2003 PIP-D Cases

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APMG
Case 31

- 4 mo boy with increasing head circumference, full fontanel, and vomiting
- Large enhancing medial temporal lobe lesion with adjacent cyst
- Angiogram with no tumor blush
- 3.5x2.5x2 cm lobulated reddish-gray firm mass with central cyst
Desmoplastic Infantile Astrocytoma/Ganglioglioma
DCA/G

- Temporal lobe with enhancing mural nodule
- Desmoplasia with spindle cells predominating
- Neuronal and astrocytic differentiation
- Increased cellularity with MF
- Occ necrosis with pseudopalisading
- Focal GFAP and +/- synaptophysin
Quick Facts

- Cerebrum most common location for CNS tumors in first 2 years of life
- Astrocytoma most common primary CNS tumor during first 2 years of life
Radiographic Features of Low Grade Tumors

- Cystic architecture
- Intracortical location
- Skull deformation/erosion
- Solid enhancement
## Pediatric Posterior Fossa Tumors

<table>
<thead>
<tr>
<th>Age</th>
<th>Tumor</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-2 yrs</td>
<td>Teratoma (20% of all)</td>
</tr>
<tr>
<td></td>
<td>Atypical teratoid/rhabdoid tumor</td>
</tr>
<tr>
<td></td>
<td>Desmoplastic infantile</td>
</tr>
<tr>
<td></td>
<td>astrocytoma/ganglioglioma</td>
</tr>
<tr>
<td>&gt;2 yrs</td>
<td>Pilocytic astrocytoma</td>
</tr>
<tr>
<td></td>
<td>Medulloblastoma</td>
</tr>
<tr>
<td></td>
<td>Ependymoma</td>
</tr>
</tbody>
</table>
Case 32

- 88 F with diffuse abdominal pain and progressive nausea and vomiting
- 11 months s/p distal pancreatectomy for Stg. I moderately differentiated adenoCA
- Exploratory laparotomy for lysis of adhesions and removal of right ovarian mass
- 140 gm 9x6x4.9 cm ovay with necrotic solid tumor
- Tumor cells + for CK7, CK20, Villin, CDX-2
Metastatic Pancreatic Adenocarcinoma
Ovarian Metastases

- 6-17% of all ovarian carcinomas
- Autopsy failed to reveal primary in 14%
- Laparotomy identified primary in 4/25 cases
Ovarian Metastases*

- Breast
- Colon
- Carcinoid
- Stomach

*Descending frequency
Favor Metastatic Carcinoma

- Surface implants
- Size <10 cm
- Multiple discrete tumor nodules
- Bilaterality
- Variation in growth pattern from each nodule
- Signet ring cells
- Vascular invasion
- Metastatic tumor may precede identification of extra-ovarian primary site by several years
Immunoperoxidase Not Helpful

- Positive for both mucinous ovarian CA and metastatic pancreatic CA
  - CK7+
  - CK20+
  - CSX-2+
  - Villin+
Mucinous Borderline Tumors

- May be indistinguishable from metastases
- Minimal atypia
- Overt features of invasion may be lacking
Case 33

- 65F with mild upper abdominal pain
- CT with large multicystic tumor in body and tail of pancreas
- FNA with light brown fluid of low viscosity with scattered polygonal cells with hyperchromatic nuclei-low levels of CEA and amylase
- Partial pancreatectomy with 25 cm tumor with thick white bosselated capsule
- Multicystic tumor with central radiating scar and cysts 1-2cm
Serous Microcystic Adenoma
Serous Microcystic Adenoma

- >60 yrs
- F>M
- Pancreas body and tail
- Multiloculated cyst, usually <2cm each
- Central sunburst scar and septal calcification
- Fluid low levels of CEA, contrast to mucinous
- Mutation of von Hippel-Lindau gene (3p)
Uncommon Variants

- **Serous Oligocystic (Macrocystic) adenoma**
  - VHL associated
  - Incompletely circumscribed

- **Solid Serous Adenoma**
  - Hyaline septa separate solid sheets of clear cells
DDX

- Mucinous cystic tumors
- Neuroendocrine tumors
- Solid-pseudopapillary tumor
- Lymphangioma
- Metastatic renal cell CA
Metastatic Renal Cell CA-Clear Cell Type

- Intracellular lipid
- PAS+
- PAS D-
- Chromogranin A-
- EMA+
- CEA-
Solid-Pseudopapillary Neoplasm

- Adolescent women
- Low grade malignancy
- A1AT+
- Progesterone Receptor+
Case 34

- 46F with lower abdominal pain, pelvic mass, menometrorrhagia
- Hysterectomy with solitary, poorly circumscribed 8cm mass
- Soft, variegated, gray yellow tumor with necrosis
Leiomyosarcoma
Leiomyosarcoma

- 1% malignant uterine corpus tumors
- Postmenopausal (average 52 yrs)
- Unrelated to endometrial CA risk factors
- Signs and symptoms similar to leiomyoma
- Usually solitary
- 2/3 intramural
- Range 6-9 cm
Prognosis

- Intraperitoneal spread to LN, lung, liver, bone, kidney

- Recurrence
  - 57% hematogenous
  - 20% pelvic
## Necrosis

<table>
<thead>
<tr>
<th>Necrosis</th>
<th>Defn.</th>
<th>LM</th>
<th>LMS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hyaline (Infarctive)</td>
<td>Necrosis separated from viable tumor by zone of connective tissue</td>
<td>+++</td>
<td>+/-</td>
</tr>
<tr>
<td>Tumor cell</td>
<td>Abrupt transition viable cells to necrotic cells without interposed zone</td>
<td>Rare</td>
<td>+++</td>
</tr>
<tr>
<td>Ulcerative</td>
<td>Ulcerated surface of tumor</td>
<td>+++</td>
<td>+++</td>
</tr>
<tr>
<td>Tumor Cell Necrosis</td>
<td>Atypia</td>
<td>MF/10 hpf</td>
<td>DX</td>
</tr>
<tr>
<td>---------------------</td>
<td>----------------</td>
<td>-----------</td>
<td>-----------</td>
</tr>
<tr>
<td>Present</td>
<td>Diffuse moderate to severe</td>
<td>Any level</td>
<td>LMS</td>
</tr>
<tr>
<td>Present</td>
<td>None to mild</td>
<td>&gt;10</td>
<td>LMS</td>
</tr>
<tr>
<td>Present</td>
<td>None to mild</td>
<td>&lt;10</td>
<td>LMS (r/o LM infarct)</td>
</tr>
<tr>
<td>Tumor Cell Necrosis</td>
<td>Atypia</td>
<td>MF/10 hpf</td>
<td>DX</td>
</tr>
<tr>
<td>---------------------</td>
<td>-------------------------------</td>
<td>-----------</td>
<td>-----------------------------------------</td>
</tr>
<tr>
<td>Absent</td>
<td>Diffuse moderate to severe</td>
<td>&gt;10</td>
<td>LMS</td>
</tr>
<tr>
<td>Absent</td>
<td>Diffuse moderate to severe</td>
<td>&lt;10</td>
<td>Atypical LM with LRR</td>
</tr>
<tr>
<td>Absent</td>
<td>None to mild</td>
<td>&lt;10</td>
<td>LM</td>
</tr>
<tr>
<td>Absent</td>
<td>None to mild</td>
<td>&gt;10</td>
<td>LM-mitotically active</td>
</tr>
<tr>
<td>Absent</td>
<td>Focal moderate to severe</td>
<td>&gt;10</td>
<td>LM with limited experience</td>
</tr>
<tr>
<td>Absent</td>
<td>Focal moderate to severe</td>
<td>&gt;15</td>
<td>STUMP</td>
</tr>
</tbody>
</table>
Adenosarcoma

- Biphasic neoplasm
- Recurrence 25-40%
- Metastasis rare <5%
Atypical Polypoid Adenomyoma

- Benign
- Usually diagnosed on EMC
- Mimics endometrioid adenocarcinoma with myometrial invasion
Case 35

- 67M with abdominal pain
- Mass in right lobe of liver
- Partial hepatectomy with 7cm well circumscribed nodule, tan, focally hemorrhagic
Hepatocellular Carcinoma
<table>
<thead>
<tr>
<th>Country</th>
<th>Incidence</th>
<th>Age Decade</th>
<th>Sex M:F</th>
</tr>
</thead>
<tbody>
<tr>
<td>China</td>
<td>80/100,1000</td>
<td>4-5th</td>
<td>8:1</td>
</tr>
<tr>
<td>USA</td>
<td>5/100,000</td>
<td>7-8th</td>
<td>2:1</td>
</tr>
<tr>
<td>Tumor</td>
<td>Hepatocytes</td>
<td>Other</td>
<td></td>
</tr>
<tr>
<td>----------------</td>
<td>------------------------</td>
<td>----------------------------</td>
<td></td>
</tr>
<tr>
<td>HCC</td>
<td>Uniform &gt;3 layers</td>
<td>Acinar growth occ.</td>
<td></td>
</tr>
<tr>
<td>Adenoma</td>
<td>Uniform &lt;3 layers</td>
<td>No bile ducts</td>
<td></td>
</tr>
<tr>
<td>FNH</td>
<td>Nodular aggregates</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>with thin fibrous</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>septae</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Fibrolamellar HCC</td>
<td>Large polygonal cells</td>
<td>Lamellar fibrosis</td>
<td></td>
</tr>
<tr>
<td></td>
<td>with eos. granular</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>cytoplasm</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Quick Facts

- Hepatocellular adenoma associated with steroid use
- Fibrolamellar hepatocellular carcinoma usually in non-cirrhotic liver young adult
Case 36

- 62M with 30lb weight loss and early satiety for 5 months
- Ulcerated mass in distal lesser curvature
- 5cm elevated, ulcerated mass in antrum with grossly involved lymph node
Adenocarcinoma, Intestinal Type
Quick Facts

- Carcinoid tumors are indolent when arising in autoimmune gastritis vs. sporadic

- Adenocarcinoma, diffuse type, may be familial due to germline E-cadherin mutations

- Adenocarcinoma, intestinal type, most strongly linked to H. pylori
Case 37

- 2yo boy with palpable right sided abdominal mass
- 13cm tumor in upper pole of right kidney with variegated, focally necrotic cut surface
Wilm’s Tumor
Blastema predominant
Nephroblastoma

- Most common GU childhood tumor
- 1-3 yrs
- Stage and histology most important prognostic features
- Anaplasia is unfavorable histology
Histopathology

- Triphasic epithelial, stromal, blastemal
  - Blastemal predominance if >33%
- VIM+, CK+/-
- MIC2 negative
<table>
<thead>
<tr>
<th>Tumor</th>
<th>Histology</th>
<th>Other</th>
</tr>
</thead>
<tbody>
<tr>
<td>CCSK</td>
<td>Round to spindled nuclei, optically clear cytoplasm, stroma with prominent arborizing capillaries</td>
<td>Mets to bone</td>
</tr>
<tr>
<td>MRTK</td>
<td>Prominent eosinophilic cytoplasm with eccentric nuclei</td>
<td>EMA and desmin+ Central PNET association</td>
</tr>
<tr>
<td>CMN</td>
<td>Uniform spindle cells with macro/microcysts</td>
<td>t(12;15)</td>
</tr>
</tbody>
</table>
Case 38

- 69F s/p grade 3 endometrioid endometrial adenocarcinoma, presents with abdominal pain
- Right adrenal mass by CT scan
- 3 cm tan nodule in adrenal gland
- Tumor
  - + CK20 and CD45
  - - EMA, pankeratin, S100, MART-1, calretinin, synaptophysin
Diffuse Large B-cell Lymphoma (Anaplastic variant)
Diffuse Large B-cell Lymphoma (Anaplastic variant)

- 30-40% of adult NHL
- 6-7th decades
Immunoperoxidase

- Pan B cell markers
- Surface Ig 75%
- Bcl-2 +
- Ki-67+ but usually <99%
Mind Your Ts and Bs

- B cell lymphomas with CD30 are not anaplastic large cell lymphomas
- Distinction from anaplastic T or null cell lymphomas
<table>
<thead>
<tr>
<th>DDX</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>HD-lymphocytic depleted</td>
<td>CD15+ CD30+ CD45-</td>
</tr>
<tr>
<td>DLBCL-anaplastic var.</td>
<td>CD20+ CD45+ EMA-</td>
</tr>
<tr>
<td>Adrenocortical CA</td>
<td>Inhibin+ VIM+</td>
</tr>
<tr>
<td>Metastatic melanoma</td>
<td>S100+ VIM+</td>
</tr>
</tbody>
</table>
Case 39

- 43M with second heart transplant
- First heart transplant occurred after idiopathic illness leading to cardiac failure
- 740gm 15x13x9 cm heart
- Epicardium diffusely granular with adhesions
- RV 0.9cm, LV 2.0cm
- Coronary arteries uniformly narrowed and patent
Severe Acute Cellular Rejection and Chronic Vasculopathy
## DDX

<table>
<thead>
<tr>
<th>Condition</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acute cellular rejection</td>
<td>Most common, Cell mediated, First 3 months post transplant</td>
</tr>
<tr>
<td>Humoral rejection</td>
<td>DIF shows complement or Ig in vessel walls, Beyond immediate post-op period</td>
</tr>
<tr>
<td>PTLD</td>
<td>Preneoplastic/neoplastic, Viral oncogenesis and immunosuppression (EBV)</td>
</tr>
</tbody>
</table>
Case 40

- 13F with type I diabetes mellitus presents with right infra-auricular mass gradually enlarging for previous 2 years
- Mass more prominent during episodes of upper respiratory infection but otherwise asymptomatic
- Parotidectomy with 1.9 cm circumscribed tan nodule
Acinic Cell Adenocarcinoma
## Acinic Cell CA

<table>
<thead>
<tr>
<th>Tumor type</th>
<th>Overall</th>
<th>Children</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>AdenoCA, NOS</td>
<td>Mucoepidermoid Acinic cell CA</td>
</tr>
<tr>
<td></td>
<td>Mucoepidermoid</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Acinic cell CA</td>
<td></td>
</tr>
</tbody>
</table>
Acinic Cell CA

- Parotid gland most common site
- F:M 3:2
- 2-8\textsuperscript{th} decades
- 3\% bilateral but most common bilateral malignant neoplasm
- Recurrence in 1/3
- Death 15\%
Acinar

Non specific glandular

Intercalated duct-like

Clear

Vacuolated
Histopathology

- Solid
- Microcystic
- Papillary-cystic
- Follicular
- Abundance of lymphocytic stroma
- Undifferentiated
### DDX

<table>
<thead>
<tr>
<th>condition</th>
<th>description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acinic Cell CA</td>
<td>May be bilateral</td>
</tr>
<tr>
<td>Polymorphous LGAC</td>
<td>Monomorphous cells in cords and trabeculae</td>
</tr>
<tr>
<td></td>
<td>Minor salivary glands</td>
</tr>
<tr>
<td>Mucoepidermoid CA</td>
<td>Most common malignant salivary gland in children</td>
</tr>
</tbody>
</table>