Retroperitoneal/Abdominal Masses

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UCSF Postgraduate Course in General Surgery
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San Francisco, CA
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Retroperitoneal Mass
Case presentation

• 53-year-old woman
  • April 2007: right LE pain
    • MRI: herniated disc; steroid injections

  • Dec. 2007: abdominal pain; CT: retroperitoneal mass

Retroperitoneal Mass
Case presentation

• Case presentation
• Differential diagnosis
• Preoperative considerations: work-up and planning
• Radiation therapy
• Advanced disease
• UCSF experience
Retropertitoneal Mass
Case presentation

• 53-year-old woman
  • Dec. 2007: CT-guided core needle biopsy
  • Spindle cell neoplasm:
    • No mitotic figures, no necrosis, SMA +, desmin +
    • Smooth muscle neoplasm, uncertain malignant potential
  • Deemed unresectable elsewhere
  • Adriamycin, ifosfamide, mesna for 2 cycles
  • March 2008: UCSF for second opinion

Retropertitoneal Mass
Case presentation

• Initial impression at UCSF
  • Leiomyosarcoma arising from the IVC
  • Appears to be low-grade
  • Only chance for cure is surgical resection
  • Wide margins not feasible
  • May need right nephrectomy
  • Appears to be anterior and away from right femoral nerve

Retropertitoneal Mass
Case presentation

• 53-year-old woman
  • Preoperative consultation with vascular surgeon (C.E.)
  • Preoperative consultation with radiation oncology (A.G.)
  • Right ureteral stent placed by urology (K.G.)

Retropertitoneal Mass
Case presentation

Resection of RP neoplasm with en-bloc infrarenal IVC
Retroperitoneal Mass
Case presentation

- Right ureter
- Aorta
- PTFE bypass graft

IORT
- Lead protection of right kidney and ureter
- 15 Gy via 9 cm cone

53-year-old woman
- Pathology: 11.5 cm, leiomyosarcoma, grade I, margins negative
- Adjuvant external beam radiation therapy

2 years postoperative: no evidence of disease
Retroperitoneal Mass
Preoperative considerations

- Contiguous organ removal
  - Kidney, colon, pancreas, IVC, iliac vessels, nerve root
- Proximity to ureter
- Proximity to femoral nerve
- Anticipated close surgical margins

- Preoperative alpha-blockade (functioning paraganglioma)
- Preoperative angioembolization

Retroperitoneal Mass
Differential diagnosis

- Malignant
  - Sarcoma
  - GIST
  - Lymphoma
  - Germ cell tumor

- Benign lesions
  - Lipoma
  - Peripheral nerve sheath tumor
  - Teratoma
  - Paraganglioma

- Desmoid

Retroperitoneal Mass
Differential diagnosis

- Malignant
  - Sarcoma
  - GIST
  - Lymphoma
  - Germ cell tumor

- Benign lesions
  - Lipoma
  - Peripheral nerve sheath tumor
  - Teratoma
  - Paraganglioma
43-year-old man with abdominal pain for 6 months

s/p resection RP neoplasm, en-bloc ileocecectomy, right nephrectomy
pathology: 8.5 cm fibromatosis (desmoid tumor)

38-year-old man with back, hip, thigh pain for 2 years

s/p resection RP neoplasm
pathology: 8.1 cm benign peripheral nerve sheath tumor (schwannoma)
44-year-old man with HTN (4 anti-hypertensive medications) and abdominal pain.

Retroperitoneal Mass
Differential diagnosis

Multifocal functional paraganglioma of the retroperitoneum and bladder

30-year-old man with enlarging right abdominal mass for 1 year

Preoperative embolization of a functional paraganglioma of the retroperitoneum
30-year-old man with enlarging right abdominal mass for 1 year s/p resection RP neoplasm with en-bloc IVC → PTFE tube graft pathology: 18 cm leiomyosarcoma (grade 1), arising from IVC

**Retroperitoneal Mass**

**Differential diagnosis**

What is the preferred imaging modality to evaluate a retroperitoneal mass?

**Retroperitoneal Mass**

**Imaging**

When might a PET scan be useful for the evaluation of patients with cancer?

- **CT:** retroperitoneal/intra-abdominal
- **MRI:** suspect nerve root involvement
- **¹⁸FDG-PET:** ???

• clinical uses evolving:
  1) diagnosis (evaluate solitary pulmonary nodules)
  2) staging (recurrent disease, nodal disease for epithelioid or angiosarcomas)
  3) prognostic assessment
  4) monitoring response to therapy

64-year-old woman
- 15 months prior underwent resection of RP sarcoma, left nephrectomy, splenectomy, colectomy
- pathology: 34 cm well-differentiated liposarcoma
- surveillance CT: recurrent RP mass

PET/CT revealed: numerous hypermetabolic masses

64-year-old woman • s/p re-resection of recurrent RP liposarcoma, IORT
- pathology: dedifferentiated liposarcoma
- adjuvant EBRT
- NED > 4 years postoperatively

46-year-old woman who developed abdominal pain after a fall
46-year-old woman who developed abdominal pain after a fall. The next step in her management should be:

A) CT-guided biopsy
B) measure plasma-free metanephrines then biopsy
C) measure plasma-free metanephrines and surgical evaluation

Soft Tissue Sarcoma

Biopsy

• None: resectable retroperitoneal/intra-abdominal

*If it will change management:
• Fine-needle aspiration: recurrent or metastatic disease
• Core-needle biopsy: equivalent to incisional biopsy
• Incisional biopsy: less common

• Experienced pathologist


Retroperitoneal Sarcoma

Treatment

• Challenges
  • Large size
  • Proximity to/invasion of adjacent structures
    • bowel, vessels, nerves, bones, kidney, ureter, bladder
  • Complete resection difficult
  • High local recurrence rate/poor survival
**Soft Tissue Sarcoma**

**Surgery**

- Principals of surgery
  - Optimal margins and oncologic control
  - Maximal function and minimal morbidity
  - Limb sparing generally preferable

- Consider preoperative cytotoxic therapy (chemotherapy, RT), if unable to achieve the above.

NCCN 2005.

**Retroperitoneal Neoplasm**

**Intraoperative neurophysiological monitoring**


**Soft Tissue Sarcoma**

**Primary retroperitoneal classification system**

<table>
<thead>
<tr>
<th></th>
<th>Low grade</th>
<th>High grade</th>
<th>5-year survival (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Complete resection</td>
<td>I</td>
<td>II</td>
<td>I</td>
</tr>
<tr>
<td>Incomplete resection</td>
<td>III</td>
<td></td>
<td>II</td>
</tr>
<tr>
<td>Distant metastasis</td>
<td>III</td>
<td></td>
<td>III</td>
</tr>
<tr>
<td></td>
<td>IV</td>
<td></td>
<td>IV</td>
</tr>
</tbody>
</table>


**Retroperitoneal Sarcoma**

**Outcome**

<table>
<thead>
<tr>
<th></th>
<th>complete resection</th>
<th>local recurrence</th>
<th>metastasis</th>
<th>survival</th>
</tr>
</thead>
<tbody>
<tr>
<td>Low grade</td>
<td>I 67%</td>
<td>41%</td>
<td>21%</td>
<td>54%</td>
</tr>
<tr>
<td>High grade</td>
<td>II 65%</td>
<td>57%</td>
<td>33%</td>
<td>46%</td>
</tr>
<tr>
<td>Complete resection</td>
<td>III 54%</td>
<td>42% **</td>
<td>22% **</td>
<td>37%</td>
</tr>
<tr>
<td>Incomplete resection</td>
<td>IV 88%</td>
<td>54%</td>
<td>11%</td>
<td>54%</td>
</tr>
<tr>
<td>Distant metastasis</td>
<td>III 78%</td>
<td>42%</td>
<td>15%</td>
<td>45%</td>
</tr>
<tr>
<td></td>
<td>IV 95%</td>
<td>45%</td>
<td>ND</td>
<td>52%</td>
</tr>
</tbody>
</table>

*primary and recurrent    ** > 5-years

Lewis et al. (MSKCC)\(^1\) 67% 41% 21% 54%
Stoeckle et al. (France)\(^2\) 65% 57% 33% 46%
von Dalen et al. (Netherlands)\(^3\) 54% 42% ** 22% ** 37%
Gronchi et al. (Milan)\(^4\) 88% 54% 11% 54%
Hassan et al. (Mayo)\(^5\) 78% 42% 15% 45%
Erzen et al. (Slovenia)\(^6\) 95% 45% ND 52%
Pawlik et al. (MDACC/Toronto)\(^7\) 95% 40% 15% 61%

\(^1\)Ann Surg. 1998
\(^2\)Cancer 2001
\(^3\)Ann Surg Oncol. 2001
\(^4\)Ann Surg Oncol. 2004
\(^5\)Ann Surg. 2004
\(^6\)Ann Surg Oncol. 2005
\(^7\)Ann Surg Oncol. 2006.
Retroperitoneal Sarcoma
Radiation therapy: IORT

- Rationale: 5-year local recurrence rates 37-75%
- > 55 Gy necessary to control microscopic residual disease
- Prohibitive toxicity to small intestine, liver, kidneys
- Single-dose IORT = 1.5-2.5 same total dose of EBRT
  - 15 Gy IORT + 45 Gy EBRT = 75-87.5 Gy EBRT


Intraoperative radiotherapy (IORT)
NCI randomized controlled trial

<table>
<thead>
<tr>
<th>IORT/low dose EBRT</th>
<th>high dose EBRT</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>(n = 15)</td>
<td>(n = 20)</td>
<td></td>
</tr>
<tr>
<td>median survival (mo.)</td>
<td>45</td>
<td>52</td>
</tr>
<tr>
<td>local recurrence (%)</td>
<td>40</td>
<td>80</td>
</tr>
<tr>
<td>median time to local recurrence (mo.)</td>
<td>&gt;127</td>
<td>38</td>
</tr>
<tr>
<td>enteritis (%)</td>
<td>13</td>
<td>50</td>
</tr>
<tr>
<td>peripheral neuropathy (%)</td>
<td>47</td>
<td>0</td>
</tr>
<tr>
<td>(moderate to severe)</td>
<td></td>
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</tr>
</tbody>
</table>


Retroperitoneal Sarcoma
Intraoperative radiotherapy (IORT)

52-year-old man with testicular CA ~30 yrs ago, s/p orchiectomy, EBRT

Retroperitoneal Sarcoma
Intraoperative radiotherapy (IORT)

52-year-old man with testicular CA ~30 yrs ago, s/p orchiectomy, EBRT s/p resection RP neoplasm, en-bloc diaphragm, IORT pathology: 12.3 cm undifferentiated pleomorphic sarcoma
A 60-year-old woman with increasing abdominal girth for 5 years

s/p resection RP neoplasm
pathology: 48 cm (17.4 kg) well differentiated liposarcoma

60-year-old woman with increasing abdominal girth for 5 years

A 60-year-old woman underwent resection of a RP well differentiated liposarcoma. Her most likely cause of death in the future will be due to:

A) lung metastases
B) liver metastases
C) multifocal bowel obstruction

Role of incomplete resection?

• No significant difference in survival between patients whose disease is unresectable and those who undergo incomplete resection

Complete resection is goal for curative intent

• Possible roles for debulking surgery
  • Palliation of symptoms
  • Unresectable retroperitoneal liposarcoma

**Soft Tissue Sarcoma**

**Advanced disease**

- Consider metastasectomy
- Chemotherapy
- Radiotherapy
- Ablative procedures
- Palliative surgery
- Supportive care

NCCN 2005.

A 52-year-old woman underwent a TAH/BSO for uterine leiomyosarcoma. Eight months later developed lung and liver metastases. She received 7 cycles of gemcitabine and docetaxel and had stable disease.

**Soft Tissue Sarcoma**

**Advanced disease**

A 52-year-old woman underwent a TAH/BSO for uterine leiomyosarcoma. Eight months later developed lung and liver metastases. She received 7 cycles of gemcitabine and docetaxel and had stable disease.

**Soft Tissue Sarcoma**

**Advanced disease**

Institution      | Years     | N      | Overall Survival | Disease-free survival | Recurrence |
----------------|-----------|--------|------------------|-----------------------|------------|
MSKCC            | 1982-2000 | 56     | 30%              | 20%                   | 84%        |
MDACC            | 1996-2005 | 66*    | 27%              | 16%                   | 67%        |

*Resection and/or ablation

### UCSF Experience

#### Malignant

<table>
<thead>
<tr>
<th>Tumor Type</th>
<th>No. of Patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Liposarcoma</td>
<td>23/43 (53)</td>
</tr>
<tr>
<td>Leiomyosarcoma</td>
<td>8/43 (19)</td>
</tr>
<tr>
<td>Metastatic Colon Ca</td>
<td>1/43 (2)</td>
</tr>
<tr>
<td>Undiff Pleomorphic Sarcoma</td>
<td>1/43 (2)</td>
</tr>
<tr>
<td>Renal Cell Sarcoma</td>
<td>1/43 (2)</td>
</tr>
<tr>
<td>Solitary Fibrous Tumor</td>
<td>1/43 (2)</td>
</tr>
<tr>
<td>Chondrosarcoma</td>
<td>1/43 (2)</td>
</tr>
<tr>
<td>Ewing's sarcoma</td>
<td>1/43 (2)</td>
</tr>
</tbody>
</table>

#### Benign

<table>
<thead>
<tr>
<th>Tumor Type</th>
<th>No. of Patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Schwannoma</td>
<td>4/17 (24)</td>
</tr>
<tr>
<td>Paraganglioma</td>
<td>2/17 (12)</td>
</tr>
<tr>
<td>Lipoma</td>
<td>2/17 (12)</td>
</tr>
<tr>
<td>Myxoma</td>
<td>2/17 (12)</td>
</tr>
<tr>
<td>Fibroma</td>
<td>1/17 (6)</td>
</tr>
<tr>
<td>Desmoid tumor</td>
<td>1/17 (6)</td>
</tr>
<tr>
<td>Epithelioid tumor</td>
<td>1/17 (6)</td>
</tr>
<tr>
<td>Capillary hemangioma</td>
<td>1/17 (6)</td>
</tr>
<tr>
<td>Teratoma</td>
<td>1/17 (6)</td>
</tr>
<tr>
<td>Muscle vasc with thrombus</td>
<td>1/17 (6)</td>
</tr>
<tr>
<td>Hematoma</td>
<td>1/17 (6)</td>
</tr>
</tbody>
</table>

#### UCSF Experience

### Complete resection

<table>
<thead>
<tr>
<th>Tumor Type</th>
<th>No. of Patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kidney</td>
<td>18/56 (32)</td>
</tr>
<tr>
<td>Colon</td>
<td>9/56 (16)</td>
</tr>
<tr>
<td>Pancreas</td>
<td>4/56 (7)</td>
</tr>
<tr>
<td>Small Bowel</td>
<td>3/56 (5)</td>
</tr>
<tr>
<td>Adrenal</td>
<td>3/56 (5)</td>
</tr>
<tr>
<td>Bladder</td>
<td>2/56 (4)</td>
</tr>
<tr>
<td>Liver</td>
<td>1/56 (2)</td>
</tr>
<tr>
<td>Spleen</td>
<td>1/56 (2)</td>
</tr>
<tr>
<td>Diaphragm</td>
<td>1/56 (2)</td>
</tr>
<tr>
<td>Iliac wing</td>
<td>1/56 (2)</td>
</tr>
</tbody>
</table>

### Major vascular resection

<table>
<thead>
<tr>
<th>Tumor Type</th>
<th>No. of Patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>IVC</td>
<td>4/56 (7)</td>
</tr>
<tr>
<td>Iliac artery, vein</td>
<td>1/56 (2)</td>
</tr>
</tbody>
</table>

### Posterior laminotomy

<table>
<thead>
<tr>
<th>Tumor Type</th>
<th>No. of Patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>IVC</td>
<td>3/56 (5)</td>
</tr>
</tbody>
</table>

### Preoperative embolization

<table>
<thead>
<tr>
<th>Tumor Type</th>
<th>No. of Patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>IVC</td>
<td>2/56 (4)</td>
</tr>
</tbody>
</table>

### UCSF Experience

#### 5-year Overall Survival

<table>
<thead>
<tr>
<th>Institution</th>
<th>Years</th>
<th>N</th>
<th>Overall Survival</th>
</tr>
</thead>
<tbody>
<tr>
<td>MSKCC</td>
<td>1982-1997</td>
<td>161</td>
<td>37%</td>
</tr>
</tbody>
</table>

Retroperitoneal Mass
Summary

• Complete and safe resection feasible
• Preoperative planning important
• Anticipate potential intraoperative findings
• Large size → difficult resection and high risk local recurrence
• IORT and EBRT when anticipate close focal margin