Clinical presentation and diagnosis of primary gastrointestinal lymphomas

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INTRODUCTION

The gastrointestinal (GI) tract is the predominant site of extranodal lymphoma involvement. Primary lymphomas of the GI tract are rare, while secondary GI involvement is relatively common. Despite their rarity, primary lymphomas of the GI tract are important since their evaluation, diagnosis, management, and prognosis are distinct from that of lymphoma at other sites and other cancers of the GI tract.

The definition of primary GI lymphoma has differed among authors, but typically refers to a lymphoma that predominantly involves any section of the GI tract from the oropharynx to the rectum [1,2]. While the disease typically involves a single primary site, multiple sites within the GI tract may be involved, as can local and distant lymph nodes. The vast majority are non-Hodgkin lymphomas (NHLs), although Hodgkin lymphoma has been reported [3,4].

GI lymphomas typically present with nonspecific signs and symptoms attributable to the site of involvement. This topic review will discuss the salient clinical features and diagnostic evaluation of GI lymphomas. The management of GI lymphoma is presented separately. (See "Treatment of extranodal marginal zone lymphoma of mucosa associated lymphoid tissue (MALT lymphoma)".)

OVERVIEW

Distribution — The GI tract is the predominant site of extranodal non-Hodgkin lymphomas (NHLs) [5]. Primary NHLs of the GI tract are rare, accounting for only 1 to 4 percent of malignancies arising in the stomach, small intestine, or colon [6]. In contrast, secondary GI involvement is relatively common, occurring in approximately 10 percent of patients with limited stage NHL at the time of diagnosis, and up to 60 percent of those dying from advanced NHL [5,7]. (See "Epidemiology, clinical features, and types of small bowel neoplasms".)

The two largest studies of GI lymphoma reported the following sites of involvement in Greek and German populations [8,9]:

- Stomach – 68 to 75 percent
- Small bowel (including duodenum) – 9 percent
- Ileo-cecal region – 7 percent
- More than one GI site – 6 to 13 percent
- Rectum – 2 percent
- Diffuse colonic involvement – 1 percent

However, the distribution of primary GI lymphomas varies among populations:

- In the United States (US), gastric lymphoma is the most common extranodal site of lymphoma. The vast majority of these lesions are either extranodal marginal zone B cell lymphoma of mucosa associated lymphoid tissue (MALT) type or diffuse large B cell lymphoma.

- Primary small intestinal lymphoma, while uncommon in Western countries, accounts for up to 75 percent of primary GI lymphomas in the Middle East and Mediterranean basin.

- The incidence of Burkitt lymphoma (BL) in Africa is approximately 50-fold higher than it is in the US. The classic GI presentation is that of an obstructing lesion in the terminal ileum. (See "Epidemiology, clinical manifestations, pathologic features, and diagnosis of Burkitt lymphoma".)

Predisposing conditions — Several conditions that predispose to GI lymphoma have been identified. These include:

- ***Helicobacter pylori* infection** — *H. pylori* infection is highly associated with the development of MALT lymphoma of the stomach and, to a lesser degree, other sites in the GI tract. (See "Association between Helicobacter pylori infection and gastrointestinal malignancy", section on 'Gastric lymphoma'.)

- **Autoimmune diseases** — A variety of autoimmune diseases, including rheumatoid arthritis, Sjögren's disease, systemic lupus erythematosus, and granulomatosis with polyangiitis, have been associated with an increased risk of lymphoma. Immunosuppressive therapy, rather than the disease itself, is thought to be responsible for the increased risk. (See "Clinical presentation and initial evaluation of non-Hodgkin lymphoma", section on 'Past medical history'.)

- **Immunodeficiency and immunosuppression** — Congenital immunodeficiency syndromes (eg, Wiskott-Aldrich syndrome, severe combined immunodeficiency syndrome, ataxia-telangiectasia, X-linked agammaglobulinemia) and acquired immunodeficiency (eg, HIV infection, iatrogenic immunosuppression) are associated with an increased incidence of B cell lymphoma. Lymphomas occurring in this setting tend to be aggressive and widespread at the time of diagnosis. Although most such patients have secondary GI tract involvement, primary involvement of the stomach and small bowel has been reported [10-16]. (See "HIV-related lymphomas: Epidemiology, risk factors, and pathobiology" and "HIV-related lymphomas: Clinical manifestations and diagnosis".)

- **Celiac disease** — Patients with gluten-sensitive enteropathy (celiac sprue) are at increased risk of developing enteropathy-associated T cell lymphoma (EATL) [17]. Population-based studies suggest that celiac disease is also associated with an increased risk of B cell lymphoma [18]. The extent to which coincident autoimmune or inflammatory disorders contribute to this risk is unknown [17,18]. (See "Clinical manifestations, pathologic features, and diagnosis of enteropathy-associated T cell lymphoma".)

  EATL is an uncommon sequela of enteropathy, occurring in approximately 5 percent of patients with celiac sprue followed for a 30-year period [19,20]. Ulcerative enteritis, another complication of long-standing celiac sprue, is probably a variant of EATL. (See "Management of celiac disease in adults", section on 'Ulcerative jejunitis and intestinal lymphoma'.)

- **Inflammatory bowel disease** — An association between inflammatory bowel disease (IBD) and lymphoma has been described in several reports, most of which were retrospective, small, and derived from tertiary referral centers, raising the possibility of referral bias. In contrast, large population-based studies have found relative risks ranging from 0.4 to 2.4 [4,21-27]. Considering the data in aggregate, they do not support an increased risk of lymphoma in patients with IBD compared with the general population [28].

Whether the risk of lymphoma is increased in patients with IBD treated with classic immunosuppressive agents (eg, azathioprine, 6-mercaptopurine) or tumor necrosis factor-alpha inhibitors (eg, etanercept, infliximab) remains an unresolved concern [29,30]. Although most studies published on immunosuppressive agents to date have found no increased risk, they lack sufficient power to provide conclusive data. A meta-analysis of six cohort studies suggested a fourfold increased risk of lymphoma in IBD patients treated azathioprine or 6-MP [31]. A possible association between tumor necrosis factor-alpha inhibitors and lymphoma is presented separately. (See "Tumor necrosis factor-alpha inhibitors: Risk of malignancy", section on 'Lymphoma risk'.)

- **Nodular lymphoid hyperplasia** — Nodular lymphoid hyperplasia, also known as follicular lymphoid hyperplasia, is a benign condition that has been implicated as a possible risk factor for primary lymphomas of the small intestine (picture 1). In children, nodular lymphoid hyperplasia tends to have a benign course and usually regresses spontaneously; in adults, however, the condition is often associated with immunodeficiency (eg, common variable immunodeficiency or selective IgA deficiency) and giardiasis, and the prognosis is less certain. Curiously, although the association between nodular lymphoid hyperplasia with humoral immunodeficiency and primary intestinal lymphoma remains controversial, the evidence is stronger for an association of nodular lymphoid hyperplasia with GI lymphoma in the absence of immunodeficiency [32-37].

ESOPHAGEAL LYMPHOMA

Primary esophageal lymphoma is very rare, accounting for less than 1 percent of primary GI lymphomas. More commonly, lymphoma may involve the esophagus as an extension of mediastinal or gastric involvement. Only case reports and series of primary esophageal lymphoma have been reported in the literature [38]. Primary esophageal lymphoma appears to more commonly involve the distal esophagus. Most patients are asymptomatic or present with complaints of dysphagia or odynophagia. There is a diverse appearance on imaging and the diagnosis is made by endoscopic biopsy in most cases [39].

GASTRIC LYMPHOMA

Epidemiology — The stomach is the most common extranodal site of lymphoma and accounts for 68 to 75 percent of GI lymphomas [8,9]. Primary gastric lymphoma accounts for 3 percent of gastric neoplasms and 10 percent of lymphomas [40]. Gastric lymphoma reaches its peak incidence between the ages of 50 to 60 years. There is a slight male predominance.

Clinical features — Patients with gastric lymphoma typically present with nonspecific symptoms frequently seen with more common gastric conditions, such as peptic ulcer disease, gastric adenocarcinoma, and nonulcer dyspepsia. The most common presenting symptoms include (table 1) [8,41-44]:

- Epigastric pain or discomfort (78 to 93 percent)
- Anorexia (47 percent)
- Weight loss (25 percent)
- Nausea and/or vomiting (18 percent)
- Occult gastrointestinal bleeding (19 percent)
- Early satiety

Systemic B symptoms (fever, night sweats) are seen in 12 percent of patients. Weight loss is frequently due to local compromise of GI structures and is not always considered a B symptom in this setting. Hematemesis and melena are uncommon. The duration of symptoms preceding the diagnosis is quite variable, ranging from a few days to six years.

The physical examination is often normal but may reveal a palpable mass and/or peripheral lymphadenopathy when the disease is advanced. Laboratory studies also tend to be normal at presentation. Anemia or an elevated erythrocyte sedimentation rate may be present in selected cases [41-43].

Diagnostic evaluation — The diagnosis of gastric lymphoma is usually established during upper endoscopy with biopsy. Laparotomy and laparoscopy are typically reserved for patients with complications such as perforation or obstruction. (See "Treatment of extranodal marginal zone lymphoma of mucosa associated lymphoid tissue (MALT lymphoma)".)

Findings on upper endoscopy are diverse and may include any of the following [45,46]:

- Mucosal erythema (picture 2)
- A mass or polypoid lesion with or without ulceration
- Benign-appearing gastric ulcer (picture 3)
- Nodularity
- Thickened, cerebroid gastric folds (picture 4A-B)

Multiple biopsies should be obtained from the stomach, duodenum, gastroesophageal junction and from abnormal appearing lesions. An endoscopic ultrasound should determine the depth of invasion and the presence of perigastric nodes [47-51].

The pattern seen on endoscopic ultrasound (EUS) may correlate with the type of lymphoma that is present. In one series, for example, superficial spreading or diffuse infiltrating lesions on EUS were seen with MALT lymphoma, while mass-forming lesions were typical of diffuse large B cell lymphoma [50].

Pathologic evaluation is required for the determination of lymph node involvement. EUS alone has suboptimal accuracy in distinguishing benign from malignant lymph nodes [47-50]. When combined with endoscopic biopsy, however, overall accuracy approaches 90 percent (versus 66 percent for EUS alone) [52]. Even higher accuracy rates may be achievable if flow cytometry is performed [53]. Thus, caution is warranted in the interpretation of findings using EUS or CT alone.

Pathology — The diagnosis of gastric lymphoma may be suggested by endoscopic and imaging findings but must be confirmed by biopsy. Both suspicious-appearing lesions and normal-appearing mucosa should be biopsied since gastric lymphoma can occasionally present as multifocal disease with involvement of tissue that appears to be unaffected on initial visualization [54]. (See "Clinical presentation and initial evaluation of non-Hodgkin lymphoma" and "Clinical features, diagnosis, and staging of gastric cancer", section on 'Upper endoscopy with biopsy'.)

Endoscopists should aim to attain the largest biopsy specimen possible. Conventional pinch biopsies may miss the diagnosis, since gastric lymphoma can infiltrate the submucosa without affecting the mucosa; this problem is most likely to occur when no obvious mass is present. Jumbo biopsies, snare biopsies, biopsies within biopsies ("well technique"), and needle aspiration can all serve to increase the yield in such cases. EUS-guided fine needle aspiration biopsy (FNAB) [55-57] or endoscopic submucosal resection [58] may provide even greater diagnostic capability.

The vast majority (greater than 90 percent) of gastric lymphomas are approximately equally divided into two histologic subtypes [8,9]:

- Extranodal marginal zone B cell lymphoma of mucosa (gut)-associated lymphoid tissue (MALT) type (previously called MALToma, MALT-type lymphoma, or MALT lymphoma) (38 to 48 percent) (table 2). (See "Clinical manifestations, pathologic features, and diagnosis of extranodal marginal zone lymphoma of mucosa associated lymphoid tissue (MALT)".)

- Diffuse large B cell lymphoma (45 to 59 percent). (See "Epidemiology, clinical manifestations, pathologic features, and diagnosis of diffuse large B cell lymphoma".)

The remaining cases of gastric lymphoma may represent any histology, but the most commonly seen are [8,9]:

- Mantle cell lymphoma (1 percent). (See "Mantle cell lymphoma: Epidemiology, pathobiology, clinical manifestations, diagnosis, and prognosis".)
- Follicular lymphoma (0.5 to 2 percent). (See "Clinical manifestations, pathologic features, diagnosis, and prognosis of follicular lymphoma".)
- Peripheral T cell lymphoma (1.5 to 4 percent). (See "Clinical manifestations, pathologic features, and diagnosis of peripheral T cell lymphoma, not otherwise specified".)

LYMPHOMA OF THE SMALL INTESTINE

Approximately 30 percent of GI lymphomas occur in the small intestine. These lymphomas may be broadly categorized into three main groups:

- Immunoproliferative small intestinal disease (IPSID, also called alpha heavy chain disease, Mediterranean lymphoma, Seligmann disease) lymphoma is a variant of extranodal marginal zone lymphoma of MALT which secretes alpha heavy chains. (See "The heavy chain diseases", section on 'Alpha HCD'.)
- Enteropathy-associated T cell lymphoma (EATL), also called intestinal T cell lymphoma, is a tumor that is highly associated with gluten-sensitive enteropathy. (See "Clinical manifestations, pathologic features, and diagnosis of enteropathy-associated T cell lymphoma".)
- Other western-type non-IPSID lymphomas (eg, diffuse large B cell lymphoma, mantle cell lymphoma, Burkitt lymphoma, follicular lymphoma [59]).

Epidemiology — The epidemiologic features of small intestine lymphomas differ with the population studied:

- In the Middle East and Mediterranean basin, primary small intestinal lymphoma, usually of the IPSID type, accounts for up to 75 percent of primary GI lymphomas. There is a male predominance with a median age at presentation of 25 years [60]. Low socioeconomic status, poor sanitation, and high rates of endemic parasitic infestation and infantile infectious enteritis are common among high-risk populations. Genetic factors (eg, linkage with HLA-Aw19, -B12, -A9 haplotypes), as well as Campylobacter infection, have also been implicated. (See "Clinical manifestations, pathologic features, and diagnosis of extranodal

marginal zone lymphoma of mucosa associated lymphoid tissue (MALT)", section on 'Campylobacter jejuni'.)

- Although uncommon, EATL is most common in areas with a high incidence of gluten-sensitive enteropathy (celiac sprue), such as the Western part of Ireland and Northern Europe [61]. EATL is most commonly found in adult males with a median age at diagnosis in the sixth decade [19,20,61]. Ulcerative enteritis, another complication of long-standing celiac sprue, is probably a variant of EATL. (See "Clinical manifestations, pathologic features, and diagnosis of enteropathy-associated T cell lymphoma", section on 'Epidemiology' and "Epidemiology, pathogenesis, and clinical manifestations of celiac disease in adults".)

- There is a small absolute increase in the incidence of intestinal lymphoma among patients with inflammatory bowel disease (eg, Crohn colitis, ulcerative colitis), especially those receiving immunosuppressive agents [62,63]. The majority of these lymphomas are of B cell origin and many demonstrate infection with Epstein-Barr virus [64]. (See "Clinical manifestations, diagnosis, and prognosis of Crohn disease in adults", section on 'Cancer risk'.)

- In industrialized nations, small intestinal lymphomas are rare, not of the IPSID type, and typically occur in middle-aged men.

Clinical features — The clinical presentation of patients with lymphoma of the small intestine differs according to the histologic tumor type. The duration of symptoms prior to diagnosis ranges from a few days to a few years. The different features of IPSID and non-IPSID-associated lymphoma are summarized in the table (table 3). Abdominal pain is a frequent symptom of all types, occurring in approximately two-thirds of patients [60].

- Patients with IPSID typically present with abdominal pain, chronic diarrhea, malabsorption, severe weight loss, clubbing, and ankle edema. Unusual manifestations include lactose intolerance [65], enteroenteric fistulae [66], ascites, fever, hypocalcemia, steatorrhea, and organomegaly [67].

- Patients with EATL often present with acute bleeding, obstruction, or perforation [19,20]. Clinical deterioration of celiac disease, despite compliance with a gluten-free diet, should raise suspicion of the possible presence of lymphoma. Conversely, since celiac disease may be undiagnosed at the time of presentation of the intestinal lymphoma, it has been suggested that patients with a T cell lymphoma and/or a gut primary localization should be tested for the presence of underlying celiac disease [18]. (See "Clinical manifestations,

pathologic features, and diagnosis of enteropathy-associated T cell lymphoma", section on 'Clinical features'.)

- Patients with other non-IPSID lymphomas have a more nonspecific presentation, which may include abdominal pain, GI bleeding, intestinal obstruction or perforation, obstructive jaundice, and/or a palpable abdominal mass [59,60,68]. In contrast, patients with primary intestinal follicular lymphoma are often diagnosed during endoscopy performed for symptoms unrelated to the lymphoma [69].

Diagnostic evaluation — The diagnostic evaluation of a suspected lymphoma of the small intestine may include a contrast-enhanced computed tomography (CT), positron emission tomography (PET), contrast radiography, conventional endoscopy, and capsule endoscopy. CT and/or contrast radiography are usually the initial diagnostic modalities performed. Diagnostic laparotomy with resection of the involved bowel is appropriate if there is obstruction, perforation, or major bleeding at presentation. (See "Treatment of extranodal marginal zone lymphoma of mucosa associated lymphoid tissue (MALT lymphoma)".)

Computed tomography — Findings on CT are varied and may include the following [39,70]:

- Multiple, large tumors
- Bowel segments with lumen that is narrowed, enlarged, or both
- Mesenteric nodal masses resulting in the "hamburger" or "sandwich" sign in which nodal masses (the bun) surround the mesenteric vessels (the meat of the hamburger)

Of particular concern are bowel wall segments with homogenous thickening, greater than 2 cm, with a normal or enlarged lumen. The presence of any of the above findings necessitates further evaluation and biopsy.

Positron emission tomography — Findings on PET in patients with lymphoma of the small intestine vary depending upon the pathologic subtype. In general, PET scanning detects clinically aggressive non-Hodgkin lymphoma variants as well as most Hodgkin lymphoma subtypes, while its overall usefulness in the clinically indolent lymphomas remains unclear. The use of PET in the staging of lymphoma is discussed in more detail separately. (See "Pretreatment evaluation and staging of non-Hodgkin lymphomas", section on 'Positron emission tomography (PET)'.)

Of the histologies most commonly seen in the small intestine, diffuse large B cell lymphoma (DLBCL), mantle cell lymphoma (MCL), Burkitt lymphoma (BL), and enteropathy associated T cell lymphoma (EATL) are typically fluorodeoxyglucose (FDG)-avid [71]. In contrast, marginal zone lymphoma (MZL) and follicular lymphoma (FL) have variable FDG-avidity.

There is a paucity of data regarding the use of PET in the evaluation of patients with small bowel lymphoma in particular. Importantly, interpretation of FDG-avidity in the GI tract may be complicated by the presence of FDG activity reflecting normal metabolic activity in the bowel or increased FDG activity related to inflammatory conditions (eg, Crohn disease), infections, or other bowel pathology [72]. The following studies have addressed the use of PET in patients with GI lymphoma:

- In a retrospective study of 33 patients with GI tract lymphoma, six of which were in the small bowel, focal nodular or diffuse hypermetabolic activity was universally seen in the GI tract of patients with biopsy-proven DLBCL (16 patients), MCL (6 patients), and peripheral T cell lymphoma (3 patients) [73]. In contrast, FDG activity was variable in patients with MZL (5 patients), FL (2 patients), and small lymphocytic lymphoma (1 patient).

- In a prospective study, patients with biopsy-proven extranodal MZL underwent PET scans as part of their initial staging studies [74]. The study was closed early when none of the first 10 consecutive patients demonstrated FDG activity.

We typically incorporate PET imaging into the pretreatment evaluation of patients with DLBCL of the GI tract. The use of PET in the pretreatment evaluation of patients with other GI lymphoma subtypes is controversial.

Endoscopy — Although technically difficult, endoscopic evaluation of the small bowel with biopsy of lesions can be diagnostic. Endoscopic approaches depending on the site of involvement. Proximal small bowel lesions may be detected by "push" enteroscopy, while lesions in the distal small bowel may be assessed with intubation of the terminal ileum during colonoscopy.

Capsule endoscopy is another useful technique for evaluating the small bowel in patients with suggestive clinical presentations or suspicious radiographic findings. Unlike other endoscopic approaches, however, capsule endoscopy does not permit tissue sampling [75]. (See "Wireless video capsule endoscopy".)

Different histologic variants of lymphoma present with typical findings on endoscopy:

- Patients with EATL of the jejunum typically demonstrate large circumferential ulcers without overt tumor masses. Biopsies of the involved mucosa demonstrate lymphoma, while biopsies of the normal-appearing mucosa usually show villous atrophy characteristic of celiac disease.

- Patients with MCL may demonstrate typical small nodular or polypoid tumors (2 mm to more than 2 cm in size), with or without normal intervening mucosa referred to as "lymphomatous polyposis" (picture 5).

- Patients with primary intestinal FL most commonly present with multiple small (1 to 5 mm) polypoid lesions in the descending part of the duodenum, with some cases demonstrating clustering around the ampulla of Vater [69]. The lesions are solitary in approximately 15 percent of cases and may grossly resemble an adenoma. Other segments of the small bowel (eg, jejunum and/or ileum) are involved in approximately 17 percent of cases.

Laparotomy — Exploratory laparotomy should be performed when the lesion is not accessible via endoscopy or when endoscopic biopsies are unavailable or non-diagnostic [19,20]. Obstructing lesions also require laparotomy.

Laboratory studies — Laboratory studies are typically normal in patients with small bowel lymphoma with the exception of alpha heavy chain paraproteinemia in IPSID-associated disease. Alpha heavy chain paraproteinemia (alpha heavy chain disease) is present in up to 70 percent of IPSID-associated lymphomas [60]. The paraproteinemia may diminish or disappear as IPSID progresses from an early prelymphomatous stage to frank lymphoma [76]. (See "The heavy chain diseases", section on 'Alpha HCD'.)

Pathology — The diagnosis of small intestinal lymphoma is dependent on the pathological review of an adequate biopsy specimen. The main histologic subtypes are:

- Immunoproliferative small intestinal disease lymphoma (IPSID), which is a variant of extranodal marginal zone lymphoma of MALT, also called alpha heavy chain disease, Mediterranean lymphoma, Seligmann disease). (See "The heavy chain diseases", section on 'Alpha HCD'.)

- Enteropathy-associated T cell lymphoma (EATL), also called intestinal T cell lymphoma. (See "Clinical manifestations, pathologic features, and diagnosis of enteropathy-associated T cell lymphoma".)

- Other western-type non-IPSID lymphomas:

  - Diffuse large B cell lymphoma (see "Epidemiology, clinical manifestations, pathologic features, and diagnosis of diffuse large B cell lymphoma", section on 'Diagnosis')

  - Mantle cell lymphoma (see "Mantle cell lymphoma: Epidemiology, pathobiology, clinical manifestations, diagnosis, and prognosis")

- Burkitt lymphoma (see "Epidemiology, clinical manifestations, pathologic features, and diagnosis of Burkitt lymphoma")

- Follicular lymphoma (see "Clinical manifestations, pathologic features, diagnosis, and prognosis of follicular lymphoma", section on 'Duodenal-type FL')

COLORECTAL LYMPHOMA

Epidemiology — Colorectal lymphoma is uncommon, accounting for approximately 3 percent of the GI lymphomas and 0.3 percent of large intestinal malignancies [8,9]. There is a male predominance.

Clinical presentation — Patients with colorectal lymphoma can present with abdominal pain, overt or occult bleeding, diarrhea, intussusception (image 1A-B), or rarely, bowel obstruction (table 1) [2,8,42,77-81].

Endemic Burkitt lymphoma is a disease of childhood with a peak incidence at about eight years of age. GI manifestations are infrequent but may include obstruction or intussusception. In contrast, sporadic Burkitt lymphoma exhibits a wider age distribution, with only 50 percent of cases affecting children. It often presents with abdominal pain (picture 6) and obstructive symptoms caused by ileocecal involvement. (See "Epidemiology, clinical manifestations, pathologic features, and diagnosis of Burkitt lymphoma".)

Diagnostic evaluation — Colonoscopy with biopsy is the principal diagnostic modality for colorectal lymphomas. Colonoscopic findings may include diffuse mucosal nodularity, colitis-like changes with induration and ulceration, or a mass with or without ulceration (picture 7A-B) [82]. Multiple biopsies of all lesions should be performed using standard techniques. Computed tomography (CT) and barium enema may suggest the diagnosis with areas of focal mucosal nodularity (image 2), polypoid masses (image 3), mucosal fold thickening, extracolonic masses, or circumferential thickening (image 4 and image 5) [39,83].

Typical small nodular or polypoid tumors (2 mm to more than 2 cm in size), with or without normal intervening mucosa, can be seen by colonoscopy (picture 5) or enteroscopy in mantle cell lymphoma. Locoregional mesenteric lymphadenopathy is often present at the time of diagnosis, and involvement of liver, spleen, bone marrow, and peripheral nodes may be evident early in the course of the disease.

Pathology — The diagnosis of colorectal lymphoma is dependent on the histologic evaluation of an adequate biopsy specimen. The most common histologies seen in this region include:

- Mantle cell lymphoma (see "Mantle cell lymphoma: Epidemiology, pathobiology, clinical manifestations, diagnosis, and prognosis")
- Burkitt lymphoma (see "Epidemiology, clinical manifestations, pathologic features, and diagnosis of Burkitt lymphoma")
- Follicular lymphoma (see "Clinical manifestations, pathologic features, diagnosis, and prognosis of follicular lymphoma")
- Diffuse large B cell lymphoma (see "Epidemiology, clinical manifestations, pathologic features, and diagnosis of diffuse large B cell lymphoma", section on 'Diagnosis')

SUMMARY

- **Description** – The gastrointestinal (GI) tract is the most common site of primary extranodal lymphoma. Nearly all cases of primary GI lymphoma are non-Hodgkin lymphomas (NHL).
- **Distribution** – Primary GI lymphomas can involve any portion of the GI tract. In addition to a primary site, there may be involvement of other GI tract sites and/or local or distant lymph nodes. (See 'Distribution' above.)
- **Predisposing factors** – Predisposing conditions include *Helicobacter pylori* infection, autoimmune diseases, immunodeficiency/immunosuppression, celiac disease, inflammatory bowel disease, and nodular lymphoid hyperplasia. (See 'Predisposing conditions' above.)
- **Presentation** – Symptoms, evaluation, and pathology vary by disease and site:
  - **Stomach** – The most common primary GI site, which often presents non-specifically (eg, epigastric pain/discomfort, anorexia, early satiety, weight loss, nausea/vomiting, occult GI bleeding) and is usually diagnosed via upper endoscopy with biopsy. Nearly all cases are either extranodal marginal zone lymphoma of mucosa associated lymphoid tissue (MALT) or diffuse large B cell lymphoma. (See 'Gastric lymphoma' above.)
  - **Small intestine** – Second most common GI site, with symptoms, presentation, and diagnosis that may vary according to associated conditions, such as immunoproliferative small intestinal disease (IPSID; a variant of MALT lymphoma), celiac disease (enteropathy-associated T cell lymphoma [EATL]), or neither (table 3). Diagnosis may be suggested on computed tomography but requires biopsy

confirmation. Most common histologic types include MALT/IPSID-related lymphoma, EATL, diffuse large B cell lymphoma (DLBCL), mantle cell lymphoma, Burkitt lymphoma, and follicular lymphoma. (See 'Lymphoma of the small intestine' above.)

- **Colorectal** – Uncommon site that may present with abdominal pain, overt or occult bleeding, diarrhea, intussusception, or bowel obstruction, generally diagnosed by colonoscopy with biopsy. Most common diagnoses are DLBCL, mantle cell lymphoma, and Burkitt lymphoma. (See 'Colorectal lymphoma' above.)

- **Esophageal** – Least common GI site, usually involving the distal esophagus, with asymptomatic presentation or dysphagia/odynophagia. Diverse appearance on imaging and diagnosis made by endoscopic biopsy. (See 'Esophageal lymphoma' above.)

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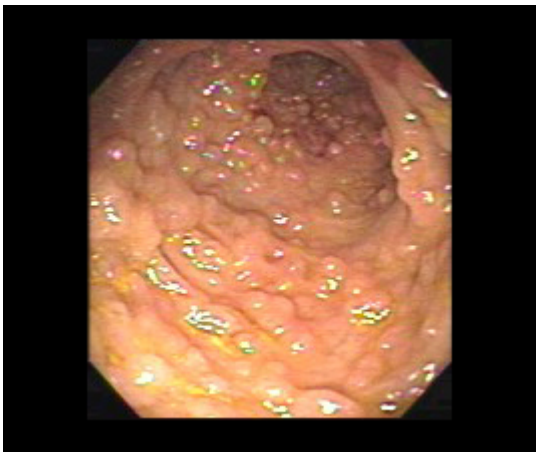
Topic 4743 Version 26.0

GRAPHICS

Nodular lymphoid hyperplasia



Nodular mucosa of the terminal ileum seen at colonoscopy in a patient later proven to have common variable immunodeficiency (CVID). Nodularity was also visible in the proximal colon.

Reproduced with permission from: Eric D Libby, MD.

Graphic 80570 Version 3.0

Symptoms in gastrointestinal lymphoma according to involved site

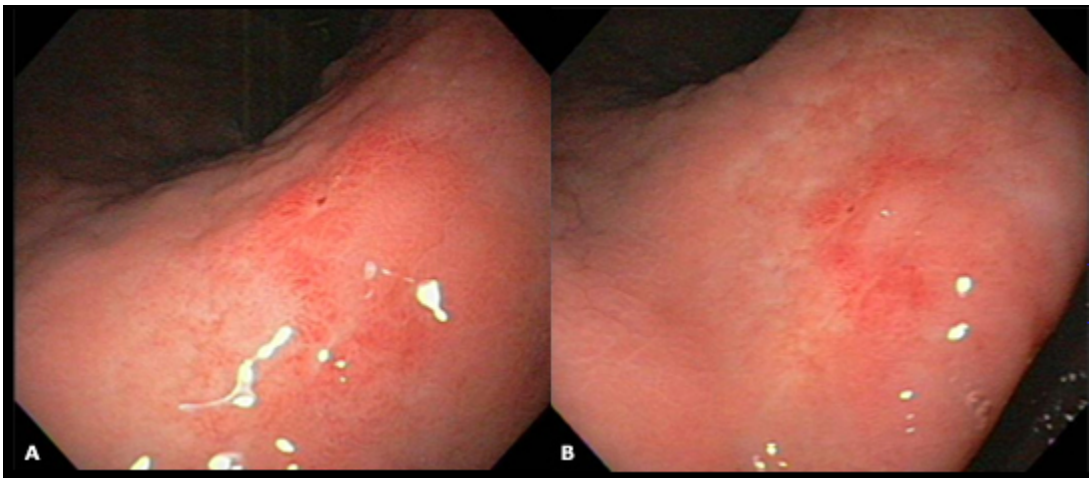
Symptom	Stomach (n=277)	Small bowel (n=32)	Ileocecal (n=26)	Multiple sites (n=24)
Pain	78	75	77	58
Loss of appetite	47	41	23	58
Weight loss	24	34	15	25
Bleeding	19	6	12	8
Vomiting	18	31	8	21
Night sweats	11	12	19	46
Diarrhea	4	12	19	29
Constipation	3	25	23	12
Fever	2	6	8	4
Perforation	2	9	-	-
Ileus	-	38	19	4
No symptoms	4	-	-	-

This table shows the percent of patients with the listed symptom at each of the four major sites of disease.

Data from Koch P, et al. J Clin Oncol 2001; 19:3861.

Graphic 68593 Version 2.0

Gastric marginal zone lymphoma of mucosa associated lymphoid tissues (MALT) on endoscopy

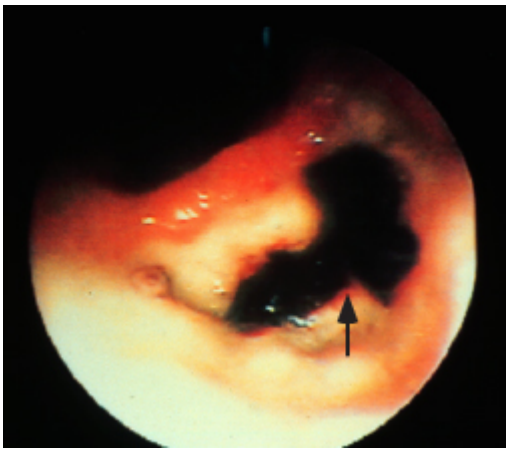


Upper endoscopy showing a superficial ulceration with mucosal thickening in the incisura with surrounding erythema.

Courtesy of John K. Kwon, MD and Harry Anastopoulus, MD.

Graphic 81847 Version 4.0

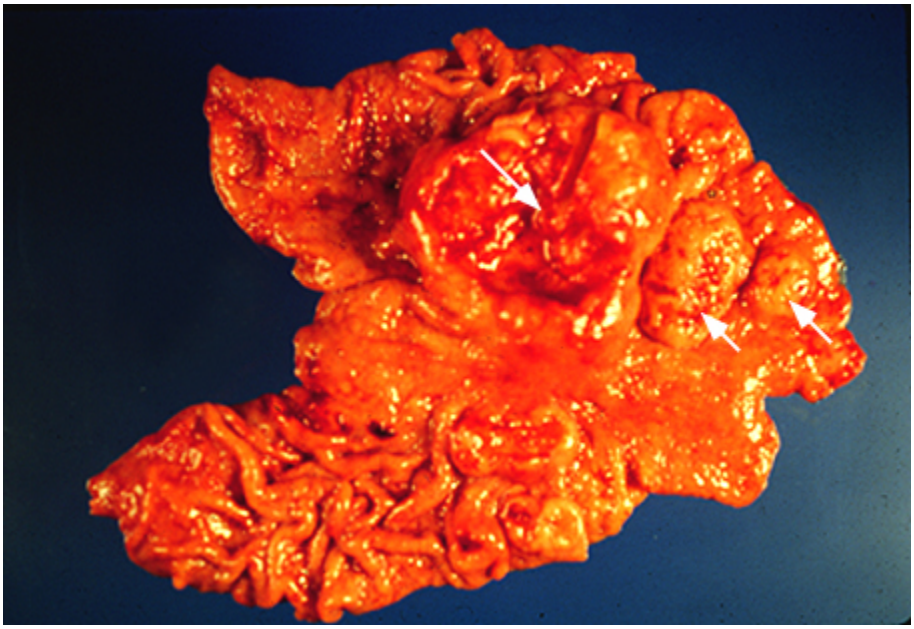
Gastric marginal zone lymphoma of mucosa associated lymphoid tissue (MALT) on endoscopy



Endoscopic view of a deep benign appearing ulcer crater in the gastric antrum (arrow) with a large blood clot indicating recent hemorrhage. Histology was consistent with a marginal zone lymphoma.

Graphic 70267 Version 4.0

Gastric large cell lymphoma



Gastric large cell lymphoma manifested as carcinoma-like ulcerative lesion (large arrow) and several variably sized nodules (small arrows) in adjacent mucosa.

Courtesy of Paul C Schroy, III, MD.

Graphic 53702 Version 2.0

Gastric large cell lymphoma

Longitudinal whole mount section on a gastric large cell lymphoma with annular ulcerations and full- thickness infiltration of the gastric wall.

Courtesy of Paul C Schroy III, MD.

Graphic 65490 Version 2.0

Histologic scoring of gastric marginal zone lymphoma

Grade	Description	Histologic features
0	Normal	Scattered plasma cells in lamina propria; no lymphoid follicles
1	Chronic active gastritis	Small clusters of lymphocytes in lamina propria; no lymphoid follicles; no LELs
2	Chronic active gastritis with florid lymphoid follicle formation	Prominent lymphoid follicles with surrounding mantle zone and plasma cells; no LELs
3	Suspicious lymphoid infiltrate in lamina propria, probably reactive	Lymphoid follicles surrounded by small lymphocytes that infiltrate diffusely in lamina propria and occasionally into epithelium
4	Suspicious lymphoid infiltrate in lamina propria, probably lymphoma	Lymphoid follicles surrounded by CCL cells that infiltrate diffusely in lamina propria and into epithelium in small groups
5	Marginal zone lymphoma	Presence of dense diffuse infiltrate of CCL cells in lamina propria with prominent LELs

CCL: centrocyte-like lesion; LEL: lymphoepithelial lesion.

Adapted from: Wotherspoon AC, Dogliogi C, Diss TC, et al. Lancet 1993; 342:575.

Graphic 78881 Version 4.0

Clinical features of the different types of small intestinal lymphomas

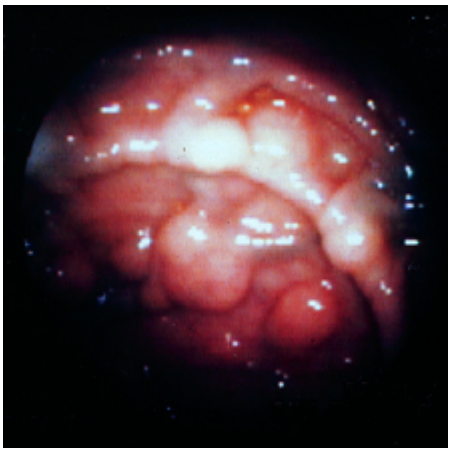
Feature	IPSID-associated lymphoma	Non-IPSID-associated lymphoma
Median age	25 years	37 years
Gender	Primarily males	Slight male predominance
Symptoms and signs	Abdominal pain Chronic diarrhea Malabsorption Severe weight loss Clubbing Ankle edema	Abdominal pain Palpable abdominal mass Bleeding Intestinal obstruction Intestinal perforation
Paraprotein	Usually present	Usually absent

IPSID: Immunoproliferative small intestinal disease

Adapted from: Salem, P, El-Hashimi, L, Allam, C, et al, Cancer 1987; 59:1670.

Graphic 76348 Version 2.0

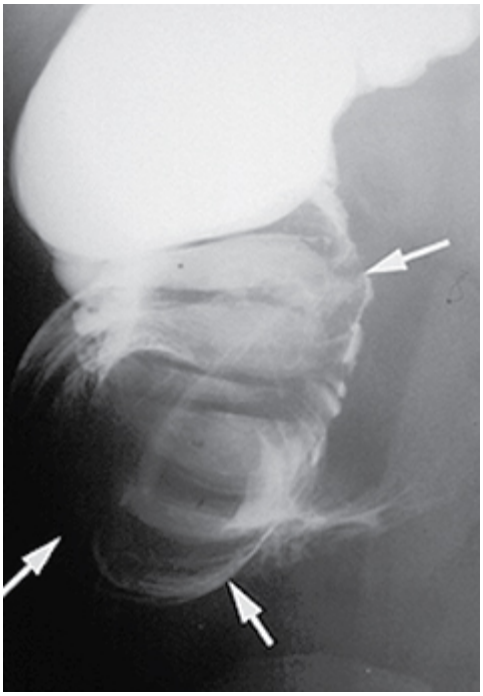
Mantle cell lymphoma polypoid lesions on colonoscopy



Colonoscopic view of multiple polypoid lesions in the proximal colon. Histology was consistent with a mantle cell lymphoma.

Graphic 64338 Version 2.0

Intussusception of ileal lymphoma

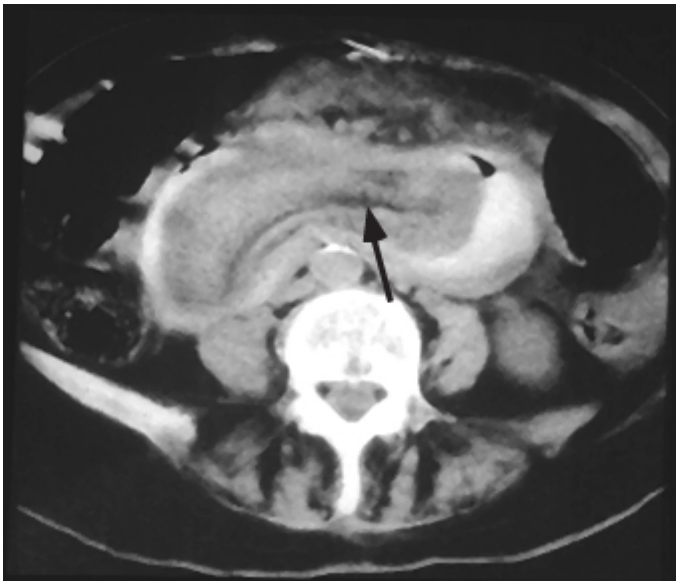


Barium enema shows a large soft tissue mass in the cecum (arrows) caused by intussusception of a lymphoma arising in the terminal ileum.

Courtesy of Jonathan Kruskal, MD, PhD.

Graphic 64048 Version 2.0

Intussusception of small bowel lymphoma

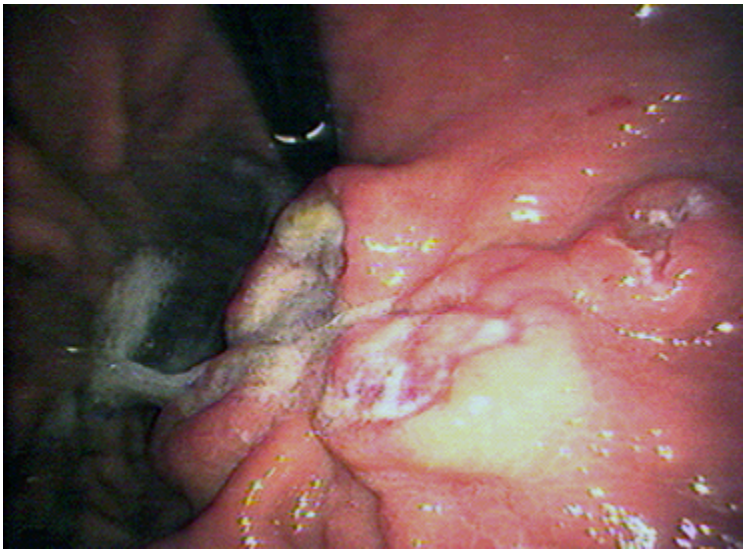


Computed tomography scan of the abdomen demonstrates a large mass in the lumen of a distended loop of small bowel. Note mesenteric fat in the center of this intraluminal mass (arrow).

Courtesy of Jonathan Kruskal, MD, PhD.

Graphic 75825 Version 3.0

Burkitt lymphoma on endoscopy

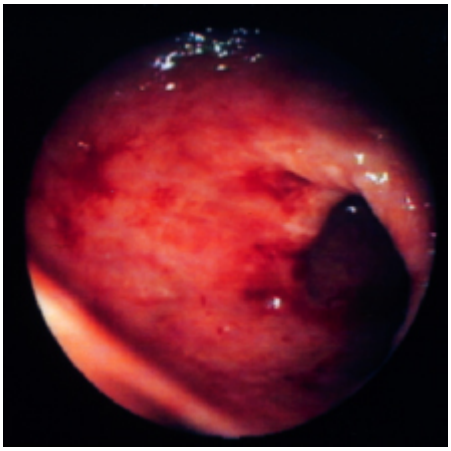


Lobulated, ulcerated protuberances seen on retroflexed view during endoscopy in a patient with abdominal pain. Biopsy revealed them to be Burkitt lymphoma.

Courtesy of Eric D Libby, MD.

Graphic 69159 Version 2.0

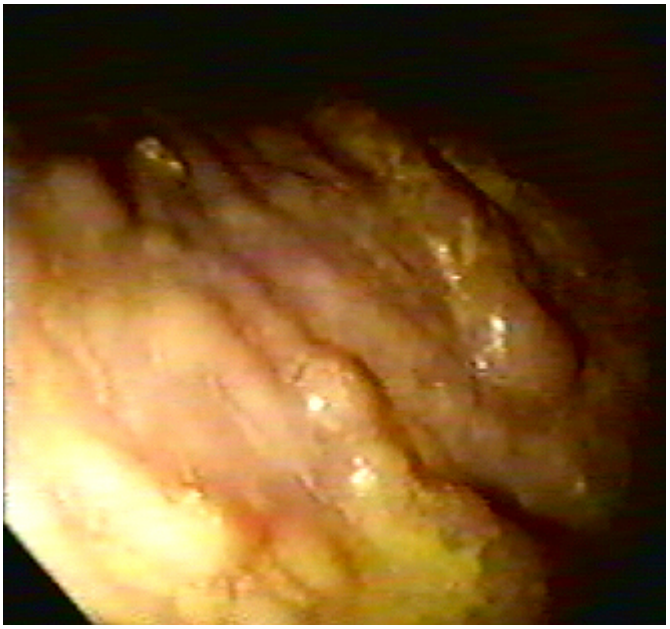
Non-Hodgkin's lymphoma of the colon



Colonoscopic view of a friable, indurated mucosa with scattered areas of ulceration suggestive of inflammatory bowel disease. Histology was consistent with a non-Hodgkin's lymphoma.

Graphic 67755 Version 1.0

Non-Hodgkin lymphoma of rectum on endoscopy

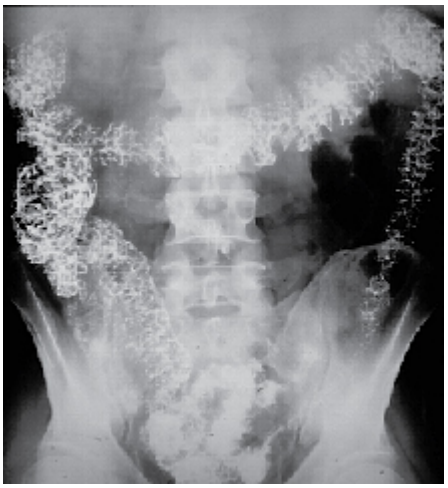


Endoscopic view of the rectum in a 95-year-old patient who presented with weight loss and anemia. The biopsy revealed non Hodgkins lymphoma.

Courtesy of Alfonso Sandoval, MD.

Graphic 72196 Version 3.0

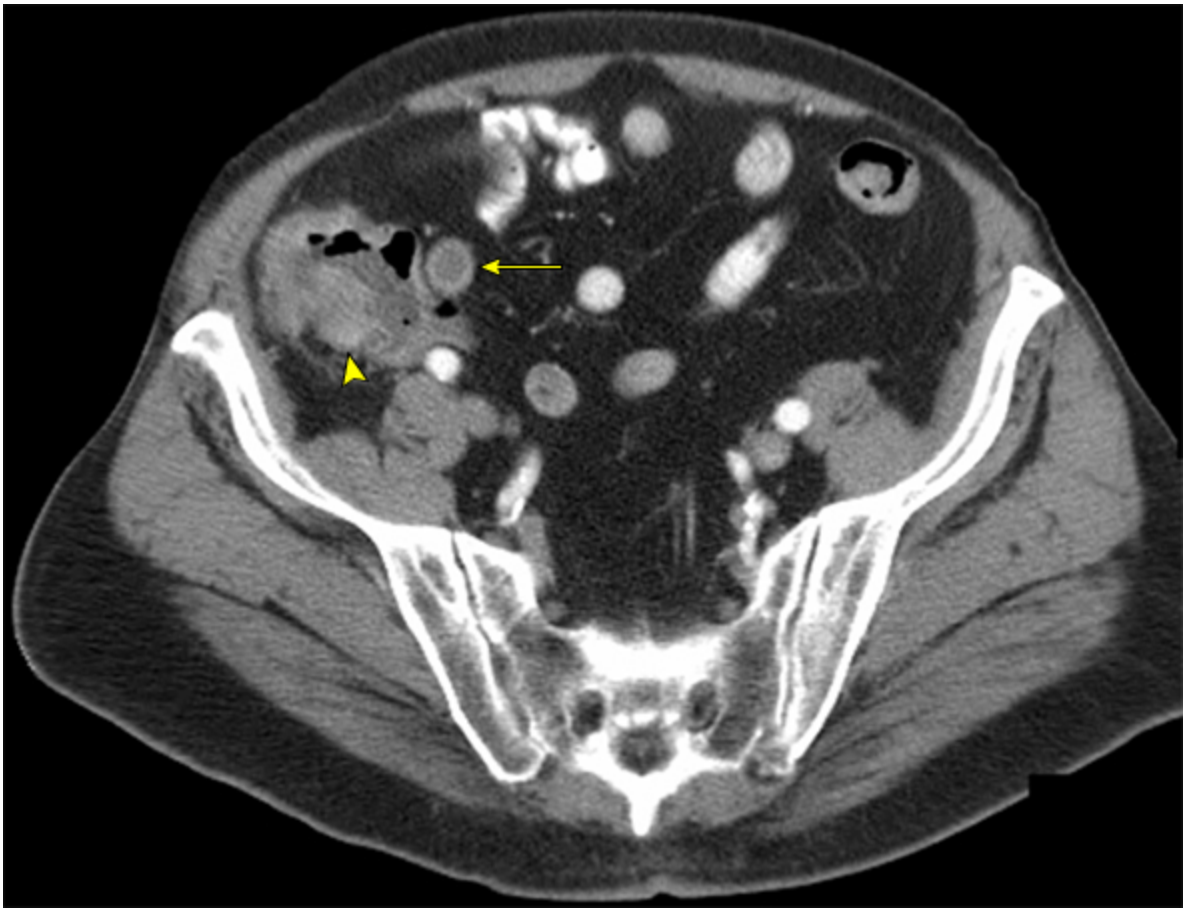
Non-Hodgkin's lymphoma of the colon and terminal ileum



Post-evacuation film from a barium enema shows diffuse nodularity of the ileum and entire colon. Histology was consistent with a non-Hodgkin's lymphoma.

Graphic 55178 Version 3.0

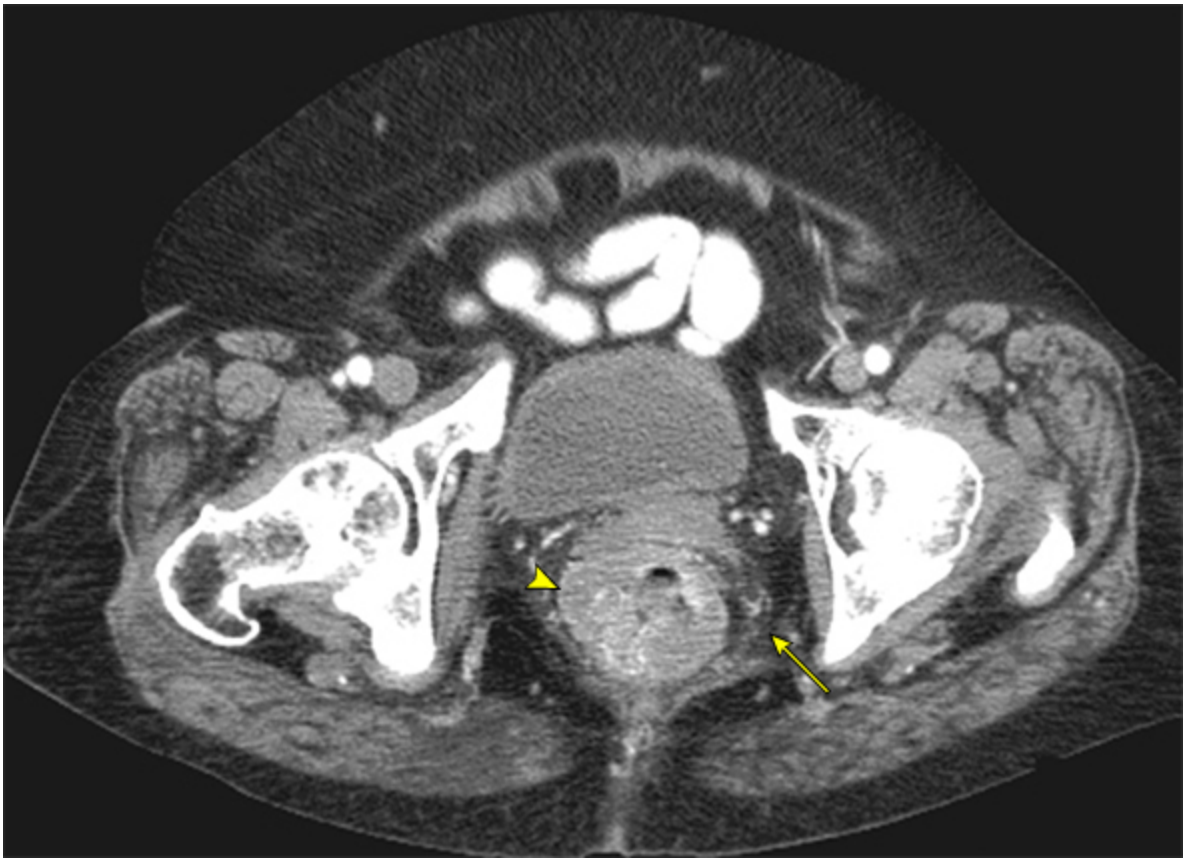
Polypoid form of colorectal B cell lymphoma on CT scan



Computed tomography (CT) scan of the abdomen at the level of the iliac crests in a patient with colorectal B cell lymphoma. There is a polypoid enhancing mass of the cecum (arrowhead) as well as enhancement and thickening of the ileum (arrow) suggesting involvement with lymphoma.

Graphic 88881 Version 3.0

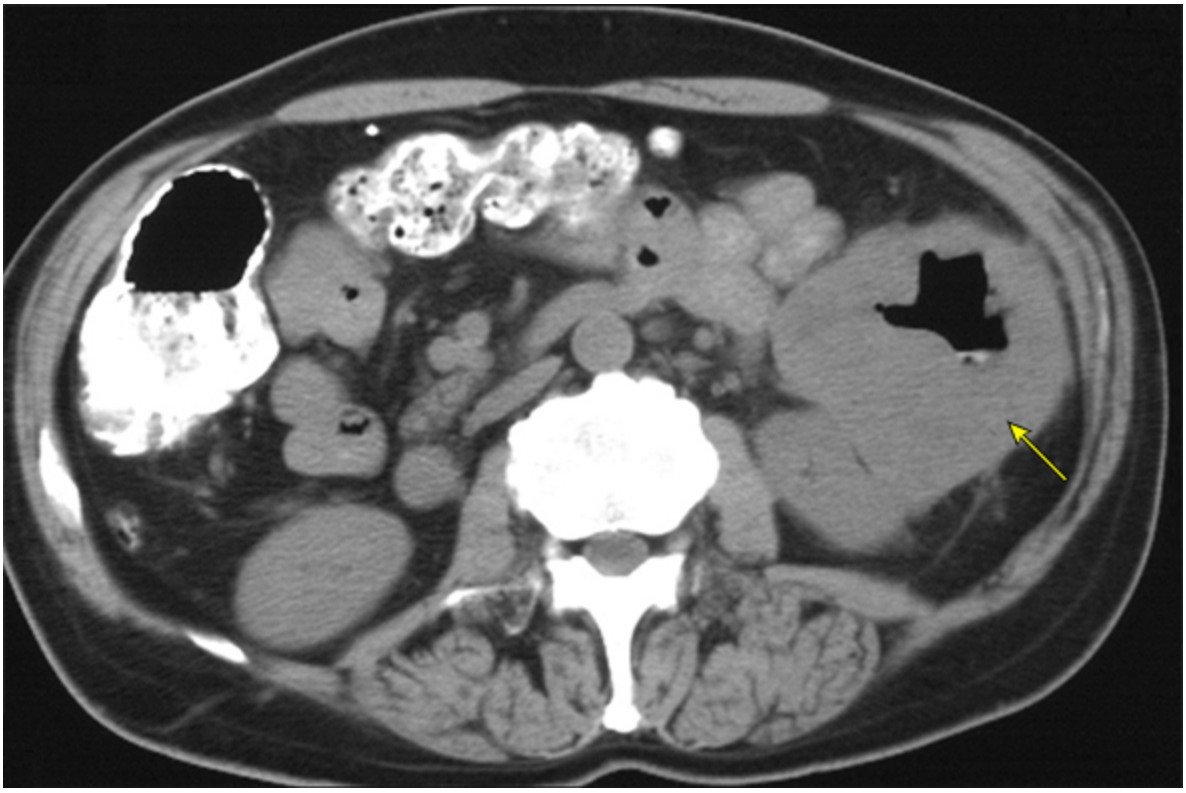
B cell lymphoma of the rectum on CT



A computed tomography (CT) scan of the abdomen and pelvis at the level of the acetabuli in a patient with rectal B cell lymphoma. There is diffuse irregular, circumferential thickening of the rectum (arrowhead) with mild perirectal induration (arrow).

Graphic 88884 Version 3.0

Non Hodgkin lymphoma of the descending colon on CT scan



Computed tomography (CT) scan of the abdomen and pelvis at the level of the mid-descending colon in a patient with non Hodgkin lymphoma. There is well defined, bulky, circumferential thickening of the colonic wall (arrow), without significant induration or obstruction despite the unusually large size of the tumor.

Graphic 88883 Version 3.0

Contributor Disclosures

Arnold S Freedman, MD Other Financial Interest: Bayer [Lymphoma DSMB]. All of the relevant financial relationships listed have been mitigated. **Andrew Lister, MD, FRCP, FRCPath, FRCR** Equity Ownership/Stock Options: AbbVie [Lymphoma]; AstraZeneca [Lymphoma]; GSK [Lymphoma]; Johnson & Johnson [Lymphoma]; Novartis [Lymphoma]; Pfizer [Lymphoma]. Consultant/Advisory Boards: BerGenBio [Lymphoma]; Merck [Data monitoring committee, Lymphoma]; Regeneron [R1979-ONC-1625 study]. All of the relevant financial relationships listed have been mitigated. **Alan G Rosmarin, MD** No relevant financial relationship(s) with ineligible companies to disclose.

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