Retroperitoneal Sarcoma: Challenges for the Surgeon

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SON Update October 22, 2011
Disclosure

- Nothing to disclose (there is no money in sarcoma work!)
Case Presentation

- 57 yo man felt mass in LUQ
- Imaging 23 cm adrenal carcinoma? Diff sarcoma
- Referred to community urologist
- Referred on to urologic oncologist because of potential technical challenge of surgery
- Pheo w/u (negative)
- CT guide biopsy arranged (hallway consultation)
CT images
CT Images
Work-up

- Core biopsy = liposarcoma
- Referred for surgical oncology opinion
- CT chest - no metastases
- Discussed at sarcoma conference
- High grade features on imaging tho’ path did not show high grade
- Technically feasible for preop XRT
- Planned surgical resection including left nephrectomy, distal pancreatectomy and splenectomy post XRT
Final Pathology

Surgical specimen including left nephrectomy, distal pancreatectomy and splenectomy

47 x 25 x 15 cm high grade dediff liposarcoma

Margins clear! (both pathologist and surgeon were surprised)
Retroperitoneal Sarcoma

- 1-2% of all solid malignancies
- RPS are still uncommon constituting about 10% of all soft tissue sarcoma
- Peak 5th decade; Equal M:F
- 1/3 of RP masses are sarcomas

Windham, RPS. Cancer Control 2005 12(1):36-43

- Majority of retroperitoneal soft tissue tumours are malignant (even if they don’t look it)
- Best management is a function of the diagnosis
Differential Diagnosis

- Sarcoma
- Neural -schwannoma, ganglioneuroma, paraganglioma
- Lymphoma
- Adrenal (adenoma, carcinoma, myelolipoma)
- Renal (carcinoma, angiomyolipoma)
- Metastatic nodes- testicular, nongerm cell
Histology of Sarcomas

- Most common are liposarcomas
  - May not have substantial fatty component on imaging but often there is some asymmetry in the amount or character of the retroperitoneal fat on the involved side

- Leiomyosarcoma
  - Typically vascular origin- IVC, renal vessels

- MFH
Transition over Time

2009

2011
# Tumour Staging

## Table 1

<table>
<thead>
<tr>
<th>Classifications</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Histological grade (G)</strong></td>
</tr>
<tr>
<td>G1</td>
</tr>
<tr>
<td>G2</td>
</tr>
<tr>
<td>G3</td>
</tr>
<tr>
<td><strong>Primary site (T)</strong></td>
</tr>
<tr>
<td>T1</td>
</tr>
<tr>
<td>T1a</td>
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<tr>
<td>T1b</td>
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<tr>
<td>T2</td>
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<tr>
<td>T2a</td>
</tr>
<tr>
<td>T2b</td>
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</tbody>
</table>

N.B. Retroperitoneal and pelvic sarcomas are classified as deep tumors.
# Staging Sarcoma

<table>
<thead>
<tr>
<th>Stage</th>
<th>Classification</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>IA</td>
<td>G1, T1, N0, M0</td>
<td>Grade 1 tumor, &lt;5 cm in diameter no regional lymph nodes and/or distant metastases</td>
</tr>
<tr>
<td>IB</td>
<td>G1, T2, N0, M0</td>
<td>Grade 1 tumor, 5 cm or more in diameter, no nodes and/or metastases</td>
</tr>
<tr>
<td>IIA</td>
<td>G2, T1, N0, M0</td>
<td>Grade 2 tumor, &lt;5 cm in diameter, no nodes and/or metastases</td>
</tr>
<tr>
<td>IIB</td>
<td>G2, T2, N0, M0</td>
<td>Grade 2 tumor, 5 cm or more in diameter, no nodes and/or metastases</td>
</tr>
<tr>
<td>IIIA</td>
<td>G3, T1, N0, M0</td>
<td>Grade 3 tumor, &lt;5 cm in diameter, no nodes and/or metastases</td>
</tr>
<tr>
<td>IIIB</td>
<td>G3, T2, N0, M0</td>
<td>Grade 3 tumor, 5 cm or more in diameter, no nodes and/or metastases</td>
</tr>
<tr>
<td>IIIC</td>
<td>G1–3, T1, 2, N1, M0</td>
<td>Tumor of any grade and/or size, with regional involved nodes, but no metastases</td>
</tr>
<tr>
<td>IIV</td>
<td>G1–3, T3, N0,</td>
<td>Tumor of any grade invading bone vessels/nerves,</td>
</tr>
</tbody>
</table>
Nearly all retroperitoneal sarcomas are >5 cm and are deep by definition.

Nearly all are therefore Stage IIB (large, low-grade, and deep) or stage III (large, high-grade and deep).

Distinction between these two made only on the basis of histologic grade.

Overall about 1/3 are low grade.
Presentation

- Usually huge unless found incidentally
- Symptoms: vague discomfort or protrusion or GI due to mass effect
- Median 4 months of symptoms before diagnosis
- May present with neurologic/MSK symptoms in the lower extremity
  - Cancer 2005, 104, 669-75
- Occasionally unexpected finding at laparotomy for other disease
- Sometimes intraoperative consult from another service (gyne/urology)
Management of the Retroperitoneal Mass

- Core biopsy should be used for tissue diagnosis after all functional investigations (if needed) are done
- Biopsy via the retroperitoneal approach
- It is usually not possible to excise the biopsy tract
- Pathology review is often necessary
- Early referral for consultation is extremely helpful
Staging

- Chest CT to r/o mets
- CT abdomen /pelvis is usually adequate
- Occasionally MRI if there is a question about vascular involvement
- PET not generally helpful
- Differential renal scan if concern re adequacy of residual renal function
Determinants of Prognosis

- The major tumour factors that affect survival are the tumor grade and resectability.
- Patients who have had a successful complete resection and also have low-grade tumors have the best survival rates.
Surgeon Beware!

- A large mass is not an indication for an emergency operation, no matter how anxious the patient (family, referring MD, radiologist, neighbours...) may be.
- Ask yourself, are there any contraindications to getting a tissue diagnosis first? What are the cons?
- A thoughtful approach is more likely to result in the best possible outcome.
Surgical Management

- 75% of complete resections involve resection of at least one adjacent organ (usually kidney, colon or adrenal)
- Need to be prepared (other specialists?)
- Even if invasion is not apparent, resection is required of contiguous organs to achieve a clear margin
- Malignant pseudocapsule gives false impression of a margin
- Resection at the level of pseudocapsule is assoc’ed with up to 80% LR
Sarcoma Pseudocapsule
Successful Surgery

- Complete resection rate between 65–99%
- Highest in centres with high volume
- Complete resection has been shown to improve survival
- Incomplete resection is ineffective with no benefit except in very low grade tumours
- More likely to achieve complete resection at first surgery
Unexpected RP Masses

- Do not perform an incisional biopsy as this contaminates the peritoneal cavity.
- Core biopsy may be acceptable if hemostasis can be assured and contamination of the peritoneal cavity avoided.
- Tissues should not be mobilized to expose the tumour for biopsy purposes.
Adjuvant Treatment in RPS

- Preoperative multidisciplinary conference should be the goal for all RPS patients.
- In BC, preoperative radiation for high-grade tumours or low-grade tumours where wide excision is not feasible or for locally recurrent tumours.
- Radiation has not been studied in RCT, so practice varies in different centres.
Evidence for XRT

- One randomized trial using IORT showed improved local control.
- Several retrospective and prospective studies suggest improved local control.
- Some evidence that XRT delays, but does not prevent, local recurrence.
- Decreased LRR and time to LR with no change is OS.

- Stoeke, Cancer 2001:(92), 359.
So, why not just give XRT postoperatively?

- Radiation can’t be given after the mass is out due to toxicity to fixed bowel in the operative field (due to adhesions)
- The postop radiation field is much larger with dose limiting toxicity to adjacent organs
What about chemotherapy?

- In high-grade disease, administration of adriamycin and ifosfamide may yield partial responses in up to 50% of patients with increased overall survival.
- Complete responses are seen in less than 10% of patients.

Chemotherapy and RPS in BC

- Reserved primarily for metastatic setting
- Selected use in very fit patients
  ‘neoadjuvantly’ as sequential therapy followed by XRT for large, high grade tumours
Metastatectomy in Sarcoma

- Most common site of distant metastases is lung
- 25% prolonged relapse free survival even with resection of multiple pulmonary metastases
Despite ‘complete’ resections, 5- and 10-year survival rates are only 51% (11-63%) and 36% (10-50%) respectively.

Better with increased magnitude of resection (43% at 10yrs).

Most frequent recurrence is in the surgical bed.

Most recurrences occur within 2 years but can be very delayed with low grade disease.
Outcomes after Local Recurrence

- Local recurrences may be suitable for re-excision.

- Median survival following resection of local recurrence is 60 months vs 20 months without surgery
  - Windham, Cancer Control 2005, 12(1) 36-43

- Re-operative surgery is generally palliative and should be offered for symptom control
Outcomes after Local Recurrence

- Cures following re-excision of lesions that were not treated with primary wide local excision have been reported.
- Prolonged palliation can be achieved for low grade tumours.
- Generally no value in high grade tumours with equivalent median survival to non-operative patients.
Follow-up Recommendations

- Frequency of follow-up dictated by the completeness of resection and tumour stage.

- CT or MRI every 3–4 months for 2 years, then every 4–6 months for 3–5 years, and every 12 months thereafter

- Follow up for greater than 5 years is recommended as marked delay in appearance of recurrent disease can occur
So how are we doing in BC?

- Review 2000-2009 BCCA Registry and CAIS database
- Coding for RP tumour identified 228 patients diagnosed with retroperitoneal tumours
- 82 of these were retroperitoneal sarcomas for which outcome data was available
BC Outcome Data

- Of the 82 patients for whom outcome data was available:
- 5 year OS 56.6% (comparable to literature)
- 41 referred prior to resection and 41 after
- 40/41 vs 27/41 had complete resections
- 34/41 vs 18/41 were alive on follow-up
- p<0.05 for both
Overall Survival based on Referral Pattern

5-year OS: 70.7% (95% CI 45.3, 85.9) vs. 46.7% (95% CI 29.6, 62.1)
Recurrence-Free Survival based on Referral Pattern

5-year RFS: 43.2% (95% CI 18.8, 65.6) vs. 20.8% (95% CI 6.6, 40.4)

(n=61)
Referring Patients with Undiagnosed Retroperitoneal Masses

- Refer to Sarcoma Clinic at BCCA (where they will be triaged to the General Surgical Oncology Group)
- Refer directly to the General Surgical Oncology Group
- We are happy to review images and advise
Bartlett E and Yoon SS, Current Treatment for the Local Control of Retroperitoneal Sarcomas, JACS, 2011 September 213(3): 436-445 (collective review)

Included in your package!
Questions ??