

Retroperitoneal Sarcomas

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Case Presentation

Chief Complaint:

“I feel a golf ball size mass in my belly”

HPI:

- 63 y/o AA female
- c/o a self-palpated mass in her right abdomen, appreciated one month prior to presentation
- interval increase in size
- + early satiety and decreased appetite
- denied nausea and vomiting or change in bowel habits.
- history was negative for constitutional symptoms; no fever or weight loss

Retroperitoneal Sarcoma

History

PMHx: Hypertension

PSHx : Hysterectomy (fibroid uterus)
Right breast mass excision

Allergies: NKDA

Social Hx: non-contributory

Meds: Atenolol , Avapro

Physical Exam

Pt was A&O, healthy appearing, non-cachectic woman in NAD

Vitals: Temp: 98 BP: 146/79 HR: 72 R:18

Abdomen: soft, NT/ND, +BS

+ palpable, non-tender mass measuring approximately 7-8 cm in the right upper quadrant

Diagnostic Evaluation

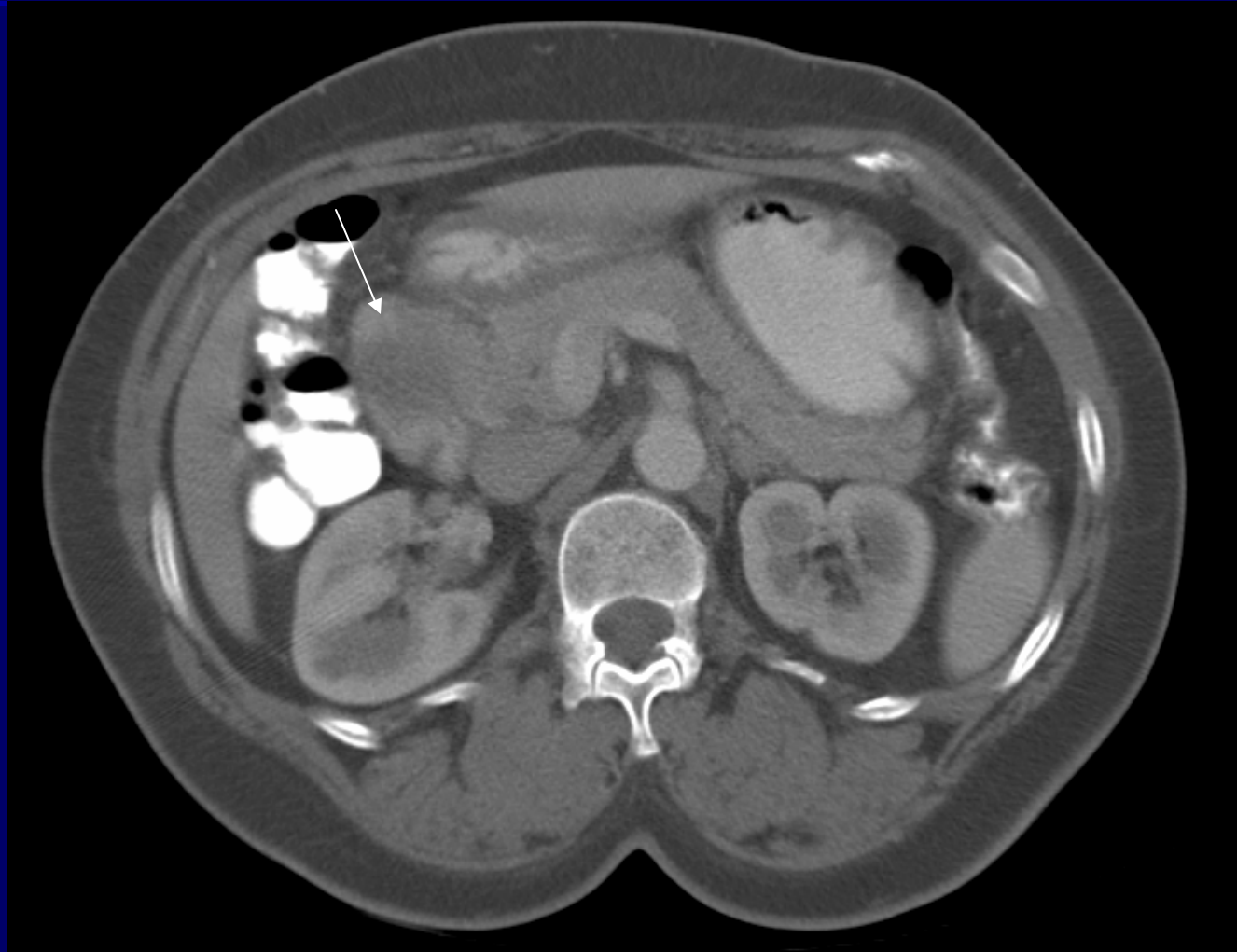
- Pt was referred for a CT scan of Abdomen and Pelvis (6/13/05)

Findings:

large heterogenous soft tissue mass in the right upper abdomen with central necrosis and punctate calcifications; measuring approximately 9 x 8 cm and thought to arise from the duodenum

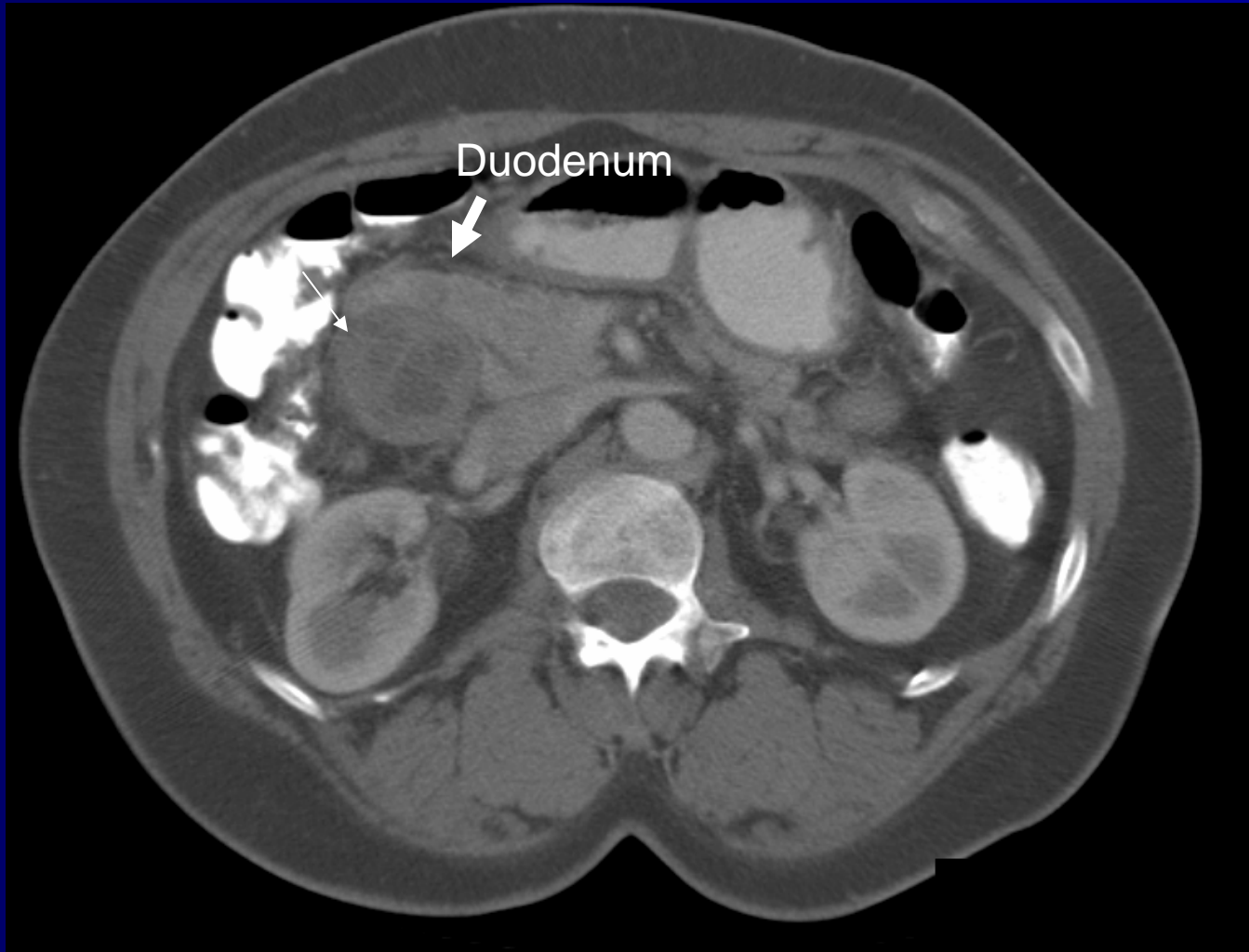
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CT Scan Abdomen and Pelvis



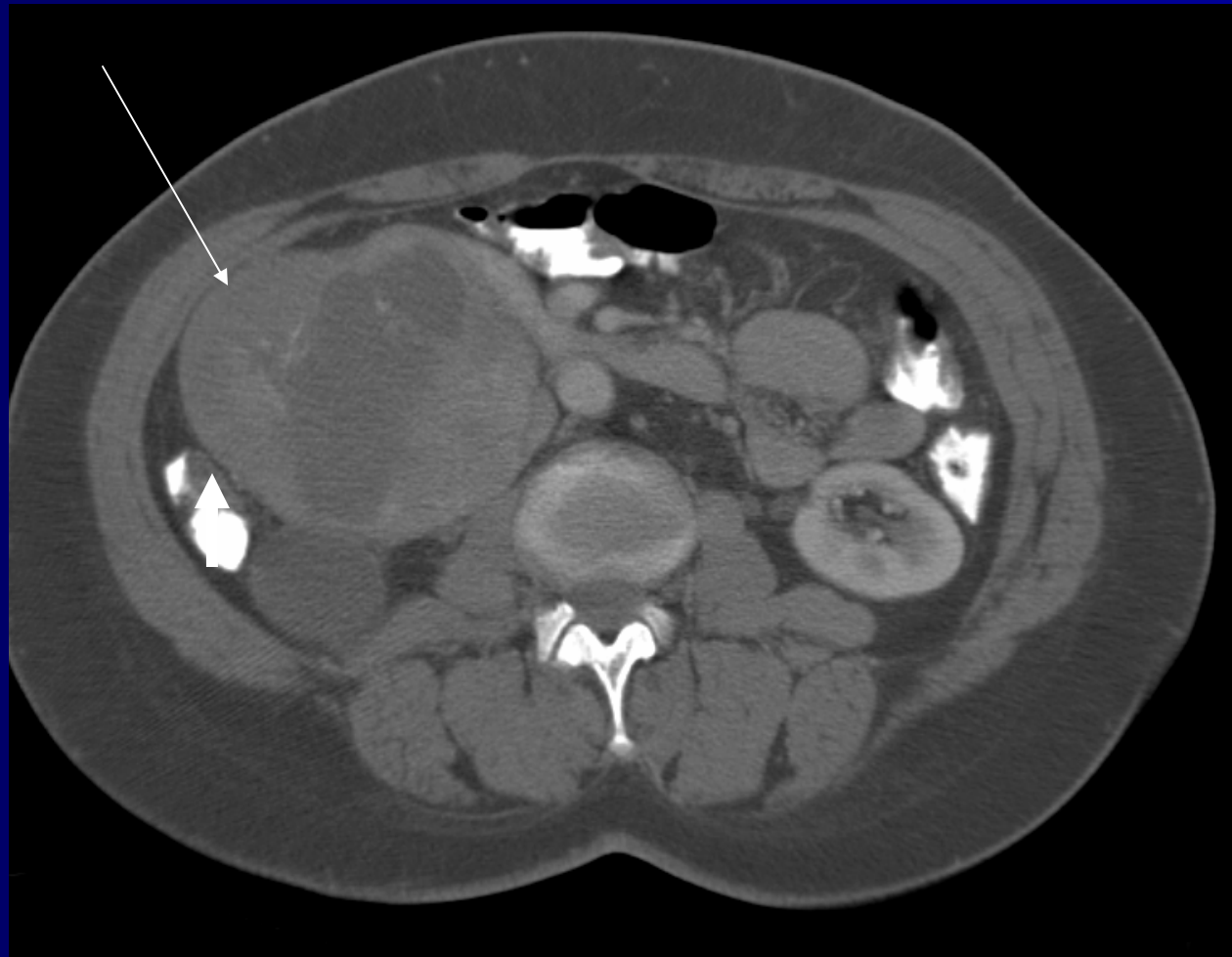
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CT Scan Abdomen and Pelvis



Retroperitoneal Sarcoma

CT Scan Abdomen and Pelvis



Evaluation Con't

- Pt scheduled for EGD and colonoscopy (6/21/05)

EGD: Extrinsic compression of the 1st and 2nd portions of the duodenum; no other abnormalities; normal mucosa

Colonoscopy: Normal

Surgical Referral

Patient was referred for surgical consultation

Planned operative exploration (6/30/05)

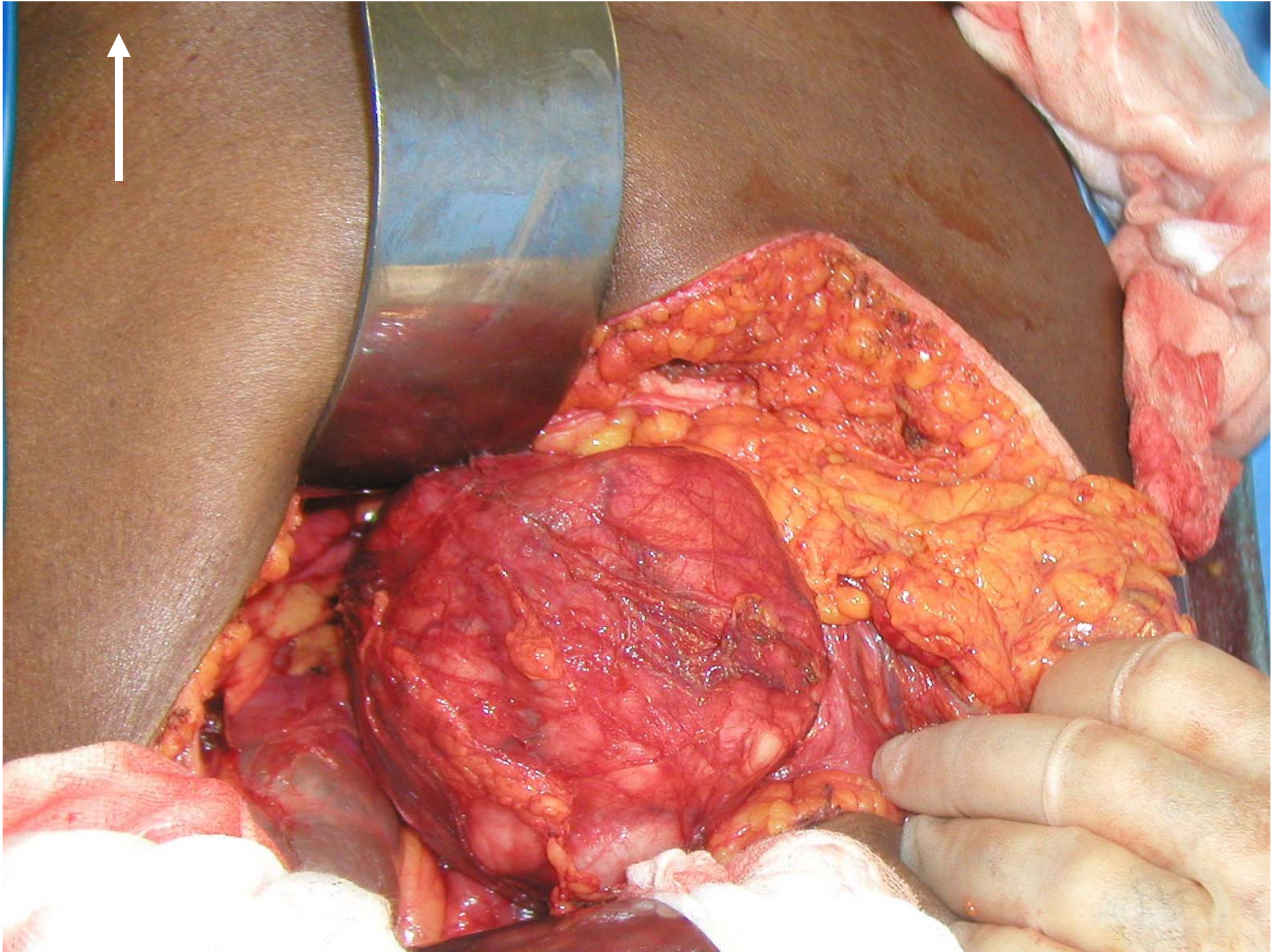
Pre-Op Diagnosis: retroperitoneal mass

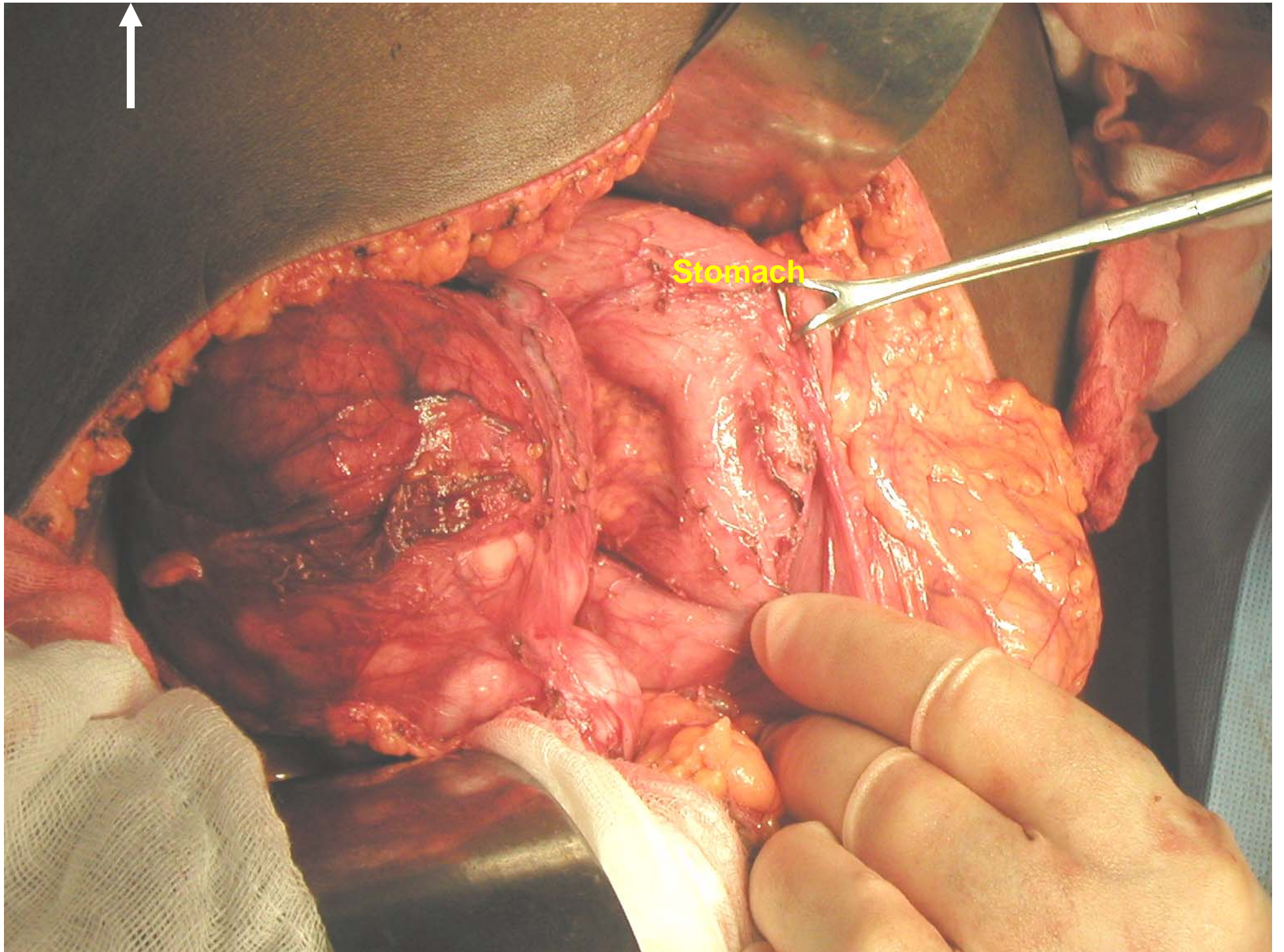
Planned Procedure: Whipple procedure

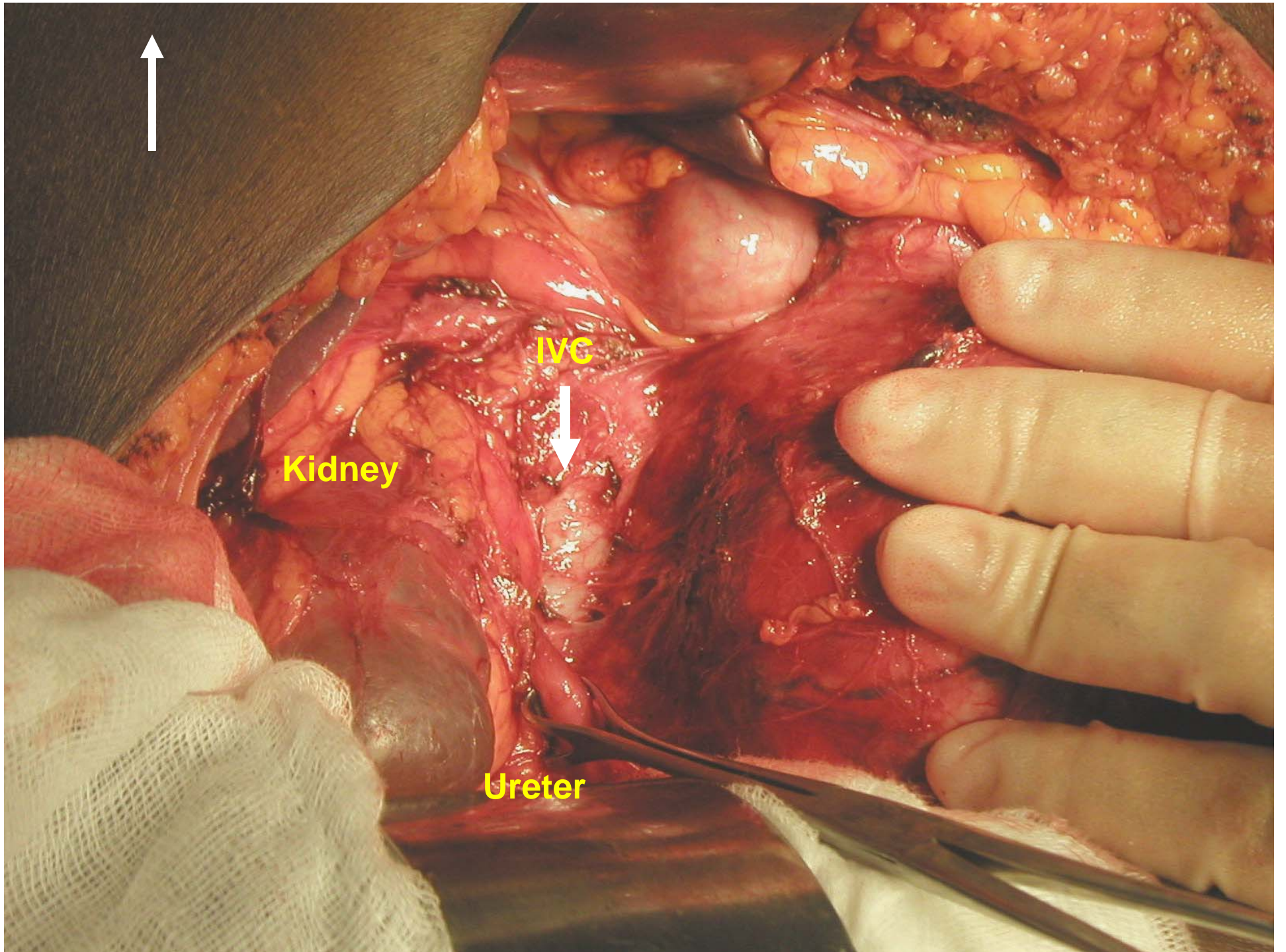
Approach: Chevron incision

Intra-operative Findings

Large, firm retroperitoneal mass, measuring approximately 9 x 11 x 8 cm was identified without attachments to surrounding viscera







Kidney

IVC

Ureter

Pathology

- High Grade Leiomyosarcoma, Grade 3 with extensive tumor necrosis
- Margins of specimen free of tumor
- T2B, Nx, Mx
- Smooth Muscle Actin (SMA) + + +
- CD117 -

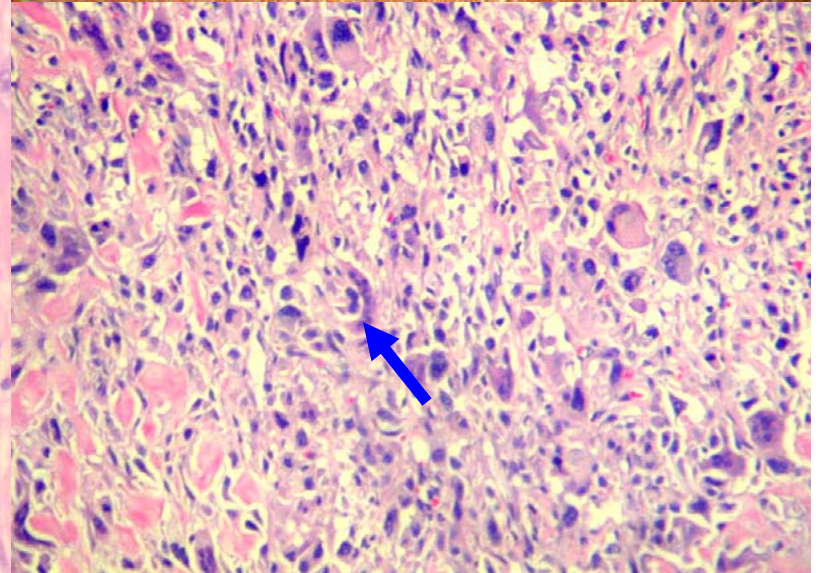
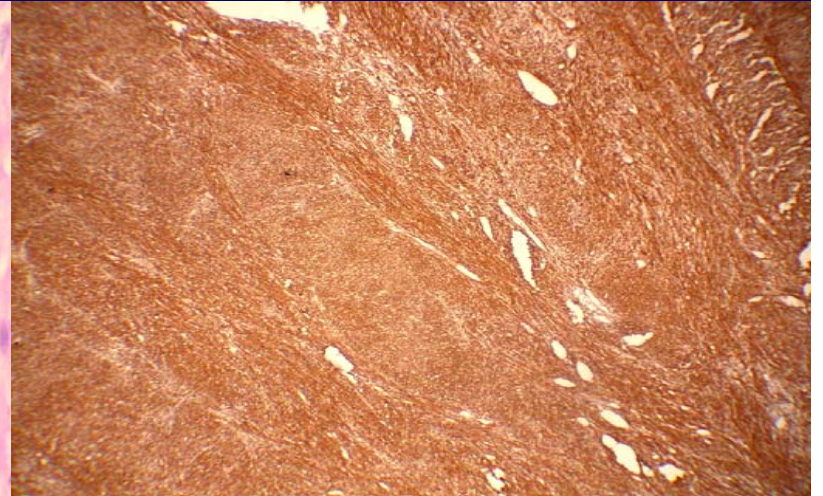
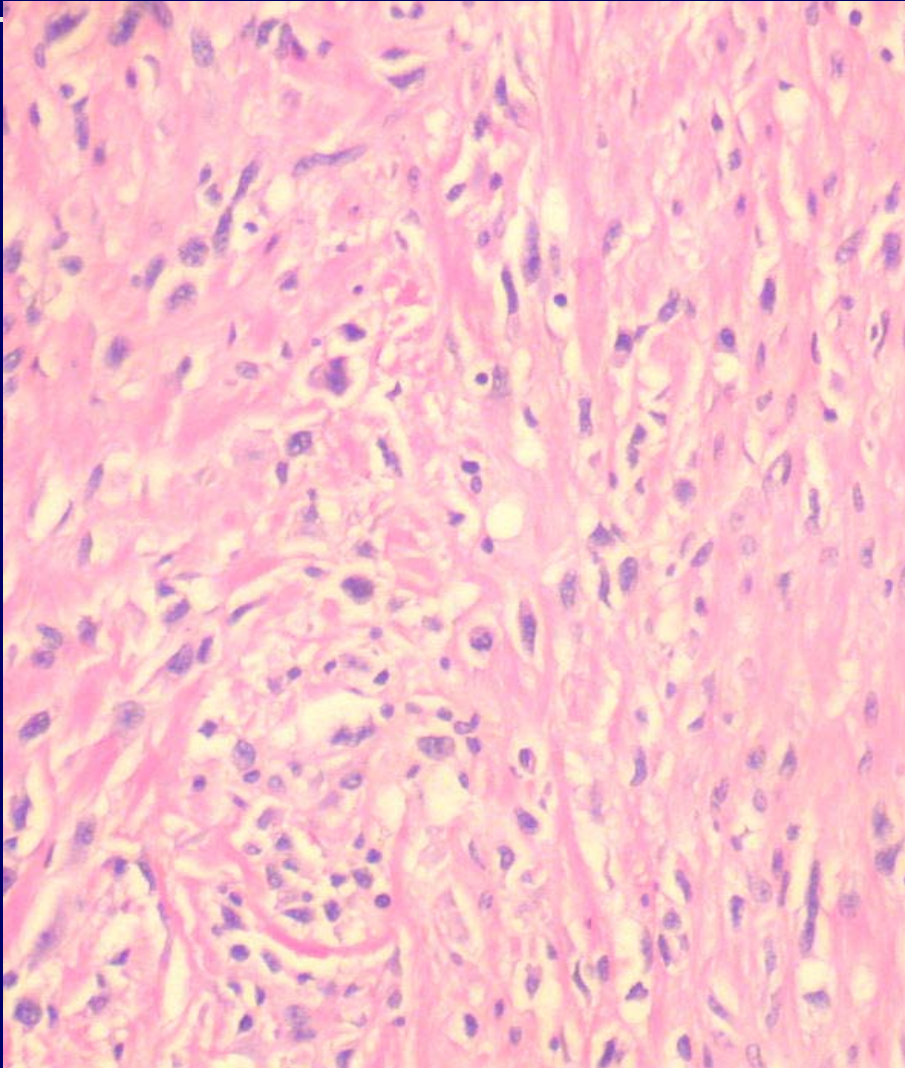
Retroperitoneal Sarcoma

Gross Pathology



Retroperitoneal Sarcoma

Pathology



Post-Operative Course

Patient underwent complete resection of retroperitoneal mass without en-bloc resection of adjacent viscera. Pt was monitored in recovery room overnight and then transferred to regular floor. Pt had uneventful post op recovery and was discharged home on POD #6

Retroperitoneal Sarcomas

Introduction

- Soft tissue sarcomas are rare; approximately 8,600 new cases diagnosed annually
- One third of malignant tumors that arise in the retroperitoneum are sarcomas
- RP sarcomas arise from mesenchymal cells, which are usually located in muscle, fat and connective tissues
- Median age for patients who present with RPS is in the sixth decade of life; male to female ratio is equal

Windham CT, Pister P. Retroperitoneal Sarcomas. Cancer Control 2005. 12 (1): 36-43

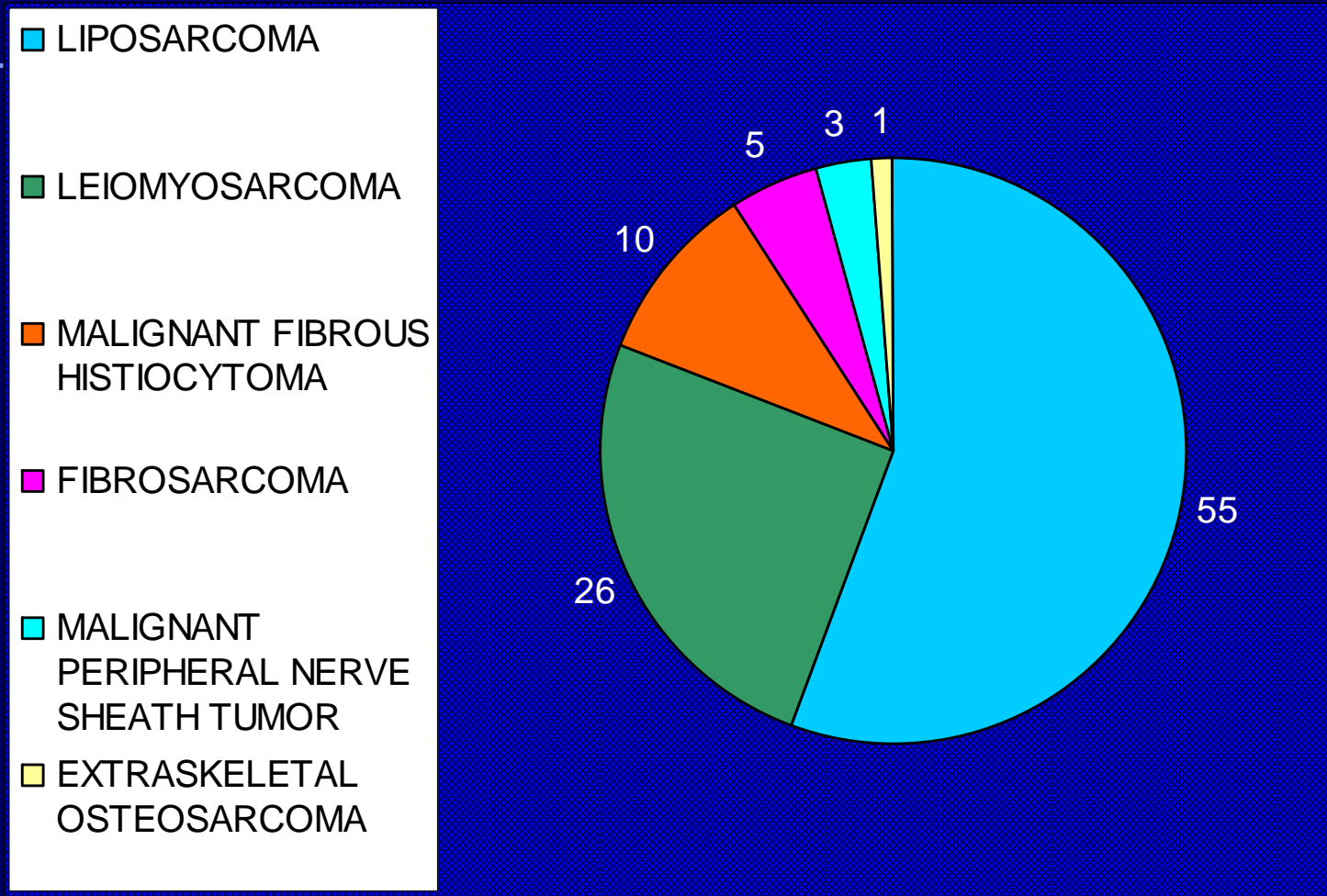
Frequency

- Thigh, buttock and groin region – 46%
- Upper extremity – 13 %
- Head and neck region – 9%
- Torso – 18%
- Retroperitoneal tissues – 13%

Lawrence, W Jr, Donegan, WL, Natarajan, N et al. Adult soft tissue sarcomas. A pattern of care survey of the ACS. Ann Surg 1987; 205:349.

Retroperitoneal Sarcoma

Histologic Types



Hassan I, Park SZ, Donohue JH, et al. Operative Management of Primary Retroperitoneal Sarcomas. Ann Surg 2004; 239: 244-250

Clinical Manifestations

- Asymptomatic abdominal mass (80%)
- Symptoms related to mass effect or local invasion (pain, gastrointestinal obstruction, early satiety and weight loss)
- Neurological and musculoskeletal symptoms referable to the lower extremity
- Median duration of symptoms before diagnosis is 4 months

Mendenhall W, Zlotecki RA, Hochwald SN, et al.

Retroperitoneal Soft Tissue Sarcoma. Cancer 2005; 104: 669-75

Diagnosis

CT scan of the Abdomen and Pelvis

- Assessment of tumor location and relation to adjacent viscera
- Identification of metastatic lesions in the liver or peritoneal cavity

MRI

****Pre-operative tissue diagnosis of resectable retroperitoneal masses is not required**

Staging

| Grade and TNM | Description | | T1a | T1b | T2a | T2b |
|---------------|--|----------|-----|-----|-----|-----|
| G1 | Well differentiated | | | | | |
| G2 | Moderately differentiated | G1 or G2 | IA | | IB | IIA |
| G3 | Poorly differentiated | G3 or G4 | IIB | | IIC | III |
| G4 | Undifferentiated | N1 | IV | | | |
| T1 | Tumor ≤5 cm in largest dimension | M1 | | | | |
| T1a | Superficial to deep fascia | | | | | |
| T1b | Deep to deep fascia (includes retroperitoneal, intrathoracic, and most head and neck tumors) | | | | | |
| T2 | Tumor >5 cm in largest dimension | | | | | |
| T2a | Superficial to deep fascia | | | | | |
| T2b | Deep to deep fascia (includes retroperitoneal, intrathoracic, and most head and neck tumors) | | | | | |
| N1 | Regional nodal metastasis | | | | | |
| M1 | Distant metastasis | | | | | |

| 5-Yr Survival | |
|---------------|-------|
| Stage | % |
| I | 86 |
| II | 72 |
| III | 52 |
| IV | 10–20 |

Surgical Considerations

- Retroperitoneal sarcomas are often unusually large at diagnosis ¹

<5cm = 6%

5-10cm = 25%

10cm = 60%

- Anatomic relations to major vascular structures and vital organs makes resection difficult; significantly impacts ability to obtain negative surgical margins and subsequent local recurrence rates

Mendenhall W, Zlotecki RA, Hochwald SN, et al.

Retroperitoneal Soft Tissue Sarcoma. Cancer 2005; 104: 669-75

Surgical Resection

The standard of care for patients with localized, resectable retroperitoneal sarcomas is surgical resection with gross and microscopically negative margins

Complete surgical resection frequently requires en-bloc resection of adjacent viscera

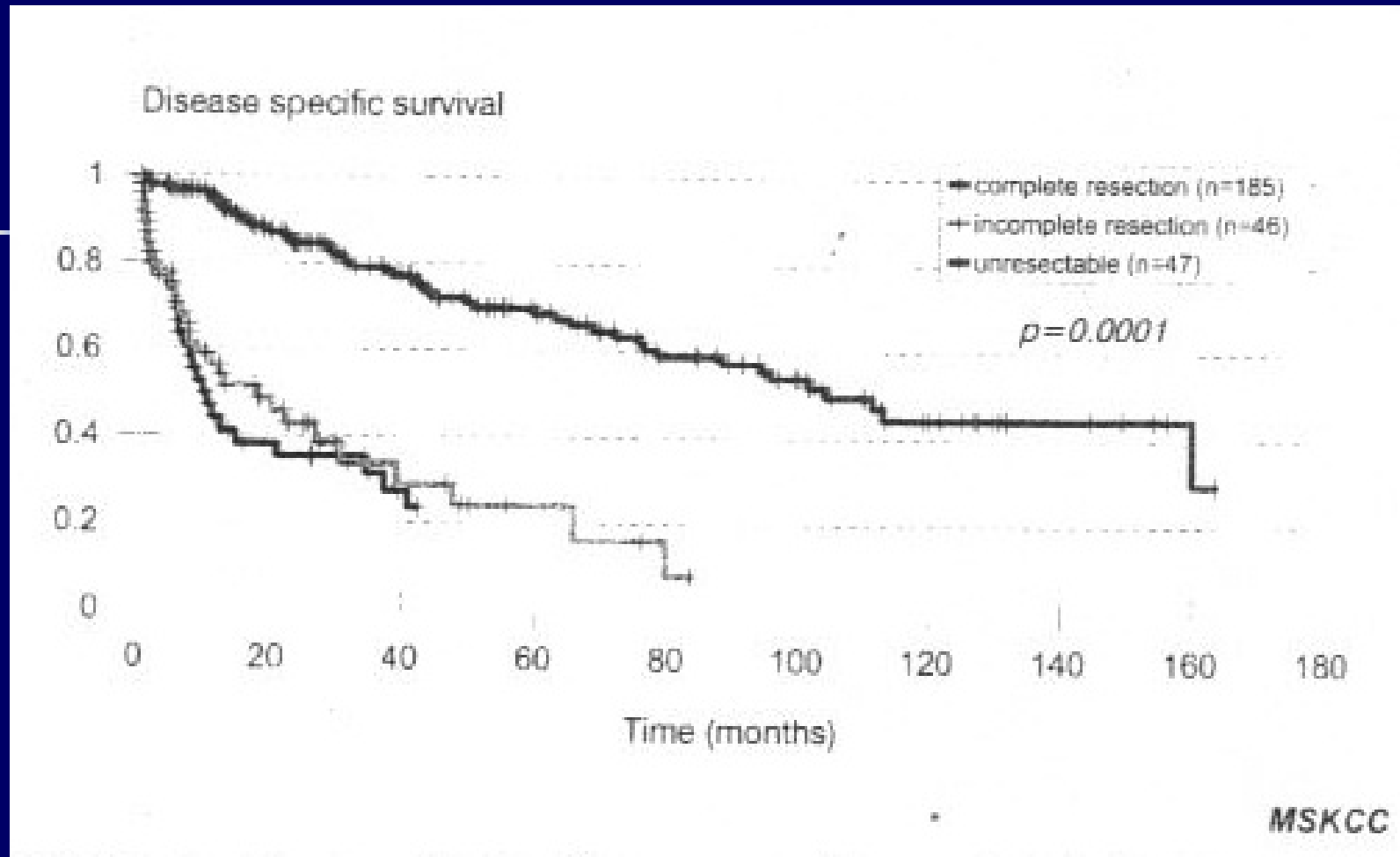
Pisters, P and O' Sullivan, B. Retroperitoneal sarcomas: combined modality treatment approaches. Curr. Opin. Oncol. 2002; 14: 400-405.

Frequency of adjacent organ resection

| | |
|--------------------|-----|
| Kidney | 36% |
| Colon | 22% |
| Spleen | 10% |
| Pancreas | 9% |
| Small Intestine | 6% |
| Stomach | 6% |
| Inferior Vena Cava | 3% |

Hassan I, Park SZ, Donohue JH, et al. Operative Management of Primary Retroperitoneal Sarcomas. Annals of Surgery. 2004; 239 (2): 244-250.

Retroperitoneal Sarcoma



Lewis J, Leung D, Woodruff J et al. Retroperitoneal soft-tissue sarcoma: analysis of 500 patients treated and followed at a single institution. Annals of Surgery 1998; 228(3): 355-365. (MSK)

Surgical Resection

- Complete surgical resection rates range from 62-86%
- The primary pattern of treatment failure after surgery is local recurrence
- Local recurrence rates range from 32-82%

Pisters, P, O'Sullivan B. Retroperitoneal sarcomas: combined modality treatment approaches. Curr Opin Oncol 2002, 14:400-405

Retroperitoneal STS: analysis of 500 patients treated and followed at a single institution

(Lewis, JJ, Leung,D, Woodruff, JM, et al. Ann Surg 1998. 228(3): 355-365

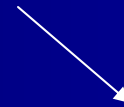
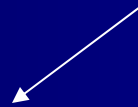
500 Patients



278 primary disease



231 resectable



185 complete resection 46 incomplete resection

** Local recurrence rate in the 231 patients who underwent resection was 40% at 5 years

Adjuvant Radiotherapy

The addition of adjuvant radiation therapy to surgical resection is associated with

- a reduced risk of local recurrence
- a longer recurrence-free interval
- no impact on overall survival

Stoeckle E, Corson JM, Demetri GD et al. Prognostic factors in retroperitoneal sarcoma: a multivariate analysis of a series of 165 patients of the French Cancer Center Federation Sarcoma Group. Cancer 2001; 92:359

Pre-operative Radiotherapy

ADVANTAGES:

- The gross tumor volume is readily definable for accurate treatment planning
- The tumor displaces radiosensitive viscera outside the treatment field
- The radiation dose believed to be biologically active is lower in the pre-operative setting
- Tumor is treated in situ prior to potential contamination of the abdominal cavity that occurs with surgery
- No adhesions and tethering of bowel to the tumor bed; decreases toxicity to radiosensitive bowel

Pisters, P and O' Sullivan, B. Retroperitoneal sarcomas: combined modality treatment approaches. Curr. Opin. Oncol. 2002; 14: 400-405.

Intra-operative Radiotherapy

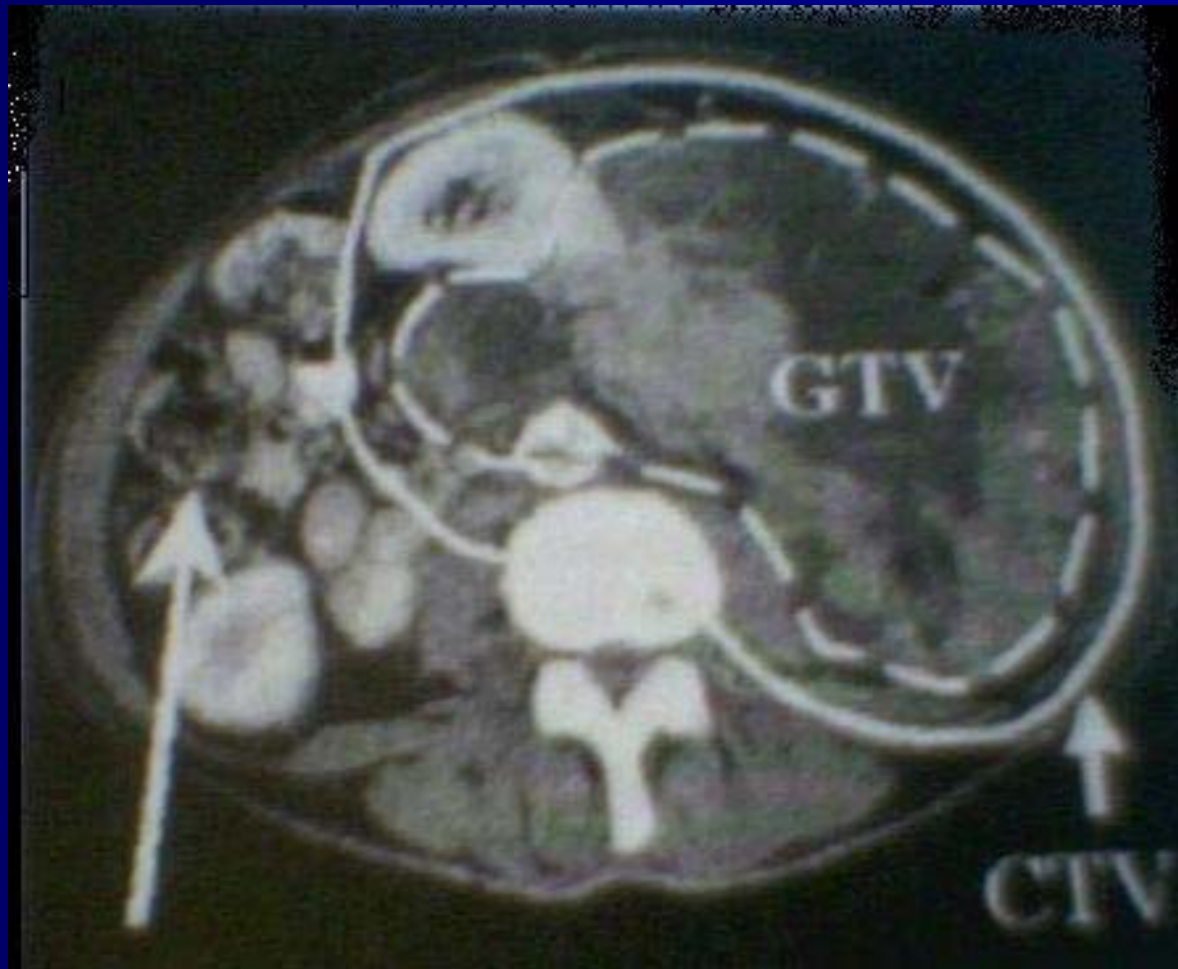
- Radiation dose can be targeted to the specific regions of the operative field that are believed to be at highest risk for harboring residual microscopic disease ¹
- Dose to tumor bed/ dose to normal tissue ratio is maximized ¹
- IORT (EBRT or brachytherapy) increases in field tumor control but not influence recurrence-free or overall survival rates ²

¹ Pisters, P and O' Sullivan, B. Retroperitoneal sarcomas: combined modality treatment approaches. *Curr. Opin. Oncol.* 2002; 14: 400-405.

² Windham CT, Pister P. Retroperitoneal Sarcomas. *Cancer Control* 2005. 12 (1): 36-43

Retroperitoneal Sarcoma

Intra-operative Radiotherapy



Chemotherapy ???

*Mendenhall W, Zlotecki RA, Hochwald SN, et al.
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669-75*

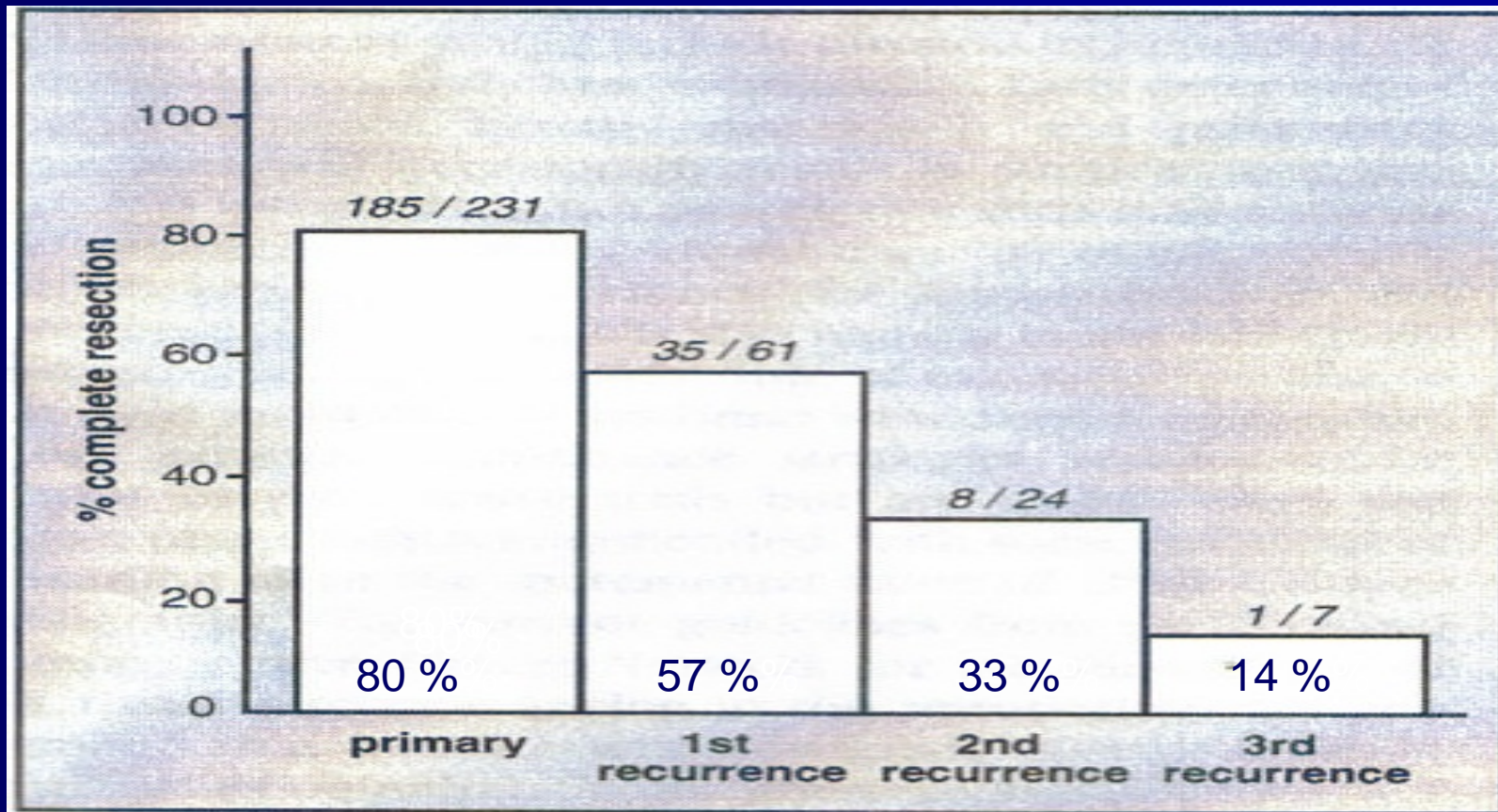
Management of Local Recurrence

- Patients with first local recurrence and no metastases → **perform re-exploration**
- Median survival after local recurrence in patients following resection is **60 months**
- Median survival after local recurrence in patients without resection is **20 months**

Lewis J, Leung D, Woodruff J et al. Retroperitoneal soft-tissue sarcoma: analysis of 500 patients treated and followed at a single institution. 1998; 228(3): 355-365. (MSK)

Retroperitoneal Sarcoma

Local Recurrence



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Management of Metastatic Disease

- Distant recurrence after resection is largely grade-dependent; high grade lesions have the highest risk for distant failure; cumulative incidence is 32%
- Increased risk of metastatic disease with positive gross and microscopic margins of resection
- Distant recurrences usually occur in the liver and lung; hematogenous dissemination

*Windham CT, Pister P. Retroperitoneal Sarcomas.
Cancer Control 2005. 12 (1): 36-43*

Management of Metastatic Disease

- **Pulmonary Metastases:**
 - median survival duration of 6-12 months
 - resection of multiple pulmonary metastases is associated with prolonged relapse-free survival in approximately 25% of patients
- **Hepatic Metastases:**
 - Survival rates are less than those observed for resection of pulmonary metastases
 - Median survival duration was 30 months for patients who underwent resection vs. 11 months for those who did not.

Surveillance

Goal: early detection of local recurrence, hepatic and pulmonary metastases

Physical Exam

CXR

CT Scan of Abdomen and Pelvis

Pisters, P and O' Sullivan, B. Retroperitoneal sarcomas: combined modality treatment approaches. Curr. Opin. Oncol. 2002; 14: 400-405.

Surveillance Guidelines

National Comprehensive Cancer Network Guidelines:

Low Grade Disease:

Physical exam and chest/abdomen/pelvis CT scan every 3-6 months for 2-3 years; then annually

High Grade Disease:

Physical exam and chest/abdomen/pelvis CT scan every 3-4 months for 3 years; then every 6 months for 2 years; then annually

Pisters, P and O' Sullivan, B. Retroperitoneal sarcomas: combined modality treatment approaches. Curr. Opin. Oncol. 2002; 14: 400-405.

Summary

- Patients with RPS often present with large, locally advanced tumors
- The most important factor in long-term success in the treatment of primary RP sarcomas is complete surgical resection
- Wide surgical resection with microscopically negative margins is usually not possible; local recurrence rates are high
- Radiation therapy is useful for local control; no effect on overall survival; chemotherapy has no proven efficacy

QUESTIONS ???