
Photoclinic



Figure 1. CT scan of the abdomen.

A 38-year-old woman was referred to our gastrointestinal (GI) clinic. Her chief complaint was an epigastric pain since six months prior to referral. The pain was not positional and did not aggravate with activity. It was colicky in pattern lasting for 15 – 45 minutes and occurring at four-hour intervals, which was exacerbated after meal. She had 18 kg weight loss, vomiting and nausea, fever, chills, and anorexia

since six months ago. She noted that her symptoms are reduced and she also gained weight (about 7 kg) in the past two months. Her medical history was negative for diabetes mellitus, hypertension, and hyperlipidemia. Physical examination revealed a mild tenderness of the abdomen and clubbing of her fingers. Laboratory data were in normal range. Abdominal ultrasonography was reported normal. Upper GI endoscopy and colonoscopy were normal too. An image from spiral computed tomography (CT) is shown in Figure 1.

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What is Your Diagnosis?

See the page 272 – 273 for the diagnosis

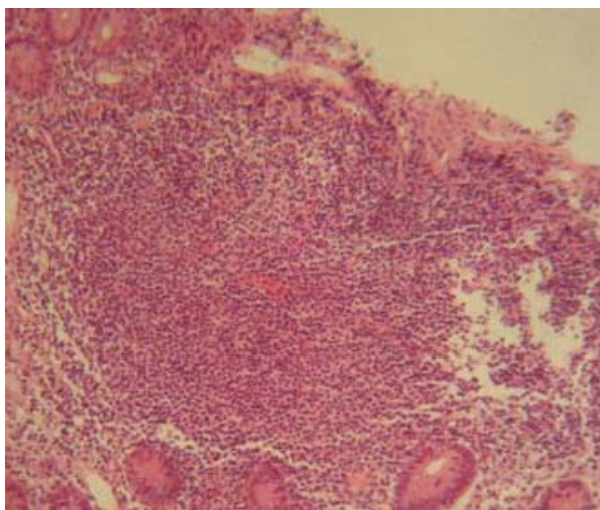


Figure 2. Histopathology: hematoxylin and eosin staining.

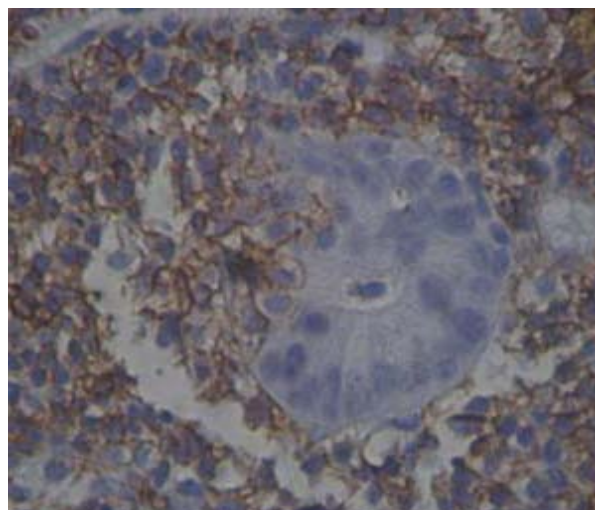


Figure 3. Histopathology immunohistochemical staining.

Although the GI tract is the most common site for extranodal lymphoma, primary follicular lymphoma of the GI tract (PFLGIT) is a rare and poorly-defined disease, especially low-grade follicular B-cell lymphomas.¹ Primary GI lymphoma is a heterogeneous and relatively rare disease, which accounts for 11 – 34% of all non-Hodgkin's lymphomas, with some regional variability in the frequency and histological type.² Lymphomas in this setting usually have the B-cell phenotype and are dominated by diffuse large cell subtype. Histological subtypes of indolent lymphomas are closely related to the involved site and are dominated by mucosa-associated lymphoid tissue (MALT) type, which is primarily localized in the stomach.^{2,3}

Prompt diagnosis and treatment result in lower mortality rate and prevalence. PFLGIT is rare and constitutes <7% of all non-Hodgkin's lymphomas at this location.⁴

Strikingly, 50% of PFLGIT show an endoscopic aspect of multiple lymphomatous polyposis. These findings are important clinically, because mantle cell lymphoma has poor prognosis and requires aggressive treatment. Therefore, immunophenotyping using immunostaining for CD5, CD10, CD20, and bcl2, and molecular biology studies looking for the presence of IgH/bcl2 or IgH/bcl1, are advisable to different-

iate the diagnosis of lymphomatous polyposis from that of follicular lymphomas.⁵

Since PFLGIT is rare, it is difficult to provide a definitive therapeutic approach. The prognosis, however, seems not to be different from nodal follicular lymphoma with an indolent course even in the absence of specific treatment. Surgery and chemotherapy should be indicated only on the basis of clinical symptoms.

Indeed, 33% of patients who were treated with surgery, chemotherapy, and/or radiotherapy relapsed after a mean time of 31 months, which was not different from the mean time for progression of the disease (37.5 months) in patients who did not receive any treatment.⁶

Follicular lymphoma of the GI tract predominantly presents in females. The clinical course is generally indolent and the endoscopic appearance may be similar to lymphomatous polyposis, which indicates immunophenotyping and even molecular biology studies for diagnosis. Therapeutic interventions are not indicated unless clinical symptoms are present or the disease is progressive. Duodenum is the most common site of the disease, although some cases with the involvement of colon and stomach have been reported.⁷ Multifocal disease may also be seen. The prognosis of these patients may be similar to follicular lymphoma diagnosed in nodal sites. There is no consensus on management, although

some patients may be managed with resection or with a watch-and-wait approach.^{6,7}

In our patient, the symptoms were indolent and subsided without any treatment despite diffuse involvement of the small bowel. Small bowel wall thickness was detected by CT, and immunohistochemical staining showed CD10+, bcl2+, CD20+, and CD5-. Pathology and immunohistochemical CD 10 pictures are shown in Figures 2 and 3.

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