

# Malignant Lymphoma Of The Duodenum Presenting With Melaena And Obstructive Jaundice At The Same Time – A Case Report

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## Abstract

We report in the rare case of a 36 year old man who presented to our endoscopic unit with duodenal large cell lymphoma presenting with gastrointestinal bleeding and obstructive jaundice at the same time.

## INTRODUCTION

The gastrointestinal (GI) tract is the predominant site of extranodal lymphomas. Primary GI lymphomas are rare, while secondary involvement is relatively common. The definition of primary GI lymphomas 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. About 16% of GI lymphomas, or approximately 5% of non-Hodgkin's lymphomas (NHL), in general, are primarily located in the intestine and differ from gastric lymphomas in clinical features, pathology, treatment, and prognosis. The most of primary small intestine lymphomas (PSIL) occurs in the ileocecal region while duodenum remains the most infrequent site. PSIL typically present with nonspecific signs and symptoms attributable to the site of involvement, which may include abdominal pain, GI bleeding, intestinal obstruction or perforation, and/or a palpable abdominal mass. PSIL is characterized by younger age at presentation; less localized stage of disease and more aggressive histological type, mainly diffuse large B-cell type (DLCL). In our case, we describe a patient with primary duodenal diffuse large B-cell lymphoma who presented initially with signs of acute hemorrhage and obstructive jaundice.

## CASE REPORT

A 36-year-old man was admitted to the General Hospital of Zadar in October 2009, with a 3-day history of weakness, anorexia, abdominal pain and bloody diarrhea.

Physical examination disclosed a pale, poorly nourished man

with signs of acute hemorrhage. There was no superficial lymphadenopathy, abdomen was flat and soft. The liver was palpable and extended 3 cm below the costal margin and the spleen wasn't palpable. Rectal examination demonstrated black tarry stool.

The RBC count was  $2,92 \times 10^{12}/L$ , Hb level 83 g/L and Hct 0,24. The WBC count was  $9,8 \times 10^9$  with 75% segmented neutrophils and 14% lymphocytes. The level of total protein was 6.0, total bilirubin level 51,4, and other values included AST 269, ALT 569, alkaline phosphatase 436, gamma-GT 891 and LDH 501. Serum level of carbohydrate antigen 19-9 was mildly elevated at 43.0 U/mL, and CEA and AFP were within normal limits.

Aspiration of bone marrow cells revealed no abnormality.

Abdominal ultrasonography revealed multiple hypoechoic focal lesions of the liver and spleen.

MSCT of abdomen showed multiple nodose lesions, size up to 43 mm, almost isodense to liver and spleen parenchyma. The gallbladder was distended with normal wall thickness and no stone. Common bile duct was fusiformly dilated to 14 mm in diameter. There was no significant lymphadenopathy or free fluid.

Duodenal endoscopy revealed edematous swelling of the mucosa and spontaneously bleeding polypoid lesion at the descending part of duodenum (region of papilla Vateri), and a mucosal biopsy was performed.

Histopathological and immunohistochemical examination of

a biopsy specimen showed diffuse large cell lymphoma. The lymphoid tumor cells had round to occasionally irregular nuclei with one or more nucleoli and increased mitotic rate, positive for B-cell surface marker CD20, and bcl-6, and negative for cyclin D1, CD3 and CD10. Proliferation marker Ki-67 was expressed in >70%.

Chest CT scans showed no evidence of lymph node enlargement in the mediastinum.

The patient was diagnosed as stage IV according to the Modified Ann Arbor staging and classified into the high intermediate risk group because the IPI score was 3.

Because our patient was considered inoperable, and bilirubin level wasn't high, chemotherapy R-CHOP protocol (rituximab-cyclophosphamide, doxorubicin, vincristine and prednisone) was started. The patient received a total of 6 cycles of chemotherapy. During immunochemotherapy patient had grade 1 of leucopenia and no other adverse event; liver function tests become normal. After six cycles of therapy, ultrasound showed reduction of tumor mass, normal bile duct in diameter. Control gastroduodenoscopy revealed great regression of the lesions, and no sign of bleeding.

## **DISCUSSION**

Primary malignant duodenal tumors are uncommon accounting for only 0.3% of all gastrointestinal tumors but about 50% of all small intestinal malignancies. Although the GI tract is the predominant site of extranodal lymphomas, primary lymphomas of the GI tract are rare. Primary lymphomas of the small intestine (PSIL) account for 5% of all NHLs, 16-17% of all GI lymphomas and 16%-38% of all small bowel malignancies.<sup>5,6</sup> The distribution of PSIL varies among populations, while uncommon in Western countries, accounts for up to 75 % of primary GI lymphomas in the Middle East and Mediterranean basin. The incidence of lymphoma in the segments of the small intestine varies directly with the amount of lymphoid tissue present; most lymphomas occur in the ileocecal region while duodenum remains the most infrequent site. Duodenal tumors pose diagnostic difficulties not only due to their rarity, but their non-specific signs and symptoms and the fact that duodenum is usually overlooked during upper gastrointestinal endoscopies.<sup>3</sup>

It is sometimes difficult to know whether patients with advanced lymphoma have primary or secondary GI involvement. According to Dawson et al., the following

criteria should be met before labelling a lymphoma of GI tract origin: (1) absence of palpable superficial lymphadenopathy; (2) absence of enlarged lymphnodes in mediastinum on CXR; (3) no grossly demonstrable involvement beyond the affected segment of the intestine and its regional mesenteric lymphnodes at the time of diagnosis; (4) normal white cell and differential count; (5) non involvement of liver and spleen.<sup>1</sup> It is clear that this restrictive definition could exclude primary GI lymphoma with widespread dissemination. Hermann et al. have suggested that primary GI lymphomas are those with predominant GI lesions or that presented initially with symptoms related to GI tract involvement. Although our case doesn't fulfill Dawson's criteria, it was considered primary intestinal lymphoma due to initial signs and symptoms.

Despite their rarity, PSIL are important since their evaluation, diagnosis, management and prognosis are distinct from that of lymphoma at other sites and other cancers of the intestine. PSIL arises focally from lymphoid tissue and the remaining small bowel is uninvolved. The main histologic subtypes are: Diffuse large B-cell lymphoma, Mantle cell lymphoma, Burkitt lymphoma, Follicular lymphoma. PSIL or the «Western» type of small intestinal lymphoma must be separated from the «Mediterranean» type of small intestinal lymphoma, which is a manifestation of immunoproliferative small intestinal disease (IPSID) and is characterized by proliferation of gut-associated B-lymphoid cells affecting virtually the whole length of the small intestine. The clinical presentation of patients with lymphoma of the duodenum differs according to the location of the tumor, the degree of obstruction, the histologic tumor type and the rapidity of growth. Lymphomas causing obstruction result in early satiety, vomiting, and postprandial pain. Jaundice may be seen in lesions that involve periampullary region. Ulcerating lesions may result in bleeding, melena, hematemesis, anemia, and other typical ulcer symptoms. Diffuse large B-cell type (DLCL), the most common histologic type of PSIL, is characterized by younger age at presentation, less localized stage of disease and aggressive behavior.

Treatment of PSIL remains controversial. Surgery has an important role in local control of the disease and related complications such as bowel obstruction, bleeding or perforation. However, resection rarely eradicates lymphoma and many patients require further treatment either with

chemotherapy or radiotherapy. Patients not fit for surgery, but who are able to tolerate anthracyclin chemotherapy, should be given chemotherapy and possibly adjuvant radiotherapy. This continues to be a matter of debate.<sup>7</sup>

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