

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

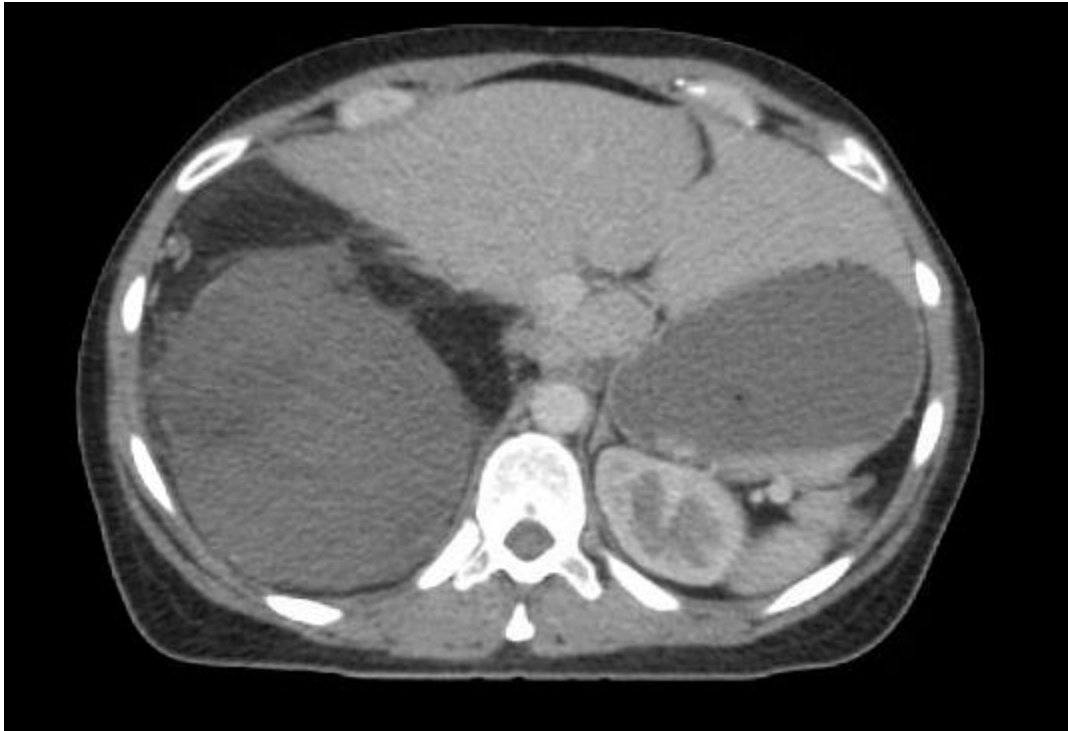
History and Physical

- 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.

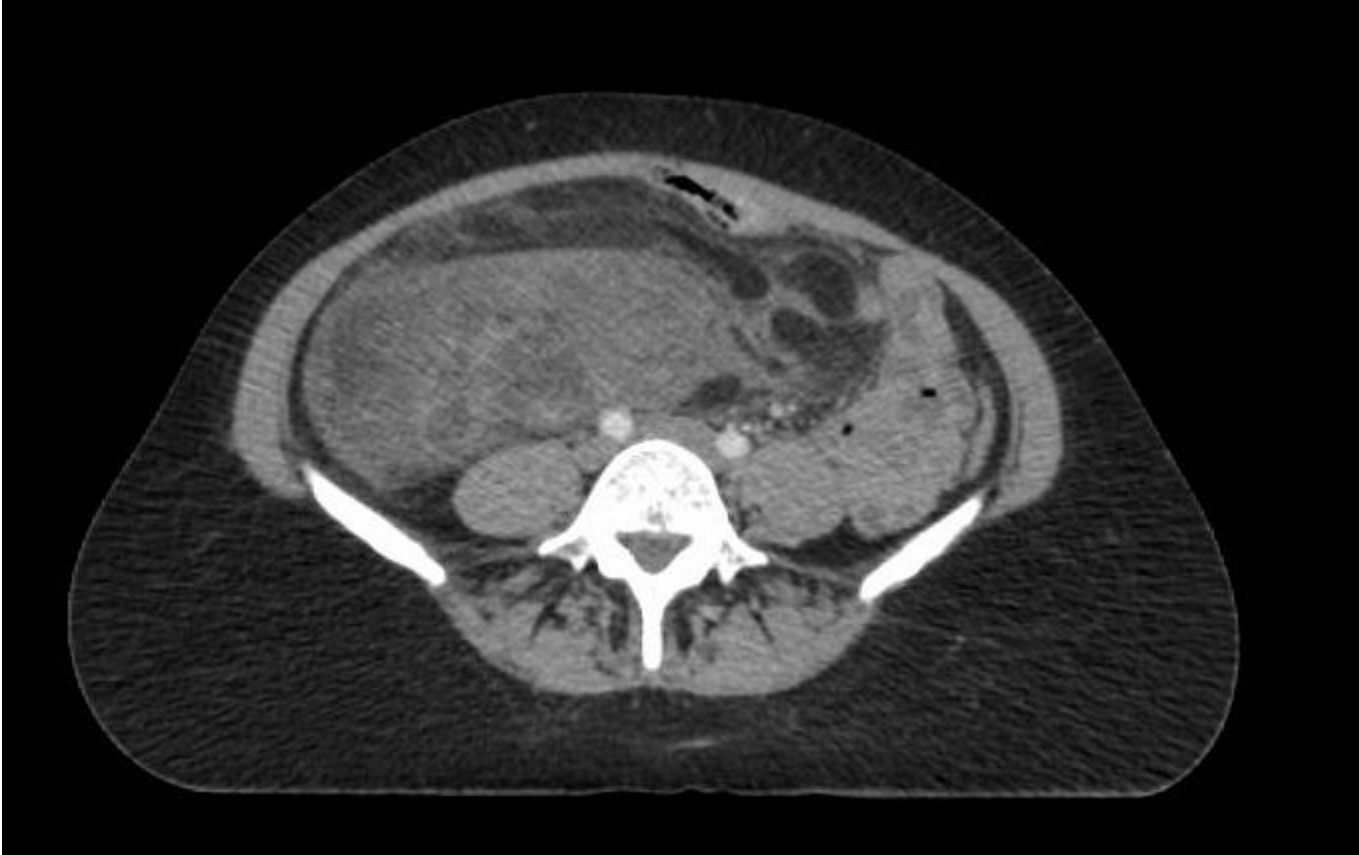
CT



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 2014¹
- 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 agreement², but high risk area CTV contours were quite variable³.

1. Roeder et al (2014)
2. Baldini et al (2015) (ref 4)
3. Baldini et al (2015) (ref 3)

Discussion with surgeon

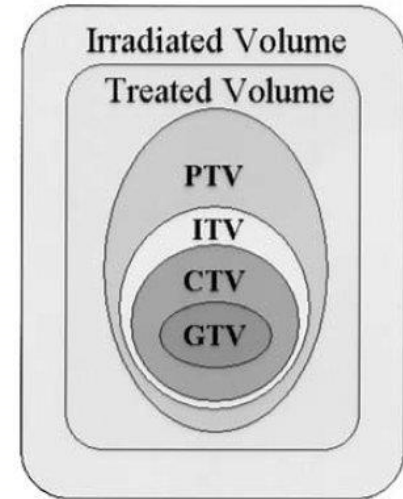
- Multidisciplinary discussion can establish at risk areas and other organs of concern
- Potential surgical considerations
 - Nephrectomy may be necessary
 - Partial liver resection may be necessary
 - Areas concerning for positive margins

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

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

EDIT CTV	
Bone	0mm
Bowel and Air Cavity	5mm
Renal and Hepatic Interfaces	2mm
Skin Surface	3-5mm
If extending inferiorly through inguinal canal	Add 3cm distally

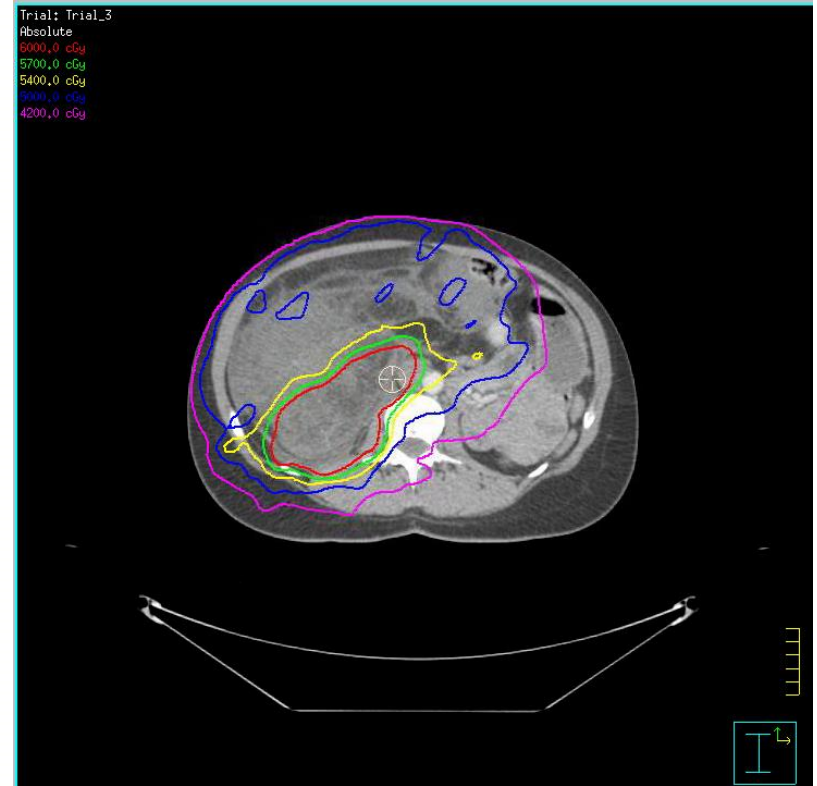
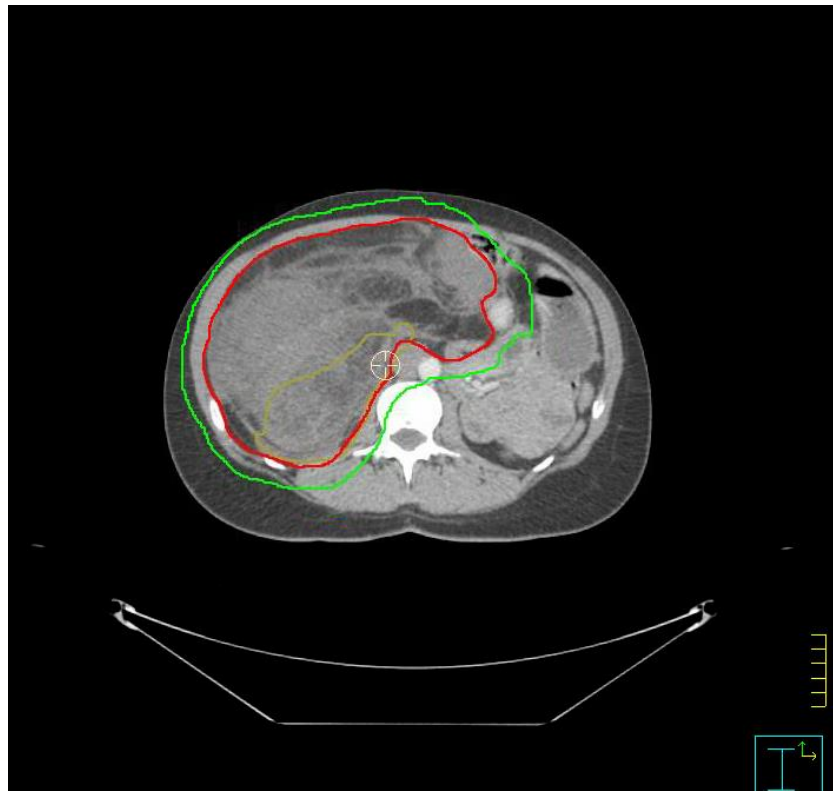
Treatment Details

- 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

Organs at Risk

Organ	Constraint
Liver	Mean Dose < 26 Gy
Stomach and Duodenum	V45 ≤ 100%, V50 < 50%, Max 56 Gy
Kidney: if one will be resected	V18 < 15% remaining kidney
Kidney: if both will remain	Mean dose ≤ 15 Gy, V18 ≤ 50%
Spinal Cord	Max Dose 50 Gy
Small & Large Bowel (Bowel Bag)	V45 ≤ 195 cc
Rectum	V50 < 50%
Testicles	V3 < 50%, Max Dose < 18 Gy
Ovary	Max Dose ≤ 3 Gy
Femoral Head	Max Dose < 50 Gy, V40 < 64%

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

1. Baldini, EH et al. (2015) Treatment guidelines for preoperative radiation therapy for retroperitoneal sarcoma: preliminary consensus of an international expert panel. IJROBP 92 (3), pp. 602-12.
2. Roeder, F et al. (2014) Clinical phase I/II trial to investigate preoperative dose-escalated intensity-modulated radiation therapy (IMRT) and intraoperative radiation therapy (IORT) in patients with retroperitoneal soft tissue sarcoma: interim analysis. BMC Cancer Aug 27 (14), pp. 617.
3. Baldini, EH et al. (2015) Retroperitoneal sarcoma (RPS) high risk gross tumor volume boost (HR GTV Boost) contour delineation agreement among NRG sarcoma radiation and surgical oncologists. Ann. Surg. Onc. 22, 9, pp. 2846-2852.
4. Baldini, EH et al. (2015) Retroperitoneal sarcoma target volume and organ at risk contour delineation agreement among NRG sarcoma radiation oncologists. IJROBP 92 (5), pp. 1053-1059.
5. Mendenhall, WM et al. (2005) Retroperitoneal soft tissue sarcoma. Cancer 104 (4), pp. 669-675.