Surgical Treatment in Retroperitoneal Sarcoma

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Retroperitoneal Sarcoma

Histologic Types

- **LIPOSARCOMA**
- **leiomyosarcoma**
- **malignant fibrous histiocytoma**
- **fibrosarcoma**
- **malignant peripheral nerve sheath tumor**
- **extraskeletal osteosarcoma**

Pie chart showing the distribution of different histologic types of sarcoma.
Retroperitoneal Sarcoma

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

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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>
</tr>
</thead>
<tbody>
<tr>
<td>G1</td>
<td>Well differentiated</td>
<td>IA</td>
<td>IB</td>
<td>IIA</td>
<td></td>
</tr>
<tr>
<td>G2</td>
<td>Moderately differentiated</td>
<td>IIA</td>
<td></td>
<td>IIC</td>
<td>III</td>
</tr>
<tr>
<td>G3</td>
<td>Poorly differentiated</td>
<td></td>
<td></td>
<td></td>
<td>III</td>
</tr>
<tr>
<td>G4</td>
<td>Undifferentiated</td>
<td>N1</td>
<td>M1</td>
<td></td>
<td>IV</td>
</tr>
</tbody>
</table>

Tumor size:
- T1: ≤5 cm in largest dimension
- T1a: Superficial to deep fascia
- T1b: Deep to deep fascia (includes retroperitoneal, intrathoracic, and most head and neck tumors)
- T2: >5 cm in largest dimension
- T2a: Superficial to deep fascia
- T2b: Deep to deep fascia (includes retroperitoneal, intrathoracic, and most head and neck tumors)

Regional nodal metastasis:
- N1

Distant metastasis:
- M1

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>
Characters

- Large
- Firm retroperitoneal mass
- Attachment
- Adhesion
- Encasement
Work-up

• Colonoscopy
• Endoscopy
• Endo sonography
• Hysteroscopy
• Vaginoscopy
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CT Scan Abdomen and Pelvis
Retroperitoneal Sarcoma

CT Scan Abdomen and Pelvis
Retroperitoneal Sarcoma

CT Scan Abdomen and Pelvis
Retroperitoneal Sarcoma

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.
# Retroperitoneal Sarcoma

## 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>
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Disease specific survival

- Complete resection (n=185)
- Incomplete resection (n=46)
- Unresectable (n=47)

p = 0.0001

Time (months)
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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%
Retroperitoneal STS: analysis of 500 patients treated and followed at a single institution

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**
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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
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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
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**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\(^2\)
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Chemotherapy ????
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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**
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Local Recurrence

- Primary: 80% (185/231)
- 1st recurrence: 57% (35/61)
- 2nd recurrence: 33% (8/24)
- 3rd recurrence: 14% (1/7)
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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
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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.
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**Surveillance**

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

Physical Exam
CXR
CT Scan of Abdomen and Pelvis
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**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