Retroperitoneal Sarcoma: Challenges for the Surgeon

Rona Cheifetz

SON Update October 22, 2011

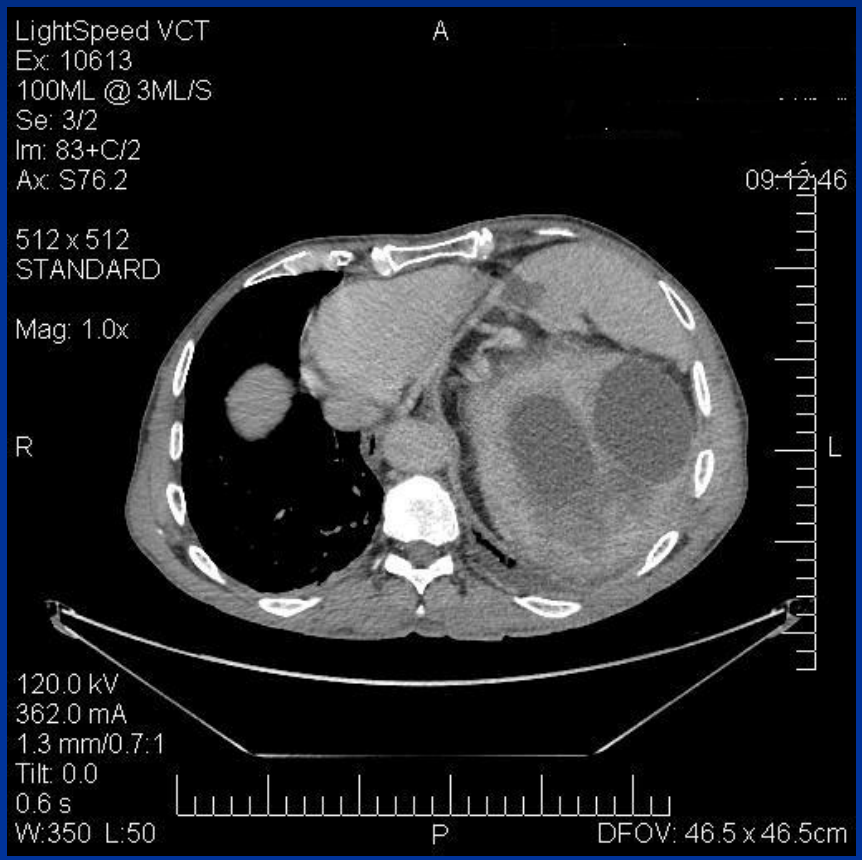
Disclosure

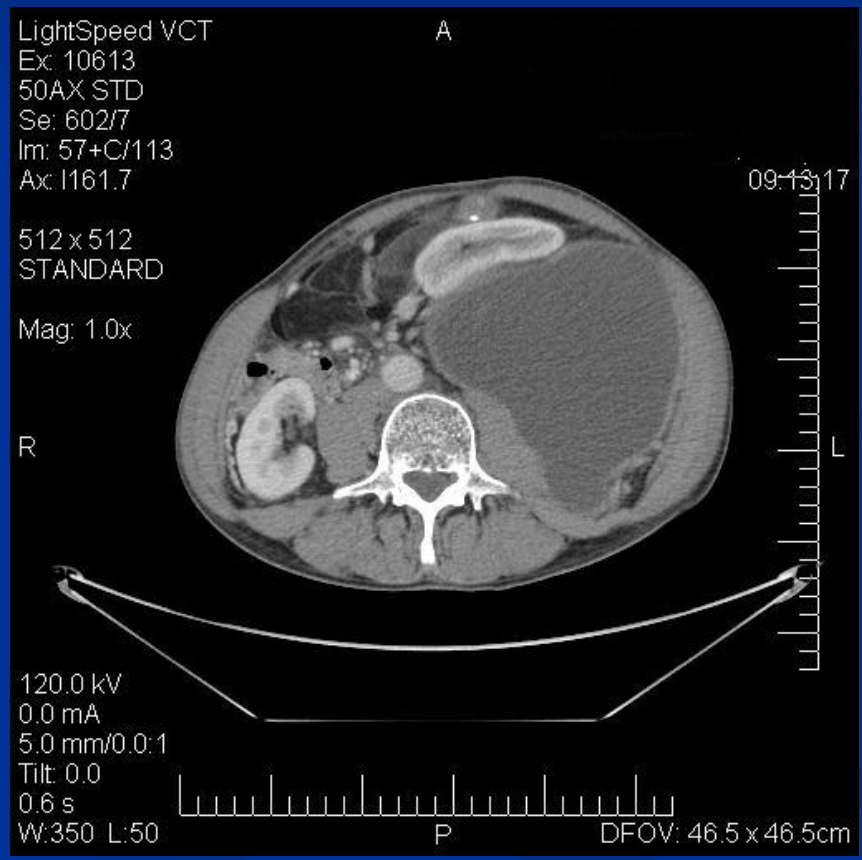
- Nothing to disclose (there is no money in sarcoma work!)

Case Presentation

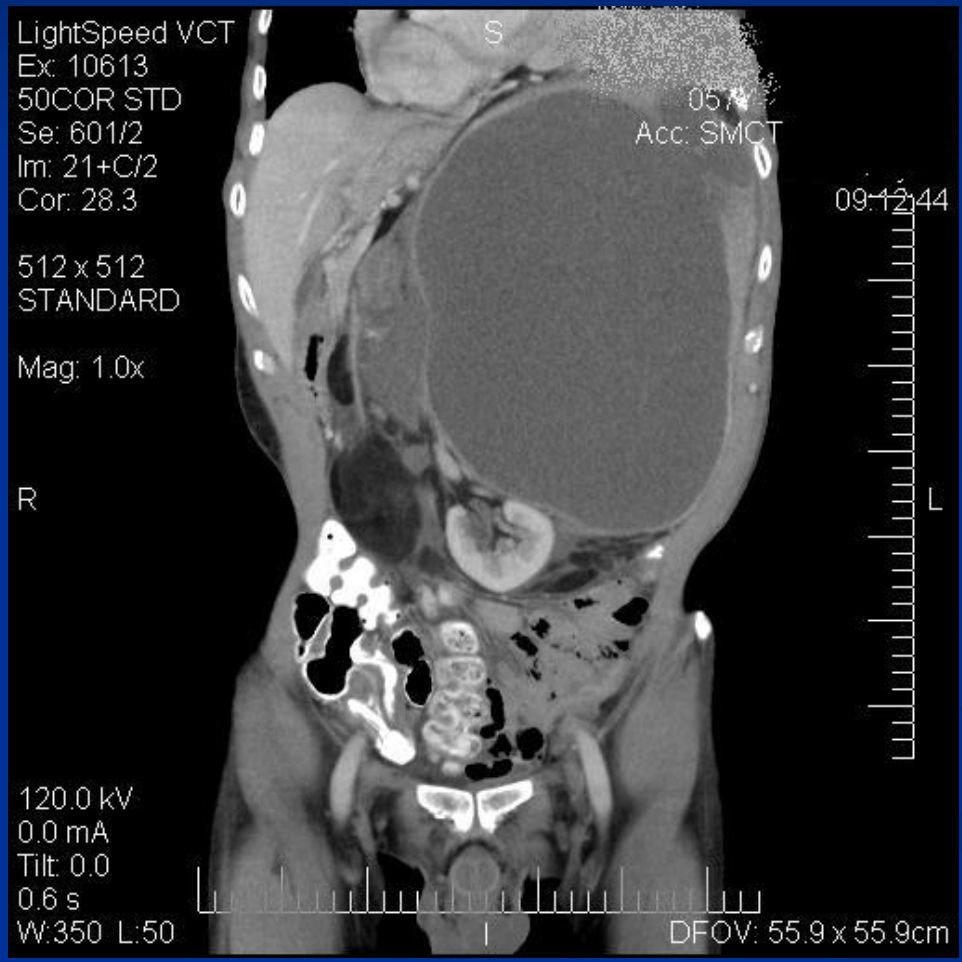
- 57 yo man felt mass in LUQ
- Imaging 23 cm adrenal carcinoma? Diff sarcoma
- Referred to community urologist
- Referred on to urologic oncologist because of potential technical challenge of surgery
- Pheo w/u (negative)
- CT guide biopsy arranged (hallway consultation)

CT images


LightSpeed VCT
Ex: 10613
100ML @ 3ML/S
Se: 3/2
Im: 83+C/2
Ax: S76.2
512 x 512
STANDARD
Mag: 1.0x
A
09:12:46
R
L
120.0 kV
362.0 mA
1.3 mm/0.7:1
Tilt: 0.0
0.6 s
W:350 L:50
P
DFOV: 46.5 x 46.5cm


LightSpeed VCT
Ex: 10613
50AX STD
Se: 602/7
Im: 57+C/113
Ax: I161.7
512 x 512
STANDARD
Mag: 1.0x
A
09:13:17
R
L
120.0 kV
0.0 mA
5.0 mm/0.0:1
Tilt: 0.0
0.6 s
W:350 L:50
P
DFOV: 46.5 x 46.5cm

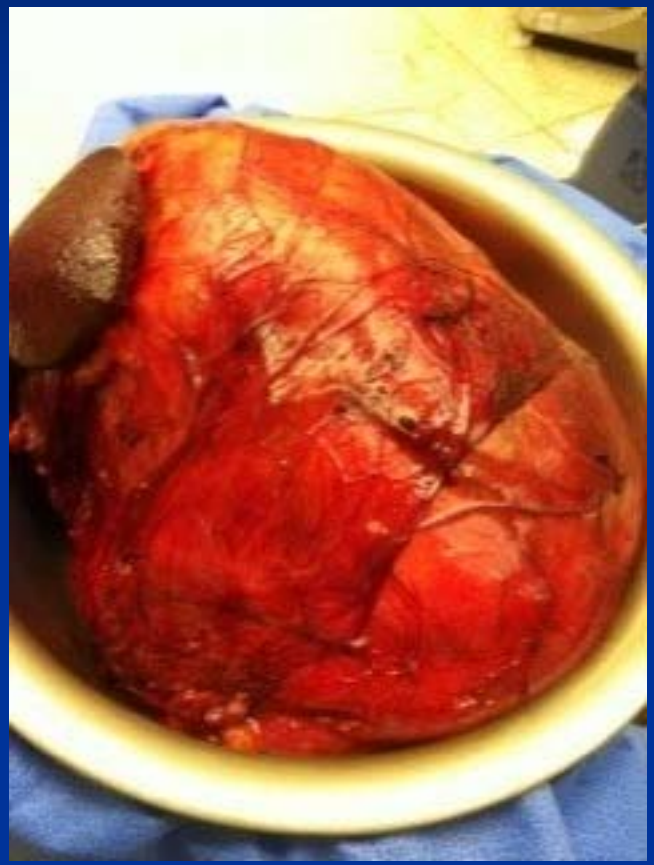
CT Images


LightSpeed VCT
Ex: 10613
50COR STD
Se: 601/2
Im: 21+C/2
Cor: 28.3
512 x 512
STANDARD
Mag: 1.0x
R
S
Acc: SMCT
09:12:44
L
120.0 kV
0.0 mA
Tilt: 0.0
0.6 s
W:350 L:50
I
DFOV: 55.9 x 55.9cm

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 25x 15cm high grade dediff liposarc

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

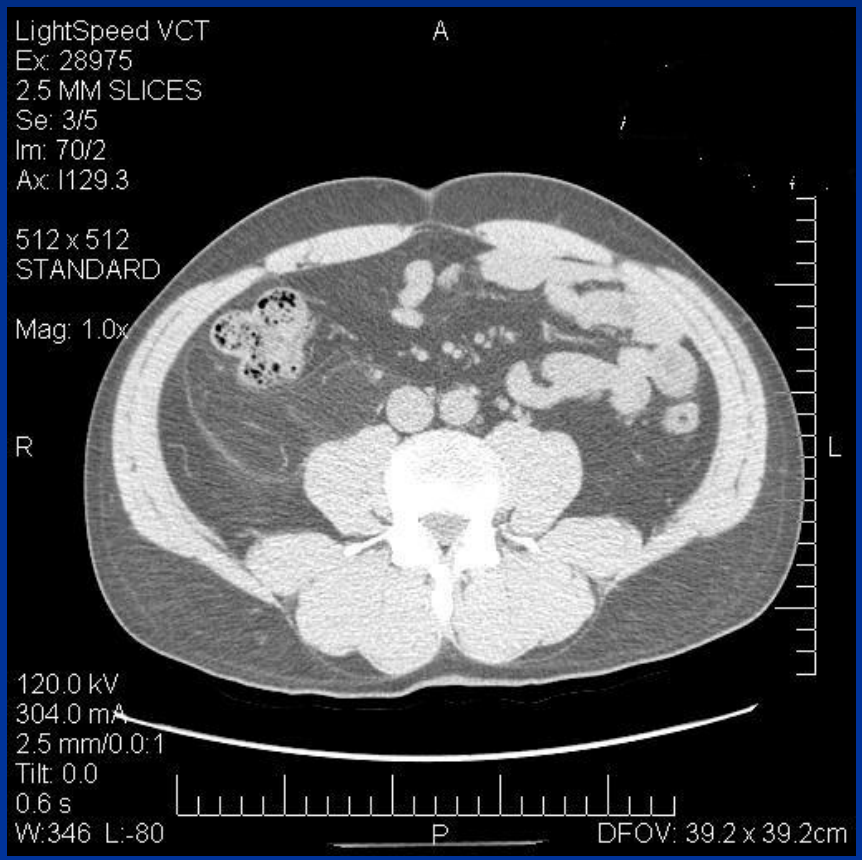
Differential Diagnosis

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

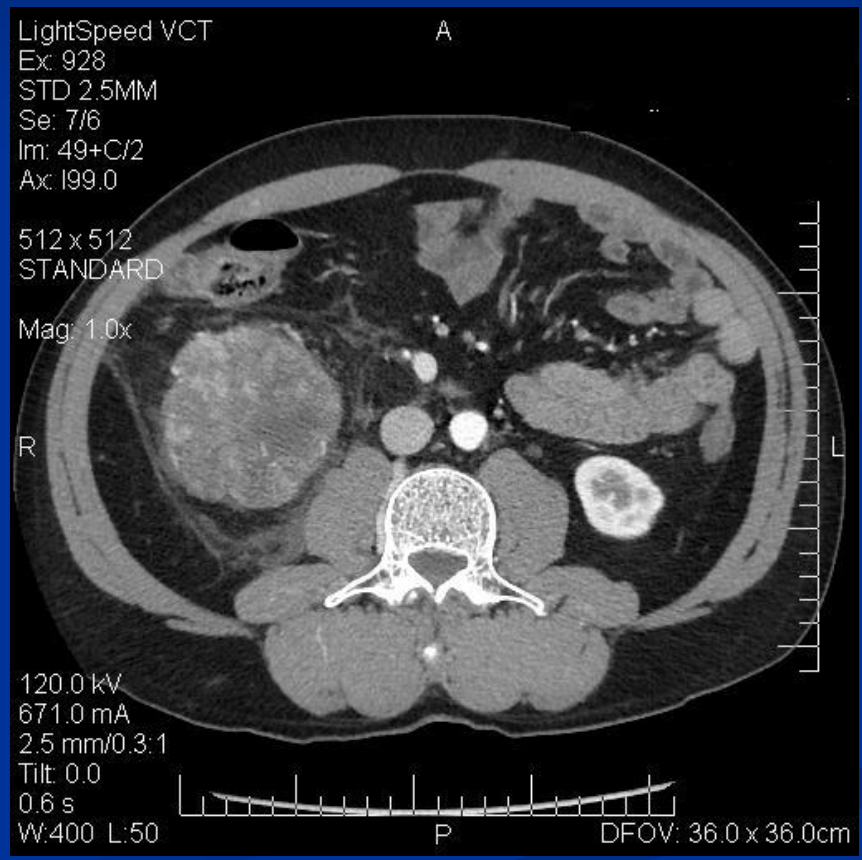
Histology of Sarcomas

- 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
- Leiomyosarc
  - Typically vascular origin- IVC, renal vessels
- MFH

Transition over Time



2009



2011

Tumour Staging

Table 1
Classifications

Histological grade (G)	
G1	Well differentiated
G2	Moderately well differentiated
G3	Poorly or very poorly differentiated
Primary site (T)	
T1	Tumor less than 5 cm in diameter
T1a	Superficial tumor
T1b	Deep tumor
T2	Tumor 5 cm or more in diameter
T2a	Superficial tumor
T2b	Deep tumor

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

Staging Sarcoma

Table 2

American Joint Committee staging of soft tissue sarcomas

Stage	Classification	Description
IA	GI, T1, N0, M0	Grade 1 tumor, <5 cm in diameter no regional lymph nodes and/or distant metastases
IB	GI, T2, N0, M0	Grade 1 tumor, 5 cm or more in diameter, no nodes and/or metastases
IIA	G2, T1, N0, M0	Grade 2 tumor, <5 cm in diameter, no nodes and/or metastases
IIB	G2, T2, N0, M0	Grade 2 tumor, 5 cm or more in diameter, no nodes and/or metastases
IIIA	G3, T1, N0, M0	Grade 3 tumor, <5 cm in diameter, no nodes and/or metastases
IIIB	G3, T2, N0, M0	Grade 3 tumor, 5 cm or more in diameter, no nodes and/or metastases
IIIC	G1–3, T1, 2, N1, M0	Tumor of any grade and/or size, with regional involved nodes, but no metastases
I	G1–3, T3, N0,	Tumor of any grade invading bone vessels/nerves,

Usual Stage at Diagnosis

- 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.

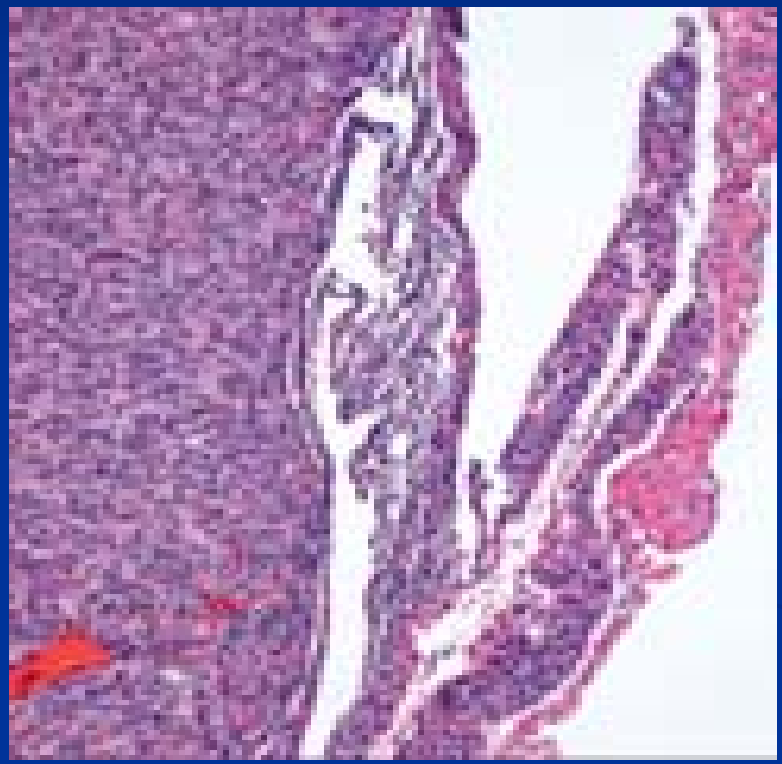
Surgeon Beware!

- 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'd 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.
  - **Raut CP, Pisters PW. *J Surg Oncol.* 2006;94(1):81–7.**

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

Prognosis

- 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

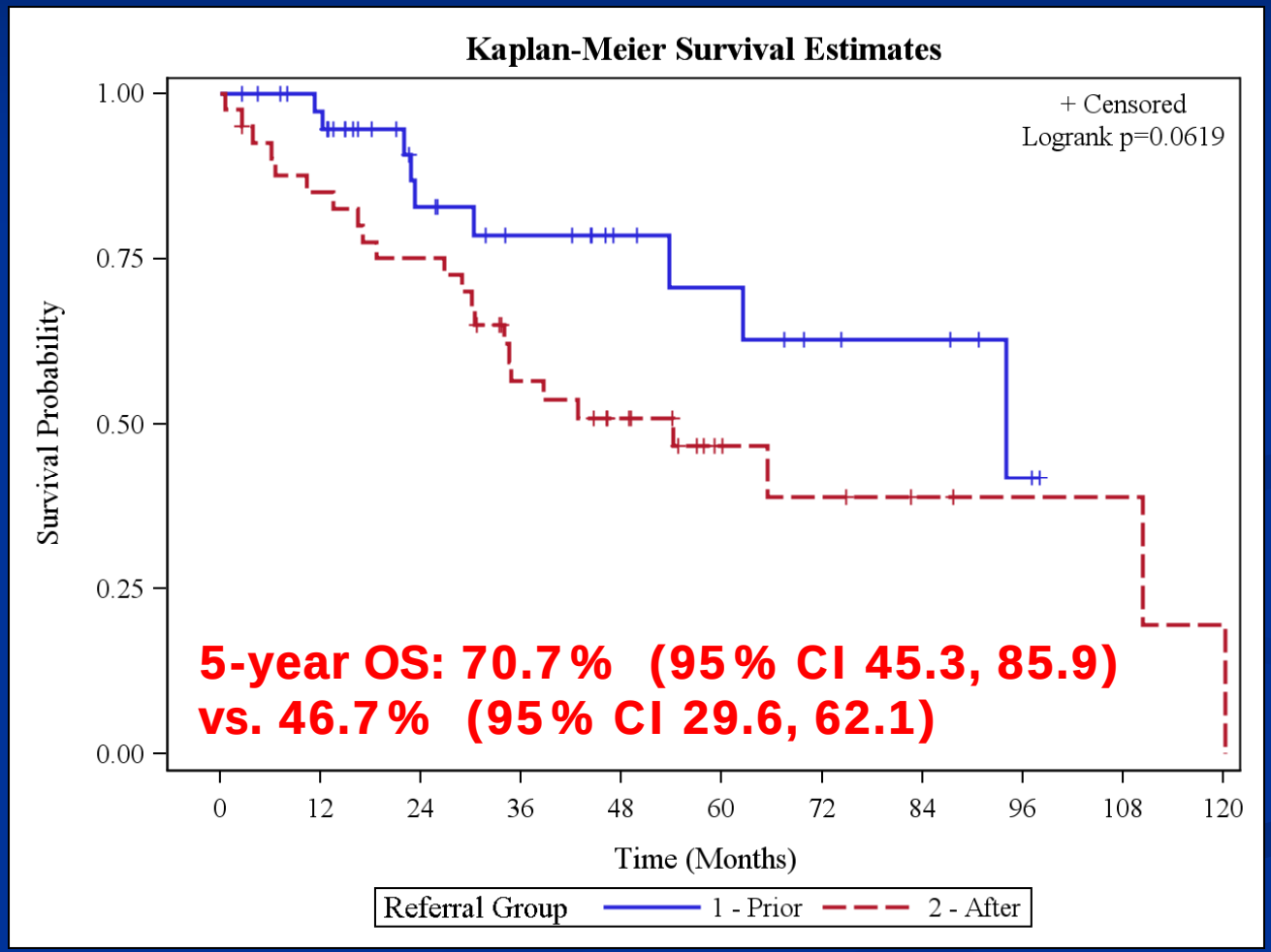
So how are we doing in BC ?

- 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

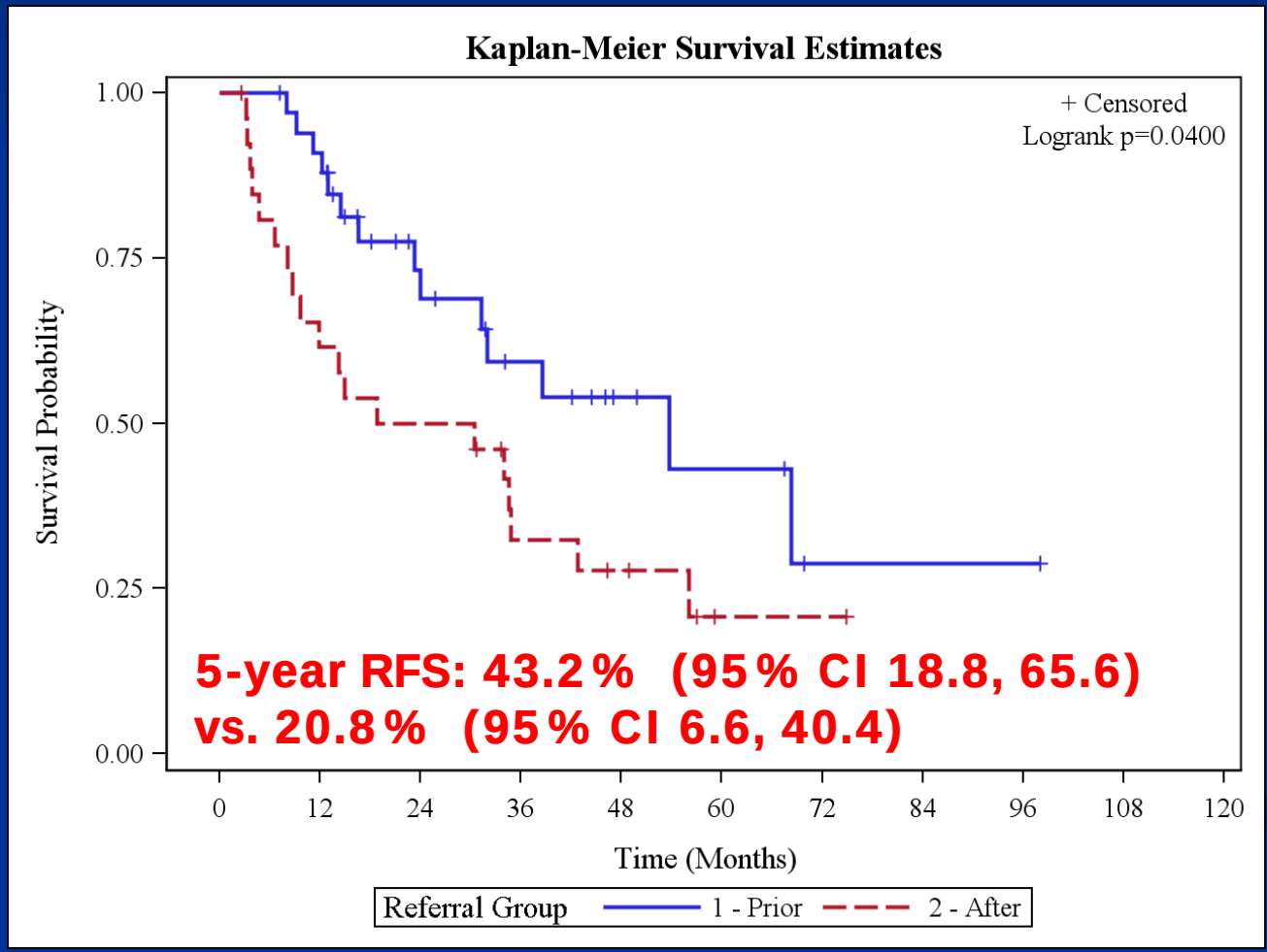
- Of the 82 patients for whom outcome data was available:
- 5 year 0S 56.6% (comparable to literature)
- 41 referred prior to resection and 41 after
- 40/41 vs 27/41 had complete resections
- 34/41 vs 18/41 were alive on follow-up
- $p<0.05$ for both

Overall Survival based on Referral Pattern



(n=82)

Recurrence-Free Survival based on Referral Pattern



(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

Reading Reference

- 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 ??