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A Rare Case of Biliary Duct Lymphoblastic Lymphoma

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Abstract

Background: Lymphoblastic lymphoma (LBL) presenting with obstructive jaundice cases are rare. It is rarely considered in the differential diagnosis of patients with obstructive jaundice.
Case description: A young woman complained of jaundice, tea-colored urine, pale stools, and lost weight. The abdominal examination showed mild tenderness in the epigastric area. Laboratory showed elevation of liver function, bilirubin, and alkaline phosphatase. The radiological examination both in abdominal ultrasound and magnetic resonance cholangio-pancreatography suggested a possible differential diagnosis between of lymphoma and cholangiocarcinoma. She underwent surgery for open biopsy from biliary duct and showed lymphoblastic lymphoma. She was received combination of chemotherapy. She had significant improvement of her quality of life.
Conclusion: The diagnosis of biliary duct lymphoblastic lymphoma is essential but can be very challenging. Clinical diagnosis is based on symptoms and signs, while laboratory testing is non-specific and occasionally misleading. However, direct imaging and biopsy guidance is highly recommended.

Keywords: Biliary Duct; Lymphoblastic Lymphoma; Obstructive Jaundice

1. Introduction

Lymphoblastic lymphoma (LBL) is responsible for about 2% of all cases of non-Hodgkin's lymphoma (NHL). Among all causes of malignant obstruction of the biliary tract, NHL accounts for 1%-2% of cases Approximately 90% are derived from immature T-cells, with precursor B-cell LBL (B-LBL) accounting for just 10% of total LBL cases (Chaudhari et al., 2013).

The term LBL is used when the disease is confined to a mass lesion, including lymph nodes and extra nodal organs with no or minimal involvement of the bone marrow. Tumour cell markers were almost always positive for B-LBL, including CD19, CD22, and CD79a B cells. In many cases, tumour cells were also positive for CD10, CD24, PAX5, and TdT (Kim et al., 2014).

Lymphoblastic lymphoma is a high-grade type of lymphoma with a Disease-Free Survival of between 55%-95% (Brown et al., 2020). An initial approach to the patient, including a comprehensive history, physical examination, and appropriate laboratory testing, is essential to differentiate biliary duct lymphoma form other causes of obstructive jaundice because treatment and prognosis are fundamentally different (Constantinescu et al., 2018).

2. Case Description

Ms. S, 19 years old, came to Dr. Soetomo general hospital with a complaint of jaundice all over her body for 1 month. The patient reported tea-colored urine and pale stools. She also had fever most of the days and lost 5 kg in 8 months. She had no history of hepatitis, diabetes mellitus and hypertension. She had no previous history of travelling and consumption of particular drugs. The physical examination showed that the patient was in good general condition with stable vital signs. The height and weight were 153 cm and 52 kg, respectively. The head and neck examination revealed icterus as shown in figure 1a, while the abdominal examination showed mild tenderness in the epigastric region to the right upper quadrant of the abdomen. There was no evidence of liver or spleen enlargement and signs of liver chronicity. On laboratory examination, anemia was found with Hemoglobin (Hb) 8.0 g/dL, elevation of liver function aspartate aminotransferase (AST) 116 U/I, alanine aminotransferase (ALT) 93 U/I, also increasing of direct and total bilirubin were 10.1 mg/dL and 11.11 mg/dL respectively. She also had hypoalbuminemia 2.9 g/dL. Furthermore, HBsAg and anti-HCV were non-reactive, while Ca19-9, AFP, and ALP were 1.2 U/ml, 1.1 ng/mL, and 637 IU/L.

Abdominal ultrasound showed chronic parenchymal liver disease bilateral obstruction of the biliary tract in intrahepatic bile duct (IHBD), which can be a differential diagnosis of (1) Cholangiocarcinoma, (2) Mass of the head of the pancreas, and (3) Gastrointestinal Tumor (GIST) that extends to the common bile duct (CBD) or head of the pancreas. An abdominal magnetic resonance cholangio-pancreatography (MRCP) examination was done for the patient as shown in figure 1b. The results of abdominal magnetic resonance imaging (MRI), MRCP with and without contrast showed an enhancing solid mass with a necrotic area +/- 4.6 x 4.3 x 10 cm from the confluence of common hepatic duct (CHD) to distal CBD obstructing the proximal CHD. This causes mild dilatation of right and left IHBD, blocks and causes mild dilatation of the pancreatic duct and gallbladder hydrops with multiple subcentimeter lymph nodes in the mesenteric, para-aortic, aortic cava with suspicion of portal vein thrombus leading to a differential diagnosis of 1) lymphoma, and 2) cholangiocarcinoma type II. Patients with obstructive jaundice were suspected of having cholangiocarcinoma or biliary lymphoma and hypoalbuminemia. Subsequently, the patient was referred for a laparotomy biopsy before undergoing percutaneous transhepatic biliary drainage (PTBD).

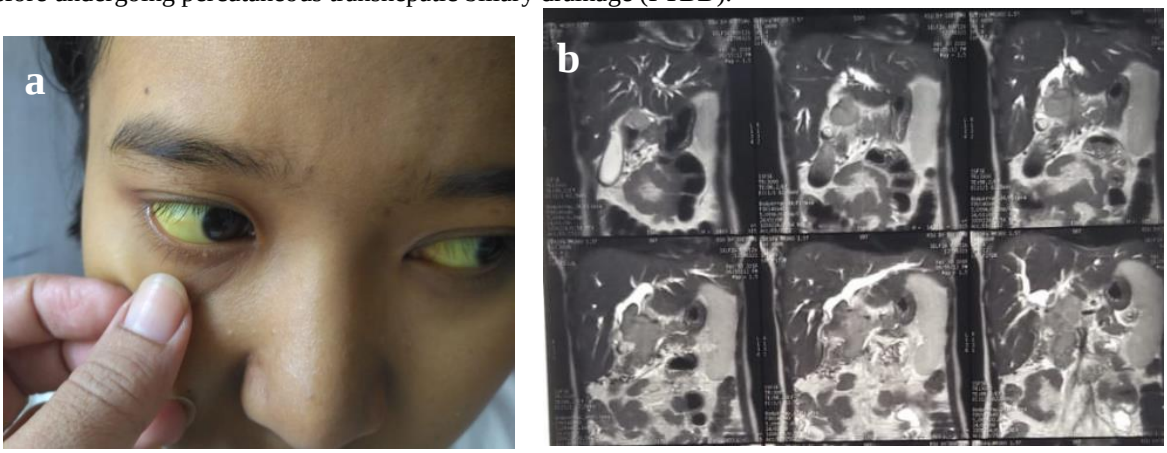

Fig 1. a) Clinical picture of the patient, jaundice of the sclera. b) Magnetic resonance cholagio-pancreatography (MRCP) before patient underwent chemotherapy.

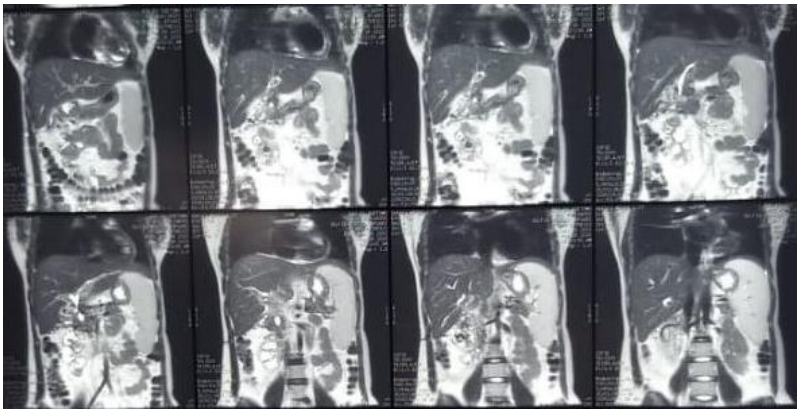

Figure 2. Magnetic resonance cholagiopancreatography (MRCP) after patient underwent 6 cycles of chemotherapy .

The results of an open biopsy of the common bile duct obtained a differential diagnosis, including (1) non-Hodgkin's lymphoma and (2) poorly differentiated carcinoma. The results of immunohistochemical examination with panCK antibodies were negative on tumor cell cytoplasm, CD45: positive on tumor cell cytoplasm, CD20: negative on the tumor cell membrane, CD3: negative on the tumor cell membrane, Ki67: 80% proliferation index, Tdt: diffuse positive in the tumor cell nucleus, CD 79a: positive on some tumor cell membranes. The patient was diagnosed with B-Lymphoblastic lymphoma Bile Duct, stage III BE, and later received chemotherapy with a combination of cyclophosphamide, doxorubicin, vincristine, and prednisone, with six cycles every 21 days.

After six cycles of chemotherapy, the complaints of fever and jaundice were no longer present. Furthermore, the appetite was improved, the weight increased by 3 kg in the past four months, and complaints of urination and defecation were within normal limits. Anemia was recovered to Hb 10.5 g/dL with thrombocytopenia 81,000, normal liver function AST 36 U/I, ALT 34 U/I were discovered during the patient's evaluation. Direct and total bilirubin levels were normal, at 0.12 mg/dL and 0.5 mg/dL, respectively.

The results of the post-chemotherapy evaluation on abdominal magnetic resonance cholangio-pancreatocraphy (MRCP) showed that a solid mass with necrotic area inside the border is not clear, edges are irregular, 2.5 x 3 x 7.7 cm in size from the confluence of common hepatic duct (CHD) to distal common bile duct (CBD) with hypointense predominant around the hilar bifurcation, and there was homogenous contrast enhancement. A mass obstructs the CHD at its proximal end, resulting in mild dilatation of the right and left IHBD, pressing into the duodenum, with prominent blood vessels surrounding the mass. Compared with the previous contrast MRCP, a residual mass of 2.5 x 3 x 7.7 cm was obtained from CHD confluence to distal CBD (predominant around the hilar bifurcation) with details of expansion and compression above (reduced impression). Lymphmnode was a subcentimeter in the paraaortic. The patient was diagnosed with biliary duct B-lymphoblastic lymphoma stage III BE post-chemotherapy with regimen combination of cyclophosphamide, doxorubicin, vincristine, and prednisone (CHOP) for 6 cycles with partial response.

3. Discussion

Jaundice increases bilirubin concentration, causing clinical manifestations at the sclera and skin. It can occur secondary to obstruction to the flow of the bile ducts, either wholly or partially, and is related to liver disorders that affect the digestive tract (Briggs and Peterson, 2007).

Based on several case reports in the patient with obstructive jaundice, clinical presentations include itching all over the body, yellowish discoloration of the sclera, dark urine (tea color), white-gray stools, weight loss, and complaints of malaise (Zakaria et al., 2017). Ultrasound investigations help find biliary

dilatation, differentiating extrahepatic cholestatic or obstructive jaundice from intrahepatic causes (Briggs and Peterson, 2007). Magnetic resonance cholagio-pancreatography (MRCP) as a follow-up modality after ultrasonography provides excellent detail of the pancreatic and biliary tract (Blakeborough and Thomas, 2003). Several case reports have shown that the appearance of biliary duct lymphoma on MRI is similar to cholangiocarcinoma. Meanwhile, there are intrahepatic and extrahepatic biliary dilation and abnormal enhancement of the bifurcation of the common hepatic duct (Durham et al., 2017).

Primary non-Hodgkin's lymphoma arising from the bile duct is extremely rare, and the reported imaging features do not differ from that of cholangiocarcinoma (Yoon et al., 2009). However, the management and prognosis for cholangiocarcinoma and lymphoma are different, and then a biopsy is indispensable to confirm the diagnosis. Treatment for cholangiocarcinoma is surgical resection or gemcitabine-based chemotherapy. On the other hand, lymphomas are more chemo-responsive using the combination of Rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone (R-CHOP) regimen. Therefore, the diagnosis carries a better overall prognosis than cholangiocarcinoma (Durham et al., 2017).

Management of LBL may use an ALL regimen, although many adults are treated with regimens for intermediate lymphomas such as cyclophosphamide, doxorubicin, vincristine, and prednisone (CHOP), where response rates range from 55% to 95% (Cortelazzo et al., 2011). The therapy response can be evaluated based on survival, objective response, subjective improvement, and life quality. Based on the International Prognostic Index, the patient's condition was classified as high risk with a 5-year Disease-Free Survival of 49%. One of the goals of cancer therapy is to enable patients to live standard life quality. Measurable tumor regression can be a determinant of treatment benefit. Tumor regression can be determined by decreasing the size. Subjective changes with acceptable quality of life are often far more important to the patient than objective responses (Skeel, 2011).

4. Conclusion

A patient was admitted to the hospital with complaints of jaundice all over the body, tea-colored urine, fever, weight loss, and pale stools. Based on the laboratory and radiology evaluation supporting the suspicion of obstructive jaundice with consideration towards malignancy, the differential diagnosis of cholangiocarcinoma and extra nodal lymphoma was obtained. Confirmation of the diagnosis by biopsy specimens greatly determines the management and prognosis of the disease. Therefore, the patient has performed a laparotomy on the bile duct resulting from lymphoblastic lymphoma. Patients underwent chemotherapy for six cycles every 21 days with a regimen of cyclophosphamide, doxorubicin, vincristine, and prednisone. It showed a reduced tumor response up to 45% compared to before. In addition, subjective changes based on complaints and the patient's life quality showed significant improvements, such as the condition before the disease.

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