51 year old female with a retroperitoneal mass

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Case Presentation

• Long standing history of right upper quadrant and back discomfort
• 2 months prior to presentation, noted worsening pain, firmness on right below rib cage
• No jaundice, weight loss, cough
• Presented to PCP for evaluation
History and Physical

**PMH:**
- Uterine fibroids, causing obstruction and DVT 2005.
- HTN
- HPLD
- Nephrolithiasis

**PSuH:**
- Cystoscopy/ureteroscopy
- Uterine artery embolization

**Fam:** Maternal aunt breast Ca age 77, maternal aunt bone Ca age 73.

**Soc:** Remote 1 year smoking history, quit 30y ago. Occasional alcohol.

**Meds:** Lisinopril, statin

**All:** Codeine, erythromycin
History and Physical

• PERFORMANCE STATUS: ECOG PS--1 (secondary to pain).
• GENERAL: Thin, well-appearing, emotional, but in no apparent distress.
• VITAL SIGNS: Temp 36.8, heart rate 95, respiratory rate 16, blood pressure 121/85.
• SKIN: Warm and dry. There are no rashes or open lesions.
• HEENT: Oropharynx is clear. Sclerae without icterus.
• LUNGS: CTAB. No wheezes, rales, or rhonchi.
• CARDIAC: Regular rate. No appreciated murmur.
• ABDOMEN: In the right upper quadrant, there is palpable hard mass. It is approximately 15 x 10 cm.
• EXTREMITIES: Full range of motion throughout. No edema.
• NEUROLOGIC: Alert and oriented times 3. Cranial nerves II-XII grossly intact.
Imaging

• Ultrasound:
  – 19cm solid mass

• CT abdomen/pelvis
  – Large mass within the right retroperitoneum with dramatic mass effect displacing the right lobe of the liver, left kidney, and gallbladder, 11.2 x 19.7 x 27 cm. Encapsulated. No obvious vascular invasion.
CT
CT
CT
CT
Pathology

• Ultrasound-guided core biopsy
• Consistent with leiomyosarcoma
• Intermediate grade (FNCLCC grade 2/3).
• Immunopathology:
  – C-KIT negative.
  – Desmin positive, 3+.
  – DOG-1 negative.
  – MDM2 negative.
  – S100 negative.
Retroperitoneal Sarcoma

- Historically, surgery is the mainstay of treatment, and gross total resection feasible in 50-67% of patients.
- The probability of local control and death at 5 years are both 50%. Local failure is the most common pattern of disease recurrence.
- Twenty to 30% of patients will develop distant metastases at 5 years.
- Post-operative radiotherapy has been shown to reduce the risk of recurrence, but has had no impact on survival.
- There is currently a trial of pre-operative radiotherapy for RP sarcoma, and Baldini and colleagues have recently published consensus guidelines on radiotherapy planning.
Treatment Considerations

• 3DCRT or IMRT recommended based on ability to meet dose constraints
• Can dose paint to areas at low and high risk for positive margins following resection
  – Consider along posterior abdominal wall, pre-vertebral space, and/or major vessels
  – Not routinely recommended outside of trial or high-volume center
IMRT dose painting high risk areas

• Prospective single-center one-armed phase I/II study interim analysis published in 2014\(^1\)
• Attempted neoadjuvant IMRT with integrated boost to 50-56Gy followed by surgery and IORT to \(~12\) Gy
• Local control \(~70\)% at 3 years, comparable to prior retrospective studies of RP sarcoma
• Study found sarcoma RadOnCs contoured the GTV, tumor CTV, and most OARs with a high level of agreement\(^2\), but high risk area CTV contours were quite variable\(^3\).

\(^1\) Roeder et al (2014)  
\(^2\) Baldini et al (2015) (ref 4)  
\(^3\) Baldini et al (2015) (ref 3)
Discussion with surgeon

• Multidisciplinary discussion can establish at risk areas and other organs of concern

• Potential surgical considerations
  – Nephrectomy may be necessary
  – Partial liver resection may be necessary
  – Areas concerning for positive margins
CT Simulation Recommendations from Consensus Guidelines

• Oral and IV contrast can be used as necessary to delineate targets/OARs
• Additional studies such as MR and PET can be fused to treatment planning CT
• 4D assessment (i.e. 4DCT) strongly recommended for tumors above the iliac crest
Targets and OARs with motion

• Internal target volume (ITV)
  – Internal GTV (IGTV)
  – Internal CTV (ICTV)

• Planning target volume (PTV)
  – Without ITV: organ motion, setup uncertainties
  – With ITV: setup uncertainties

• Planning organ at risk volume (PRV)
  – Can make a volume of OAR with motion data
Target Definition from Consensus Guidelines

- $CTV = GTV + 1.5\text{cm}$
- $ITV$ can be used as $CTV$
- $PTV$ with $CBCT = CTV + 5\text{mm}$
- $PTV$ without $CBCT = CTV + 9-12\text{mm}$

<table>
<thead>
<tr>
<th>EDIT CTV</th>
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<tbody>
<tr>
<td>Bone</td>
<td>0\text{mm}</td>
</tr>
<tr>
<td>Bowel and Air Cavity</td>
<td>5\text{mm}</td>
</tr>
<tr>
<td>Renal and Hepatic Interfaces</td>
<td>2\text{mm}</td>
</tr>
<tr>
<td>Skin Surface</td>
<td>3-5\text{mm}</td>
</tr>
<tr>
<td>If extending inferiorly through inguinal canal</td>
<td>Add 3\text{cm} distally</td>
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</tbody>
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Treatment Details

• The patient was discussed at multidisciplinary tumor board with the surgeon
• There was concern about positive margins along the spine, vessels (i.e. aorta), and posterior retroperitoneum
• Surgeon expressed plans to perform left nephrectomy and adrenalectomy up front
Treatment Details

• Motion of tumor determined to be minimal on 4DCT examination during simulation, so motion-related volumes not used

• Low-dose and high-dose targets prescribed to 50Gy and 60Gy respectively
  – High-dose target discussed with surgeon after planning prior to starting radiotherapy

• Treatment course delivered in 25 fractions using IMRT technique using integrated boost to high dose volume
Target Contours and Isodose Lines

GTV
CTV high dose
PTV low dose
<table>
<thead>
<tr>
<th>Organ</th>
<th>Constraint</th>
</tr>
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<tbody>
<tr>
<td>Liver</td>
<td>Mean Dose &lt; 26 Gy</td>
</tr>
<tr>
<td>Stomach and Duodenum</td>
<td>V45&lt;100%, V50&lt;50%, Max 56 Gy</td>
</tr>
<tr>
<td>Kidney: if one will be resected</td>
<td>V18 &lt; 15% remaining kidney</td>
</tr>
<tr>
<td>Kidney: if both will remain</td>
<td>Mean dose &lt; 15 Gy, V18 &lt; 50%</td>
</tr>
<tr>
<td>Spinal Cord</td>
<td>Max Dose 50 Gy</td>
</tr>
<tr>
<td>Small &amp; Large Bowel (Bowel Bag)</td>
<td>V45 ≤ 195 cc</td>
</tr>
<tr>
<td>Rectum</td>
<td>V50 &lt; 50%</td>
</tr>
<tr>
<td>Testicles</td>
<td>V3 &lt; 50%, Max Dose &lt; 18 Gy</td>
</tr>
<tr>
<td>Ovary</td>
<td>Max Dose ≤ 3 Gy</td>
</tr>
<tr>
<td>Femoral Head</td>
<td>Max Dose &lt; 50 Gy, V40 &lt; 64%</td>
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</tbody>
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Surgery and Follow up

• Patient had surgical resection of the mass approximately 1 month following completion of radiotherapy

• A 30 cm tumor in the right retroperitoneum invading the right diaphragm was noted intraoperatively
  – Note right kidney and right adrenal were resected
  – 20% necrosis and negative margins were noted
  – Pathology again noted to be grade II leiomyosarcoma

• Patient was recently seen in follow up approximately 6 months following diagnosis with no evidence of local or distant recurrence
References


