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.
There was a 8.7x8.3 cm cystic lesion with central septum at pancreatic tail.

Head and body were negative.
An unenhanced cystic tumor with very thin enhanced capsulation at intraperitoneum. (ant. to pancreatic body/ tail, but post. to stomach)
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-Hipple-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 be 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 uropia
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, 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, serosanguinuous, 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

- Lymphangiomias are benign hamartomatous malformations instead of true neoplasms.
- Locally invasion may occur.
- The prognosis is excellent.
Thanks for your attention~
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