Retroperitoneal Sarcomas

Rosemarie E. Hardin
Kings County Hospital
October 14, 2005
Case Presentation

**Chief Complaint:**

“I feel a golf ball size mass in my belly”

**HPI:**

- 63 y/o AA female
- c/o a self-palpated mass in her right abdomen, appreciated one month prior to presentation
- interval increase in size
- + early satiety and decreased appetite
- denied nausea and vomiting or change in bowel habits.
- history was negative for constitutional symptoms; no fever or weight loss
Retroperitoneal Sarcoma

**History**

**PMHx:** Hypertension

**PSHx:** Hysterectomy (fibroid uterus)
Right breast mass excision

**Allergies:** NKDA

**Social Hx:** non-contributory

**Meds:** Atenolol, Avapro
Physical Exam

Pt was A&O, healthy appearing, non-cachetic woman in NAD

**Vitals:** Temp: 98   BP: 146/79   HR: 72   R: 18

**Abdomen:** soft, NT/ND, +BS
+ palpable, non-tender mass measuring approximately 7-8 cm in the right upper quadrant

Retroperitoneal Sarcoma
Pt was referred for a CT scan of Abdomen and Pelvis (6/13/05)

Findings:

large heterogenous soft tissue mass in the right upper abdomen with central necrosis and punctate calcifications; measuring approximately 9 x 8 cm and thought to arise from the duodenum
Retroperitoneal Sarcoma

CT Scan Abdomen and Pelvis
Retroperitoneal Sarcoma

CT Scan Abdomen and Pelvis
Retroperitoneal Sarcoma

CT Scan Abdomen and Pelvis
Pt scheduled for EGD and colonoscopy (6/21/05)

**EGD:** Extrinsic compression of the 1\(^{st}\) and 2\(^{nd}\) portions of the duodenum; no other abnormalities; normal mucosa

**Colonoscopy:** Normal
Surgical Referral

Patient was referred for surgical consultation

Planned operative exploration (6/30/05)
Pre-Op Diagnosis: retroperitoneal mass
Planned Procedure: Whipple procedure
Approach: Chevron incision
Intra-operative Findings

Large, firm retroperitoneal mass, measuring approximately 9 x 11 x 8 cm was identified without attachments to surrounding viscera.
Pathology

- High Grade Leiomyosarcoma, Grade 3 with extensive tumor necrosis
- Margins of specimen free of tumor
- T2B, Nx, Mx
- Smooth Muscle Actin (SMA) +++
- CD117 -
Retroperitoneal Sarcoma

Gross Pathology
Pathology

Retroperitoneal Sarcoma
Retroperitoneal Sarcoma

Post-Operative Course

Patient underwent complete resection of retroperitoneal mass without en-bloc resection of adjacent viscera. Pt was monitored in recovery room overnight and then transferred to regular floor. Pt had uneventful post op recovery and was discharged home on POD #6
Retroperitoneal Sarcomas
Introduction

- Soft tissue sarcomas are rare; approximately 8,600 new cases diagnosed annually
- One third of malignant tumors that arise in the retroperitoneum are sarcomas
- RP sarcomas arise from mesenchymal cells, which are usually located in muscle, fat and connective tissues
- Median age for patients who present with RPS is in the sixth decade of life; male to female ratio is equal

Retroperitoneal Sarcoma

**Frequency**

- Thigh, buttock and groin region – 46%
- Upper extremity – 13%
- Head and neck region – 9%
- Torso – 18%
- Retroperitoneal tissues – 13%

Histologic Types

- LIPOSARCOMA: 55
- LEIOMYOSARCOMA: 26
- MALIGNANT FIBROUS HISTIOCYTOMA: 10
- FIBROSARCOMA: 3
- MALIGNANT PERIPHERAL NERVE SHEATH TUMOR: 5
- EXTRASKELETAL OSTEOSARCOMA: 1

Clinical Manifestations

- Asymptomatic abdominal mass (80%)
- Symptoms related to mass effect or local invasion (pain, gastrointestinal obstruction, early satiety and weight loss)
- Neurological and musculoskeletal symptoms referable to the lower extremity
- Median duration of symptoms before diagnosis is 4 months

Diagnosis

CT scan of the Abdomen and Pelvis
- Assessment of tumor location and relation to adjacent viscera
- Identification of metastatic lesions in the liver or peritoneal cavity

MRI

**Pre-operative tissue diagnosis of resectable retroperitoneal masses is not required**
Retroperitoneal Sarcoma

## Staging

<table>
<thead>
<tr>
<th>Grade and TNM</th>
<th>Description</th>
<th>T1a</th>
<th>T1b</th>
<th>T2a</th>
<th>T2b</th>
<th>Stage</th>
</tr>
</thead>
<tbody>
<tr>
<td>G1</td>
<td>Well differentiated</td>
<td>IIA</td>
<td>IIB</td>
<td>IIB</td>
<td>IIC</td>
<td>III</td>
</tr>
<tr>
<td>G2</td>
<td>Moderately differentiated</td>
<td>IIA</td>
<td>IIB</td>
<td>IIB</td>
<td>IIC</td>
<td>III</td>
</tr>
<tr>
<td>G3</td>
<td>Poorly differentiated</td>
<td>IIA</td>
<td>IIB</td>
<td>IIB</td>
<td>IIC</td>
<td>III</td>
</tr>
<tr>
<td>G4</td>
<td>Undifferentiated</td>
<td>IIA</td>
<td>IIB</td>
<td>IIB</td>
<td>IIC</td>
<td>III</td>
</tr>
<tr>
<td>T1</td>
<td>Tumor ≤5 cm in largest dimension</td>
<td>IIA</td>
<td>IIB</td>
<td>IIB</td>
<td>IIC</td>
<td>III</td>
</tr>
<tr>
<td>T1a</td>
<td>Superficial to deep fascia</td>
<td>IIA</td>
<td>IIB</td>
<td>IIB</td>
<td>IIC</td>
<td>III</td>
</tr>
<tr>
<td>T1b</td>
<td>Deep to deep fascia (includes retroperitoneal, intrathoracic, and most head and neck tumors)</td>
<td>IIA</td>
<td>IIB</td>
<td>IIB</td>
<td>IIC</td>
<td>III</td>
</tr>
<tr>
<td>T2</td>
<td>Tumor &gt;5 cm in largest dimension</td>
<td>IIA</td>
<td>IIB</td>
<td>IIB</td>
<td>IIC</td>
<td>III</td>
</tr>
<tr>
<td>T2a</td>
<td>Superficial to deep fascia</td>
<td>IIA</td>
<td>IIB</td>
<td>IIB</td>
<td>IIC</td>
<td>III</td>
</tr>
<tr>
<td>T2b</td>
<td>Deep to deep fascia (includes retroperitoneal, intrathoracic, and most head and neck tumors)</td>
<td>IIA</td>
<td>IIB</td>
<td>IIB</td>
<td>IIC</td>
<td>III</td>
</tr>
<tr>
<td>N1</td>
<td>Regional nodal metastasis</td>
<td>IIA</td>
<td>IIB</td>
<td>IIB</td>
<td>IIC</td>
<td>III</td>
</tr>
<tr>
<td>M1</td>
<td>Distant metastasis</td>
<td>IIA</td>
<td>IIB</td>
<td>IIB</td>
<td>IIC</td>
<td>III</td>
</tr>
</tbody>
</table>

### 5-Yr Survival

<table>
<thead>
<tr>
<th>Stage</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>86</td>
</tr>
<tr>
<td>II</td>
<td>72</td>
</tr>
<tr>
<td>III</td>
<td>52</td>
</tr>
<tr>
<td>IV</td>
<td>10–20</td>
</tr>
</tbody>
</table>
Surgical Considerations

- Retroperitoneal sarcomas are often unusually large at diagnosis:
  - <5cm = 6%
  - 5-10cm = 25%
  - 10cm = 60%

- Anatomic relations to major vascular structures and vital organs makes resection difficult; significantly impacts ability to obtain negative surgical margins and subsequent local recurrence rates.

Surgical Resection

The standard of care for patients with localized, resectable retroperitoneal sarcomas is surgical resection with gross and microscopically negative margins.

Complete surgical resection frequently requires en-bloc resection of adjacent viscera.

## Frequency of adjacent organ resection

<table>
<thead>
<tr>
<th>Organ</th>
<th>Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kidney</td>
<td>36%</td>
</tr>
<tr>
<td>Colon</td>
<td>22%</td>
</tr>
<tr>
<td>Spleen</td>
<td>10%</td>
</tr>
<tr>
<td>Pancreas</td>
<td>9%</td>
</tr>
<tr>
<td>Small Intestine</td>
<td>6%</td>
</tr>
<tr>
<td>Stomach</td>
<td>6%</td>
</tr>
<tr>
<td>Inferior Vena Cava</td>
<td>3%</td>
</tr>
</tbody>
</table>

Surgical Resection

- Complete surgical resection rates range from 62-86%
- The primary pattern of treatment failure after surgery is local recurrence
- Local recurrence rates range from 32-82%


500 Patients

278 primary disease

231 resectable

185 complete resection  46 incomplete resection

**Local recurrence** rate in the 231 patients who underwent resection was **40% at 5 years**
Adjuvant Radiotherapy

The addition of adjuvant radiation therapy to surgical resection is associated with:

- a reduced risk of local recurrence
- a longer recurrence-free interval
- no impact on overall survival

Pre-operative Radiotherapy

ADVANTAGES:

- The gross tumor volume is readily definable for accurate treatment planning
- The tumor displaces radiosensitive viscera outside the treatment field
- The radiation dose believed to be biologically active is lower in the pre-operative setting
- Tumor is treated in situ prior to potential contamination of the abdominal cavity that occurs with surgery
- No adhesions and tethering of bowel to the tumor bed; decreases toxicity to radiosensitive bowel

Intra-operative Radiotherapy

- Radiation dose can be targeted to the specific regions of the operative field that are believed to be at highest risk for harboring residual microscopic disease\(^1\)
- Dose to tumor bed/ dose to normal tissue ratio is maximized \(^1\)
- IORT (EBRT or brachytherapy) increases in field tumor control but not influence recurrence-free or overall survival rates \(^2\)

Retroperitoneal Sarcoma

**Intra-operative Radiotherapy**
Chemotherapy

Retroperitoneal Soft Tissue Sarcoma. Cancer 2005; 104: 669-75
Management of Local Recurrence

- Patients with first local recurrence and no metastases → perform re-exploration

- Median survival after local recurrence in patients following resection is **60 months**

- Median survival after local recurrence in patients without resection is **20 months**

Retroperitoneal Sarcoma

Local Recurrence

Management of Metastatic Disease

- Distant recurrence after resection is largely grade-dependent; high grade lesions have the highest risk for distant failure; cumulative incidence is 32%
- Increased risk of metastatic disease with positive gross and microscopic margins of resection
- Distant recurrences usually occur in the liver and lung; hematogenous dissemination

Management of Metastatic Disease

- **Pulmonary Metastases:**
  - median survival duration of 6-12 months
  - resection of multiple pulmonary metastases is associated with prolonged relapse-free survival in approximately 25% of patients

- **Hepatic Metastases:**
  - Survival rates are less than those observed for resection of pulmonary metastases
  - Median survival duration was 30 months for patients who underwent resection vs. 11 months for those who did not.
Surveillance

Goal: early detection of local recurrence, hepatic and pulmonary metastases

Physical Exam
CXR
CT Scan of Abdomen and Pelvis

Surveillance Guidelines

**National Comprehensive Cancer Network Guidelines:**

**Low Grade Disease:**
- Physical exam and chest/abdomen/pelvis CT scan every 3-6 months for 2-3 years; then annually

**High Grade Disease:**
- Physical exam and chest/abdomen/pelvis CT scan every 3-4 months for 3 years; then every 6 months for 2 years; then annually

Retroperitoneal Sarcoma

Summary

- Patients with RPS often present with large, locally advanced tumors.

- The most important factor in long-term success in the treatment of primary RP sarcomas is complete surgical resection.

- Wide surgical resection with microscopically negative margins is usually not possible; local recurrence rates are high.

- Radiation therapy is useful for local control; no effect on overall survival; chemotherapy has no proven efficacy.
QUESTIONS ???