Retroperitoneal Sarcoma

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Introduction

• 15% soft tissue sarcomas are in the retroperitoneum (RP)

• 1/3 RP tumors are soft tissue sarcomas
  – Ddx: lymphoma, germ cell tumor

• Most common RP sarcomas: liposarcoma, malignant fibrous histiocytoma, leiomyosarcoma
Work up

• Exclude other diagnosis, rule out metastasis

• Inquire about fevers, and night sweats on history

• Check lymph nodes, and perform a testicular exam

• Labs: LDH, AFP, AFP

• Biopsy if suspect another diagnosis, or if appears marginally unresectable, metastatic
Treatment

• Surgery - en bloc resection
  – Unresectable if
    • Extensive vascular involvement (ex. Aorta, superior mesenterics at root of mesentery)
    • Spinal cord involvement
    • Peritoneal implants - sarcomatosis
    • Distant metastasis that is unresectable
• Adjuvant therapy
  – Chemotherapy (ex. Doxorubicin, ifosfamide)
    • Poor response rate
  – Radiation (ex. external beam or intraoperative radiation)
    • Some local control but little effect on outcome
    • Marked morbidity (ex. GI obstruction, fistula, peripheral neuropathy)
  – Clinical trials on CDK4 inhibitor, trabectadin (DNA binding agent)
Prognosis

• What matters?
  – Location
  – Margin status
  – Histology
  – Others
Other predictors that decrease survival

- Age >65 years
- Recurrence and metastasis
  - Median survival 72 months for primary disease, 28 months for local recurrence, 10 months for metastasis

Case

• 51 year old male with no PMH presented to an outside hospital with complaints of intermittent left sided flank pain in September 2013. He denied weight loss, cough, shortness of breath, change in appetite, and or energy level.
Case

• PMH, PSH were non contributory
• PE
• NAD
• CV: Reg rate, no murmurs
• Respiratory: clear
• Abdomen: soft, NT, no masses palpated
• No CVA tenderness
• No Testicular masses, no inguinal adenopathy
Case

- CT guided biopsy revealed spindle cell tumor suspicious for sarcoma.
- He was referred to Kings County Hospital surgical oncology clinic.
- Testicular US and MRI of the abdomen were obtained.
Abdominal MRI

www.downstatesurgery.org
Abdominal MRI
Abdominal MRI
Case

Procedure: exploratory laparotomy, en bloc resection of retroperitoneal tumor, left nephrectomy, IVC reconstruction with ring PTFE graft and veno-veno bypass

EBL: 2500, IVF 10L, 6 Units PRBC, 3 Units FFP
Case Image 2
Case

- POD#1 Patient was extubated in SICU
- POD#2 Patient was advanced to clears, started on Aspirin 325mg
- POD#4 Transferred to the floor
- POD#7 Discharged home
Leiomyosarcoma, Actin Grade 3, 11cm, invading to the IVC and left renal vein.

T2NxMx Grade 3, Stage 3

Superior peripheral retroperitoneal margin involved by leiomyosarcoma (piece meal resection at the aorta)
1. Should neoadjuvant radiation therapy been given to this patient?

2. Was his tumor resectable?

3. Is anticoagulation necessary in patients with IVC reconstruction?
The Problem

Location

– RP sarcomas with worse prognosis than extremity sarcomas
  • Large size before clinically apparent
  • Anatomic constraints to resection
  • Less response to chemotherapy and radiation
The possible solution

- Neoadjuvant radiation may decrease size and reduce resectability of tumor as in other malignancies.

- The bulk of tumor displaces the adjacent organs out of the radiation field → less radiation associated toxicity

- Higher doses of radiation can reach the target field.
Zhou et al. Arch Surg 2010
A 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

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Abstract

Background: Local control rates in patients with retroperitoneal soft tissue sarcoma (RSTS) remain disappointing even after gross total resection, mainly because wide margins are not achievable in the majority of patients. In contrast to extremity sarcoma, postoperative radiation therapy (RT) has shown limited efficacy due to its limitations in achievable dose and coverage. Although Intraoperative Radiation Therapy (IORT) has been introduced in some centers to overcome the dose limitations and resulted in increased outcome, local failure rates are still high even if considerable treatment related toxicity is accepted. As postoperative administration of RT has some general disadvantages, neoadjuvant approaches could offer benefits in terms of dose escalation, target coverage and reduction of toxicity, especially if highly conformal techniques like intensity-modulated radiation therapy (IMRT) are considered.

Methods/design: The trial is a prospective, one armed, single center phase I/II study investigating a combination of neoadjuvant dose-escalated IMRT (50–56 Gy) followed by surgery and IORT (10–12 Gy) in patients with at least marginally resectable RSTS. The primary objective is the local control rate after five years. Secondary endpoints are progression-free and overall survival, acute and late toxicity, surgical resectability and patterns of failure. The aim of accrual is 37 patients in the per-protocol population.

Discussion: The present study evaluates combined neoadjuvant dose-escalated IMRT followed by surgery and IORT concerning its value for improved local control without markedly increased toxicity.

Trial registration: NCT01566123
The problem

- No survival benefit of debulking tumor, only palliative benefit
Positive margin decreases survival

The Problem

- IVC reconstruction with the use of graft can lead to thrombosis of the IVC.
Summary

• Surgical resection with negative margins remains the key to curative treatment and improved overall survival.

• Use of neoadjuvant radiation therapy for retroperitoneal sarcomas is currently under investigation.
Questions

• Which of the following is true regarding retroperitoneal sarcomas?
  • A. The majority of patients will present with paraneoplastic syndromes
  • B. Some patients present with symptoms of lower extremity neural compromise.
  • C. Biopsy is indicated in all patients diagnosed with a RP mass.
  • D. Leiomyosarcoma is the most common pathology
  • E. The retroperitoneum is the most common site of soft tissue sarcoma.
Questions

• Which of the following is true regarding medical therapy in patients with RP sarcoma?
  • A. RP sarcomas are highly responsive to chemotherapy.
  • B. Adjuvant radiation therapy is associated with significant morbidity in patients with RP sarcomas
  • C. Radiation enteritis is a rare complication of the adjuvant radiation therapy
  • D. Radiation therapy results in increased overall survival in these patients
  • E. Neoadjuvant radiation is not used in patients with RP sarcomas.