Management of Pancreatic Tumor

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• **CC:** abdominal mass

• **HPI:** Patient is a 67 y/o female who was found to have an incidental abdominal mass in 2008 while being worked up for uncontrolled diabetes in Trinidad. Since that time the mass has increased in size leading to abdominal discomfort, decrease in appetite, early satiety, constipation and a 20 lb weight loss.
• PMHx: DM, HTN, hypothyroidism, depression, fibroids

• PSHx: pannulectomy ’76, TAHBSO ‘78

• Allergies: NKDA

• Meds: synthroid, lantus, lisinopril, actos, cymbalta, micardis, lorazepam

• FHx: parents- DM
• Vitals: Temp 98.3° F   BP 171/81   HR 76   RR 18

• Physical Exam:
  General: AAOx3
  HEENT: NCAT, EOMI
  Chest: CTA bilaterally
  CVS: S1S2, rrr
  Abdomen: obese, (+)BS, NT, ND, palpable LUQ mass
  Extr: no edema or calf tenderness
• Labs:

CBC: 6.7/12.3/37.0/217
Chem: 137/5.1/99/29/23/1.6/302
Coags: 13.4/34/6/1.0
EKG normal sinus rhythm
- Radiologic Studies: CT Scan Abd/Pelvis
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• Intra-op:

Exploratory laparotomy, distal pancreatectomy, splenectomy and resection of pancreatic mass

• Pathology:

Low Grade Malignant Solid Pseudopapillary Neoplasm, focal vascular and capsular invasion, tumor size 14x11x7cm
• Pathology
Pathology
Pathology

- Polygonal, uniform cells, red globules
- Pseudopapillary structures
- alpha-1 antitrypsin
- Pathology

Cystic and solid components of mass

Focal tumor invasion within vessel
• account for 10-15% of pancreatic tumors

• most are benign exocrine tumors that are cystic

• signs and symptoms frequently seen include weight loss, nausea, anorexia, vomiting

• types: Serous cystadenoma
  Mucinous cystic tumors
  Intraductal Papillary Mucinous Tumor (IPMT)
  Solid-Pseudopapillary Tumor (SPT)
  Neuroendocrine tumors
Serous Cystadenoma

- account for 20-40% of cystic pancreatic tumors
- lined by a flattened epithelium with glycogen-rich cytoplasm which when found on cytologic exam is diagnostic
- benign with no malignant potential
Serous Cystadenoma

- typically large, microcystic, spherical masses that tend to have a central, calcified stellate scar
- when diagnosis is unclear or if the mass leads to symptoms, resection is indicated
Mucinous Cystic Tumors

• account for 20-40% of cystic tumors
• usually benign but have malignant potential
• two types:
  type 1: contains area of ovarian-like stroma, predilection for women and almost always found in tail
  type II: lacks ovarian stroma, occurs equally in both sexes and found anywhere in pancreas

neither type communicates with pancreatic duct
Mucinous Cystic Tumors

• lined by columnar, mucin-producing epithelium
• lesions on imaging are composed of very large cysts (macrocytic)
• resection of benign mucinous cystic tumors, approx. 50% will have a >5yr survival
Intraductal Papillary Mucinous Tumor (IPMT)

- first described in Japan in the 1980s
- affects men and women equally
- two types: can involve the major ducts (main duct variety) or the smaller ducts (branch duct variety)
- lined by columnar mucin-producing cells that develop papillary projections
Intraductal Papillary Mucinous Tumor (IPMT)

- diagnosis can be made during endoscopy if mucus is seen extruding from papillary orifice
- patients may present with episodes of pancreatitis due to obstruction of the pancreatic duct from mucin secreted by tumor cells
Intraductal Papillary Mucinous Tumor (IPMT)

- Can be classified according to PanIN scheme:
  - PanIN-1: minimal to no dysplasia
  - PanIN-2: moderate dysplasia
  - PanIN-3: severe dysplasia/carcinoma in situ - which may become locally invasive and metastasize
- resection prior to invasive malignancy is curative
Solid Pseudopapillary Tumor (SPT)

- rare neoplasm of the pancreas that was first described by Frantz in 1959
- referred to as solid and papillary epithelial tumors, papillary cystic tumors, Frantz tumors or Hamoudi tumors
- World Health Organization (WHO) classified the tumor as SPT in 1996
Solid Pseudopapillary Tumor

• comprises 1-2% of all pancreatic tumors
• strong predilection for female gender
• usually occurs in the 2nd to 4th decades of life
• benign, indolent tumor with low grade malignant potential
• symptoms: abdominal pain/discomfort
  early satiety
  nausea and vomiting
  anorexia and weight loss
Solid Pseudopapillary Tumor

- Imaging:
  - Ultrasound: heterogeneous mass with solid echogenic and cystic hypoechogenic components
  - CT scan: well-encapsulated, circumscribed retroperitoneal mass, central cystic with peripheral solid components with/without calcifications
  - MRI: well defined lesion with low signal intensity (T1) and high signal intensity (T2) images
Solid Pseudopapillary Tumor

- Histopathology: solid and cystic structure, characteristic pseudopapillary features, well-defined capsule, areas of hemorrhage and necrosis, uniform, polygonal cells

- Immunohistochemical studies: performed to confirm diagnosis, (+) for vimentin, PR, CD10 markers
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Solid Pseudopapillary Tumor

- Treatment: complete resection of tumor (primary and metastasis) is recommended as it has a low malignant potential and is well-encapsulated
- role of adjuvant therapy is unclear
- recurrence rates are low
- prognosis is relatively good with >95% survival after resection
Surgical Management of Solid Pseudopapillary Neoplasms of the Pancreas
(Franz or Hamoudi Tumors): A Large Single-Institutional Series
Reddy S., Cameron JL et al., JACS. 2009;208:950-959

• Retrospective review of surgical pancreatic database from 1970-2008
• study design was to evaluate long-term outcome of pts diagnosed with an SPT
• 37 patients were identified, 33 were female
• median age: 32
• median tumor size: 4.5cm
36 pts underwent resection of tumor

median follow-up was 4.8 yrs. (8 months-27yrs)

35 pts remained disease free following resection

Conclusion: formal surgical resection maybe performed safely and is associated with long-term survival
Cystic Lesion in the Pancreas

- **Unilocular**
  - Pancreatitis
    - ↑ str. amylase
  - No pancreatitis
    - Normal str. amylase
  - Pseudocyst
    - Consider alternative diagnosis

- **Microcyst**
  - Serous cystadenoma
    - Asymptomatic
      - Management depends on several factors (*)
    - Symptomatic
      - Surgery or cyst aspiration
      - Imaging follow-up

- **Cyst with solid component**
  - Malignant neoplasm
    - Asymptomatic
      - Surgery
    - Symptomatic
      - Imaging follow-up
      - Surgery

- **Macrocyst**
  - Mucinous cystadenoma
    - Asymptomatic
      - Management depends on several factors (*)
    - Symptomatic
      - dilated MPD (combined IPMN)
      - Surgery
Neuroendocrine Tumor: VIPoma

• arises from pancreatic islet cells that secrete VIP
• pt presents: watery diarrhea, hypokalemia, and achlorhydria
• diagnostic triad: secretory diarrhea, high circulating level of VIP (225-2000 pg/ml) and pancreatic tumor
• Tumor localization: CT scan, MRI or arteriography
• Treatment: octreotide, surgical resection: distal pancreatectomy
Neuroendocrine Tumor: Glucagonoma

- tumor of pancreatic islet alpha cells
- characteristics: DM, necrolytic migrating erythema, weight loss
- diagnosis made with elevated glucagon levels (200-2000pg/ml), pancreatic tumor and characteristic skin lesion
- Islet tumor may be seen on CT scan, MRI or angiography
- Treatment: amino acids, octreotide, surgical resection