Surgical Management of Soft Tissue Sarcoma: Retroperitoneal Masses

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

Retroperitoneal Mass
Differential diagnosis

• malignant
  • sarcoma
  • GIST
  • lymphoma
  • germ cell tumor
  • other metastatic adenopathy

Retroperitoneal Sarcoma
Case presentation

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

Retroperitoneal Mass
Differential diagnosis

• malignant
  • sarcoma
  • GIST
  • lymphoma
  • germ cell tumor/metastatic adenopathy

• benign lesions
  • lipoma
  • peripheral nerve sheath tumor
  • teratoma
  • paraganglioma
Retroperitoneal Mass
Differential diagnosis

- malignant
  - sarcoma
  - GIST
  - lymphoma
  - germ cell tumor
- desmoid
- benign lesions
  - lipoma
  - peripheral nerve sheath tumor
  - teratoma
  - paraganglioma

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.
Multifocal functional paraganglioma of the retroperitoneum and bladder


She underwent a Whipple procedure (pylorus-preserving), cholecystectomy, and extensive retroperitoneal lymphadenectomy; all gross disease removed.

Path: well-differentiated PNET, 8 cm, invasion of duodenum/peripancreatic tissues, < 2 mitoses/10 hpf; no necrosis, 47/49 lymph nodes.

43-year-old man with abdominal pain for 6 months
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)

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

53-year-old woman with abdominal pain.
Retroperitoneal Mass

Case presentation

Resection of RP neoplasm with en-bloc infrarenal IVC
Pathology: 11.5 cm leiomyosarcoma, grade I, margins negative

Retroperitoneal Mass

Case presentation

Right ureter
PTFE bypass graft
Aorta

Retroperitoneal Mass

60-year-old man with increasing abdominal girth for 8 months.
For 2 weeks, he had bilateral lower extremity edema refractory to lasix.
He gained more than 30 pounds.

Retroperitoneal Mass

60-year-old man with increasing abdominal girth for 8 months.
60-year-old man with increasing abdominal girth for 8 months. s/p resection RP neoplasm, right hemicolecction, right nephrectomy. pathology: 45 cm dedifferentiated liposarcoma (grade 2), 70 pounds

Waist size: preoperative 48 inches; postoperative 34 inches.

60-year-old man 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

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

- CT: retroperitoneal/intra-abdominal
- MRI: suspect nerve root involvement
- $^{18}$FDG-PET: ?

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

- 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
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

Retroperitoneal Mass
Role of biopsy?

46-year-old woman who developed abdominal pain after a fall
s/p resection RP neoplasm, en-bloc right adrenalectomy pathology: 10.1 cm paraganglioma (extra-adrenal pheochromocytoma)

Retroperitoneal Mass
Role of biopsy?

Retroperitoneal Mass
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.

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>70-90</td>
</tr>
<tr>
<td>Incomplete resection</td>
<td>III</td>
<td></td>
<td>40-45</td>
</tr>
<tr>
<td>Distant metastasis</td>
<td>III</td>
<td>IV</td>
<td>25</td>
</tr>
<tr>
<td></td>
<td>IV</td>
<td></td>
<td>0-15</td>
</tr>
</tbody>
</table>

Retropertitoneal Sarcoma

### Outcome

<table>
<thead>
<tr>
<th>Study</th>
<th>Complete Resection</th>
<th>Local Recurrence</th>
<th>Metastasis</th>
<th>Survival</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lewis et al. (MSKCC)</td>
<td>67%</td>
<td>41%</td>
<td>21%</td>
<td>54%</td>
</tr>
<tr>
<td>Stoeckle et al. (France)</td>
<td>65%</td>
<td>57%</td>
<td>33%</td>
<td>46%</td>
</tr>
<tr>
<td>van Dalen et al. (Netherlands)</td>
<td>54%</td>
<td>42% **</td>
<td>22% **</td>
<td>37%</td>
</tr>
<tr>
<td>Gronchi et al. (Milan)</td>
<td>88%</td>
<td>54%</td>
<td>11%</td>
<td>54%</td>
</tr>
<tr>
<td>Hassan et al. (Mayo)</td>
<td>79%</td>
<td>42%</td>
<td>15%</td>
<td>45%</td>
</tr>
<tr>
<td>Erzen et al. (Slovenia)**</td>
<td>95%</td>
<td>45%</td>
<td>ND</td>
<td>52%</td>
</tr>
<tr>
<td>Pawlik et al. (MDACC/Toronto)**</td>
<td>95%</td>
<td>40%</td>
<td>15%</td>
<td>61%</td>
</tr>
</tbody>
</table>

*primary and recurrent  ** > 5-years

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### 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

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### 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

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### Intraoperative radiotherapy (IORT)

NCI randomized controlled trial

<table>
<thead>
<tr>
<th></th>
<th>IORT/low dose EBRT (n = 15)</th>
<th>high dose EBRT (n = 20)</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Median survival (mo.)</td>
<td>45</td>
<td>52</td>
<td>NS</td>
</tr>
<tr>
<td>Local recurrence (%)</td>
<td>40</td>
<td>80</td>
<td>&lt; 0.05</td>
</tr>
<tr>
<td>Median time to local recurrence (mo.)</td>
<td>&gt;127</td>
<td>38</td>
<td>&lt; 0.05</td>
</tr>
<tr>
<td>Enteritis (%)</td>
<td>13</td>
<td>50</td>
<td>&lt; 0.05</td>
</tr>
<tr>
<td>Peripheral neuropathy (%) (moderate to severe)</td>
<td>47</td>
<td>0</td>
<td>&lt; 0.01</td>
</tr>
</tbody>
</table>

Soft Tissue Sarcoma
Advanced disease

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

NCCN 2005.

Soft Tissue Sarcoma
Resection of liver metastases

<table>
<thead>
<tr>
<th>Institution</th>
<th>Years</th>
<th>N</th>
<th>Overall Survival</th>
<th>Disease-free survival</th>
<th>Recurrence</th>
</tr>
</thead>
<tbody>
<tr>
<td>MSKCC</td>
<td>1982-2000</td>
<td>56</td>
<td>30%</td>
<td>20%</td>
<td>84%</td>
</tr>
<tr>
<td>MDACC</td>
<td>1996-2005</td>
<td>66*</td>
<td>27%</td>
<td>16%</td>
<td>67%</td>
</tr>
</tbody>
</table>

*Resection and/or ablation


Soft Tissue Sarcoma
Resection of pulmonary metastases

<table>
<thead>
<tr>
<th>Institution</th>
<th>Years</th>
<th>N</th>
<th>Overall Survival</th>
<th>Disease-free survival</th>
<th>Recurrence</th>
</tr>
</thead>
<tbody>
<tr>
<td>MSKCC</td>
<td>1982-1997</td>
<td>161</td>
<td>37%</td>
<td>?</td>
<td>65%</td>
</tr>
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Soft Tissue Sarcoma
Intraoperative radiotherapy (IORT)

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

Retroperitoneal Sarcoma
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

Retroperitoneal Mass
IORT

UCSF Experience

<table>
<thead>
<tr>
<th>Malignant Tumor</th>
<th>No. of Patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Liposarcoma</td>
<td>23/43 (53)</td>
</tr>
<tr>
<td>Leiomysarcoma</td>
<td>6/43 (15)</td>
</tr>
<tr>
<td>Malign Periph Nerve Sheath Tumor</td>
<td>5/43 (12)</td>
</tr>
<tr>
<td>Metastatic Colon Ca</td>
<td>1/43 (2)</td>
</tr>
<tr>
<td>Spindle Cell Sarcoma</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>
<tr>
<td>Lipoma</td>
<td>2/17 (12)</td>
</tr>
<tr>
<td>Schwannoma</td>
<td>4/17 (24)</td>
</tr>
<tr>
<td>Paraganglioma</td>
<td>2/17 (12)</td>
</tr>
<tr>
<td>Myelolipoma</td>
<td>2/17 (12)</td>
</tr>
<tr>
<td>Fibroma</td>
<td>1/17 (6)</td>
</tr>
<tr>
<td>Densosid 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>Muscular vessel with thrombus</td>
<td>1/17 (6)</td>
</tr>
<tr>
<td>Hematoma</td>
<td>1/17 (6)</td>
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UCSF Experience

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UCSF Experience

Complete resection: 56/60 (93)
Multi-organ resection: 31/56 (55)

- Kidney: 18/56 (32)
- Colon: 3/56 (5)
- Pancreas: 4/56 (7)
- Small Bowel: 3/56 (5)
- Adrenal: 3/56 (5)
- Bladder: 5/56 (4)
- Liver: 1/56 (2)
- Spleen: 1/56 (2)
- Diaphragm: 1/56 (2)
- Iliac wing: 1/56 (2)

Major vascular resection: 5/56 (9)
- IVC: 4/56 (7)
- Iliac artery, vein: 1/56 (2)

Posterior laminotomy: 3/56 (5)
Preoperative embolization: 2/56 (4)

Ureteral stent: 22/60 (36)
Femoral nerve monitoring: 7/60 (12)

Major complications: 10/60 (17)
- Abscess: 6/60 (10)
- Atrial fibrillation: 4/60 (7)
- Enterocutaneous fistula: 1/60 (2)
- Death: 1/60 (2)

Soft Tissue Sarcoma: Retroperitoneal Masses

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