Sarcoma

- Malignancy of Mesenchymal origin
- Incidence: 12,000/yr  median age 50
- Risk factors: Li-Fraumeni, NF, P53
- Presentation: Painless mass, abdominal discomfort
Sarcoma

• Diagnosis
  – Core Biopsy – skin incision in line
  – Excisional biopsy - <2cm
  – Incisional biopsy
  – Orientation → longitudinal

• Why biopsy?
  – Retroperitoneal/abdominal
    • r/o other lesions esp. lymphoma
  – Extremity
    • Ewing’s and Rhabdo need chemo
    • Clear cell and epithelioid need nodes
Sarcoma Staging

<table>
<thead>
<tr>
<th>Prognostic features</th>
<th>good</th>
<th>poor</th>
</tr>
</thead>
<tbody>
<tr>
<td>– Grade</td>
<td>low</td>
<td>high</td>
</tr>
<tr>
<td>– Size</td>
<td>&lt;5cm</td>
<td>&gt;5cm</td>
</tr>
<tr>
<td>– Depth</td>
<td>Superficial</td>
<td>Deep</td>
</tr>
<tr>
<td>– Metastases</td>
<td>absent</td>
<td>present</td>
</tr>
</tbody>
</table>

Stage I-III = # of poor prognostic features
Stage IV  = metastases present
Sarcoma

• Treatment
  – Wide surgical excision to negative margins (why not amputate?)
  – Adjuvant radiation for high grade tumors
  – Chemotherapy (Adriamycin Ifos) for advanced tumors

• Prognosis
  – Stage I >85% 10 yr
  – Stage II 70% 10 yr
  – Stage III 50% 10 yr
  – Stage IV 10% 10 yr
Adjuvant/neoadjuvant RT

• Consider in High grade large sarcomas
• Especially if it will aid resection to R0
• NO survival benefit
• NO benefit in Low grade
• Pre-op increases wound complications but decreases long term sequellae.
Adjuvant/Neoadjuvant Chemo

- Consider in High grade, large and adjacent to neurovascular structures
- Definite benefit in Ewing’s and some benefit in Rhabdo, and angiosarc
- No proven benefit as adjuvant in most other resected tumors R0/R1
- Save it for advanced disease
Sarcoma

- Most common site of metastases – Lungs
- Resection is best therapy for recurrent or metastatic disease
GIST

- GI Stromal Tumors are c-Kit positive tumors and respond to Gleevec (STI-571)
- Tyrosine Kinase constitutive ON mutation in exons 11, 9, 13 or 17 of the SCF receptor (CD 117) in Interstitial Cells of Cajal or PDGFα receptor
- Management includes Gleevec, Sutent or Regorafenib