CASE

Dr. Serafini
King’s County Hospital
• 29F presented with 1 week of abdominal pain, worse in the past 2 days; no weight loss/fevers/chills/vomiting / diarrhea
• PMH: DM type II, HTN, HLD
• PSH: none
• Fam hx: DM, HTN; no cancer
• Meds: metformin, simvastatin, amlodipine
• Soc hx: no tobacco / alcohol / other drugs
• VS: 99.2 / 147/88 / 99 / 20
• Obese
• NAD, AAO
• RRR
• Clear BS b/l
• Abd soft, ND, palpable mass in LUQ and epigastric area that is TTP and dull to percussion, +BS
• No CVA tenderness
• Ext: no edema
- CBC: 12.5 / 10.2 / 33.5 / 334
- BMP: 144 / 4.1 / 104 / 24 / 5 / 0.54 / 89
- LFT: 7.0 / 3.6 / 31 / 32 / 56 / 0.3
- Lipase 43, UA negative
- CXR: negative
- CT: 17 x 17 x 19 cm mass displacing the pancreas, stomach, small and large bowel
Surgery

Findings:
- Large, firm, with hemorrhagic component
- Obliterated lesser sac space and adherent to tail of pancreas
• Procedure: exploratory laparotomy, en bloc resection of retroperitoneal mass with omentum, spleen, and body and tail of pancreas
• JP drain left by pancreatic stump
• EBL: 300 ml
• Patient passed flatus on POD#3; clear liquid diet started and advanced
• Metformin restarted
• Hemophilus, pneumococcal, and meningococcal vaccines given
• Discharged home on POD#6 with JP
Pathology:
- 25 cm, 3750 g
- Retroperitoneal fibromatosis
- Invasion into pancreas
- Negative margins
- Negative for CD117, DOG1, CD34, S100, HHF-35
- Pending study on t(7;16) for fibromyxoid sarcoma
RETROPERITONEAL LIPOSARCOMAS

Introduction
Workup
Treatment
Prognosis
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
• Histological types:
  Atypical lipomatous tumor/Well-differentiated (ALT/WDLPS)
  De-differentiated
  Myxoid / Round cell
  Pleomorphic
• **Presentation**
  
  • **Asymptomatic**
    - Nearly 50% are >20 cm at time of diagnosis
  
  • **Symptoms with compression or invasion of structures**
    - Bowel obstruction
    - Neurologic symptoms from lumbar or pelvic nerves
• Metastasis
  • Liver metastasis
  • Sarcomatosis
  • Lung metastasis
WORKUP

- Exclusion of other dx
  - Hx – fever, night sweats
  - Phys exam – LN, testicular exam
  - Labs – LDH, HCG, AFP
• Rule out metastasis
  • Imaging – chest and abdominal CT
• Biopsy if suspicion of other dx, unresectable, or metastasis - implications for primary non-surgical treatment or neoadjuvant therapy
TREATMENT

- Surgery - en bloc resection
  - Unresectable if
    - Extensive vascular involvement (ex. Aorta, superior mesenterics at root of mesentary)
    - Spinal cord involvement
    - Peritoneal implants - sarcomatosis
    - Distant metastasis that is unresectable
Treatment for local recurrence is surgery although ability to achieve negative margins decreases.
• 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

- **AJCC staging**

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<th>M0</th>
<th>G1</th>
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<td>Any N</td>
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- Not very predictive of outcome
• What matters?
  • Location
  • Margin status
  • Histology
  • Others
A.) Location

- RP sarcomas with worse prognosis than extremity sarcomas
  - Large size before clinically apparent
  - Anatomic constraints to resection
  - Less response to chemotherapy and radiation
B.) Positive margin decreases survival

- No survival benefit to debulking; only palliative

• 1 cm/month rule
  • Faster growth decreases survival

C.) Different histological types with different survival

D.) 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

• Nomogram for adult primary liposarcoma

ex. 30F with 30 cm RP myxoid liposarcoma, resected with negative margins

- Age: 30
- Gender: Female
- Presentation status: Biopsy
- Primary site: Upper extremity
- Histologic variant: Myxoid
- Tumor depth: Deep
- Margins: Negative
- Total Points: 160
- 5-yr DSS Probability: 90%
- 12-yr DSS Probability: 78%
TAKE-HOME POINTS

- Workup of a RP tumor takes into account the ddx to determine management

- Complete resection is key to curative treatment and survival
QUESTIONS

ACROSS
1. Ddx of RP tumor
2. Matters for survival

DOWN
3. RP metastasis
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Thank you