Case Reports in Gastroenterology

Case Rep Gastroenterol 2011;5:578–582
DOI: 10.1159/000331137

Published online:
October 1, 2011

ISSN 1662–0631
www.karger.com/crg

Gastric Marginal Zone B Cell Lymphoma of the Duodenum

A. Ndzengue[a] R. Khurana[a] M. Mora[a] R.B. Rafal[b]
D. Trauber[a] M. Mansour[a] G.L. Posner[a] E.A. Jaffe[a]

Departments of [a]Medicine and [b]Radiology, Interfaith Medical Center, Brooklyn, N.Y., USA

Key Words
Small bowel lymphoma · Gastric lymphoma · Duodenum

Abstract
Small bowel lymphomas of the extranodal type occur in the young and are characteristically associated with malabsorption syndrome. We present the case of an elderly in whom there was no malabsorption and the duodenal tumor was a gastric type marginal zone B cell lymphoma also known as gastric mucosa-associated lymphoid tissue (MALT) lymphoma. A 73-year-old woman presented to the emergency room with 2 weeks of general weakness, recurrent vomiting containing food particles and abdominal distension. She had been diagnosed with diabetic gastroparesis 4 years prior. CT of the abdomen and pelvis was suggestive of gastric outlet obstruction but no evidence of pancreatic or duodenal mass. Endoscopy and biopsy of the tumor obstructing the distal first part of the duodenum confirmed a gastric marginal MALT lymphoma. The patient's symptoms improved with radiotherapy. Gastric MALT lymphoma, an extranodal lymphoma primarily described in the stomach, can also present in the small bowel and is not associated with malabsorption.

Introduction

Young adults who are otherwise well and present with upper gastrointestinal (GI) symptoms are not routinely investigated; however, malignancies should be ruled out in the middle-aged and the elderly. Small bowel cancers account for 2% of all GI tract cancers [1]. Lymphomas represent 8% of all small bowel malignancies [2]. Primary small bowel lymphomas occur in patients with normal white cell count, no peripheral or mediastinal lymphadenopathy and no liver or spleen involvement. They represent only 9% of all primary GI lymphomas, unlike primary gastric lymphomas which make up to 75% of this entity [3]. Most GI lymphomas are extranodal lymphomas of B cell or T cell origin [4]. B cell lymphomas of the GI tract are divided into B cell lymphoma of

Albert Ndzengue, MD
Department of Medicine, Interfaith Medical Center
1545 Atlantic Avenue, Brooklyn, NY 11213 (USA)
Tel. +1 718 613 4063

Case Rep Gastroenterol 2011;5:578–582
DOI: 10.1159/000331137
Published online: October 1, 2011

ISSN 1662–0631
www.karger.com/crg

mucosa-associated lymphoid tissue (MALT), diffuse large B cell lymphomas, mantle cell lymphomas and Burkitt and Burkitt-like variant lymphomas [3]. MALT lymphomas of the stomach represent up to 48% of all primary gastric lymphomas and are called gastric marginal zone B cell lymphomas [5]. The predominant extranodal primary lymphoma in the small bowel is immunoproliferative small bowel disease (IPSID) or alpha chain disease [6]. Both gastric MALT lymphomas and IPSID arise from postgerminal center (known as marginal zone) B cells [7]. The B cells characteristically secrete alpha heavy chains in IPSID lymphomas. Gastric MALT lymphomas and IPSID differ in their epidemiology and clinical presentations. We present the case of an elderly female where the lymphoma obstructing the duodenum was not an IPSID lymphoma but a gastric MALT lymphoma. The literature on the latter entity is further reviewed.

Case Report

A 73-year-old African-American woman with type 2 diabetes and gastroparesis presented to the emergency room with 2 weeks of general weakness, recurrent projectile vomiting after little food intake, abdominal distension, discomfort and significant weight loss. During a bout of symptoms similar to the above 20 months prior, gastric outlet obstruction had been suspected on a CT scan of the abdomen. However the patient had refused esophagogastroduodenoscopy and had not follow up at the gastroenterology clinic. Colonoscopy had been normal 2 years previously.

On this admission, her BMI was 25.4 compared to 40.5 12 months earlier on her previous visit. She was afebrile and had a pulse of 103 per minute and a blood pressure of 92/52 mm Hg. She was emaciated with dry oral mucosa. Her abdomen was distended with visible peristaltic waves. There was no abdominal tenderness. A succussion splash was elicited and bowel sounds were slightly active. A fecal occult blood test was positive.

Laboratory results showed normocytic normochromic anemia with a hemoglobin of 9.6 g/dl, hypochloremic hypokalemic metabolic alkalosis with a potassium of 2.53 mmol/l, hypomagnesemia of 1.3 mmol/l and hypoalbuminemia of 2.6 g/dl. Renal function, blood glucose and liver function tests were normal. *Helicobacter pylori* antibody was negative. A CT scan of the abdomen and pelvis (**fig. 1**) showed gastric distension suggestive of gastric outlet obstruction but no pancreatic or duodenal mass. The electrolyte imbalance was corrected. On endoscopy, 4,900 ml of brown gastric fluid mixed with food residue was aspirated and biopsies of the mass obstructing the distal first part of the duodenum were obtained. Histopathology confirmed marginal B cell lymphoma (**fig. 2**, **fig. 3**). The patient improved as the obstructing mass shrunk after radiotherapy and was discharged 3 weeks later.

Discussion

Gastric MALT lymphomas and IPSID are both extranodal marginal zone B cell lymphomas. They differ not only in their common anatomic locations but also in their epidemiology and clinical presentations. Gastric MALT lymphomas occur in women in their sixties. Upper abdominal symptoms are the common clinical features [7]. IPSID lymphomas typically occur in males around 25 years of age [6]. Common symptoms include chronic diarrhea, malabsorption, weight loss, ankle edema and clubbing [6], although nonspecific peptic ulcer-like symptoms predominate occasionally. Small bowel lymphomas may cause gastric outlet obstruction syndrome by their bulk-like effect. The oddity in our case is that the diagnosed extranodal marginal zone B cell lymphoma was of the gastric type but located in the duodenum. Our patient's age was also far from the median age of presentation of 25 years in IPSID [6]. The gastric epithelium in the duodenal tumor could be a metaplasia or dysplasia of the native epithelium [8] or a

Case Rep Gastroenterol 2011;5:578–582
DOI: 10.1159/000331137
Published online: October 1, 2011

ISSN 1662–0631
www.karger.com/crg

duplication cyst epithelium. MALT lymphomas occur in lymphoid organs or lymphoid tissues and are induced by autoimmune/infection-associated chronic inflammation [9]. They are low-grade lymphomas which tend to remain localized on the gastrointestinal epithelium [10, 11] as demonstrated in our patient. On immunohistochemistry, the cells' immunophenotype is that of B cells: CD20+, Bcl-2+, CD23–, CD10– and CD5– [9].

Upper GI symptoms such as epigastric pain, postprandial vomiting and early satiety are nonspecific [12]. Gastroparesis, chronic gastric outlet obstruction syndrome and upper gastrointestinal lymphoma all share these symptoms. The latter two entities fare worse left undiagnosed and not treated early and must be investigated with endoscopy prior to making the diagnosis of gastroparesis. Gastric MALT lymphoma can also present in the small bowel and is not associated with malabsorption as demonstrated in our case.

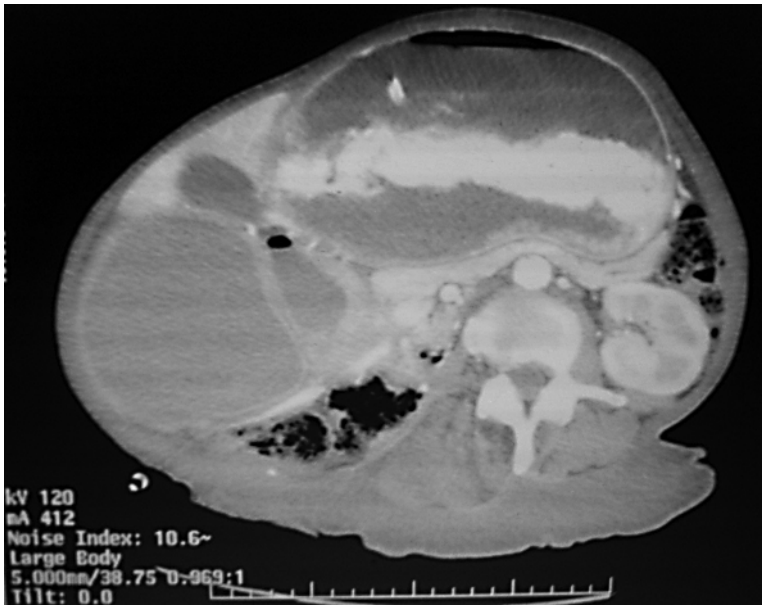

Fig. 1. CT scan of the abdomen and pelvis. Note the gross distension of the stomach and the duodenum.

Case Rep Gastroenterol 2011;5:578–582
DOI: 10.1159/000331137
Published online: October 1, 2011

ISSN 1662–0631
www.karger.com/crg

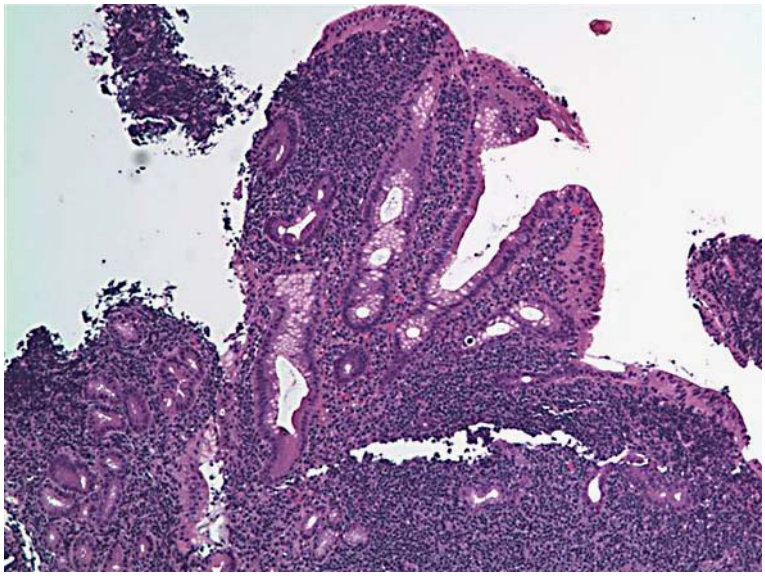

Fig. 2. The duodenal tumor. The mucosa is of gastric type. The submucosa is filled with small B lymphocytes with conspicuous nuclei. There is no evidence of an inflammatory process. H&E, ×10.

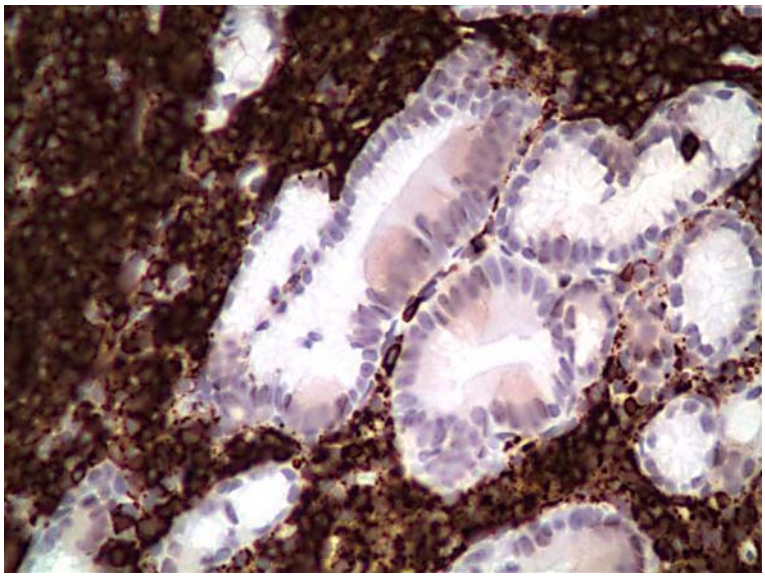

Fig. 3. The duodenal tumor. Submucosal B cells stain positive for CD43, CD20 and Bcl-2; those cells are also CD5-, CD10- and CD23-negative. A lymph node was used as control. ×40.

Case Rep Gastroenterol 2011;5:578–582
DOI: 10.1159/000331137
Published online: October 1, 2011

ISSN 1662–0631
www.karger.com/crg

References

1 Jemal A, Siegel R, Ward E, et al: Cancer statistics, 2009. CA Cancer J Clin 2009;59:225–249.

2 Bilimoria KY, Bentrem DJ, Wayne JD, et al: Small bowel cancer in the United States: changes in epidemiology, treatment, and survival over the last 20 years. Ann Surg 2009;249:63–71.

3 Harris NL, Jaffe ES, Stein H, et al: A revised European-American classification of lymphoid neoplasms: a proposal from the International Lymphoma Study Group. Blood 1994;84:1361–1392.

4 Koch P, del Valle F, Berdel WE, et al: Primary gastrointestinal non-Hodgkin's lymphoma: I. Anatomic and histologic distribution, clinical features, and survival data of 371 patients registered in the German Multicenter Study GIT NHL 01/92. J Clin Oncol 2001;19:3861–3873.

5 Cogliatti SB, Schmid U, Schumacher U, et al: Primary B-cell gastric lymphoma: a clinicopathological study of 145 patients. Gastroenterology 1991;101:1159–1170.

6 Salem P, el-Hashimi L, Anaissie E, et al: Primary small intestinal lymphoma in adults: a comparative study of IPSID versus non-IPSID in the Middle East. Cancer 1987;59:1670–1676.

7 Cavalli F, Isaacson PG, Gascoyne RD, Zucca E: MALT lymphomas. Hematology Am Soc Hematol Educ Program 2001:241–258.

8 Shabib SM, Cutz E, Drumm B, Sherman PM: Association of gastric metaplasia and duodenitis with *Helicobacter pylori* infection in children. Am J Clin Pathol 1994;102:188–191.

9 Ioachim HL, Medeiros LJ: Lymphomas of salivary glands; in: Ioachim's Lymph Node Pathology. Philadelphia, Pa., Wolters Kluwer Health/Lippincott Williams and Wilkins, 2009, pp 447–451.

10 Wotherspoon AC, Doglioni C, Diss TC, et al: Regression of primary low-grade B-cell gastric lymphoma of mucosa-associated lymphoid tissue type after eradication of *Helicobacter pylori*. Lancet 1993;342:575–577.

11 Pinotti G, Zucca E, Roggero E, et al: Clinical features, treatment and outcome in a series of 93 patients with low grade MALT lymphoma. Leuk Lymphoma 1997;26:527–537.

12 Siew LC, Huang C, Fleming J: Gastric adenocarcinoma mistakenly diagnosed as an eating disorder: case report. Int J Eat Disord 2010;43:286–288.