

B-Lymphoblastic Lymphoma of the Duodenum Producing Obstructive Jaundice: A Case Report

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Abstract:

Background: Lymphoblastic lymphoma (LBL) accounts for approximately 2% of all non-Hodgkin lymphoma (NHL), and approximately 90% are of immature T-cell lineage. Precursor B-cell LBL (B-LBL) is uncommon and accounts for less than 10% of total LBL cases and 0.3% of adult NHL.

Case Presentation: We report a case of B-cell lymphoblastic lymphoma (B-LBL) in the duodenum, causing painless obstructive jaundice as the first and only symptom, in a 21-year-old man. Duodenal endoscopy revealed a mass in the ampulla of Vater and narrowing of the second portion of the duodenum, although diagnosing lymphoma from an endoscopic biopsy was impossible.

Discussion: We performed gastrojejunal roux-en-Y anastomosis to establish a histological diagnosis and relieve the obstructive jaundice. Histological and immunohistochemical examination of the surgically resected specimen confirmed a diagnosis of B-cell lymphoblastic lymphoma.

Conclusion: Chemotherapy is the mainstay of treatment for lymphoma; however, surgery still plays an important role when the histological diagnosis cannot be established preoperatively and when complications are not amenable to nonsurgical therapy.

Keywords: B-cell lymphoblastic lymphoma, obstructive jaundice, Roux-en-y gastrojejunal anastomosis.

1. INTRODUCTION

Lymphoma is a type of malignant tumor originating in the lymphohematopoietic system. Due to the characteristics of the systemic distribution of the lymphatic system, lymphoma can invade any tissue and organ in the whole body. The main clinical manifestations are painless lymphadenopathy and hepatosplenomegaly [1].

Among all gastrointestinal malignancies, small intestinal tumors account for only 1–2% of them. Lymphoma only accounts for 4–11% of small intestinal malignancies, most of which occur in the ileum, followed by jejunum and duodenum. Duodenal lymphoma accounts for only 12% of all duodenal malignancies. In gastrointestinal lymphoma, while the stomach

accounts for most cases, duodenopapillary lymphoma is extremely rare [2]. We reported a case of primary duodenal papilla invasive B-cell lymphoma with obstructive jaundice as the first and only symptom.

B-lymphoblastic lymphoma constitutes a highly aggressive entity, rapidly progressive, and with a tendency to spread to the central nervous system. Chemotherapy is the first-line treatment for B-LBL. Intensive remission induction chemotherapy is recommended because standard chemotherapy, used in other types of lymphomas, is less effective in B-LBL [3]. Due to the rapid spread to the central nervous system, treatment protocols for B-LBL include an early central nervous system prophylaxis along with consolidation blocks

and subsequent maintenance therapy. In this report, we present a case of precursor B lymphoblastic lymphoma involving the duodenum, biliary duct, gallbladder, jejunal, and pancreas.

2. CASE PRESENTATION

A 21-year-old man presented with jaundice, abdominal pain, significant loss of weight of 5-weeks duration. Previous medical history was unremarkable, routine laboratory test with the following results: albumin 2.9 gr/dl, total bilirubin 22.2 gr/dl, indirect bilirubin 6.8 gr/dl, direct bilirubin 15.422 gr/dl, glucose 81.2 gr/dl, hemoglobin 12.1 gr/dl, platelets 607 000, leukocytes 9.5×10^9 cells/l, creatinine 0.5 gr/dl,

urea 31.4 gr/dl, alkaline phosphatase 576 gr/dl, gamma-glutamyltransferase 138 gr/dl. Jaundice with obstructive pattern is found.

Computer tomography scan showed a mass of unspecified origin located in duodenum, probably neoplastic, who causes dilatation of the intra and extrahepatic bile duct, hydrocholecyst, non-specific space-occupying lesions in the jejunal mucosa, metastasis to be ruled out, concentric thickening of the jejunal segment, possible infiltration, mesenteric lymph node growths averaging 10 mm, retroperitoneal lymph node growths averaging 12 mm. (FIGURE 1).

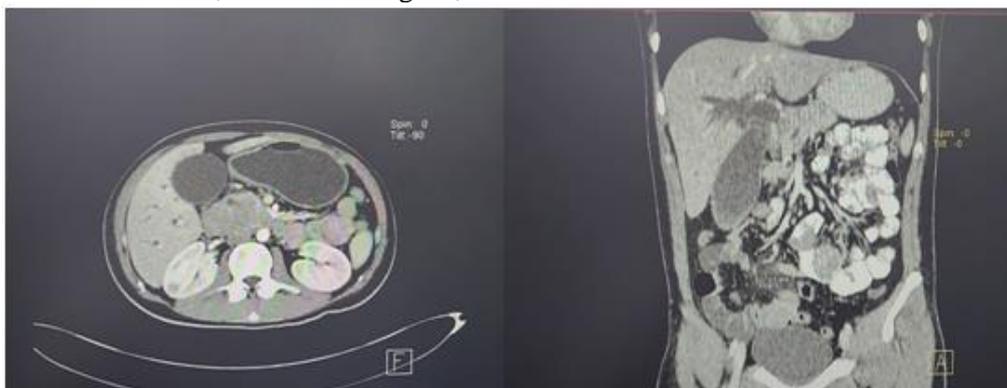


Figure1. Computed tomography at admission. A mass of unspecified origin located in duodenum, probably neoplastic, who causes dilatation of the intra and extrahepatic bile duct was observed. a) coronal section b) axial section.

a)

b)

Magnetic resonance imaging reported a tumor lesion in the head of the pancreas that causes retrograde dilation of the intra/extrahepatic bile duct and the main pancreatic duct, involving the second portion of the duodenum,

of origin to be determined, hydrocholecyst associated with bile sludge, jejuno-jejunal intussusception, associated neoplastic infiltration was not ruled out. (FIGURE 2).

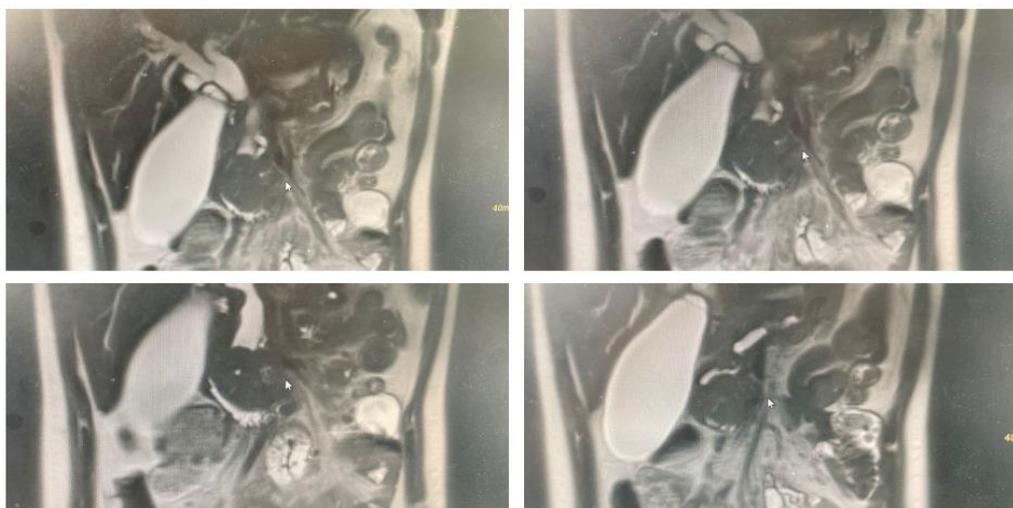


Figure2. Magnetic resonance imaging. Tumor in the head of the pancreas that causes retrograde dilation of the intra/extrahepatic bile duct and the main pancreatic duct, involving the second portion of the duodenum was observed.

As part of diagnosis of jaundice endoscopic retrograde cholangiopancreatography (ERCP) was performed with next result duodenal tumor activity with a filtering appearance not passable when passing through the duodenoscope, retentionist stomach secondary to extrinsic compression due to tumor activity.

The patient underwent emergency laparotomy, and a duodenal tumor extending to the biliary

duct, and gallbladder was observed, also we found two tumors located in jejunum, reason for which partial cholecystectomy, T tube placement, Roux-en-y gastrojejunal anastomosis, enterectomy of the two affected small bowel segments with hand-sewn end-to-end anastomosis was performed (FIGURE 3).

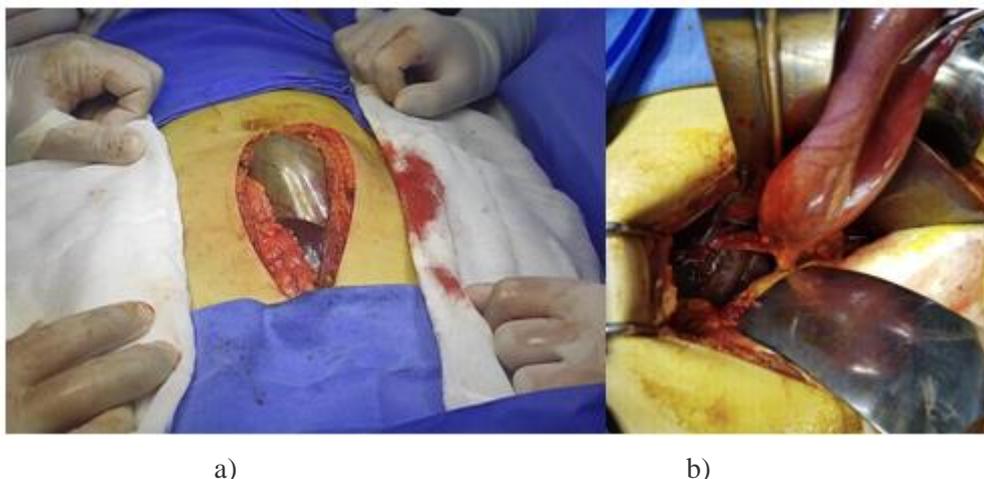


Figure3. Exploratory laparotomy. a) hydrocholecyst due to ampulla obstruction. b) duodenal tumor extending to the biliary duct.

Histopathology revealed B-LBL, affecting the entire bowel wall. On immunohistochemistry, the cells were positive for Tdt, CD10, PAX 5. The patient's postoperative course was uneventful, and he was discharged on the eighth postoperative day. Following discharge, he was referred for further treatment to the hematology.

3. DISCUSSION

Lymphoblastic lymphoma (LBL) accounts for approximately 2% of all non-Hodgkin lymphoma (NHL), and approximately 90% are of immature T-cell lineage. Precursor B-cell LBL (B-LBL) is uncommon and accounts for less than 10% of total LBL cases and 0.3% of adult NHL [3,4].

The disease presents mainly with extranodal involvement, sparing the bone marrow. The most commonly involved sites include the skin (33%), lymph nodes (22%), bone (19%), and mediastinum (5%), while hepatomegaly, splenomegaly, a leukemic picture, or involvement of the central nervous system have been reported infrequently [5]. The natural history of adult B-LBL has not been well defined because of its rarity.

Extranodal sites such as the skin and bone are often involved, but gastrointestinal lesions of

this disease are rarely encountered. Due to the infrequency, macroscopic forms of the gastrointestinal lesions have not been fully described.

They frequently present with advanced disease, B symptoms and elevated serum LDH levels. Abdominal involvement (liver and spleen) is unusual; however in this case the first symptom was jaundice⁵. Obstructive jaundice is often caused by benign diseases such as hepatolithiasis and cholangitis, and also by malignant diseases such as cholangiocarcinoma, periampullary cancer, liver cancer and pancreatic cancer, so malignant obstructive jaundice caused by duodenal papillary B-cell lymphoma is extremely rare [6].

In immunohistochemistry of B-LBL, tumor cells are always positive for B-cell markers CD19, CD79a, and CD22 and frequently express common acute lymphoblastic leukemia antigen CD10, CD 24, PAX5, and TdT in most cases. The expression of CD20, CD45, CD34, and CD99 is variable or even absent, like in this case [7].

In this report, after surgical resection and final pathologic diagnosis of B-LBL, the patient was treated with chemotherapy, standard

therapeutic option for patients with LBL is based on intensive multi-drug leukemia chemotherapy protocols [8]. These regimens contain 7-10 drugs, such as cyclophosphamide, methotrexate, prednisone, vincristine, cytarabine, thioguanine, l-asparaginase, nitrosoureas, etoposide, and anthracyclines [9]. Now, he is followed-up at the outpatient hematology clinic and in complete remission.

4. CONCLUSION

Obstructive jaundice caused by duodenal papillary lymphoma is extremely rare. Primary duodenal lymphomas present with initial nonspecific symptoms leading to delay in diagnosis. Hence, it is important to detect them at the earliest. Further studies on genetic and molecular profiling are necessary for understanding the pathogenetic mechanisms involved and for therapeutic and prognostic implications; establishment of histology, immunohistochemistry and molecular detection is essential for diagnosis and treatment. Different histological subtypes have different treatment and prognosis. Surgery plays an important role in resolving obstructive jaundice when accurate histological diagnosis cannot be made. After diagnosis, chemotherapy should play a central role in treatment.

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6. DECLARATIONS

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Ethical approval: The authors declare that patient consent was obtained, which is in accordance with the policies and standards of our institutional ethics committee. No patient-identifying information is presented in this article, thus anonymity is preserved. No animal and/or human experiments were performed.

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