Multidisciplinary management of retroperitoneal sarcomas

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Adult Soft Tissue Sarcoma

Overview

• Definition: arise from nonepithelial, extraskeletal tissues
  • fat, muscle, nerve and nerve sheath, blood vessels, other connective tissues

• Incidence: 9,220 cases per year in the United States

• Mortality: 3,560 cases per year

• Multidisciplinary management essential

Adult Soft Tissue Sarcoma Distribution

Clark et al. NEJM 2005.
Retroperitoneal Sarcoma
Differential diagnosis of retroperitoneal soft tissue mass

• malignant
  • sarcoma
  • GIST
  • lymphoma
  • germ cell tumor
Retroperitoneal Sarcoma
Differential diagnosis of retroperitoneal soft tissue mass

• malignant
  • sarcoma
  • GIST
  • lymphoma
  • germ cell tumor

• benign lesions
  • lipoma
  • peripheral nerve sheath tumor
  • teratoma
  • paraganglioma
Retroperitoneal Sarcoma
Differential diagnosis of retroperitoneal soft tissue mass

• malignant
  • sarcoma
  • GIST
  • lymphoma
  • germ cell tumor

• desmoid

• benign lesions
  • lipoma
  • peripheral nerve sheath tumor
  • teratoma
  • paraganglioma
Soft Tissue Sarcoma Imaging

- CXR/chest CT
- CT: retroperitoneal/intraabdominal
- MRI: extremity

$^{18}$FDG-PET: routine use unjustified

- potential roles:
  - recurrent disease
  - nodal disease for epithelioid or angiosarcomas
  - preoperative prognostic assessment
  - 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
Retroperitoneal Sarcoma
Imaging

PET-CT revealed: numerous hypermetabolic masses
Retroperitoneal Sarcoma
Imaging

64-year-old woman
- s/p re-resection of recurrent RP liposarcoma, IORT
- pathology: dedifferentiated liposarcoma
- adjuvant EBRT
- NED > 2 years postoperatively
Retroperitoneal Sarcoma
Differential diagnosis of retroperitoneal soft tissue mass

46-year-old woman who developed abdominal pain after a fall
Retroperitoneal Sarcoma
Differential diagnosis of retroperitoneal soft tissue mass

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 Sarcoma
Differential diagnosis of retroperitoneal soft tissue mass

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)
Soft Tissue Sarcoma

Biopsy

- None: resectable retroperitoneal/intraabdominal
- Fine-needle aspiration: recurrent or metastatic disease
- Core-needle biopsy: equivalent to incisional biopsy
- Incisional biopsy: less common

- Experienced pathologist

Soft Tissue Sarcoma
Staging

<table>
<thead>
<tr>
<th>Grade</th>
<th>T1a</th>
<th>T1b</th>
<th>T2a</th>
<th>T2b</th>
</tr>
</thead>
<tbody>
<tr>
<td>Low</td>
<td>I</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>High</td>
<td>II</td>
<td>III</td>
<td></td>
<td></td>
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</tbody>
</table>

N1

M1

5-year survival (%)
(except for)—

<table>
<thead>
<tr>
<th>Stage</th>
<th>I</th>
<th>II</th>
<th>III</th>
<th>IV</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>90</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>II</td>
<td>81</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>III</td>
<td>56</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>IV</td>
<td>10-20</td>
<td></td>
<td></td>
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</tr>
</tbody>
</table>

T1: ≤ 5 cm  a: superficial (to fascia)
T2: > 5 cm  b: deep (to fascia, retroperitoneal, pelvic)

# 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></td>
</tr>
<tr>
<td>Incomplete resection</td>
<td>III</td>
<td>III</td>
<td></td>
</tr>
<tr>
<td>Distant metastasis</td>
<td>IV</td>
<td>IV</td>
<td></td>
</tr>
</tbody>
</table>

- I: 70-90%
- II: 40-45%
- III: 25%
- IV: 0-15%

Retroperitoneal Sarcoma
Treatment

- Multidisciplinary management
  - Radiology
  - Pathology
  - Surgery
  - Radiation Oncology
  - Medical Oncology
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
Retroperitoneal Sarcoma
Differential diagnosis of retroperitoneal soft tissue mass

60-year-old woman with increasing abdominal girth for 5 years
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
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
Retroperitoneal Sarcoma
Differential diagnosis of retroperitoneal soft tissue mass

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)
38-year-old man with back, hip, thigh pain for 2 years
Retroperitoneal Sarcoma
Differential diagnosis of retroperitoneal soft tissue mass

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)
Retroperitoneal Sarcoma
Differential diagnosis of retroperitoneal soft tissue mass

30-year-old man with enlarging right abdominal mass for 1 year
Retroperitoneal Sarcoma
Differential diagnosis of retroperitoneal soft tissue mass

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 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)(^1)</td>
<td>67%</td>
<td>41%</td>
<td>21%</td>
<td>54%</td>
</tr>
<tr>
<td>Stoeckle et al. (France)(^2)</td>
<td>65%</td>
<td>57%</td>
<td>33%</td>
<td>46%</td>
</tr>
<tr>
<td>van Dalen et al. (Netherlands)(^3)</td>
<td>54%</td>
<td>42% **</td>
<td>22%**</td>
<td>37%</td>
</tr>
<tr>
<td>Gronchi et al. (Milan)(^4)^*</td>
<td>88%</td>
<td>54%</td>
<td>11%</td>
<td>54%</td>
</tr>
<tr>
<td>Hassan et al. (Mayo)(^5)</td>
<td>78%</td>
<td>42%</td>
<td>15%</td>
<td>45%</td>
</tr>
<tr>
<td>Erzen et al. (Slovenia)(^6)^*</td>
<td>95%</td>
<td>45%</td>
<td>ND</td>
<td>52%</td>
</tr>
</tbody>
</table>

*primary and recurrent  ** > 5-years

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# Retroperitoneal Sarcoma

## 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 (%)</td>
<td>47</td>
<td>0</td>
<td>&lt; 0.01</td>
</tr>
</tbody>
</table>

(moderate to severe)

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Retroperitoneal Sarcoma
Differential diagnosis of retroperitoneal soft tissue mass

52-year-old man with testicular CA ~30 yrs ago, s/p orchiectomy, XRT
Retroperitoneal Sarcoma
Differential diagnosis of retroperitoneal soft tissue mass

52-year-old man with testicular CA ~30 yrs ago, s/p orchiectomy, XRT
s/p resection RP neoplasm, en-bloc diaphragm, IORT
pathology: 12.3 cm undifferentiated pleomorphic sarcoma
Retroperitoneal Sarcoma
Intraoperative neurophysiological monitoring
Soft Tissue Sarcoma
Advanced disease

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

NCCN 2005.
Retroperitoneal Sarcoma
Summary

- Multidisciplinary management essential
  - radiology, pathology, medical oncology, radiation oncology, and surgery
- Large size → difficult resection and high risk local recurrence
- IORT and EBRT when anticipate close focal margin
- High risk of metastatic disease → need better chemotherapy
- Cooperative clinical trials needed due to rarity of disease
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.