



Personal Information

- Age: 34 y/o
- Sex: female
- Past history:
 - major systemic medical history(-)
 - surgical history(-), family history(-)
- Denied food or drug allergy



Chief Complaint

- Retroperitoneal cystic mass incidentally found at health examination center.

Present Illness

- Health checkup –
 - cystic lesion over adrenal area
- No specific symptoms
- Nephrologist -- negative finding
- Gastro-enterologist–
 - further lab and image exam



Physical Examination

- Negative finding.



Lab Data

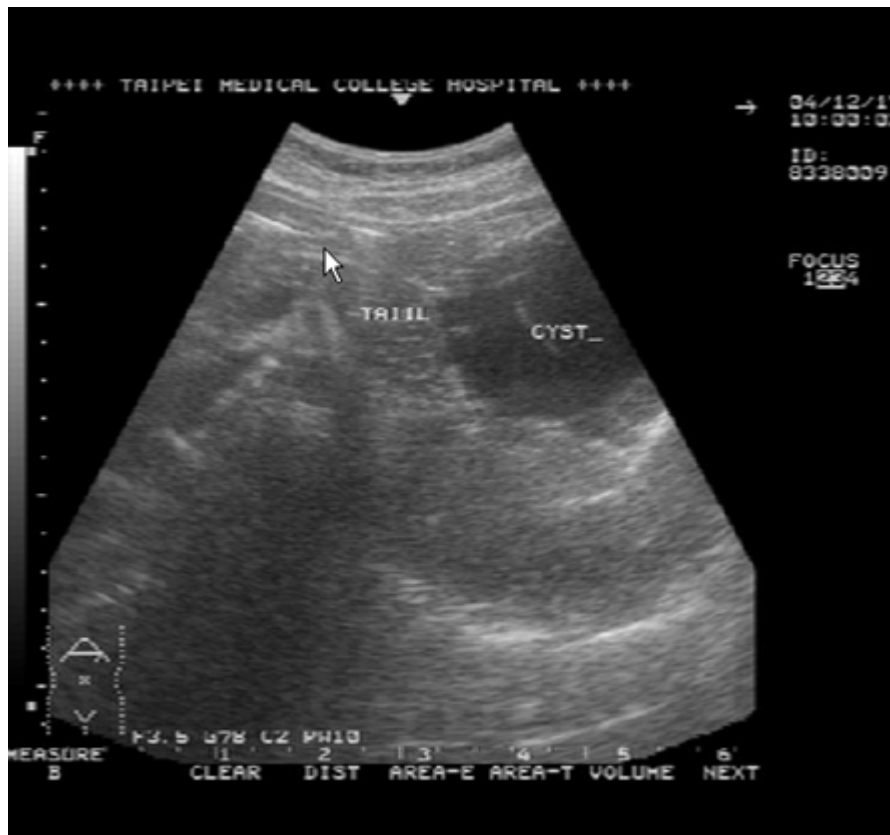
- CA-199: 2.41
- CEA: 0.1
- Other data were within normal range.

Image – CXR



- Normal lung field .
- No abnormal defect over bone or soft tissue.

Image – Abdominal Sonography



- There was a 8.7x8.3 cm cystic lesion with central septum at pancreatic tail.
- Head and body were negative.

Image – CT + contrast

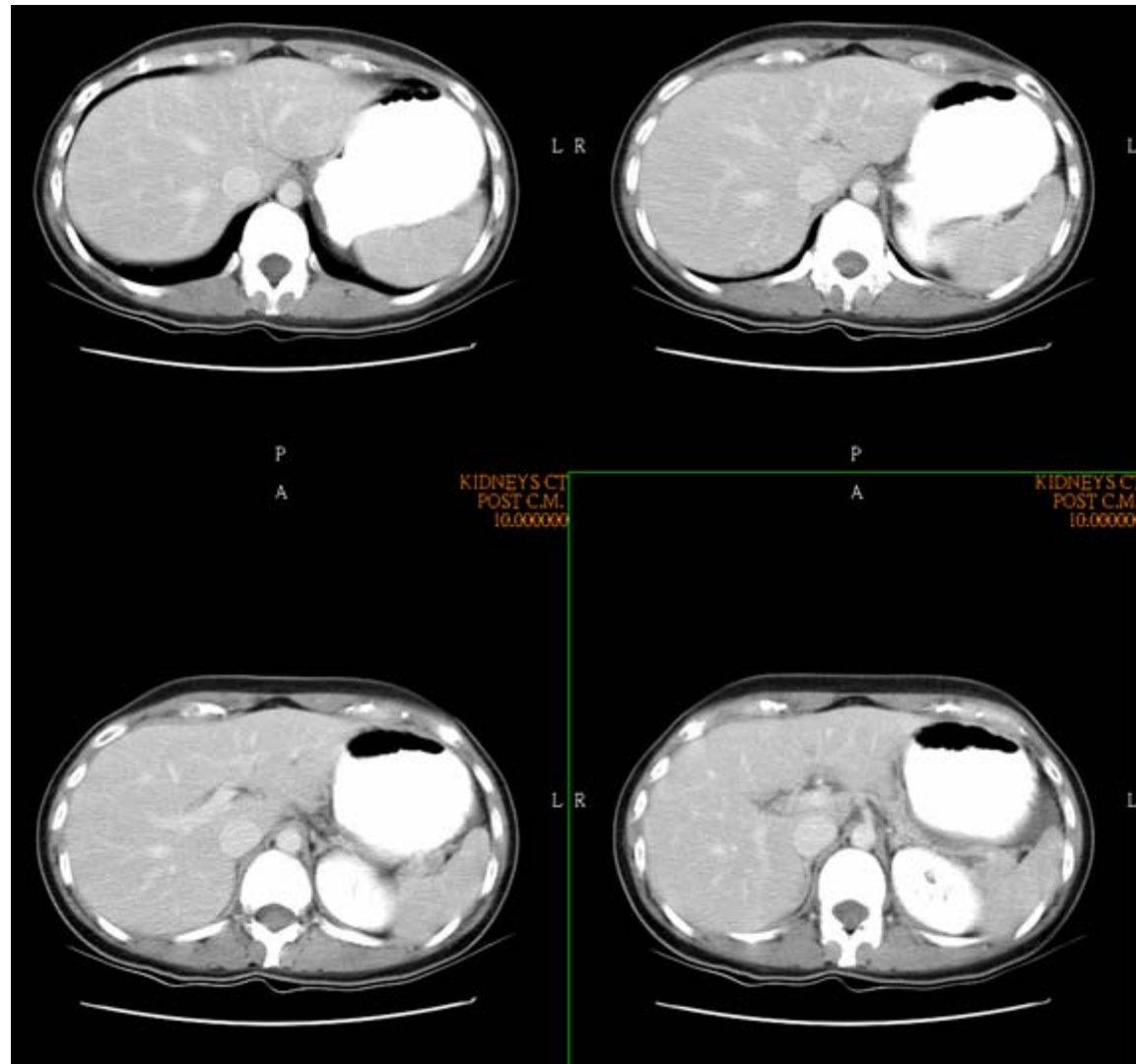
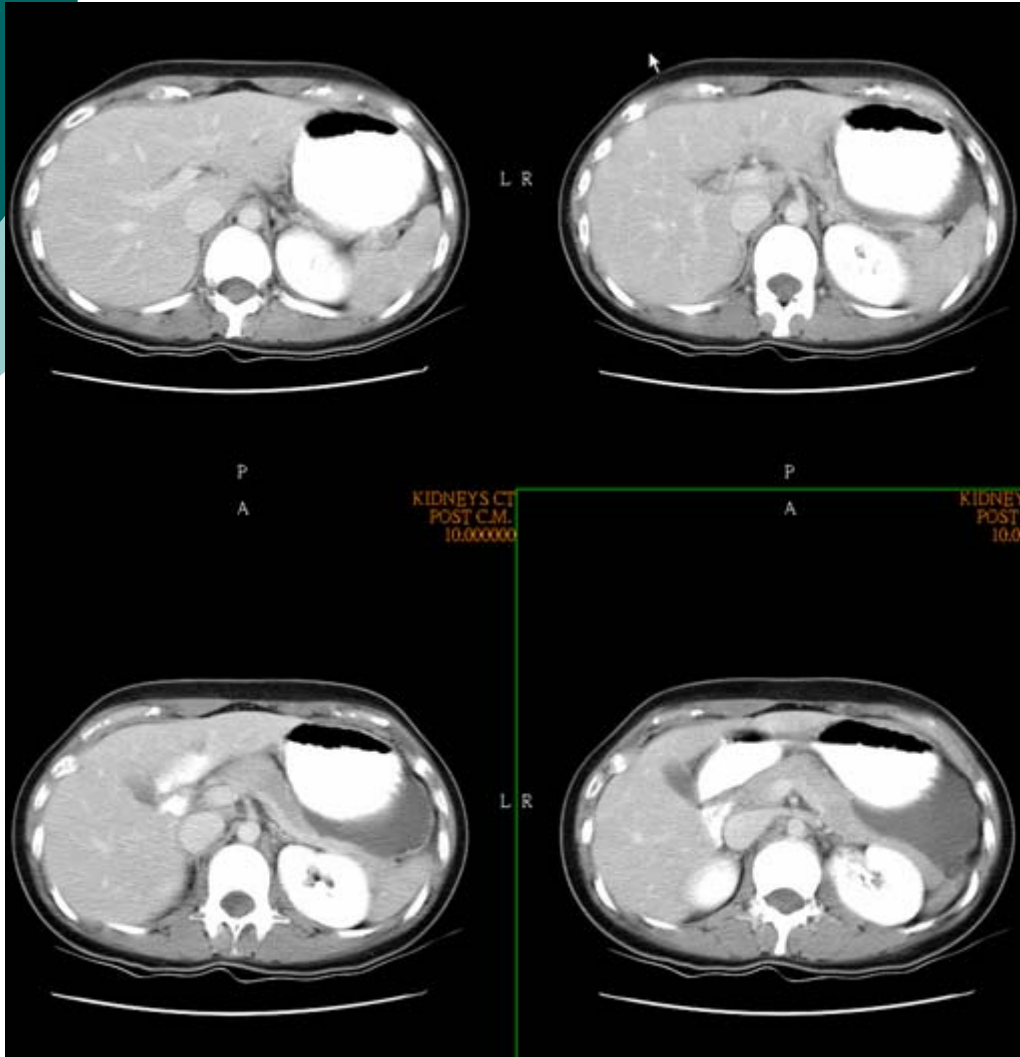
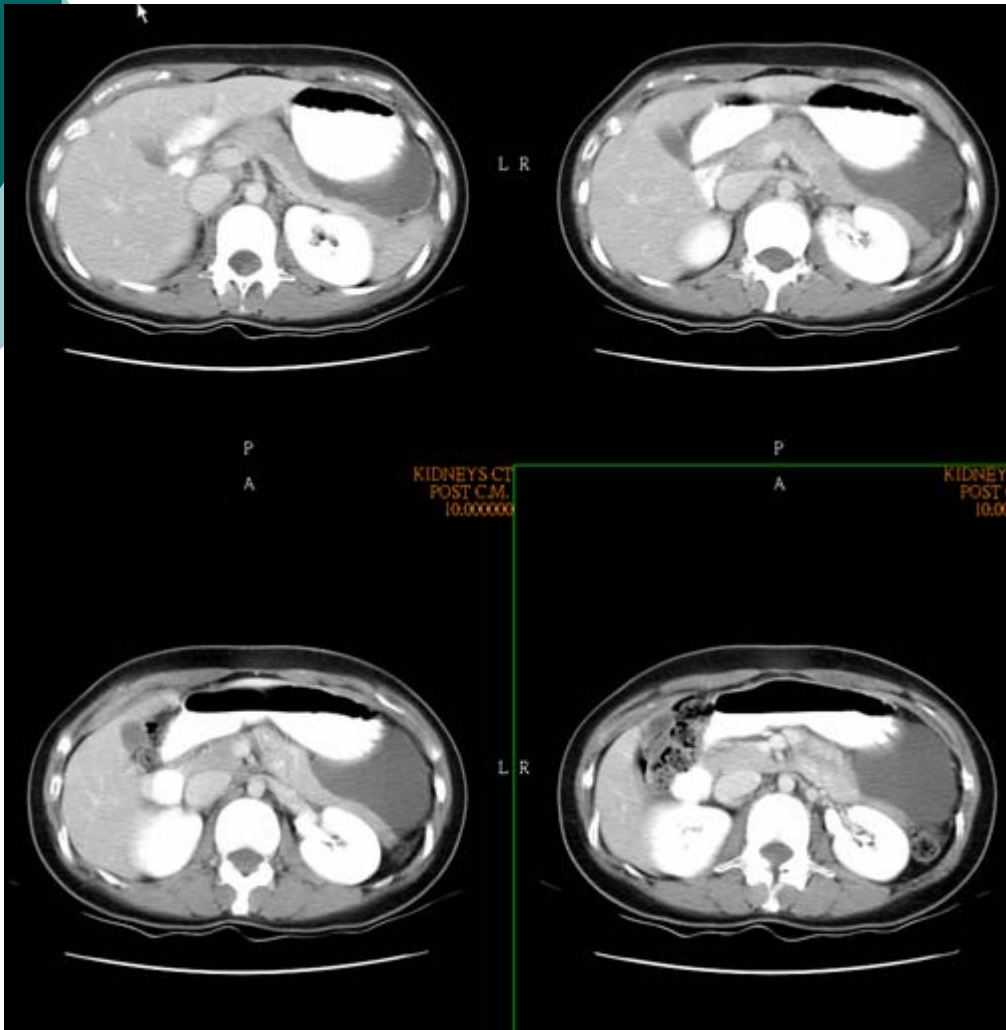


Image – CT + contrast



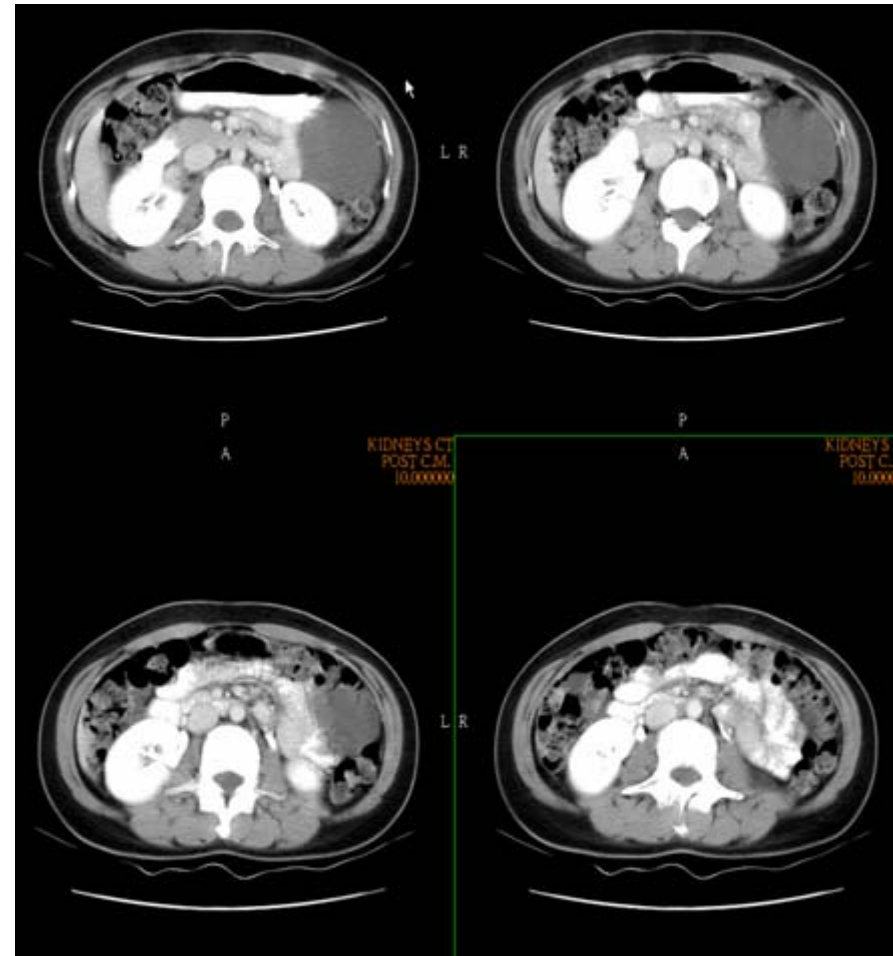
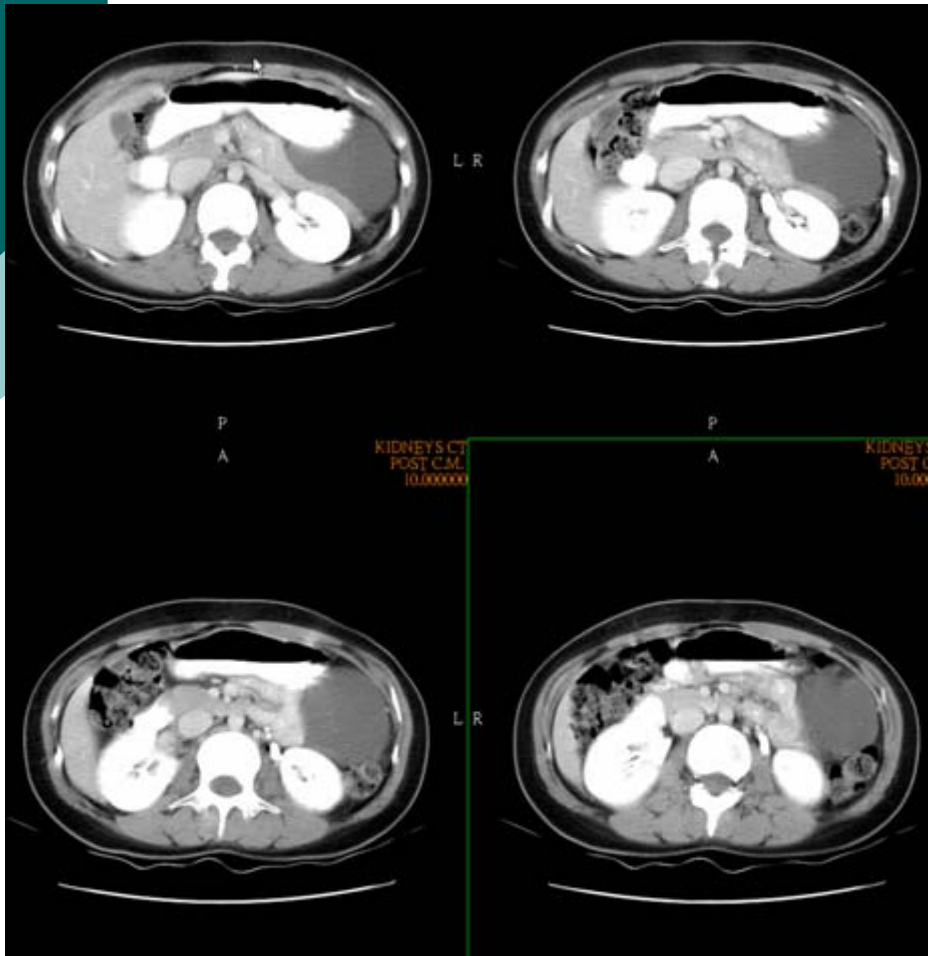
- An unenhanced **cystic tumor** with very thin enhanced capsulation at intraperitoneum. (ant. to pancreatic body/ tail, but post. to stomach)

Image – CT + contrast



- 5.8x5.6 cm in largest diameter
- **Mass effect** onto spleen and pancreas.
- No obvious focal infiltration over peri-cystic mesentery.
- LN (-)

Image – CT + contrast



CT character

- Solitary.
- Well capsulation with thin wall.
- Homogenous fluid content.
- Located at pancreas or para-pancreatic area (near tail).
- No enhancement with contrast.
- No inflammation or metastatic sign.



D/D for cystic lesion of pancreas
or para- pancreatic area

by CT

D/D by CT

1. Pancreatic pseudocyst
2. Abscess
3. Microcystic Adenoma (Cystadenoma, Serous adenoma, Glycogen-rich adenoma)
4. Lymphangioma
5. Mucinous Adenocarcinoma (Colloid Carcinoma)

D/D by CT

6. Mucinous Cystic Tumor

(Cystadenocarcinoma, Macrocystic Adenoma)

7. Metastatic tumor

8. Mesenteric cyst

9. Hemangioma

10. Congenital cystic lesion (ADPKD, Von-Hippel-Lindau disease, cystic fibrosis)

Pancreatic pseudocyst -- CT



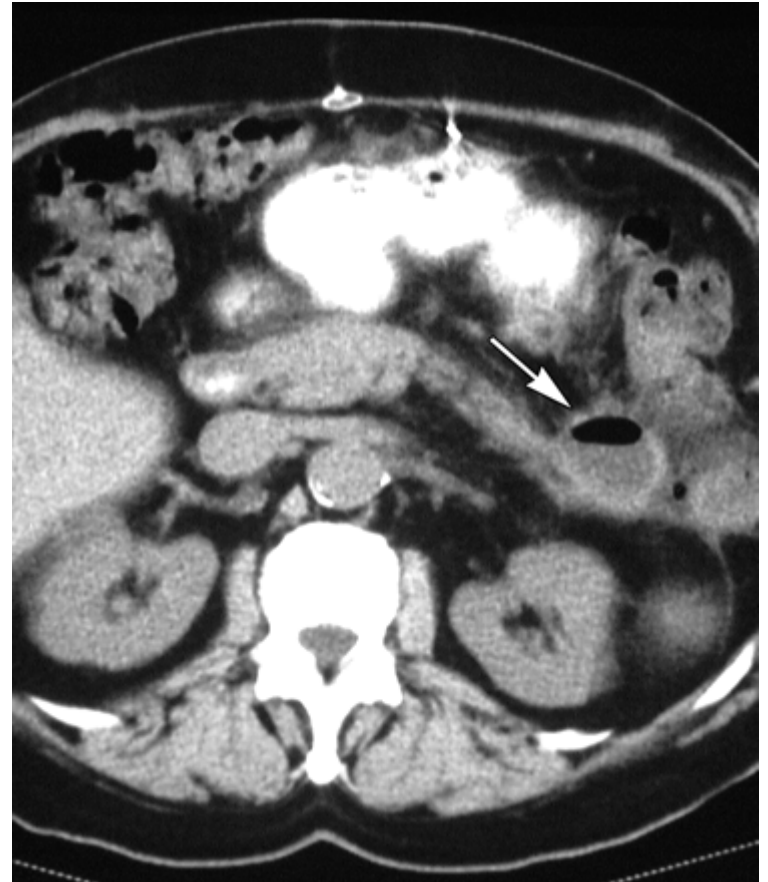
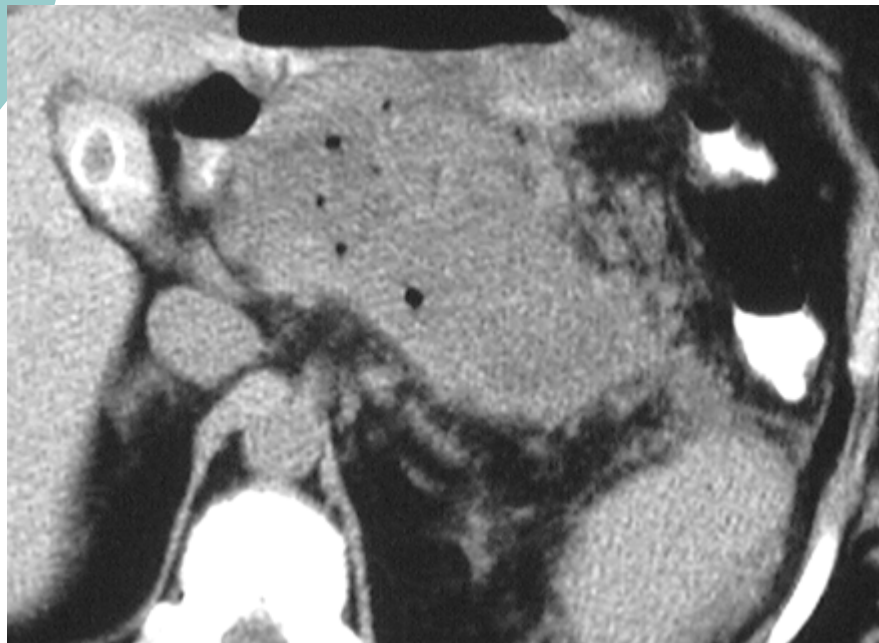
Pancreatic pseudocyst – CT character

- Well-defined wall (**fibrous tissue**, no epithelial lining), may calcified.
- Uniform, **low-attenuation fluid collection** with a thin uniform wall that enhances after administration of IV contrast material.

Pancreatic pseudocyst – CT character

- Accompanying signs of pancreatitis to a varying degree. (enlarged pancreas, inflammation surrounded)

Abscess -- CT



Abscess – CT character

- Poorly defined margins and are often suspected when gas is present in a fluid collection.
- Some rare pancreatic infections, including fungi, tuberculosis, and parasites, can have a cystic appearance.

Microcystic Adenoma -- CT



Microcystic Adenoma – CT character

- Most are < 2cm.
- Central stellate scar, with or without calcification .
- Contains intracellular glycogen but no mucin. (aspiration D/D)



Microcystic Adenoma – CT character

- Can be water, soft-tissue, or mixed density.
- Margin ranging from poorly defined to a thin well-defined capsule.
- Enhancement of cyst walls and septa ranges from moderate to marked.

Lymphangioma -- CT



Lymphangioma -- CT





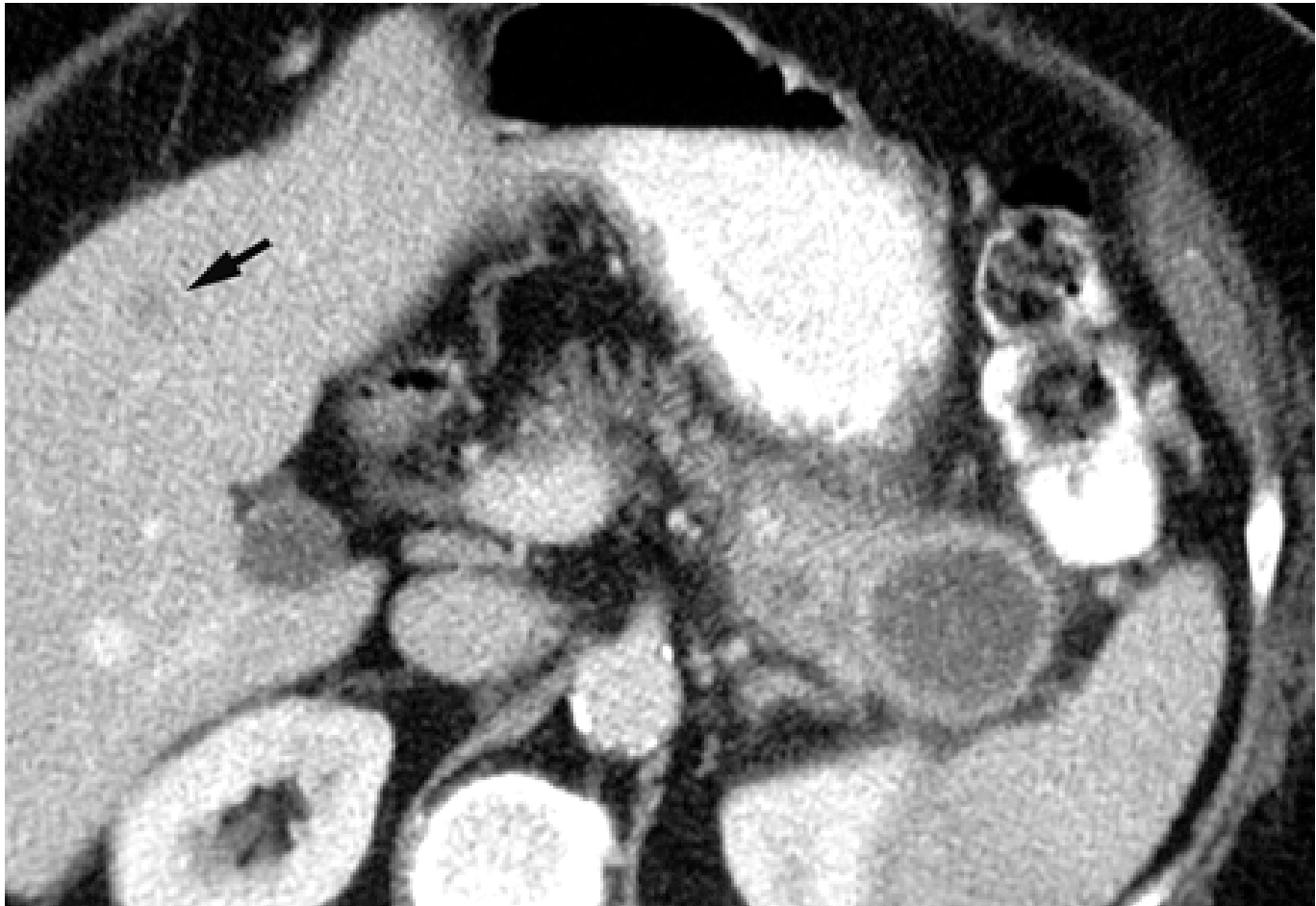
Lymphangioma – CT character

- Most often homogeneous, thin-walled, fluid-filled cysts.
- May have septa, thick walls, calcification, and internal debris.
- Have epithelial lining but do not contain keratin. (D/D from lymphoepithelial cysts)

Lymphangioma – CT character

- The lesion **displaces solid organs**, has uniform septa which slightly enhance and has contents of attenuation near that of water.

Mucinous Adenocarcinoma -- CT

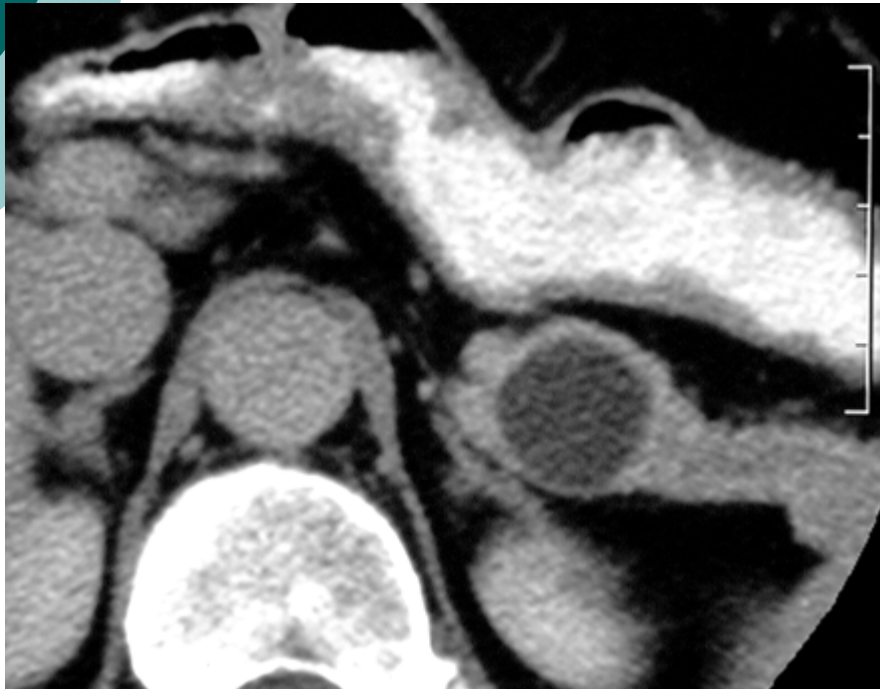




Mucinous Adenocarcinoma -- CT

- Well-defined cystic lesion.
- Enlarged pancreas and destruction of normal shape may noted.

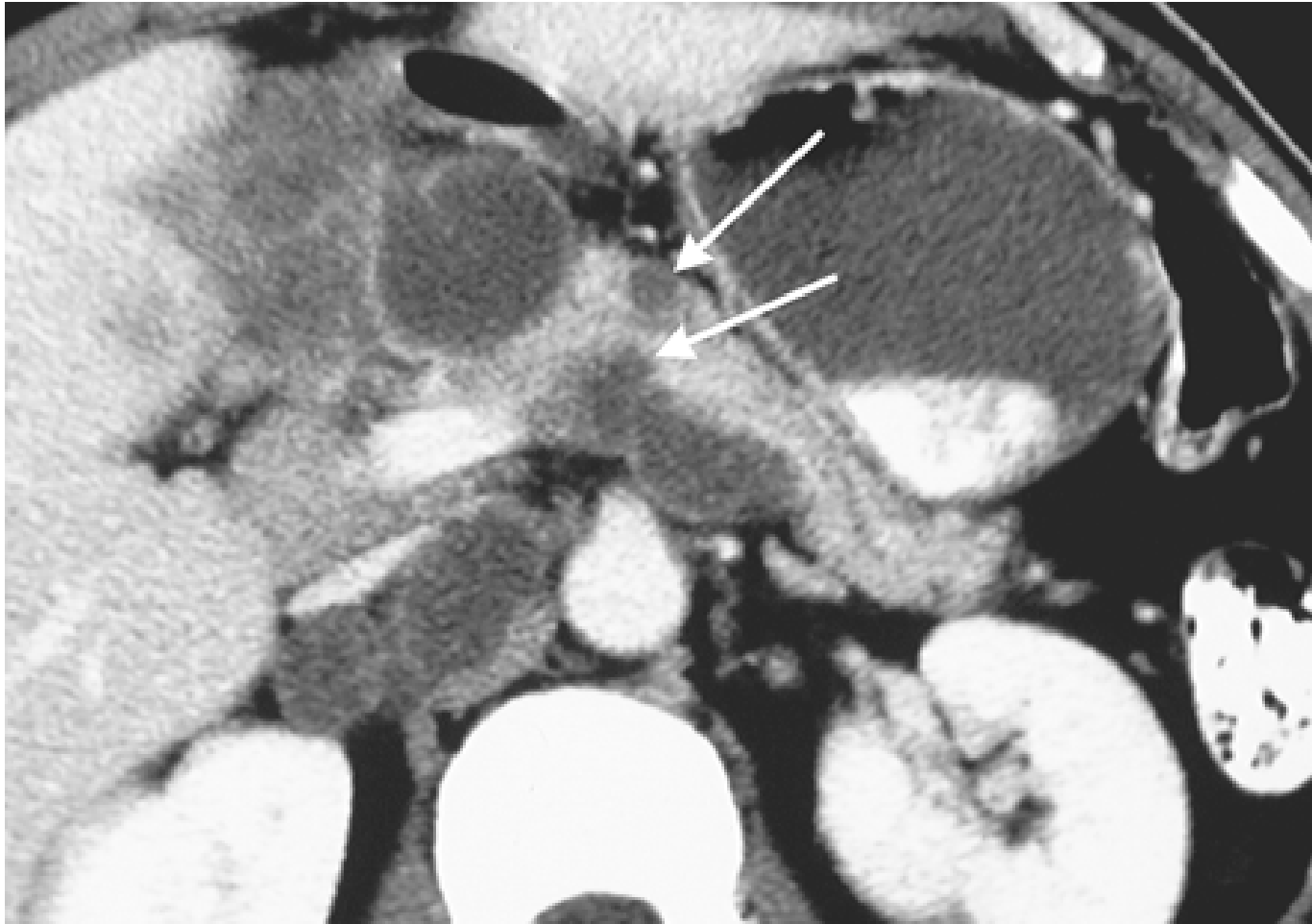
Mucinous Cystic Tumor -- CT



Mucinous Cystic Tumor -- CT

- Most lesions are located in the pancreatic body or tail .
- Most are larger than 2 cm.
- Near-water-density unilocular or multilocular cystic lesion with enhancing walls.
- Peripheral or curvilinear calcifications .

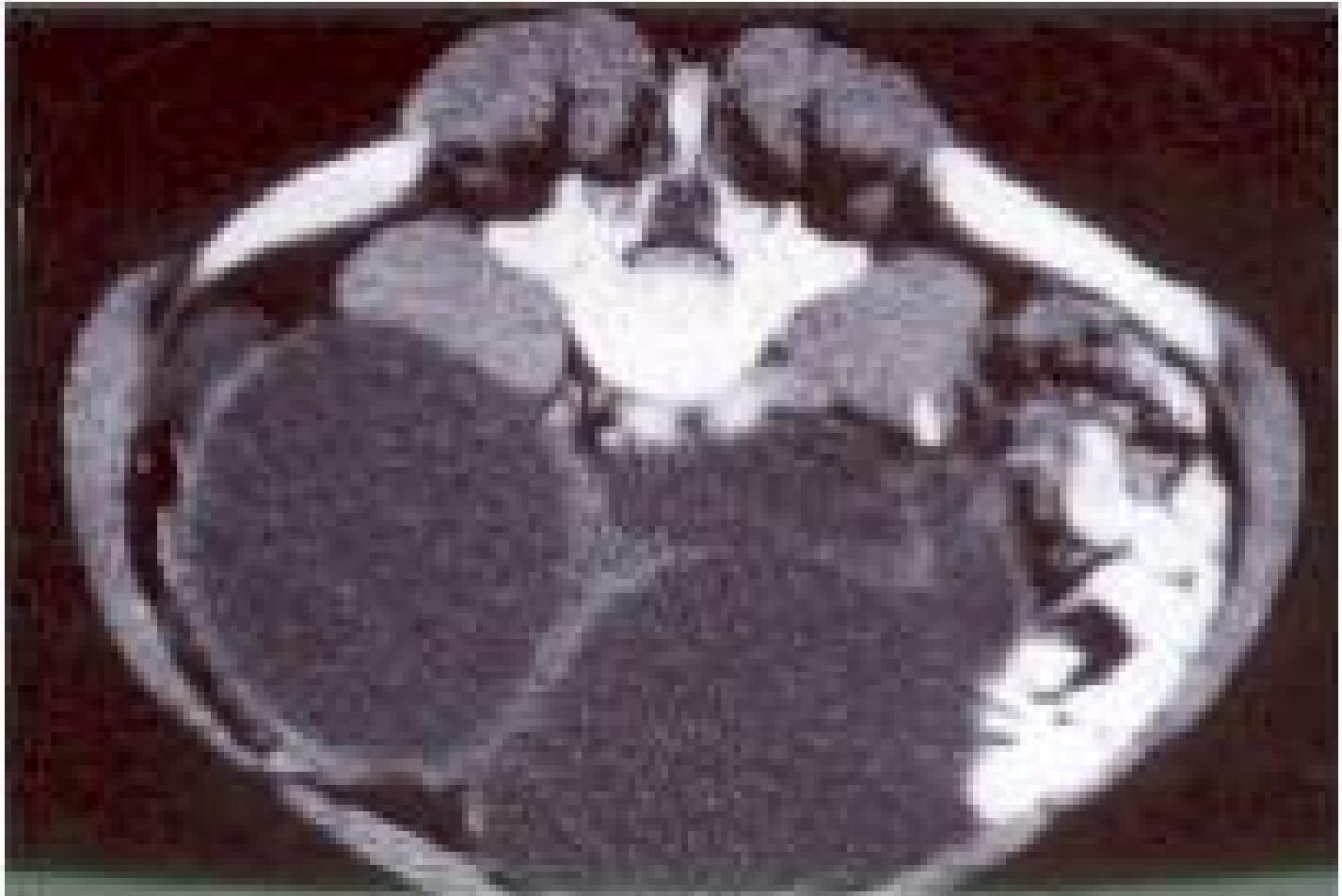
Metastatic tumor -- CT



Metastatic tumor -- CT

- Breast, lung, melanoma, and gastrointestinal tract were the most common primary sites.
- Mostly multiple lesions.
- Cystic metastases can be the result of central necrosis or cystic degeneration.

Mesenteric cyst -- CT



Mesenteric cyst -- CT

- Cysts can be unilocular or multilocular, and may contain chylous, serous or infrequently hemorrhagic fluid.
- Calcification and reactive, chronic inflammatory changes may be present within their fibrous walls.

Mesenteric cyst -- CT

- May be quite large with several **complications** : torsion, infarction, volvulus formation, perforation, infection, anemia from intracystic hemorrhage, intestinal obstruction and obstructive uropathy

Hemangioma -- CT





Hemangioma -- CT

- Enhanced with contrast media.
- Heterogenous content.

Congenital Cyst (ADPKD)





Congenital Cyst (ADPKD)

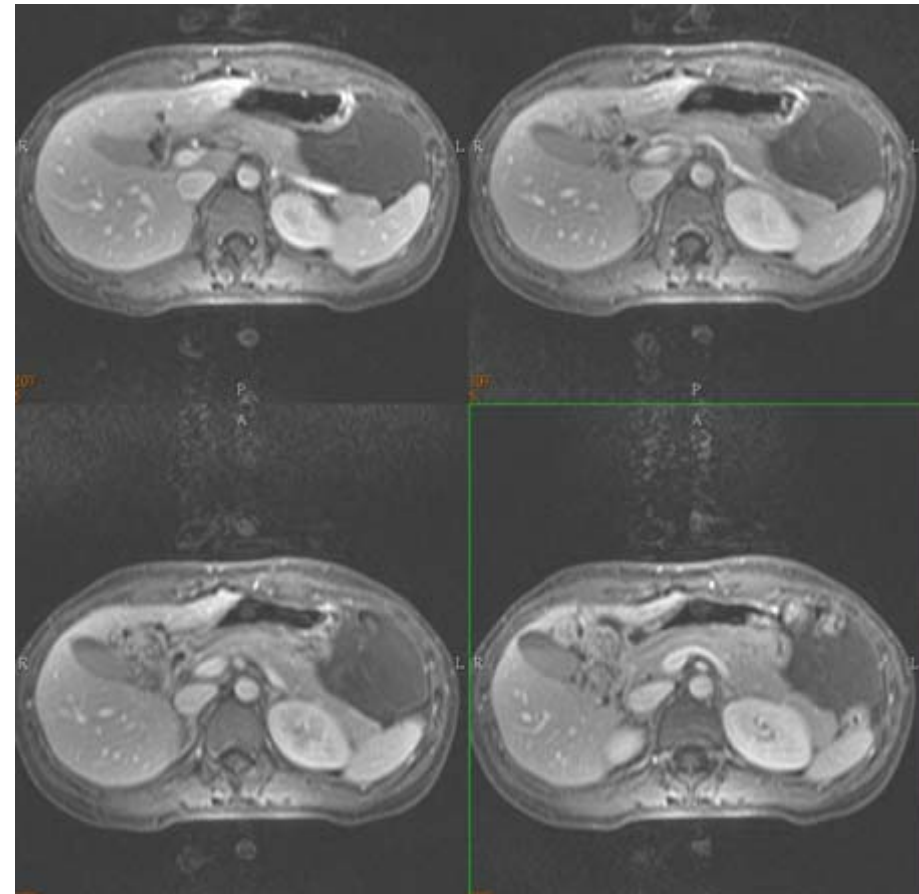
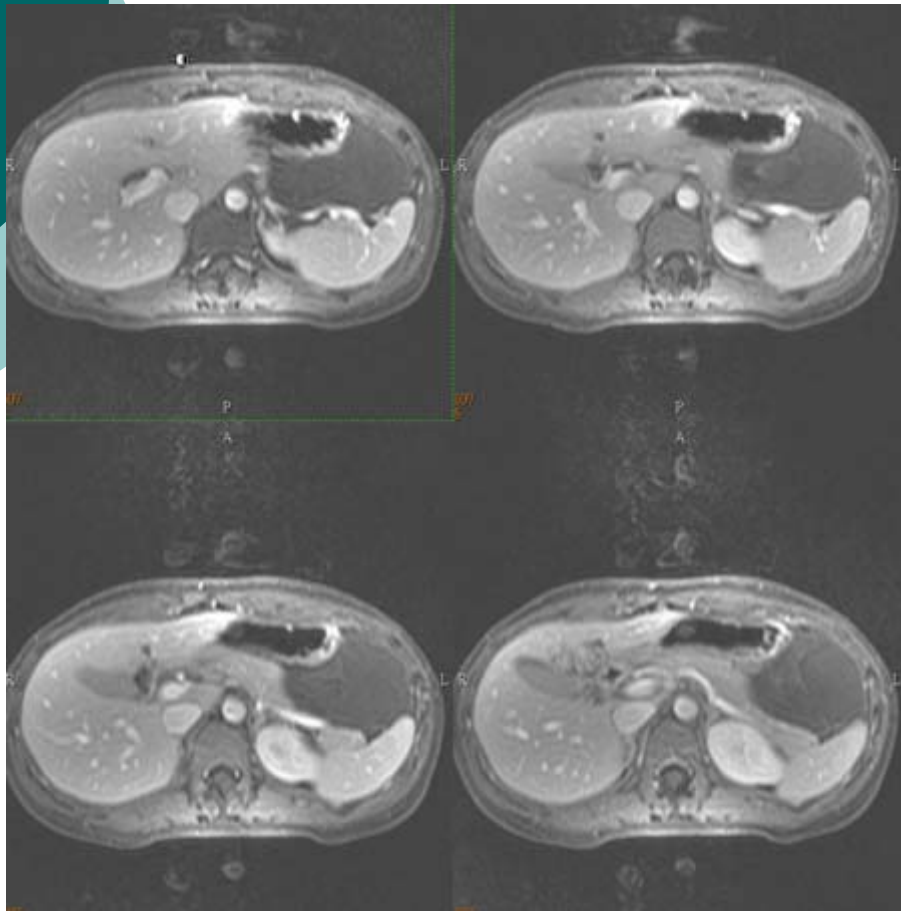
- Accompanied with other cystic lesions over different organs (Kidney, liver.....).
- Family history.



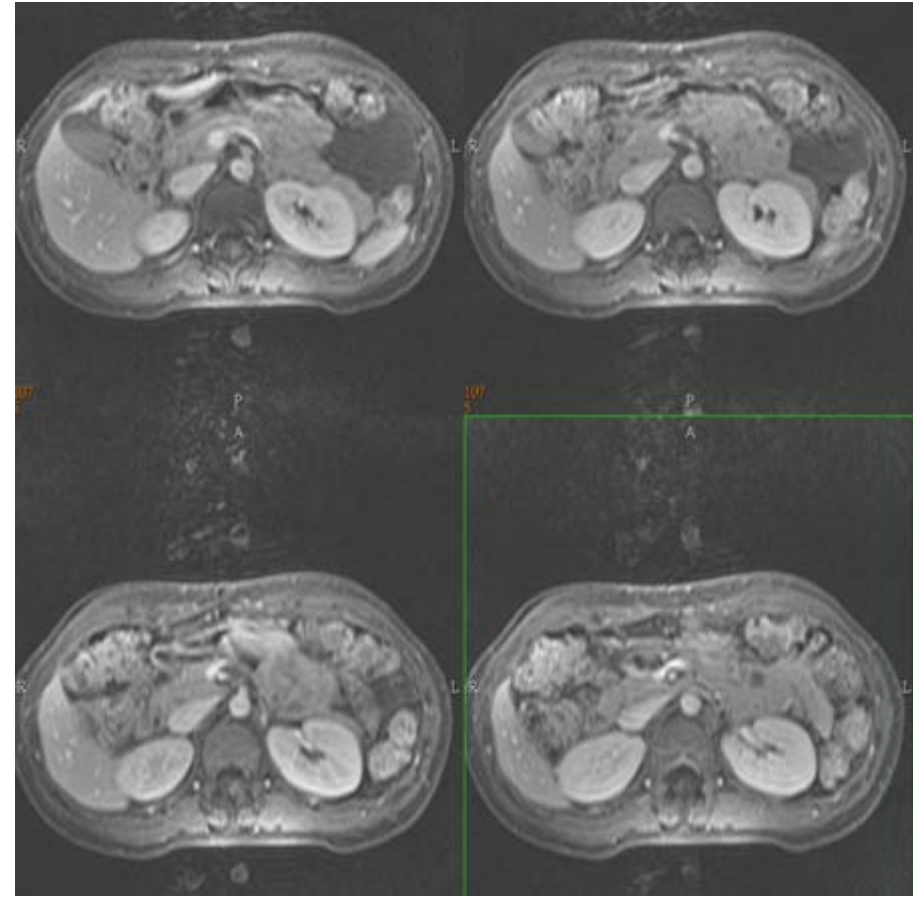
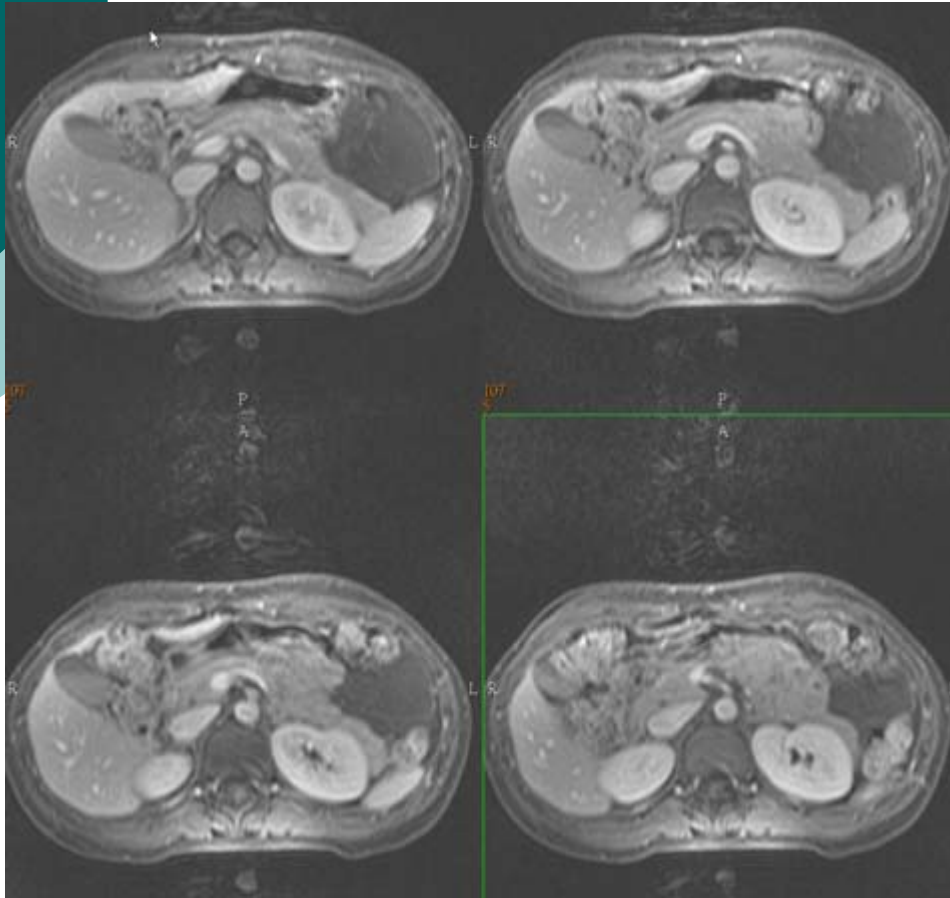
CT Impression

- Lymphangioma
- Mesenteric cyst
- Mucinous cystic tumor

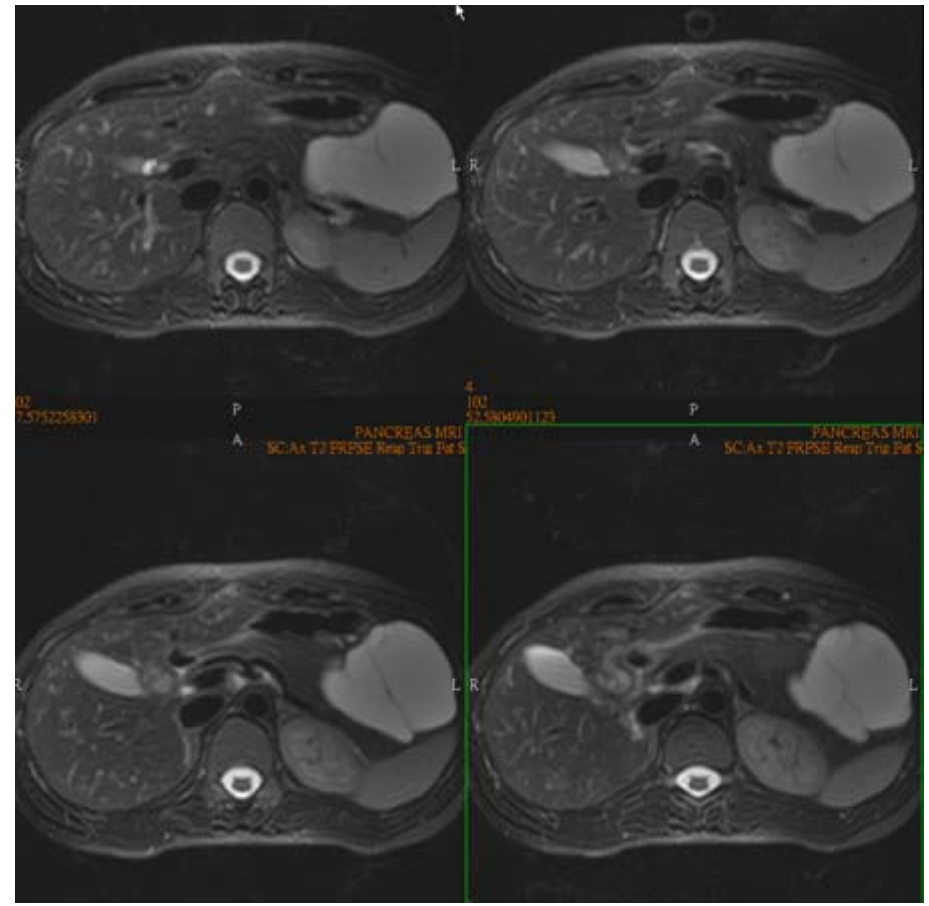
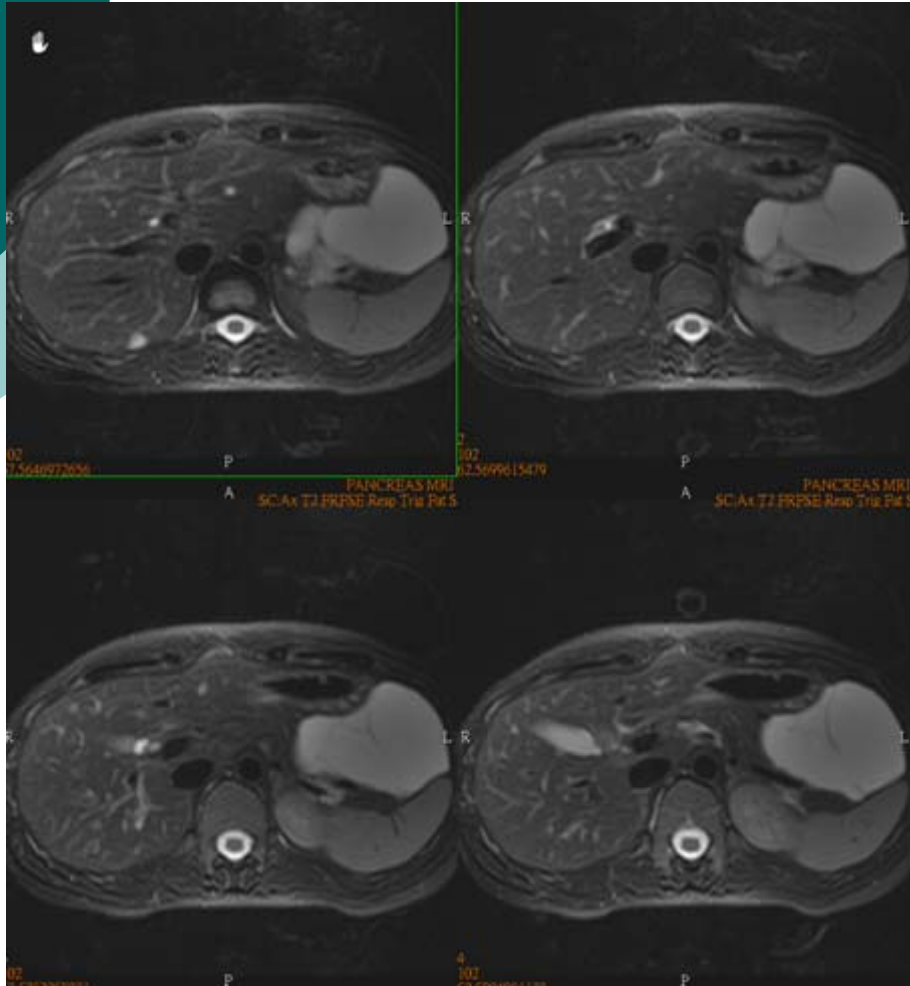
Image– MRI, T1 W



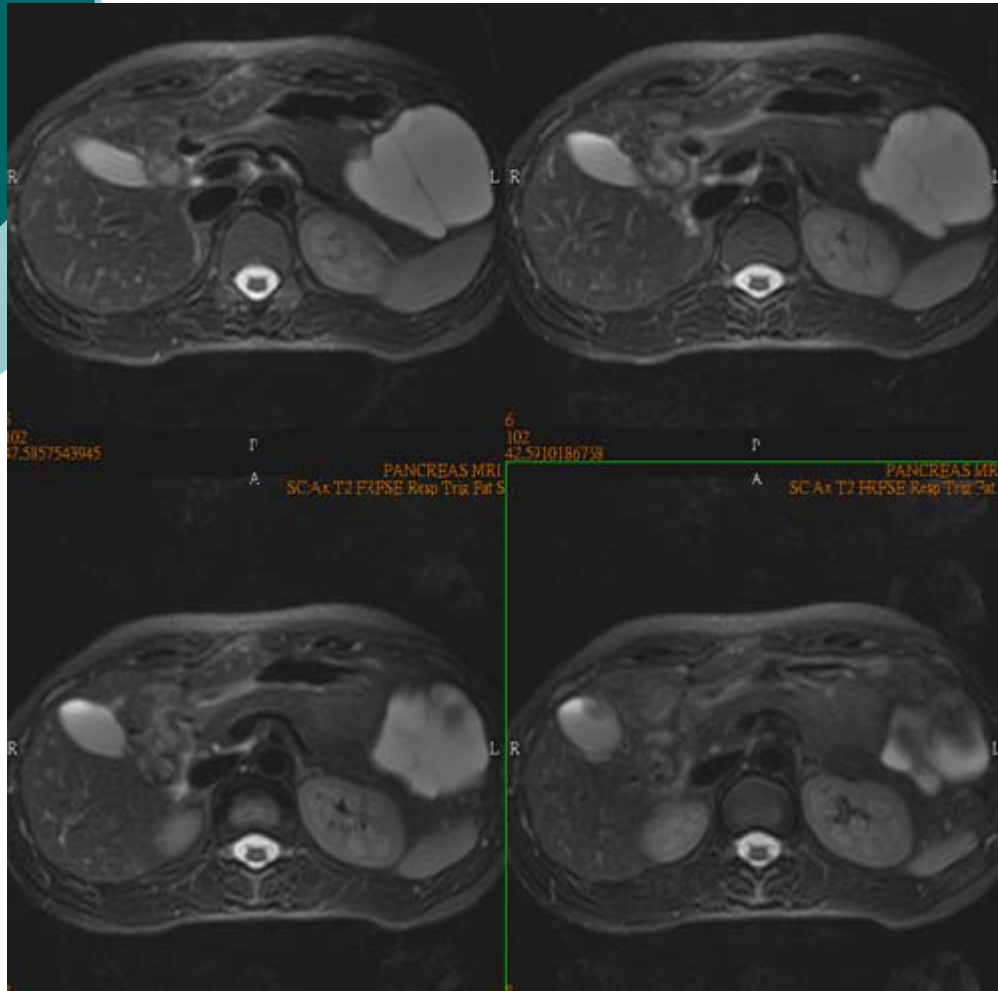
Image— MRI, T1 W



Image– MRI, T2 W



Image– MRI, T2 W



- Cystic lesion enhanced in T2 phase
- With septum

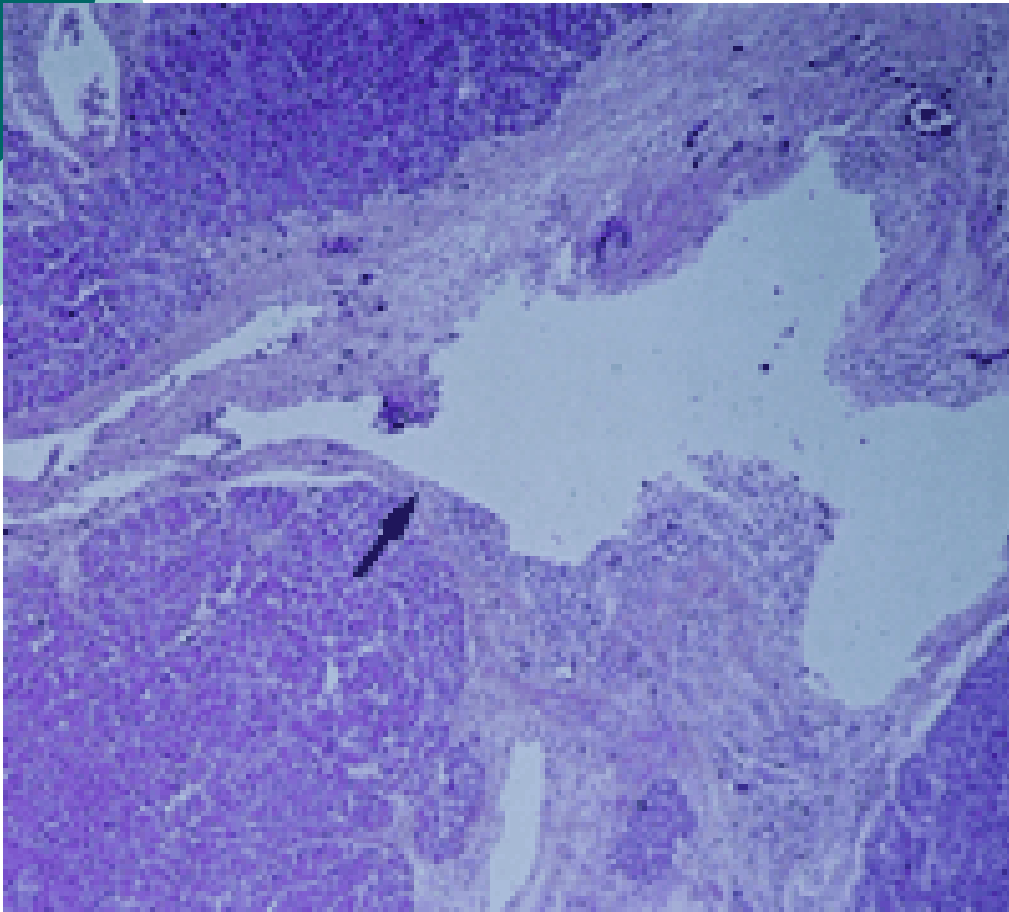
Clinical Impression

- Pancreatic cyst
 - r/o lymphangioma.
 - Mesenteric cyst
 - Mucinous cystic tumor

Final Diagnosis

- Surgical resection
- Pathology:
 - grossly measured 6x6x1.5 cm in size.
 - multiloculated cystic spaces** with thin and transparent wall and **clear fluid**.

Pathology – histologic picture



- Enlarged cystic-like space lined by endothelial cell.
- Numerous lymphocytes are present in fibrous stroma.



Pathology character

- Multiple cysts lined by endothelial cells.
- Irregularly distributed smooth muscle cells.
- Lymphoid aggregates in the wall of the cyst.

Final Diagnosis

--Cystically **dilated lymphatics** lined by flattened lining cells and **lymph-like fluid**, focal lymphocytes infiltration, and **fibrous stroma** with scattered smooth muscle fibers.

→ **Pancreas, tail, excision,**
lymphangioma



Discussion

Pancreatic Lymphangioma

Background

- Neoplasms, hamartomas, or lymphangiectasias ??
- **Malformations** arising from sequestered lymphatic channels
or
- Acquired lesions due to obstruction caused by fibrosis of lymph channels.

Background

- Developmental anomaly,
→ distension of sequestered lymphatic channels within primitive mesenchyme rather than a true neoplasm.

Background

- Most common sites: head, neck, and axilla.
- Only 5 % at mesentery, omentum, mesocolon, and retroperitoneum.
- Pancreatic lymphangiomas (1%) occur predominantly in women.
(F/M: 2:1).
Average age : 25.6 years.

Classification

- Cystic, capillary, and cavernous.
- Only **cystic** and **cavernous** types have been reported in the pancreas
- Considered to be of pancreatic origin :
 - in the pancreatic parenchyma,
 - adjacent to the pancreas,
 - connected to the organ by a pedicle



Clinical Presentations

- Abdominal pain
- Nausea
- Vomiting
- Palpable mass
- Silent , incidental finding.



Morbidity/ Mortality

- No risk of malignant transformation.
- May local invasion.
- Strong tendency for **local recurrence** unless they are completely excised.



Gross

- Soft, multiloculated cystic masses.
- Content: serous, serosanguinous, or lymphatic fluid.

Histology

- Dilated lymphatic channels , separated by thin septa.
- Cystic spaces lining: flattened or cuboidal endothelial cells.
- Aggregates of lymphocytes.
- The septa and walls: smooth muscle fascicles and collagenous connective tissue.



Image -- Sonography character

- Anechoic or hypoechoic
- Fluid-filled
- Multiseptated mass in the pancreatic region.



Image -- CT character

- Well-circumscribed
- Homogeneous cystic masses in or adjacent to the pancreas.
- Septums and thin walls may enhance after IV contrast injection.



Image -- MRI character

- Hypointense on T1-weighted image
- Hyperintense on T2-weighted image.
- Not provide new information.

Diagnosis

- History
- Imaging: sono, CT, MRI
- Fine-needle aspiration cytology.
- Definite diagnosis: pathology report.



Treatment

- No proven medical care for lymphangiomas exists.
- Treatment of choice:
complete surgical excision

Prognosis

- Lymphangiomas are **benign hamartomatous malformations** instead of true neoplasms.
- Locally invasion may occur.
- The prognosis is **excellent**.



Thanks for your attention~

Reference

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- *Acute presentation of lymphangioma of the retroperitoneum* 129 *The Ulster Medical Journal, Volume 73, No. 2, pp. 129-131, November 2004.* DH Taylor, C Loughrey.
- *Cystic Lymphangioma of the Pancreas* *American Journal of Roentgenology* *Received March 13, 2001; accepted after revision April 4, 2001.*