Session II

25-year-old Man with Obstructive Jaundice

Jae Hyun Lim, Huapyong Kang, Jung Hyun Jo, In Rae Cho, Hee Seung Lee, Jeong Youp Park, Seungmin Bang, Seung Woo Park, Si Young Song, Moon Jae Chung

Division of Gastroenterology, Department of Internal Medicine, Yonsei University of College of Medicine, Seoul, Korea

1. Case presentation

A 25-year-old man presented jaundice and right upper quadrant abdominal discomfort for 14 days. He had no medical history and vital signs were normal on admission. Physical examination revealed the patient had icteric sclera and skin. His abdomen was soft, non-distended, and there was mild direct tenderness to palpation of the right upper quadrant without rebound tenderness or guarding.

Initial laboratory studies yielded the following abnormal results: AST 86 IU/L, ALT 224 IU/L, Total bilirubin 9.5 mg/dL, Direct bilirubin 7.4 mg/dL, Gamma-glutamyltransferase 227 IU/L, CA 19-9 172.0 U/mL, and ANA Positive (1:320). Following results were normal; CEA 3.12 ng/mL, IgG Quantitation 1252 mg/dL, Subclass IgG4 564.0 mg/L.

On Abdomen-pelvic CT and MRCP, Ill-defined, hypovascular soft tissue infiltration around the hilar duct, obliterating CBD, right secondary confluence and cystic duct was noted. After that, 22 mm sized polypoid lesion was seen at cystic duct on EUS and asymmetric bile duct wall thickening from CHD to Lt. IHD was noted on ERCP with IDUS (intraductal ultrasonography). Bland looking epithelial cells were noted and IgG 4 stain was negative on CHD biopsy by ERCP.

When we reviewed his history and examined him again, a 5 cm sized, fixed, round shape mass was noted on his sternum. Multiple FDG uptakes involving liver, spleen, multiple lymph nodes, and bones were seen on PET-CT. Therefore, lymphoma was highly suspected as final diagnosis.

Finally, diffuse large B-cell lymphoma confirmed by incisional biopsy. After that, the patient treated with R-CHOP chemotherapy from 14-Feb-2018~18-Feb-2018. At discharge, his total bilirubin level was normalized, 1.3 mg/dL.

2. Diagnosis

Diffuse large B-cell lymphoma

3. Therapy and Clinical course

1st R-CHOP chemotherapy was done

Obstructive jaundice was resolved

4. Conclusion

Lymphoma should be considered as one of the differential diagnosis of patients with unusual obstructive jaundice.

Key Words: Malignancy of bile duct, Lymphoma

Case

- **M/25**
- **Chief complaint**
  RUQ abdominal discomfort, Visible jaundice
- **Duration** : 14 days
- **Present illness**
  내과적 특이 과거력 없던 25세 남자 환자가 RUQ abdominal discomfort 및 Jaundice를 주소로 타 병원 내원, CT (2018.01.18.)결과 Biliary obstruction 및 Total bilirubin 9.8 소견 보여, further evaluation 위해 본원 내원함.

Case

- **Past medical history**
  None
  No medical history
- **Social history**
  Smoking : 0.1P/d * 5yrs = 0.5PYs
  Alcohol : Non-drinker
- **Review of systems**

Generalized Weakness (-)	Fever (-)
Nausea(-)	Vomiting(-)
Visible jaundice (+)	**Abdominal discomfort, RUQ (+)**

Case

- **Physical examination**

  Mental status : Alert

  General appearance : Not ill looking appearance

  Soft and flat abdomen

  Abdomen direct tenderness (+) at RUQ

  Abdomen rebound tenderness (-)

- **Vital sign**

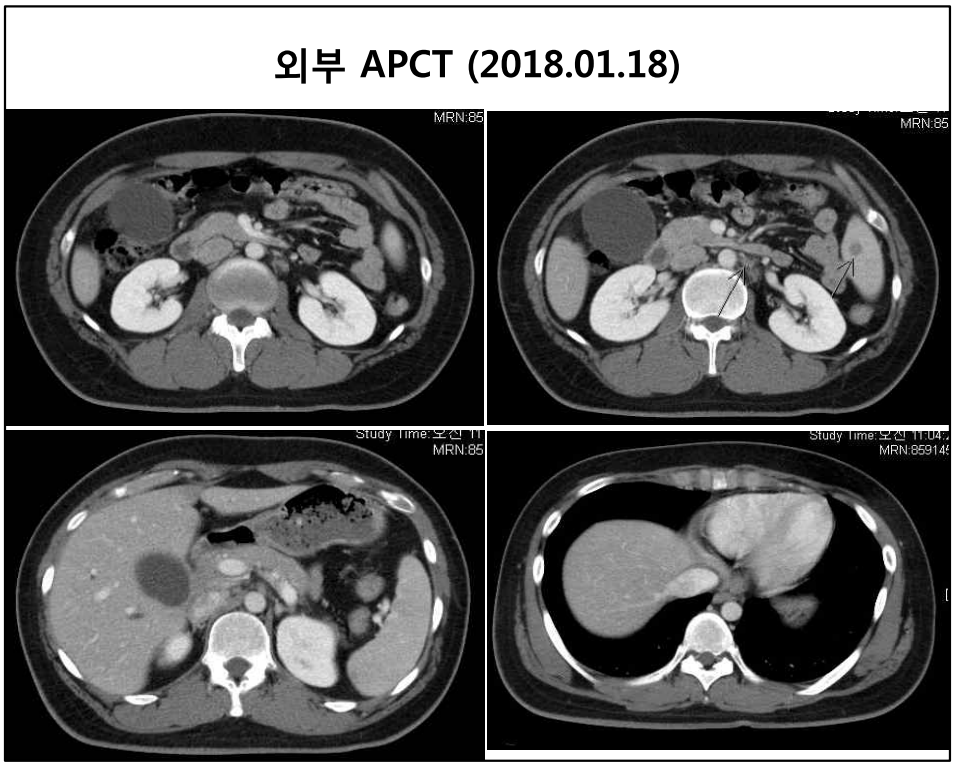
  BP 141/82 mmHg PR 84/min

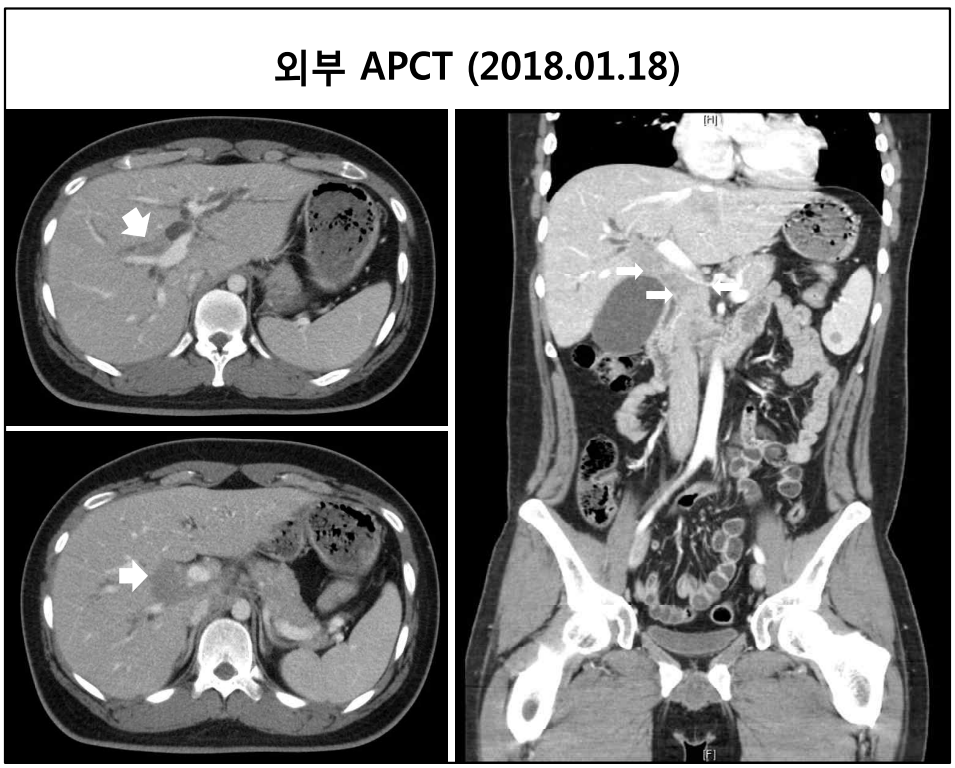
  RR 20/min BT 36.2 ºC

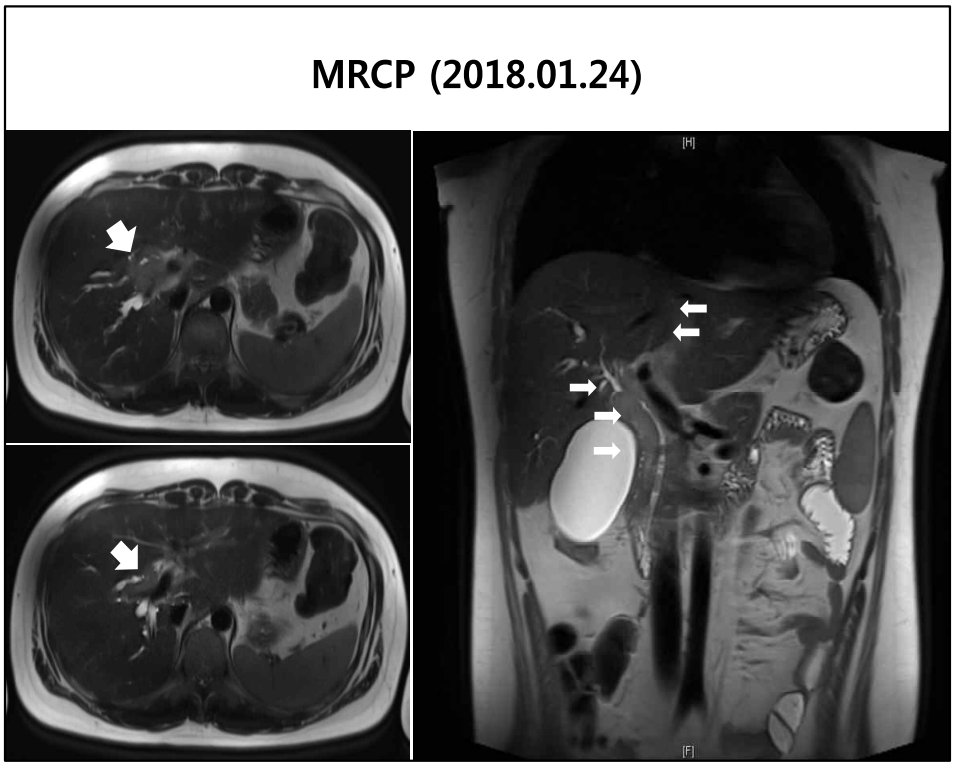
Case

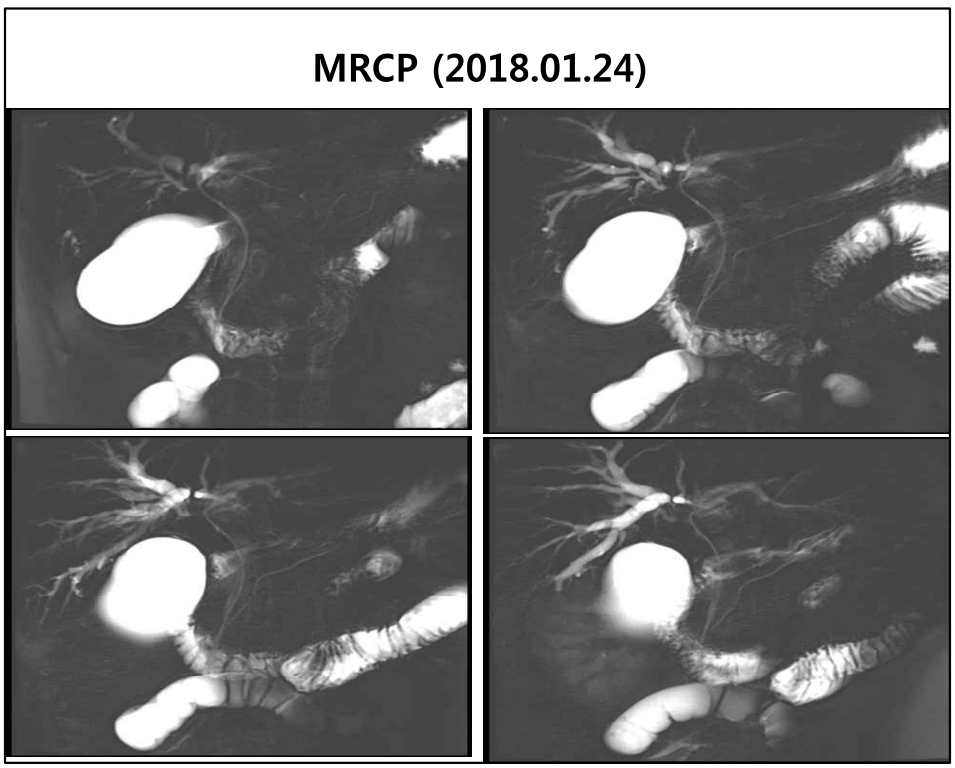
- **Laboratory study**

WBC (ANC)	4430/uL (3180/uL)
Hb	15.3g/dL
PLT count	258000/uL
Na/K/Cl/tCO2	138/4.8/97/23 mmol/L
BUN/Cr	14.2/0.55 mg/dL
Albumin	4.2 g/dL
LDH	211 IU/L
AST/ALT	**86/224 IU/L**
Total bilirubin	**9.5 mg/dL**
Direct bilirubin	**7.4 mg/dL**
Gamma-GT	**227 IU/L**
CEA	3.12 ng/mL
CA 19-9	**172.0 U/mL**
PT/aPTT	0.92/24.2
ANA	**Positive (1:320)**
P-ANCA/C-ANCA	Negative
IgG Quantitation	1252 mg/dL
Subclass IgG4	564.0 mg/L


외부 APCT (2018.01.18)


외부 APCT (2018.01.18)


MRCP (2018.01.24)
[H]
[F]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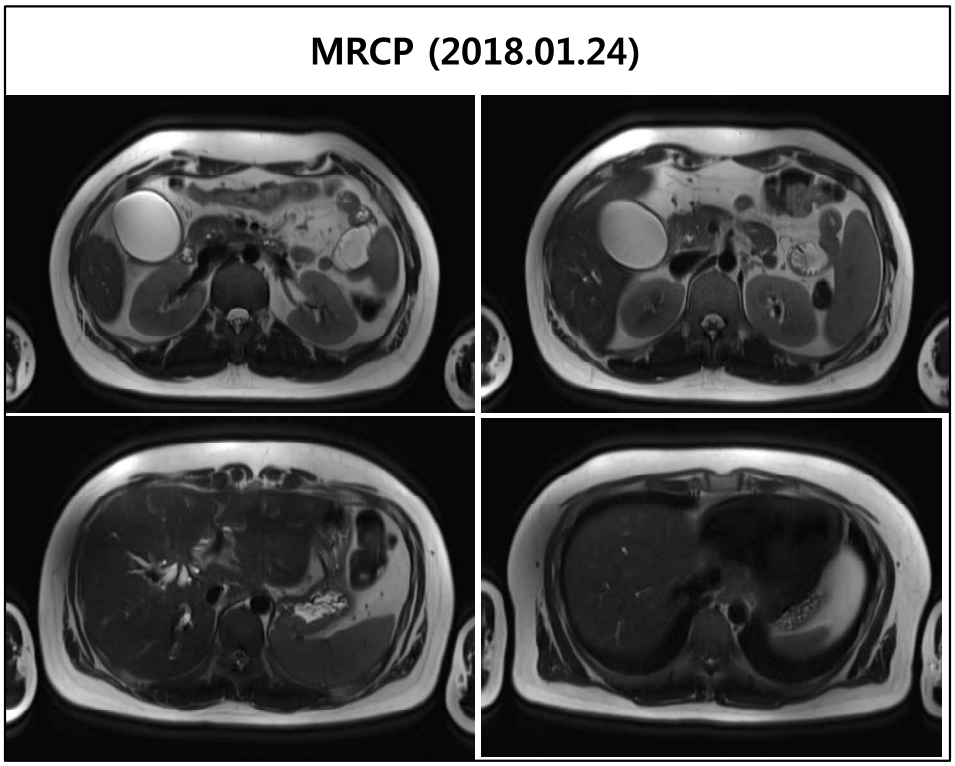
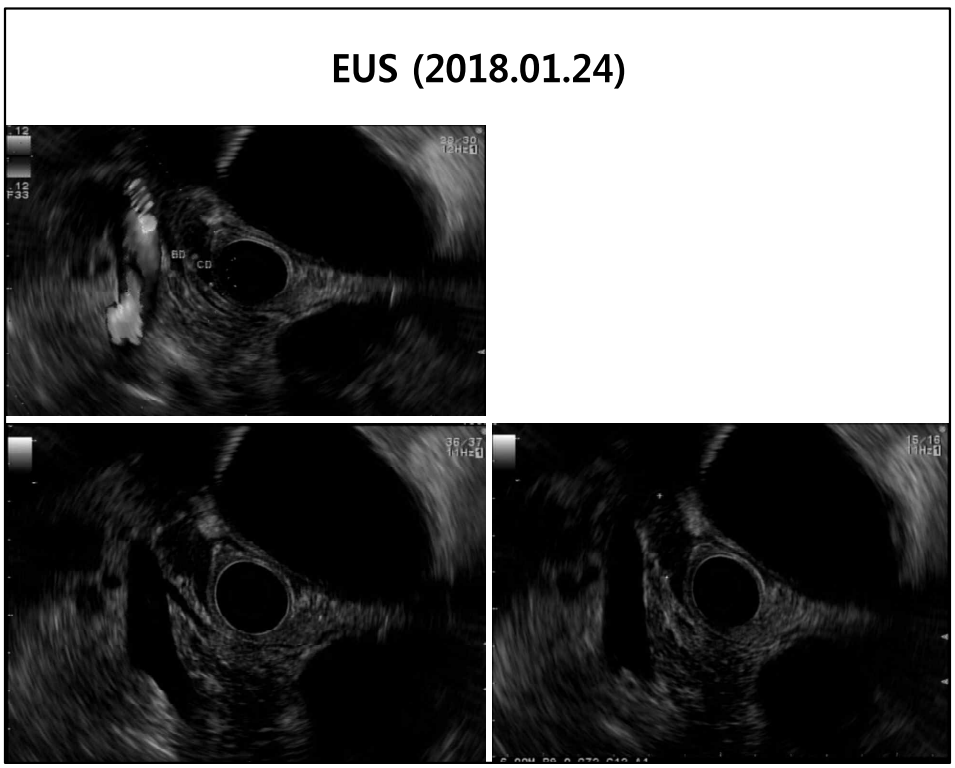
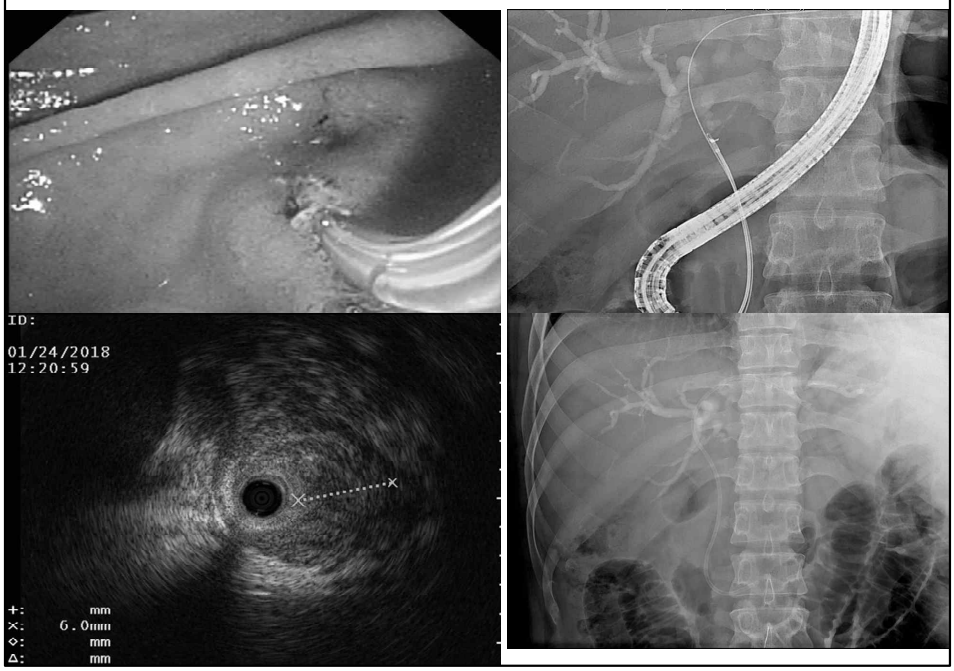
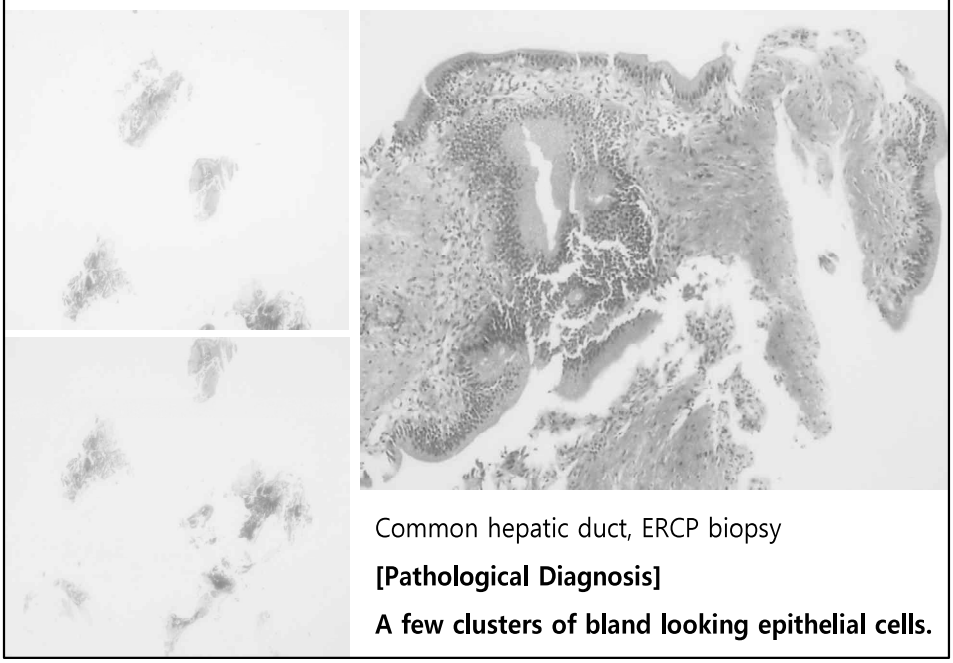

MRCP (2018.01.24)


EUS (2018.01.24)

ERCP (2018.01.24)



Pathology (2018.01.28)



Common hepatic duct, ERCP biopsy

[Pathological Diagnosis]

A few clusters of bland looking epithelial cells.

Progress

Diagnosis

R/O Lymphoma with biliary tract involvement

Plan

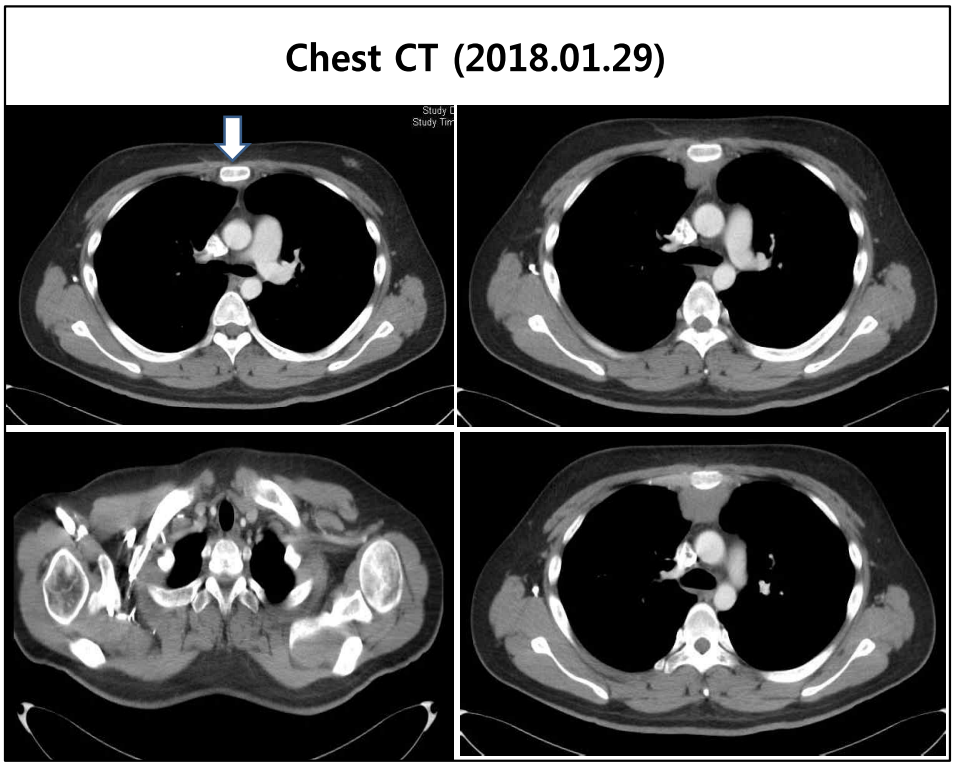
Re-biopsy

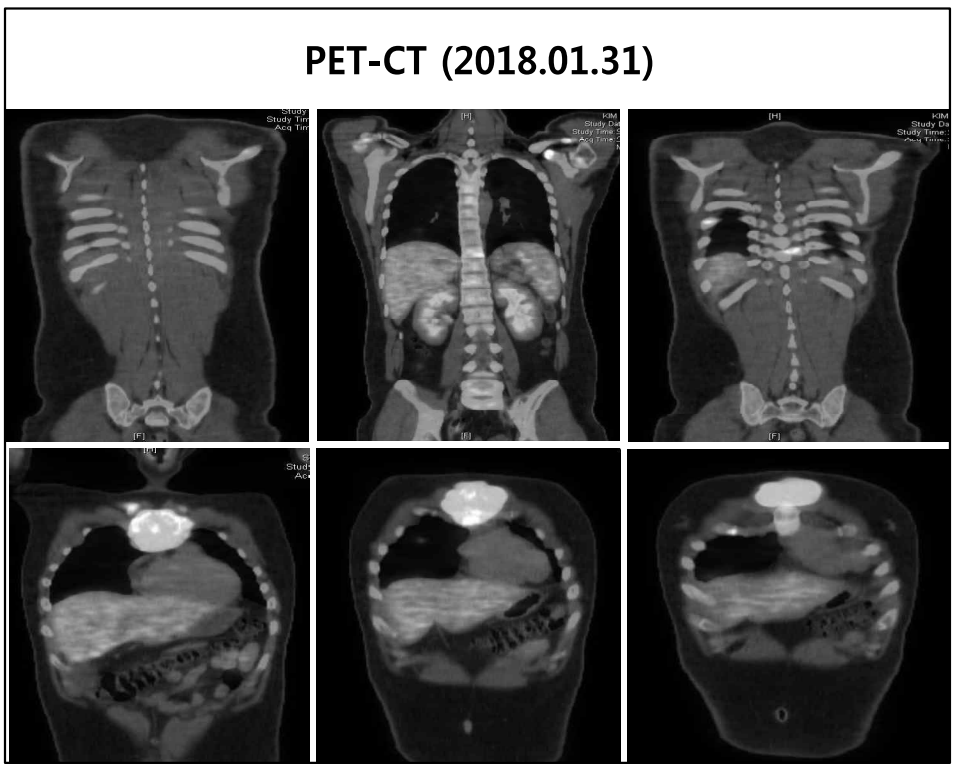
Hematology consultation

Progress

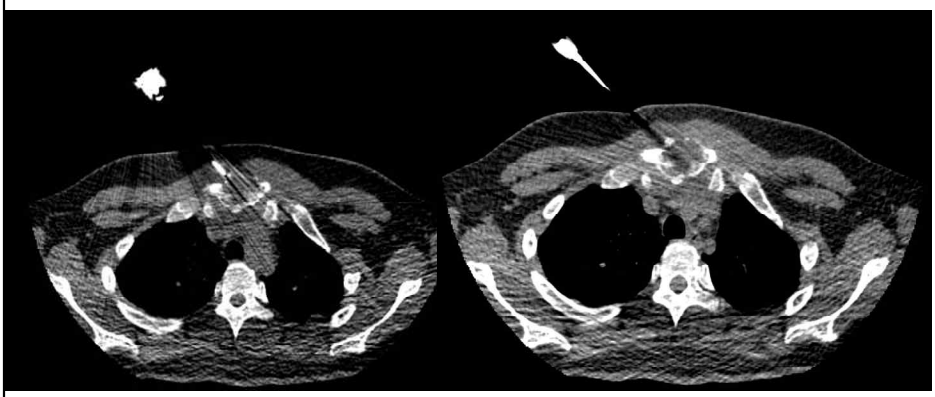
- P/E re-evaluation
  - Palpable cervical area LN (+)
  - **Palpable mass at sternal area, about 6 cm**

➢ Chest CT, PET CT evaluation


Chest CT (2018.01.29)

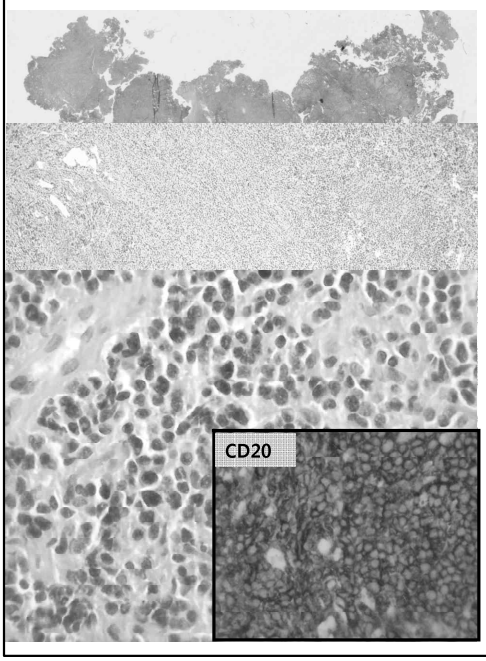

PET-CT (2018.01.31)

CT-guided sternal biopsy (2018.02.01)



Impression
Mainly blood clot with a few CD20+. CD3-, CD30-, TdT- monotomous lymphoid cells
Non-diagnostic, but B cell lymphoma suspected
Rec> re-biopsy

Incisional biopsy of sternal mass (2018.02.08)

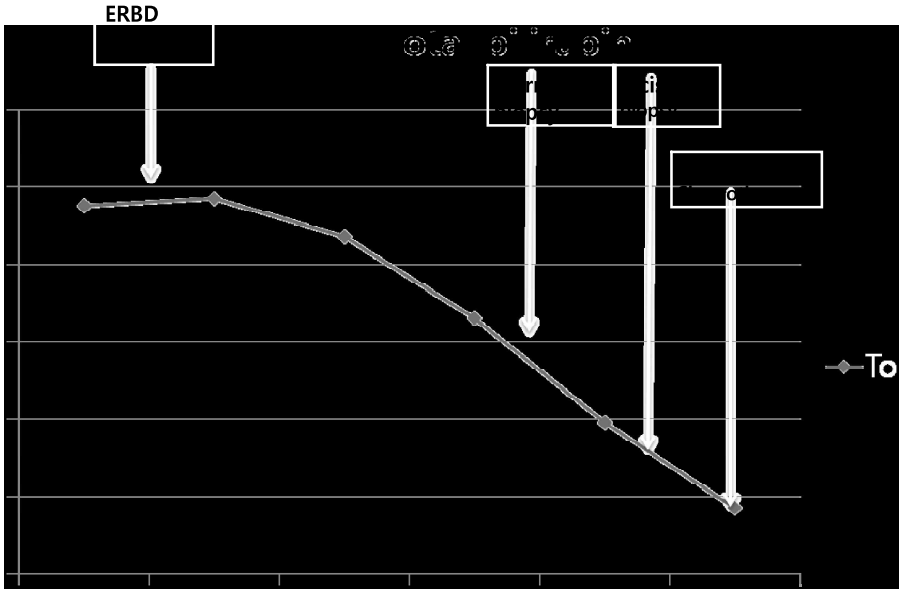


Soft tissue, anterior chest wall, incisional biopsy

[Pathological Diagnosis]
Diffuse large B cell lymphoma

Note)
1. The immunohistochemical staining results:
CD20 (L26): Positive in tumor cells
CD10 (CALLA): Negative in tumor cells
Bcl-6 : Positive in 5% of tumor cells
MUM1 : Positive in 10% of tumor cells
Bcl-2: Positive in 70% of tumor cells
C-myc : Positive in 30% of tumor cells
CD3 (T-cell): Negative in tumor cells
Ki-67 L.I.: almost 100%
2. EBER in situ hybridization: Pending
3. Favored subtype: Diffuse large B cell lymphoma, not otherwise specified, non-germinal center B cell-like type, see note
4. Immunohistochemical double expressor: Not identified

Progress



Final diagnosis

- Final diagnosis
  - **Diffuse Large B-cell Lymphoma with biliary tract involvement**

- Treatment
  - R-CHOP Chemotherapy (2018.02.14.~02.18)
    - Rituximab 375 mg/m^2, D1
    - Cyclophosphamide 750 mg/m^2, D1
    - Doxorubicin 50 mg/m^2, D1
    - Vincristine 1.4 mg/m^2, D1
    - Prednisolone 100 mg, D1-5

Jaundice and lymphoma

Jaundice concomitant with lymphoma

- primary hepatic lymphoma
- direct hepatic involvement
- extrahepatic bile duct obstruction due to various causes
  (ex. enlarged lymph nodes)
- intrahepatic cholestasis, such as vanishing bile duct syndrome caused by HD

Dig Dis Sci. 2007 Sep;52(9):2323-32. Epub 2007 Apr 4.
Biliary tract obstruction secondary to malignant lymphoma: experience at a referral center. Odemiş B1, Parlak E, Başar O, Yüksel O, Sahin B.

Dig Dis Sci. 2010 Nov;55(11):3271-7. doi: 10.1007/s10620-010-1310-6. Epub 2010 Jul 15.
Outcomes in lymphoma patients with obstructive jaundice: a cancer center experience.
Ross WA1, Egwim CI, Wallace MJ, Wang M, Madoff DC, Lee JH.

Jaundice and lymphoma

- Lymphoma - rare cause of extrahepatic obstruction.
- Approximately 1.3% of patients with lymphoma
  → extrahepatic biliary obstruction
  (0.5% for HD and 0.2–2% for NHL)
- Among all causes of malignant biliary obstruction, NHL : 1–2%
- Therefore, lymphoma, should be considered in the differential diagnosis of malignant bile duct obstruction, particularly in younger patients.

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Jaundice and lymphoma

Obstruction of the extrahepatic bile duct by lymphoma

- primary NHL arising from the extrahepatic bile duct
- primary involvement of the CBD by HD
- compression of the extrahepatic bile duct by enlarged lymph nodes
- Primary lymphoma arising from the porta hepatis
- Primary lymphoma of the ampulla of Vater
- primary lymphoma of the pancreas involving the CBD by direct extension
- GI lymphoma obstructing the bile duct at the ampulla of Vater

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Jaundice and lymphoma

- Extrahepatic biliary primary NHL's have similar features and findings with cholangiocarcinoma in terms of imaging modalities, clinical and laboratory findings
- However, in lymhomas LDH level is high and CA 19–9 level is normal.
- lymphoma arising from the bile ducts cannot be differentiated radiologically from Klatskin's tumor

International Journal of Surgery Case Reports journal
Primary extrahepatic bile duct lymphoma mimicking Klatskin's tumor, dramatic response to chemotherapy
Cakir Mikaila,*, Tuzun Sefaa, Karsidag Tamera, Savas Osman Anila, Yegen Gulcinb

Jaundice and lymphoma

- Biliary obstruction in lymphoma usually appears in the advanced stage of the disease.
- The prognosis of patients with biliary obstruction due to lymphoma appears to be generally poor.
- Chemotherapy is the mainstay of treatment for patients with biliary obstruction due to lymphoma
- Stricture frequently resolves with chemotherapy in lymphoma, the need for a permanent metal stent is less prevalent than with other more refractory causes of malignant biliary obstruction

  → should be treated with a plastic stent

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