Soft Tissue Sarcoma of the Extremity: The Case for Multidisciplinary Management

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Outline

• Diagnosis
• Treatment approaches
• Outcomes
Case Presentation

• 40 year old man
• 2 week h/o fullness in his thigh
• Avid weightlifter
• Mild tenderness
• Internist – “pulled muscle”
  – Physical Therapy
  – Ibuprofen
Case Presentation

- Represents 2 months later
- Increasing fullness despite not limiting activity
- Axial imaging performed
- Biopsy reveals sarcoma
Background

• Incidence: 10,660 cases/ year
  – 3,820 deaths
• Biologically and anatomically diverse
• Curable at an early stage
• Rarely curable when locally recurrent or metastatic
• Multidisciplinary management

Jemal et al., CA Cancer J Clin, 2011
Distribution (%)

- Extremities: 15
- Retroperitoneum: 9
- Trunk: 57
- Head and Neck: 9

Pie chart showing the distribution of locations.
## Overview

<table>
<thead>
<tr>
<th>Tissue Type</th>
<th>Histology</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fat</td>
<td>Liposarcoma</td>
</tr>
<tr>
<td>Smooth muscle</td>
<td>Leiomyosarcoma</td>
</tr>
<tr>
<td>Skeletal muscle</td>
<td>Rhabdomyosarcoma</td>
</tr>
<tr>
<td>Nerve</td>
<td>Malignant Peripheral Nerve Sheath Tumor (MPNST)</td>
</tr>
<tr>
<td>Blood vessels</td>
<td>Angiosarcoma</td>
</tr>
<tr>
<td>Connective tissue</td>
<td>Fibrosarcoma</td>
</tr>
</tbody>
</table>
Histologic Subtypes

- GIST
- Liposarcoma
- Leiomyosarcoma
- Undifferentiated Pleomorphic Sarcoma
Clinical Presentation

• Mass
  – painless
  – often large (>5 cm)

• Diagnosis
  – MRI or CT
  – Biopsy
    • Tru-cut, FNA, open
CT
Principles of the Biopsy

Biopsy should not compromise resection
NCCN Guidelines: Workup

• Essential:

All patients should be managed by a multidisciplinary team with expertise in sarcoma

NCCN Guidelines, Soft Tissue Sarcomas. v2-2012
Additional Imaging

• Possible sites of metastasis
  – Extremity: lung (70%)
    • Chest x-ray (small, low-grade)
    • CT Chest (large, high-grade)
  – Retroperitoneal/ visceral sarcomas: liver
    • CT Abdomen/ pelvis
Sites of Metastasis

# STS: Staging

<table>
<thead>
<tr>
<th>Stage</th>
<th>T</th>
<th>N</th>
<th>M</th>
<th>Grade</th>
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</thead>
<tbody>
<tr>
<td>IA</td>
<td>1a,1b</td>
<td>0</td>
<td>0</td>
<td>Low (G1)</td>
</tr>
<tr>
<td>IB</td>
<td>2a,2b</td>
<td></td>
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<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>IIA</td>
<td>1a,1b</td>
<td>0</td>
<td>0</td>
<td>Intermediate or High grade (G2 or G3)</td>
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<tr>
<td>IIB</td>
<td>2a,2b</td>
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<td></td>
<td>Intermediate (G2)</td>
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<tr>
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<td></td>
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<td></td>
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</tr>
<tr>
<td>III</td>
<td>2b</td>
<td>0</td>
<td>0</td>
<td>High (G3)</td>
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<tr>
<td></td>
<td>Any</td>
<td>1</td>
<td></td>
<td>Any</td>
</tr>
<tr>
<td>IV</td>
<td>Any T</td>
<td>Any N</td>
<td>1</td>
<td>Any</td>
</tr>
</tbody>
</table>
Surgical Management

• Mainstay of treatment for all STS of the extremity is wide local excision
  – (+/-) Radiation therapy

• Wide en bloc resection
  – 1-2 cm margins in all directions
  – Limiting factors:
    • neurovascular juxtaposition
    • Bony juxtaposition
Marginal Surgical Margin
Radical Surgical Margin
Amputation

- Radical
- Wide
- Marginal
- Intraloesional
Wide Surgical Margin
Wide Surgical Margin
FOLLOW-UP

- Evaluation for rehabilitation (OT, PT)
  - Continue until maximal function is achieved
- H&P and chest imaging (plain radiograph or chest CT)
  - every 3-6 mo for 2-3 y,
  - then every 6 mo for next 2 y,
  - then annually
- Consider obtaining postoperative baseline and periodic imaging of primary site based on estimated risk of locoregional recurrence
  (MRI, CT, consider ultrasound)

If recurrence, See Recurrent Disease (EXTSARC-6)
Final Conclusions

• Sarcomas are a rare, heterogeneous group of diseases.

• Existing challenges need to be overcome.

• Progress is being made.

• Novel therapeutic approaches are needed.