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Primary extrahepatic bile duct lymphoma mimicking Klatskin's tumor, dramatic response to chemotherapy

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ABSTRACT

INTRODUCTION: Non- Hodgkin's lymphoma (NHL) mimicking Klatskin's tumor is rare but possible. It can be confused with tumors for which major surgery is needed. Imaging modalities have similar features in Klatskin's tumor and non-Hodgkin's lymphoma.
PRESENTATION OF CASE: We herein report a patient who was first thought having a Klatskin's tumor and prepared for major surgery. A month later, obstructive jaundice developed and a second magnetic resonance-magnetic resonance cholangiopancreotography showed a doubling size of the tumor in one month. In terms of tumor behavior, lymphoma was suspected and a tru-cut biopsy was performed by interventional radiology. Pathological examinations revealed non-Hodgkin's lymphoma large B-cell type. After 4 cycles of chemotherapy by hematology-oncology department, complete cure was achieved.
DISCUSSION: Since the radiologic and clinical features are confusable in Klatskin's tumor and non-Hodgkin lymphoma. Tumor biologic behavior and other biochemical parameters together may put a suspicion in mind. Upon suspicion biopsy must be done radiologically.
CONCLUSION: Primary liver lymphoma arising from the bile ducts is extremely rare and radiologic imaging features do not differ from cholangiocarcinoma. A strong suspicion may prevent unnecessary surgery. Since the treatment of non-Hodgkin's lymphoma is mainly chemotherapy.

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1. Introduction

Non-Hodgkin's lymphoma (NHL) mimicking Klatskin's tumor is a relatively rare clinical entity. In the initial staging of systemic NHL, liver involvement is detected in 40% of patients. However, primary liver NHL is less than 1% of all NHL's [1]. In the literature, primary NHL of extrahepatic bile ducts was first reported by Nguyen in 1982 [2,3]. Features of imaging studies are not clear to differentiate NHL from cholangiocarcinoma. Major surgery is performed in most of patients with presumptive diagnosis of cholangicarcinoma [4]. In the literature only 20% of primary NHL of liver is diagnosed without surgery and patients are treated with chemotherapy [5].

The clinical picture is similar to Klatskin's tumor. Jaundice, right upper quadrant pain, weight loss are the most common complaints. Fever, nightsweats, fatigue of lymphomas can also be presented by these patient [1].

2. Presentation of Case

A 58 year- old female patient with type 2 diabetes mellitus and hypertension was checked for routine follow up with biocemical parameters and abdominal ultrasonography. A liver hilus mass with 2 cm in diameter was detected and abdominal magnetic resonance and magnetic resonance cholangiopancreotography (MRCP) was taken upon detection. MR and MRCP showed a 35 × 31 mm mass at liver hilus with intrahepatic biliary dilatation.

Laboratory findings: Total Bilirubin(TB): 0.4 mg/dl, Direct Bilirubin(DB): 0.1 mg/dl, Aspartat aminotransferase (AST): 70 U/L, Alanine aminotransferase(ALT): 137 U/L, Alkaline phospatase (ALP): 388 U/L, Lactate dehydrogenase(LDH): 237 U/L, Gama glutamyl transferase(GGT): 132 U/L, CA19–9: 51 (0–35), WBC: 5900 /mm^3 HCT: 36,Other tumor markers were within normal limits. Markers for hepatitis A, C were all negative.

The patient was thought having a Klatskin's tumor and prepared for major surgery. At that time, obstructive jaundice developed (TB: 8.4 mg/dl, DB: 5 mg/dl). A second MR-MRCP was taken (about one month later from the first MR-MRCP). A 50 × 30 mm mass extending from the left liver lobe entrance to the hilus, was detected. The mass was seperating completely central main biliary ducts and

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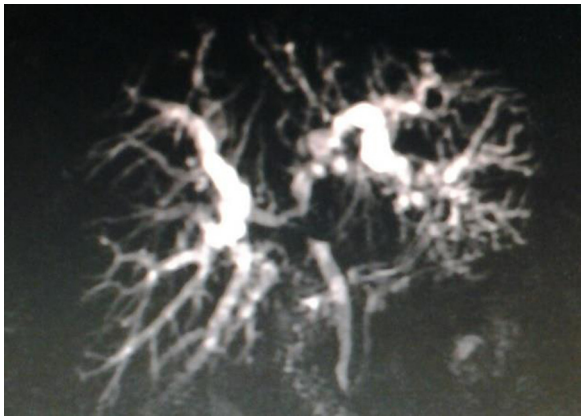


Picture 1. Seperated bile ducts.

causing dilatations of intrahepatic biliary ducts. Common hepatic duct couldn't be seen in 1 cm from the biliary confluence (Picture 1). Contrast enhanced diffusion series suggested a cholangiocarcinoma. In addition, multiple lymph nodes (the largest 20 × 10 mm) in hilus and in front of inferior vena cava, were detected intraabdominally.

Tumor mass had been doubled and suspicious lymph nodes had developed intraabdominally in one month. A type of lymphoma was suspected upon concerning biological behavior of the tumor and the patient was referred for PET-CT and biopsy after biliary drainage with 3 percuteneous transhepatic cholangiography (PTC) catheters into right anterior, right posterior and left bile ducts.

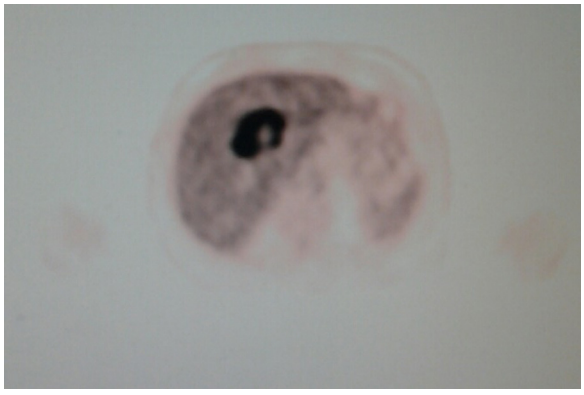
After biliary drainage the labarotuary values were TB: mg/dl, DB: 0.9 mg/dl,AST: 86 U/L, ALT: 132 U/L, ALP: 429 U/L, GGT: 115 U/L.

PET-CT at that time revealed a tumoral mass of 50 × 35 mm in dimensions with a FDG involvement of SUD MAX: 19 (Picture 2).

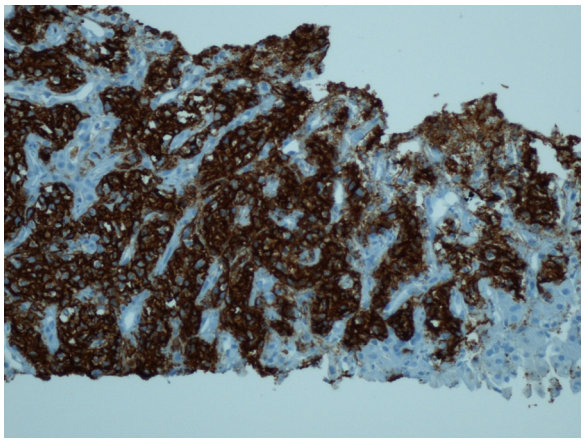
A tumor tru-cut biopsy was performed by interventional radiology department and pathological examinations revealed as highgrade large B-cell NHL with CD 20(+) staining and Ki-67 proliferation index: 60% (Picture 3).

The patient was referred to hematology and oncology department and treated with 4 cycles of R-CHOP (rituximab/cyclophosphamide/doxorubicine/vincristine/prednisolone).

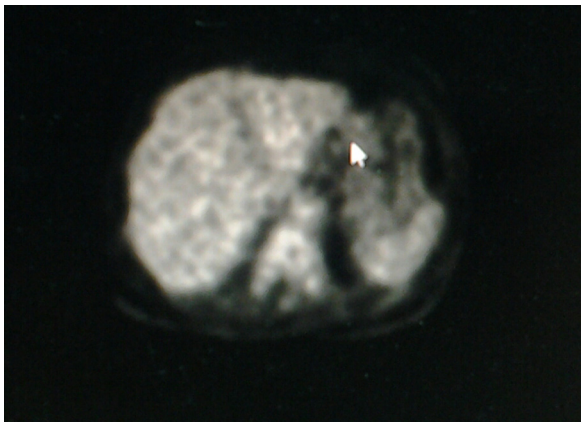
Complete regresion was achieved 5 months later with control PET-CT (Picture 4) and MR-MRCP (Picture 5). PTC catheters were clamped a week and taken out after the control laboratory values which were all within normal limits.



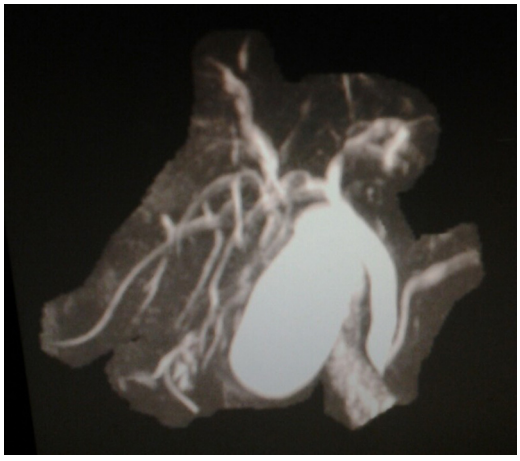
Picture 2. PET-CT of tumor.



Picture 3. Ki-67 proliferation index 60%.



Picture 4. Complete cure after chemotherapy in PET-CT.



Picture 5. Normalised anatomy of bile ducts in MR-MRCP.

The patient is followed up routinely with 4 month- intervals for a year. Recurrence has not been detected.

3. Discussion

Extrahepatic biliary primary NHL's have similar features and findings with cholangiocarcinoma in terms of imaging modalities, clinical and laboratory findings [4]. In the literature there are no results enough to make a differential diagnosis. However, in lymhomas LDH level is high and CA 19–9 level is normal. As in our

case, -when there is a rapid growing of tumor into a doubling size in one month, it must be kept in mind that a lyphoma may arise from the biliary system. Most of cases are pathologically diagnosed after major surgery and chemotherapy is continued. An accurate diagnosis can prevent unnecessary surgical intervention.

4. Conclusion

Primary liver lymphoma arising from the bile ducts cannot be differentiated radiologically from Klatskin's tumor. In terms of tumor behavior, if there is a rapid growing of tumoral mass, it must be kept in mind that it would be a lymphoma. Since the treatment is totally different in lymphomas, unneccessary surgery can be prevented upon suspicion.

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