with regard to morphology and expression of typically germ cell markers such as CD10, CD38, and BCL-6. Genetically the hallmark of these tumors are considered to be positive for t(14;18)(q32;q21) which ends up leading to an overexpression of the BCL2 oncogene (see image 2B). The BCL2 oncogene contributes to the further development of a follicular lymphoma which interferes with apoptosis^[7]. In comparison, most primary cutaneous follicular lymphomas lack t(14;18)(q32;q21)^[1]. Generally, these tumors are considered to be low grade and are confined to the GI tract; they tend to have a low tendency to disseminate^[8]. Bende *et al.* demonstrated that GI-FLs expressed alpha4beta7 integrin; a well diagnosed mucosal homing receptor that made a distinguishable factor in assessing these tumors^[9]. Accurate immunochemistry and histological classification are paramount in determining treatment options of duodenal lymphomas^[5,9].

Duodenal follicular lymphomas are generally confined to the mucosa or submucosa^[7]. Treatment regimen for a primary follicular lymphoma usually involves four treatment strategies: watch and wait, radiation therapy, rituximab monotherapy, or chemotherapy with or without radiation. The largest case study involved in low-stage GI FL ever assembled included 56 patients who were followed for an average of 77 months. Of these patients, 24 chose the option to actively have surveillance, seven of those patients showed spontaneous complete regression and 17 had stable disease; radiotherapy resulted in complete regression in all 19 patients; anti- CD20 antibody mono-

therapy achieved complete regression in four patients and stable in one of these patients. The study further proved that none of these patients required surgery or died of the disease^[7].

In our particular case, the patient’s PET SCAN, bone marrow biopsy, and clinical history for any alarm factors was all negative with his diagnosis being a stage 1E follicular lymphoma; it was optimal to watchfully monitor his disease. Therefore, it was advised that the patient have active surveillance of his lesion and follow up within 6 months for a repeat endoscopy as these tumors can cause obstructive symptoms or rarely transform to malignancy. Although, watchful waiting is the standard of care frequent surveillance is needed and physicians should be inclined to closely follow these patients.

This case highlights the necessity for clinicians to consider the indolent course of follicular lymphomas and shows that the majority of these lymphomas are found incidentally. Since these tumors can turn malignant and cause symptoms. Further studies need to determine the overall relapse rate as only small case reports have been published. Our patient had a *de novo* formation of a follicular lymphoma. Therapy is generally not required unless significant symptoms are present or until disease progression^[5,9]. The long term clinical outcome for Gastrointestinal follicular lymphomas are still unclear and there is no analysis of a large population based registry of survival for patients due to the rarity of the disease^[8]. Majority of the data known from these tumors are case series reports and further studies still need to be conducted. Perhaps as endoscopy studies continue to advance with wireless capsule endoscopy and double-balloon enteroscopy more cases of gastrointestinal lymphomas will be found. The long-term clinical outcome of GI-FL remains unclear because there has been no analysis of a large population based registry of survival for patients with GI-FL^[8].

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