

A Case of Pancreatic Neuroendocrine Tumor with Massive Cystic Degeneration Mimicking Pancreatic Pseudocyst

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1. Case presentation

55세 남자 환자로 chronic alcoholics이며, 췌장 가성 낭종 의심 하에 15개월 전부터 6개월 간격으로 APCT 진행하며 외래 추적 받던 중 췌장 낭종 크기가 증가하여 입원하였다. EUS에서 cyst wall이 매우 두꺼운 unilocular cyst 관찰되었고, 내부의 다량의 echogenic material이 동반되었다. Cyst 내에 solid nodule은 없었으며, p-duct와의 communication도 관찰되지 않았다. 혈청 CA 19-9, amylase, lipase는 모두 정상범위였다.

2. Diagnosis

Pseudocyst ⇒ Neuroendocrine Tumor of the Pancreas with Cystic Degeneration

3. Therapy and Clinical course

입원하여 EUS-guided fine needle aspiration을 시행하였고, 약 10 cc 가량의 chocolate color cystic fluid를 흡입하였다. Pseudocyst 의심 하에 EUS-guided cyst drain을 시도하였으나 cyst wall이 두꺼워 fistula dilatation이 어려웠고, 시술 중 환자가 극심한 통증을 호소하여 시술 중단하였다. EUS 시행 후 환자는 지속적인 심한 복통을 호소하였고, 외과적 응급 수술을 계획하였다. 복강경에서 EUS puncture site에 active bleeding 동반한 hemoperitoneum 소견 관찰되어 laparoscopic distal pancreatectomy를 시행하였다. 수술 후 시행한 병리 조직검사 상 Neuroendocrine tumor, G1 with cystic degeneration more than 95%로 확인되었다. 수술 후 3개월이 지났고, 주기적으로 APCT 시행하며 외래 추적 중이며, 재발의 증거는 없다.

4. Conclusion

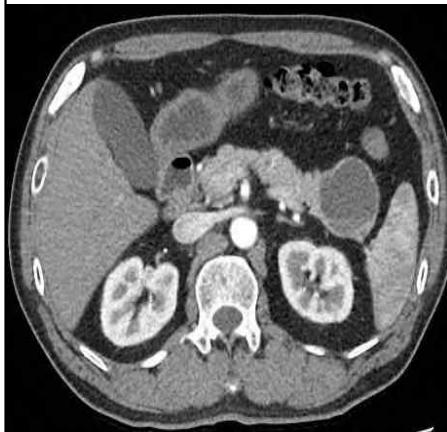
본 증례는 가성낭종으로 오인되어 EUS-guided drain까지 시도했던 췌장신경내분비종양의 케이스로, 췌장 낭성종양의 진단에 있어 고형종양의 낭성 변화 가능성을 염두에 두어야 할 것이다.

Key words: Pancreatic neuroendocrine tumor, Cystic degeneration, EUS

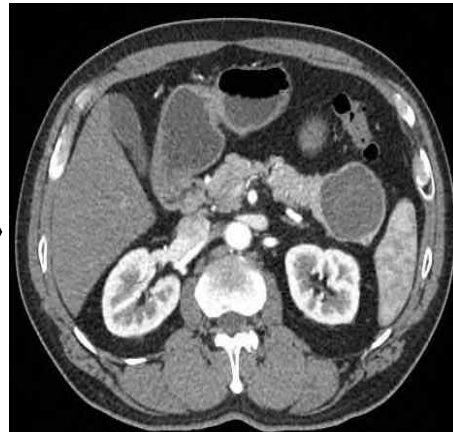
55 / M

- ▶ C.C) Increased size of a cystic mass in the pancreatic tail
- ▶ P/I) Chronic alcoholics로 기저질환 gout 있으며, 1년 전 시행한 APCT에서 우연히 발견된 r/o pancreatic pseudocyst로 6개월 간격 APCT f/u 하며 외래 추적 하던 분으로, 올해 8월 f/u한 APCT 상에서 pancreatic tail 부위의 cystic mass에 크기 증가 소견을 보여 EUS-guided pseudocyst drainage 위하여 입원하였음 .

APCT f/u

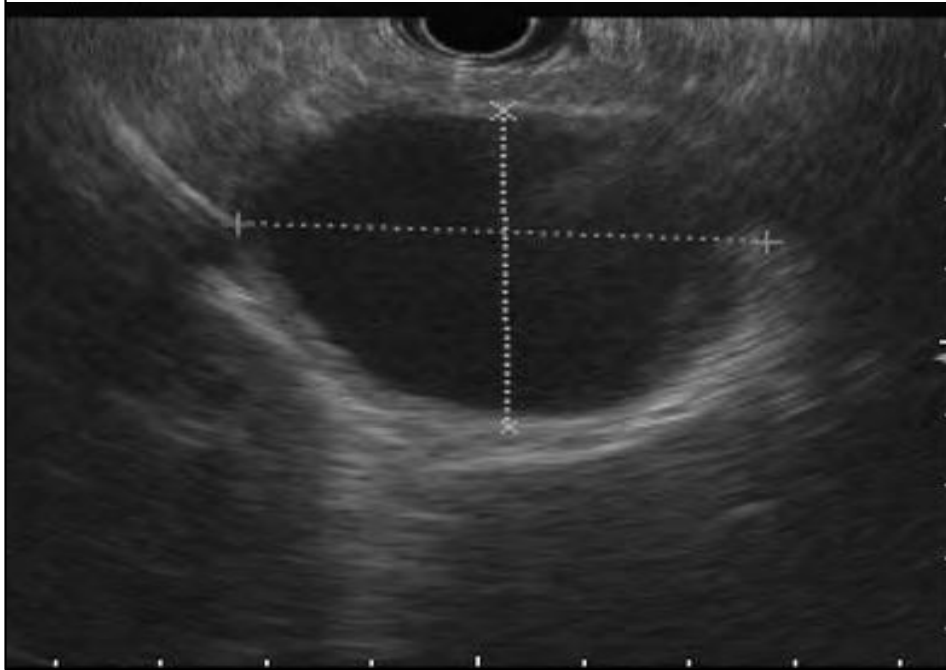


2015.05.13 4.7x4.6 cm



2016.08.01 5.3x5.0 cm

EUS (2015.7)



Past History

- ▶ HTN/DM/Hepatitis/Tbc (-/-/-/-)

Social History

- ▶ Smoking : Ex-smoker (30년 x 반 갑 = 15갑년)
- ▶ Alcohol : Chronic alcoholics (주 5회 x 소주 1병)
- ▶ Occupation : 회사원

Familial History

- ▶ 부모님 : HTN

Review of system

▶ **General**

- General weakness / Fatigue (-/-)
- Headache / Dizziness (-/-), Febrile sense/chilling (-/-)
- Night sweat (-), Myalgia (-)

▶ **Cardiovascular system**

- Chest pain (-)
- Palpitation (-)
- Orthopnea (-)
- Paroxysmal nocturnal dyspnea (-)

▶ **Respiratory system**

- Cough / Sputum / Rhinorrhea (-/-/-)
- Hemoptysis / Dyspnea / Wheezing (-/-/-)

Review of system

▶ **Gastrointestinal system**

- Abdominal pain (-)
- Anorexia / Nausea / Vomiting / Diarrhea / Constipation (-/-/-/-)
- Hematemesis/ Melena/ Hematochezia (-/-/-)
- Dyspepsia (-)

▶ **Genitourinary system**

- Frequency/Dysuria/Voiding difficulty(-/-/-)
- Nocturia / Foamy urine (-/-)

▶ **Neuromuscular system**

- Motor weakness (-)
- Numbness / Paresthesia(-/-)
- Arthralgia (-) / stiffness (-)

Physical examination

▶ Vital sign

- BP : 139/81 mmHg
- BT : 36.8 °C
- HR : 68/min
- RR : 20/min

▶ General

- Mental alert
- Not ill looking appearance

▶ HEENT

- L/R (++)/ (++), isocoric pupil
- Pinkish conjunctivae, whitish sclerae
- Jugular vein engorgement (-/-)
- Palpable Cervical lymph node (-/-)

Physical examination

▶ Chest

- Symmetric expansion without deformities
- Clear breathing sound without crackle
- Regular heartbeat without murmur

▶ Abdomen

- Soft & flat
- Normoactive bowel sound
- Abdominal tenderness / rebound tenderness (-/-)
- Palpable abdominal mass (-)
- Hepatomegaly / Splenomegaly (-/-)

▶ Back & Extremity

- Td (-), swelling (-)
- Pitting edema (-/-)

LAB (8/15)

CBC	14.8-6290-90 K
BUN/Cr (mg/dL)	12.1 / 1.01
Na-K-Cl (mEq/L)	141-4.0-107
Protein/albumin (g/dL)	7.13/4.42
AST/ALT (U/L)	20/22
CRP (mg/dL)	< 0.1

Amylase	79
Lipase	25
CA 19-9	1.6

Problem List

- ▶ #1. Increased size of a cystic mass in the pancreatic tail without ductal dilatation during 1 year follow-up (4.7x4.6 cm -> 5.3x5.0 cm)

- ▶ #2. Chronic alcoholics

Assessment

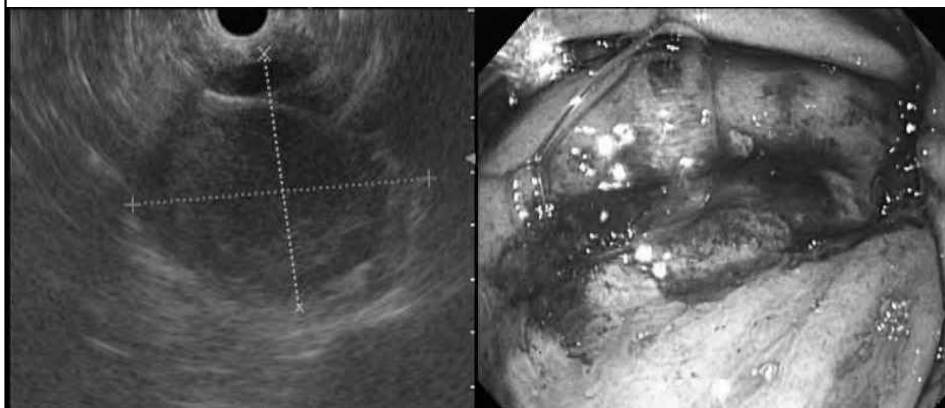
- ▶ #1. Pancreas tail cyst
 - r/o Pseudocyst
 - r/o MCN (Mucinous cystic neoplasm), less likely

Plan

- ▶ Diagnostic plan
 - EUS-FNA
 - Cytology analysis
- ▶ Therapeutic plan
 - EUS-drain

EUS-FNA (8/16)

- ▶ Pancreas tail : 58 mm sized Unilocular cyst lesion, echogenic material
- ▶ Chocolate color cystic fluid aspirated, string sign(-)
- ▶ Fistula dilatation failure due to thickened cyst wall
- ▶ Stomach: luminal bleeding (+)



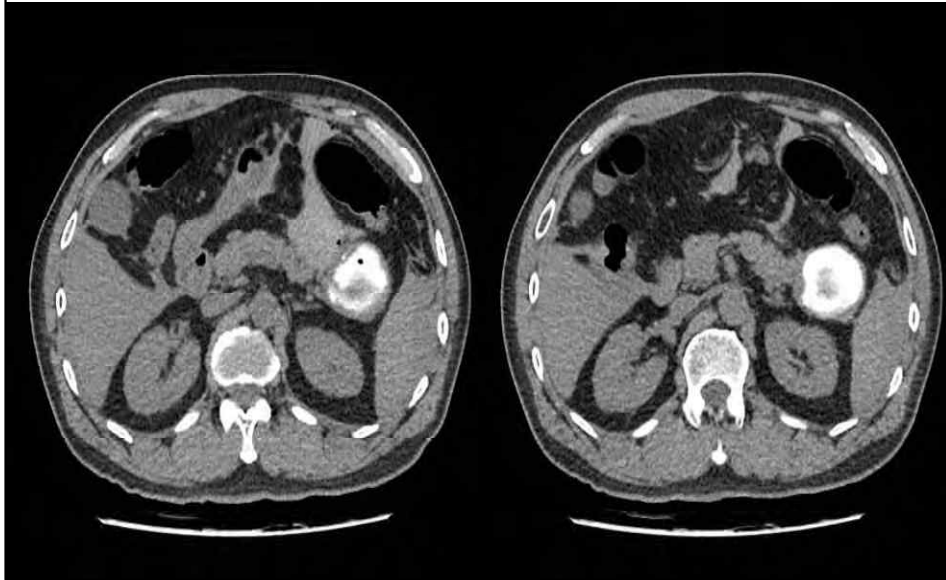
Flurography (8/16)



HD # 3 (8/17)

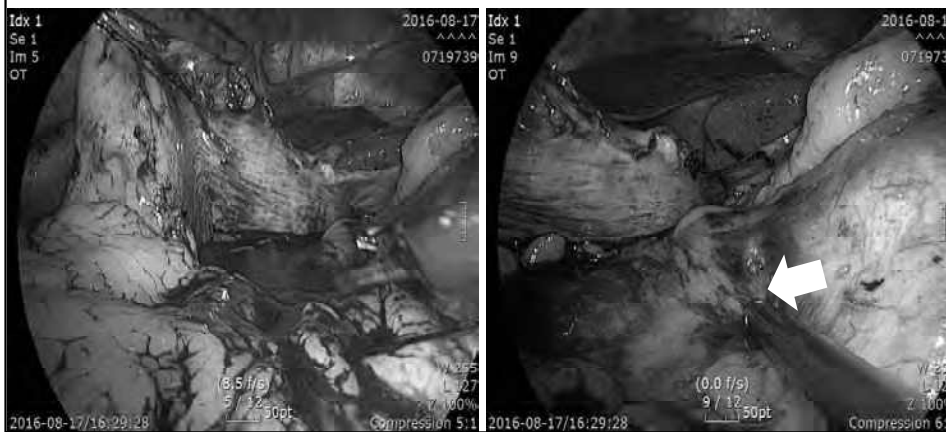
- ▶ S)
 - ▶ Abdominal pain
 - ▶ Sweating
- ▶ O)
 - ▶ 전일 저녁 V/S : 80/50-68-18-37.0 → 당일 오전 V/S : 128/76-65-20-36.8
 - ▶ Whole abdomen Td/rTd (+/+)
 - ▶ CBC 12.3-**13720**-98K
 - ▶ BUN/Cr **26.8/1.71**
 - ▶ AST/ALT 19/17, Amylase/Lipase 58/21
 - ▶ *Cytology : Negative for malignancy*
- ▶ A)
 - ▶ #1. r/o Hemoperitoneum
- ▶ P)
 - ▶ APCT
 - ▶ GS consult for emergency operation

APCT (8/17)



Surgical procedure (8/17)

- ▶ Hemoperitoneum, whole abdominal cavity
- ▶ EUS puncture site (white arrow)
- ▶ Laparoscopic distal pancreatectomy performed

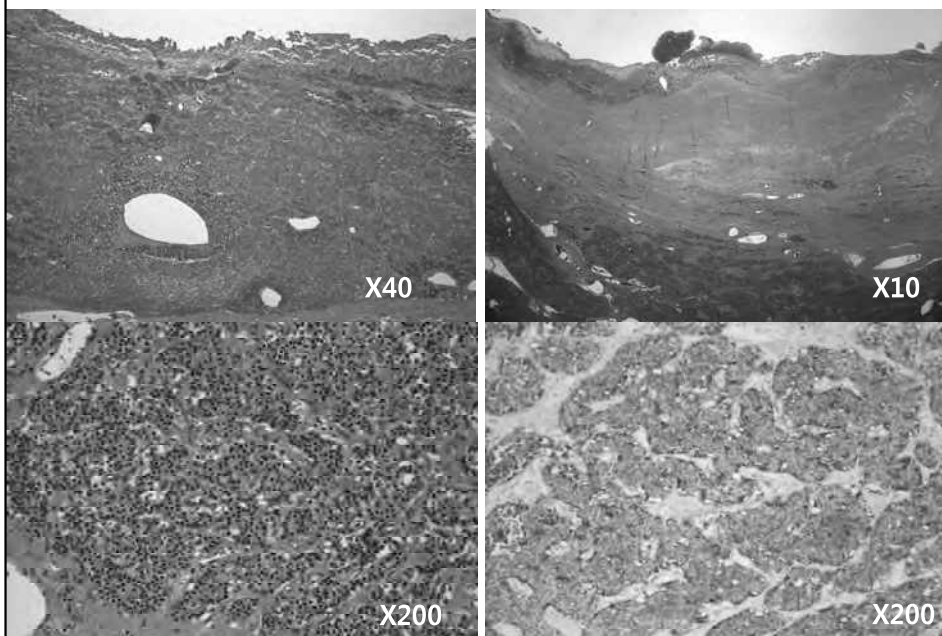


Pathologic report

- ▶ Neuroendocrine tumor, G1, pancreas tail
- ▶ Tumor size: 4x3.5 cm
- ▶ Mitotic rate: <1 per 10 HPFs
- ▶ Necrosis: present, extensive (more than 95% of tumor volume)
- ▶ Synaptophysin (+), CD56 (-), Ki-67: 1%



Microscopy



Final diagnosis

- ▶ PNET with cystic degeneration

MEMO

MEMO