

AMSER Rad Path Case of the Month:

53 year old female presenting with abdominal pain

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Patient Presentation

HPI: 53 year old female presents with one week history of dull alternating with sharp abdominal pain located in the subcostal and bilateral lower quadrants. Associated with bloating, decreased appetite, nausea, and back pain. She denies fever, chills and changes in weight.

SHx: Denies smoking, alcohol and recreational drug use.

FHx: Colon cancer (mother); prostate cancer (father); thyroid cancer (aunt and cousin)

Physical exam: Abdominal distention and tenderness to palpation

Pertinent Labs

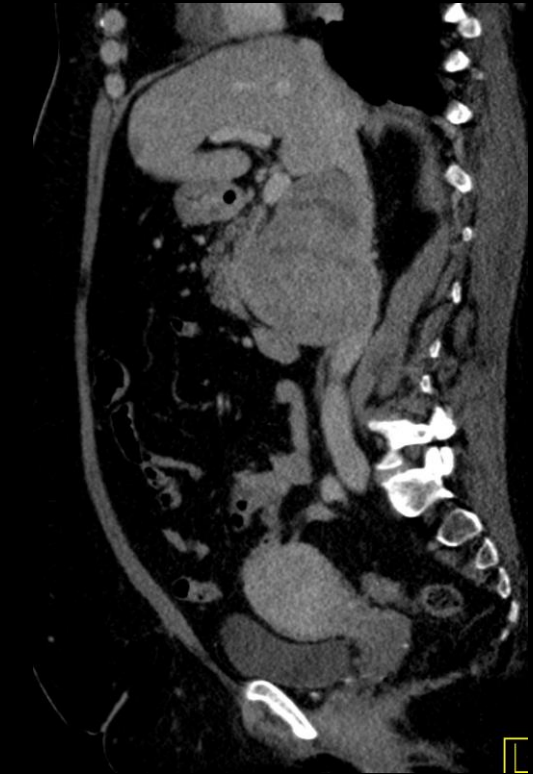
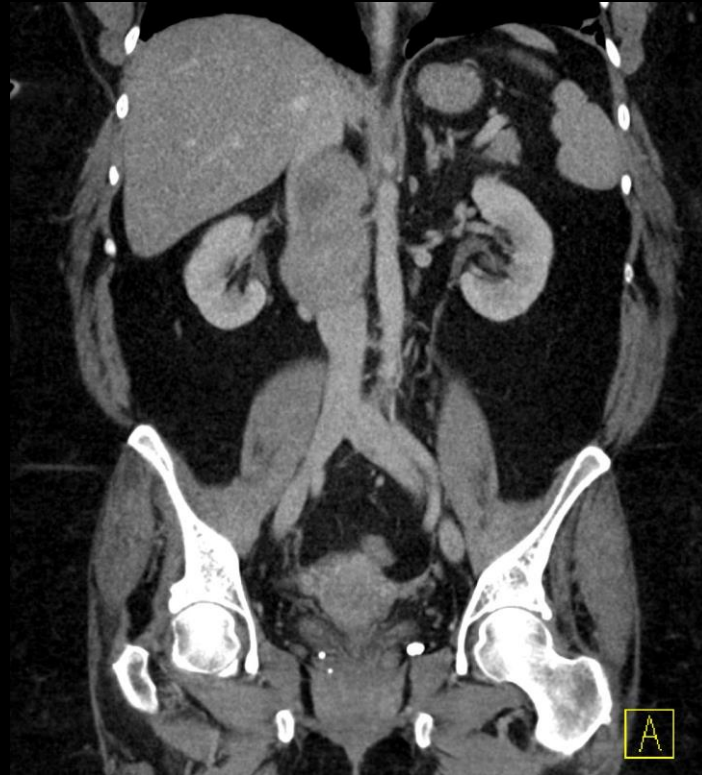
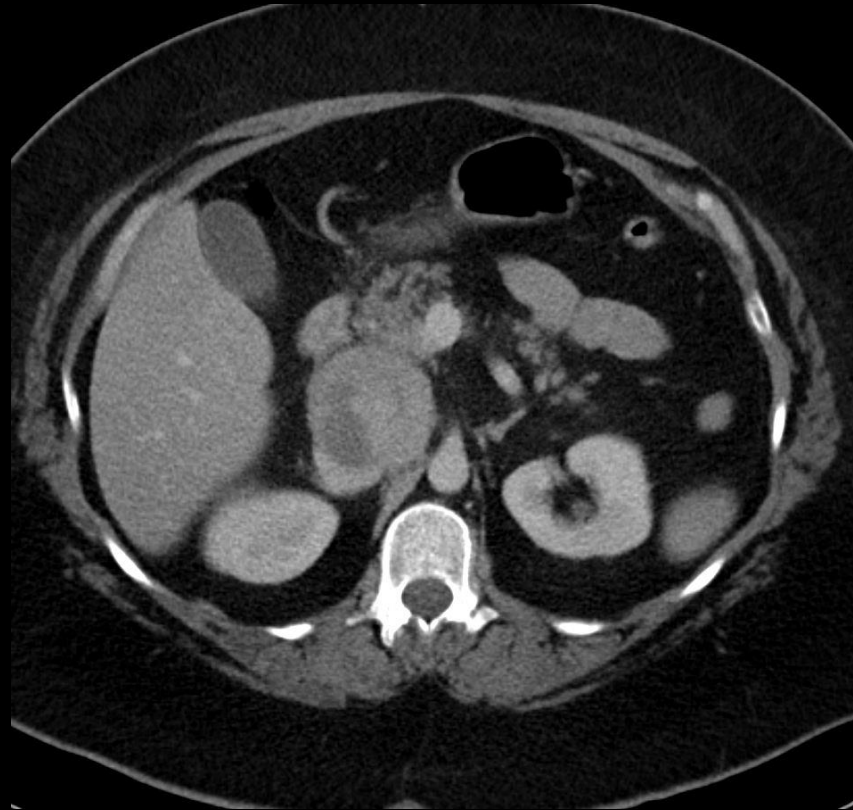
- Tumor Markers CA 15-3, CA 19-9, CA 125, and CAE all within normal limits

ACR Appropriateness Criteria

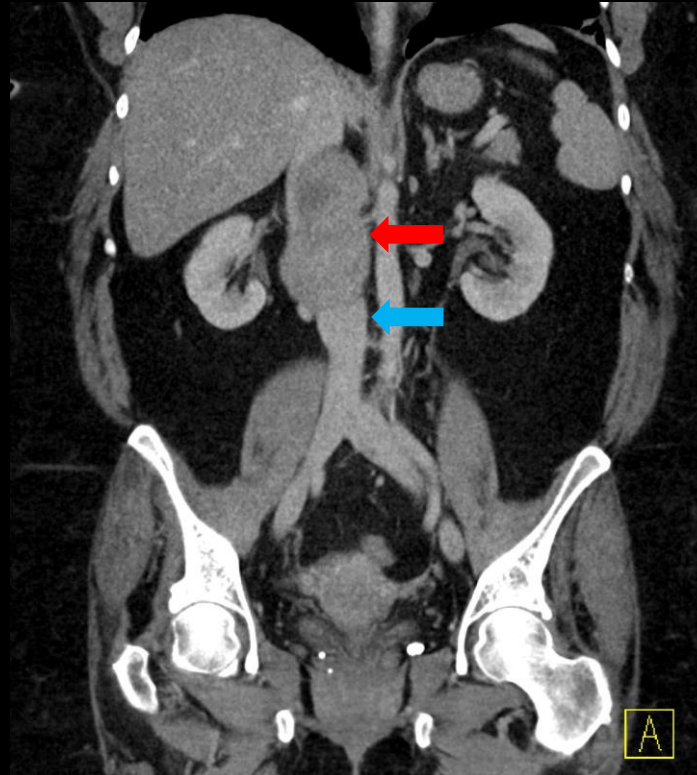
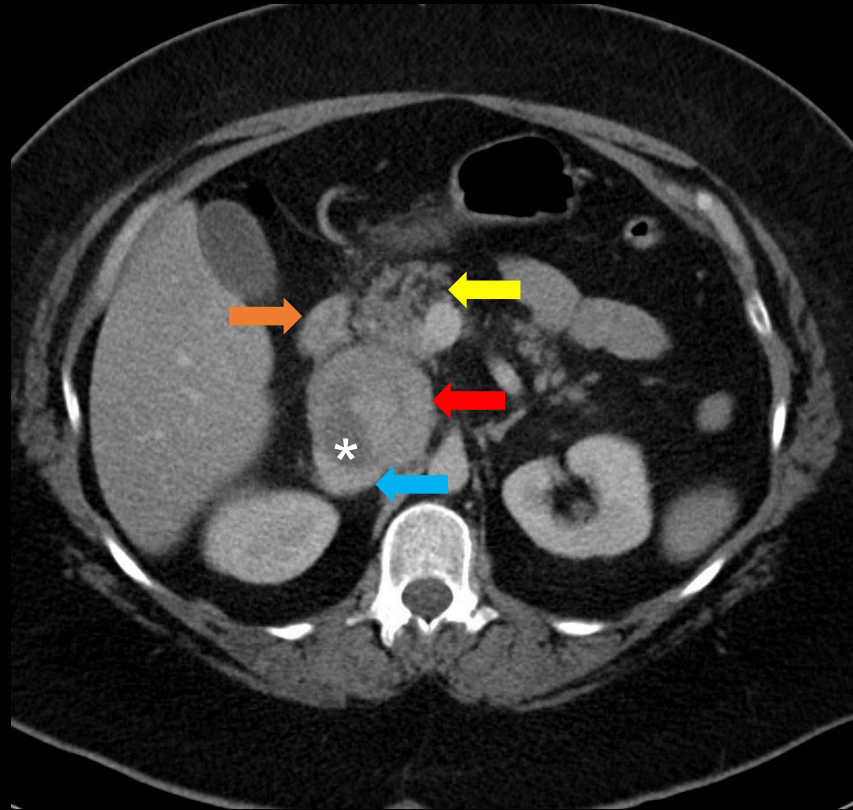
Variant 4: Acute nonlocalized abdominal pain. Not otherwise specified. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
CT abdomen and pelvis with IV contrast	Usually Appropriate	☼☼☼
CT abdomen and pelvis without IV contrast	Usually Appropriate	☼☼☼
MRI abdomen and pelvis without and with IV contrast	Usually Appropriate	○
US abdomen	May Be Appropriate	○
MRI abdomen and pelvis without IV contrast	May Be Appropriate	○
CT abdomen and pelvis without and with IV contrast	May Be Appropriate	☼☼☼☼
Radiography abdomen	May Be Appropriate	☼☼
FDG-PET/CT skull base to mid-thigh	Usually Not Appropriate	☼☼☼☼
In-111 WBC scan abdomen and pelvis	Usually Not Appropriate	☼☼☼☼
Tc-99m cholescintigraphy	Usually Not Appropriate	☼☼
Tc-99m WBC scan abdomen and pelvis	Usually Not Appropriate	☼☼☼☼
Fluoroscopy upper GI series with small bowel follow-through	Usually Not Appropriate	☼☼☼
Fluoroscopy contrast enema	Usually Not Appropriate	☼☼☼

Radiology Images (not labeled)



Radiology Images (labeled)

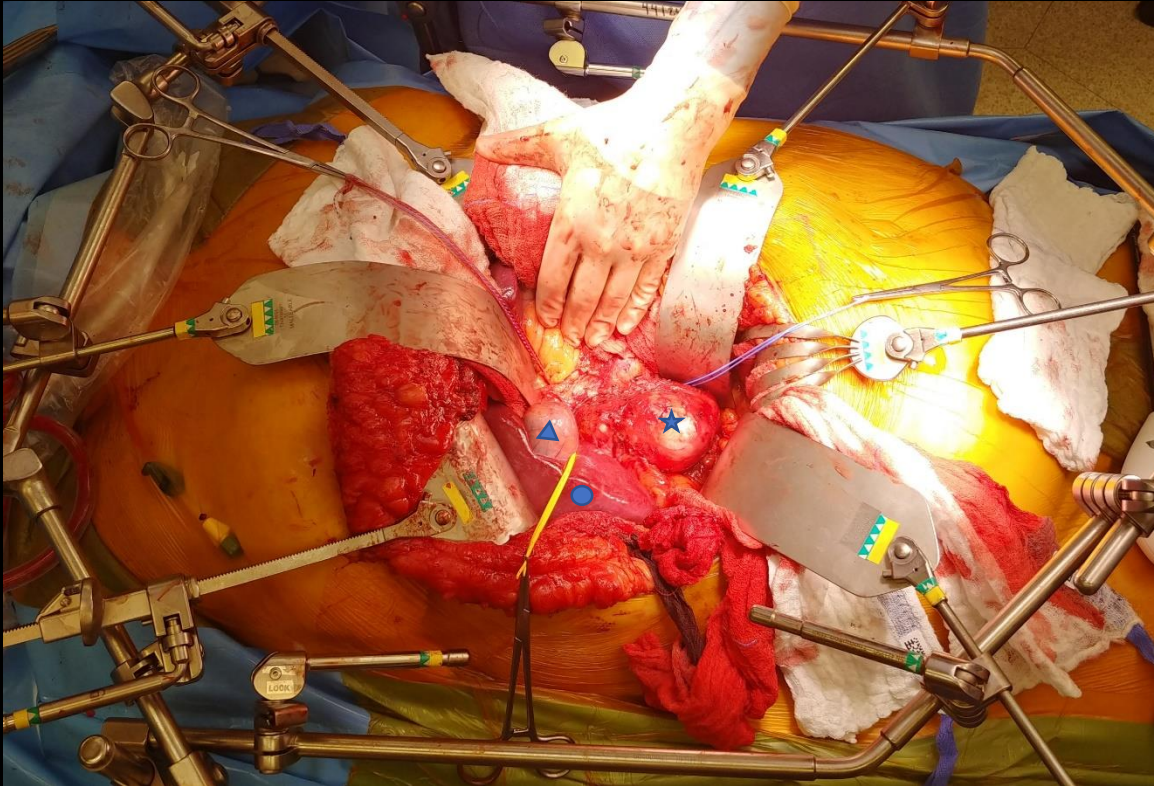


Heterogeneously enhancing retroperitoneal mass (red arrow) along the inferior vena cava (IVC) (blue) from which it may be arising. The mass displaces duodenum (orange) and pancreatic head (yellow) anteriorly. Mass is clearly separate from the kidney and adrenal glands. Areas of hypodensity (*) possibly representing necrosis.

DDX

- Leiomyosarcoma
- Lymphoma
- Metastatic tumor
- Liposarcoma
- Neurofibroma
- Schwannoma
- Ganglioneuroma
- Paraganglioma
- Teratoma

Gross Path (labeled)



Retroperitoneal mass (star) excised by open laparotomy.
Liver (circle) and gallbladder (triangle).



9.5 cm ivory pink mass and ~ 2 cm
section of anterior IVC (arrow) excised.

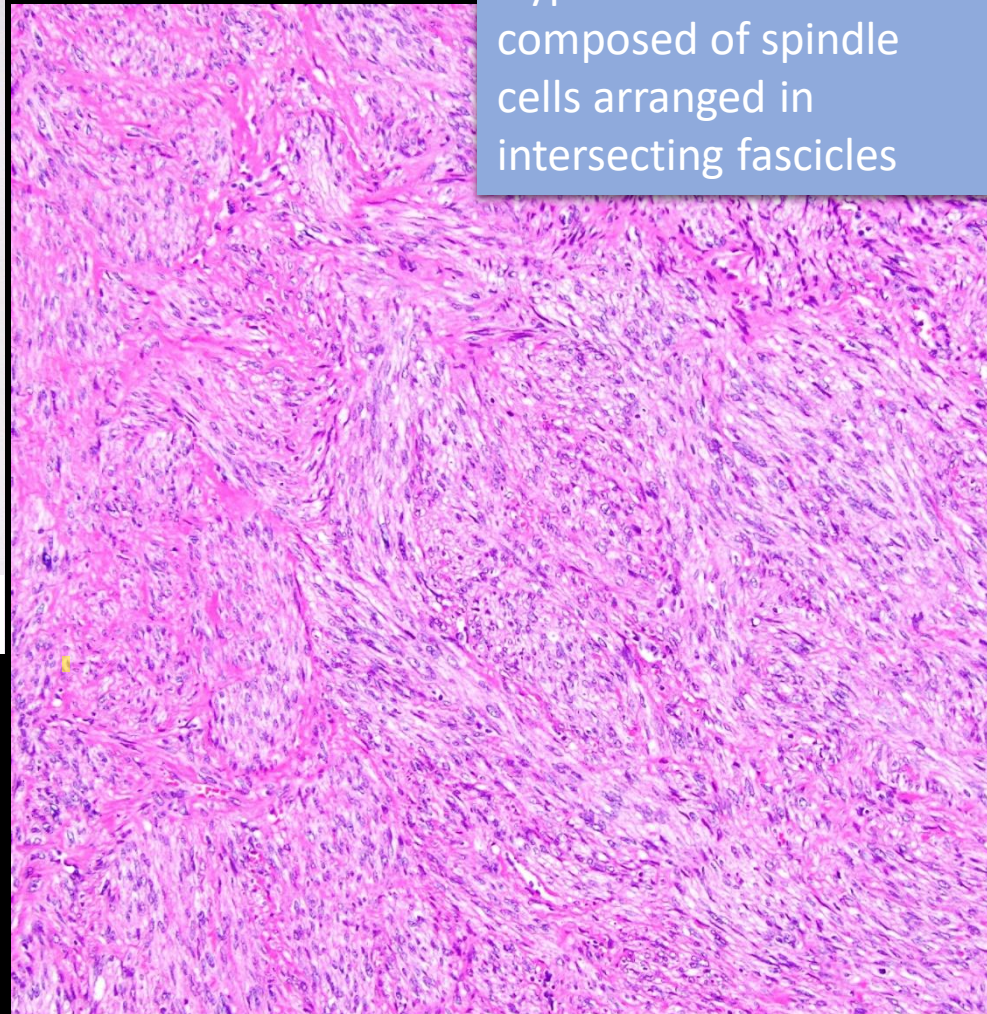
Micro Path (labeled)



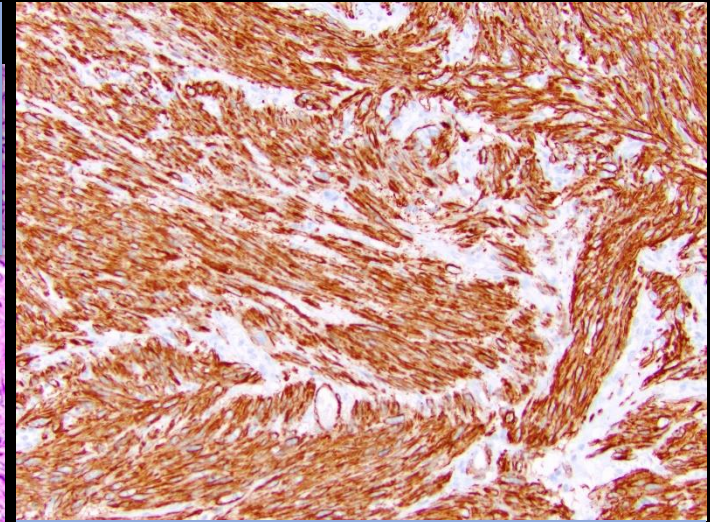
H&E 4x

Neoplastic growth (triangle) arising from smooth muscle of the IVC (star)

Hypercellular tumor composed of spindle cells arranged in intersecting fascicles



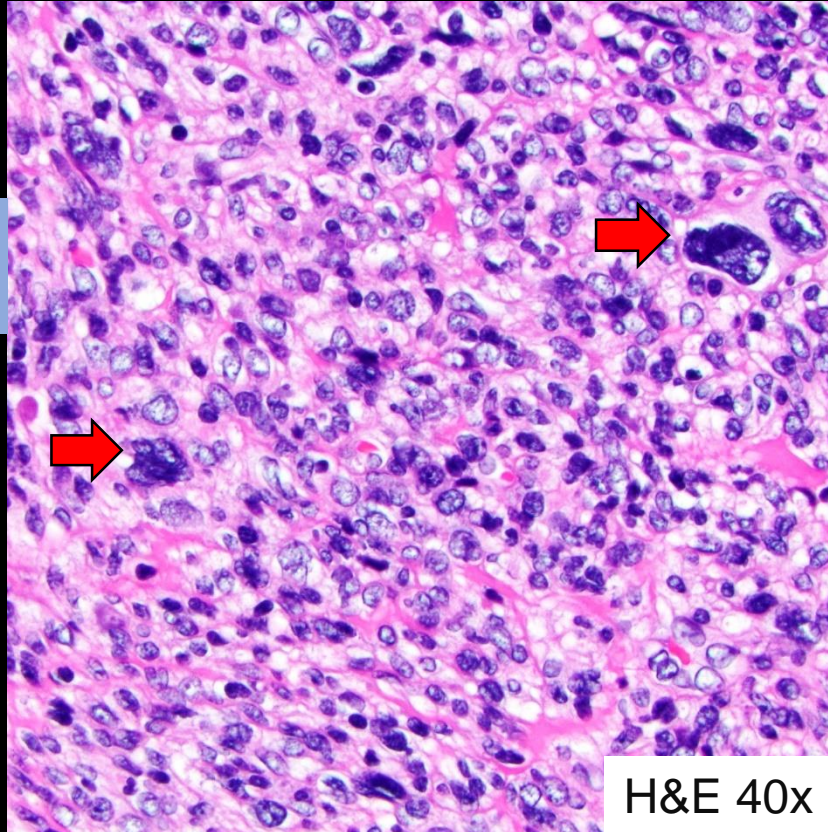
H&E 10x



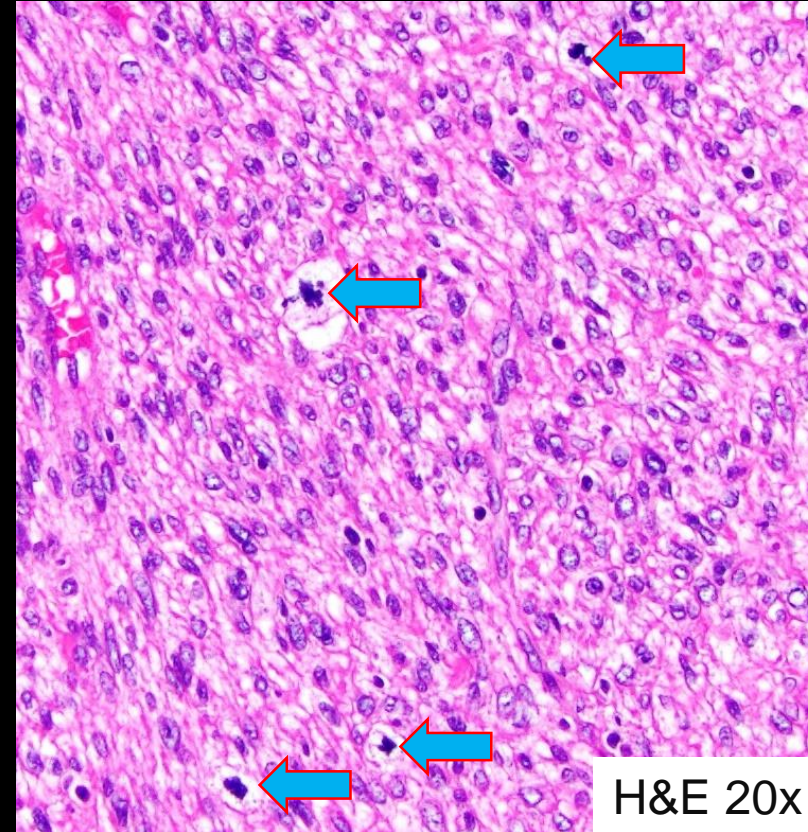
Positive immunostaining for caldesmon (shown above at 10x) suggesting smooth muscle origin; Mass was also positive for smooth muscle actin (SMA), muscle specific actin (MSA) and desmin

Micro Path (labeled)

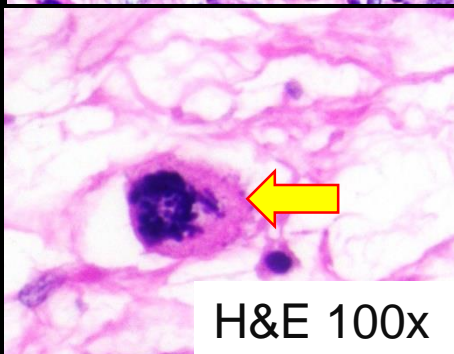
Nuclear atypia
(red arrows)



Numerous mitotic figures
(blue arrow)



Abnormal mitotic figures
(yellow arrows)



Final Dx:

Retroperitoneal Leiomyosarcoma

Case Discussion

Background

- Leiomyosarcomas are soft-tissue sarcomas composed of smooth muscle cells
- Most commonly found in adults ages 54-65; More common in women
- Account for 28% of retroperitoneal soft-tissue sarcomas; second most common after liposarcoma
- Commonly arise from large vessels of the retroperitoneum including the IVC and renal veins

Case Discussion

Clinical Features

- May grow large before compressing structures and causing symptoms; Mean size is 11.4 for non-vascular tumors and 10.4 for vascular
- Presentation varies on location and involves symptoms of compression
 - If supra-hepatic may cause Budd-Chiari syndrome presenting with hepatomegaly, ascites and jaundice
 - If at level of renal veins may cause renal dysfunction
 - If infrarenal may cause lower extremity edema

Case Discussion

Imaging Features

- Large lobulated heterogeneous retroperitoneal mass on CT
- Solid, often with a cystic or necrotic component
- Solid portions are hyperattenuating and necrotic portions are hypoattenuating on post-contrast CT
- Local invasion is common
- Hematogenous metastasis more common than lymphatic
- Most commonly spread to lungs and liver

Case Discussion

Pathologic Features

- Grossly a large, fleshy mass with whorled appearance on cut surface
- Histologically shows features of smooth muscle tissue including:
 - Spindle cells forming fascicles
 - Immunohistochemistry (+) for smooth muscle actin (SMA), desmin, and h-Caldesmon (more specific for smooth muscle)
- Usually 2 out of 3 features of malignancy listed below:
 1. Nuclear atypia/pleiomorphism
 2. Tumor necrosis
 3. >10 mitotic figures per 10 high power field

References:

- American College of Radiology. ACR Appropriateness Criteria®. Available at <https://acsearch.acr.org/list> . Accessed 7/31/2019
- Kempson, R. L., & Rouse, R. V. (2007, December 1). Leiomyosarcoma of Deep Soft Tissue, Retroperitoneum, Mesentery and Omentum. Retrieved August 09, 2019, from http://surgpathcriteria.stanford.edu/softsmoothmuscle/soft_tissue_leiomyosarcoma/
- Marko, J., and Wolfman D. J. (2018) Retroperitoneal Leiomyosarcoma From the *Radiologic Pathology Archives* RadioGraphics 2018 38:5, 1403-1420 available at <https://pubs.rsna.org/doi/abs/10.1148/rq.2018180006> . Accessed 7/31/2019