Leiomyosarcoma of the Inferior Vena Cava

Hayley Walker, Harvard Medical School, MSIII
Gillian Lieberman, MD
Overview

- Patient presentation
- Pulmonary embolism on CTA
- Retroperitoneal mass on CT
- Differential diagnosis for retroperitoneal mass
- Menu of tests
- Approach to IVC masses
- Narrowing the differential with imaging
- Leiomyosarcoma
- Impact of imaging on surgical planning
- IVC anatomy review
- Intraoperative ultrasound
- History continued
- Post-surgical follow-up
Our Patient: History of Present Illness

- The patient is a 26-year-old male who developed left leg swelling and pain in August 2011. Ultrasound revealed a left occlusive deep vein thrombosis (DVT) from the left common femoral vein through the posterior tibial vein.

- He was started on warfarin. He had no known DVT risk factors. Hematology work-up found no evidence of an acquired hypercoagulable state or inherited thrombophilia.

- In February 2012, he was admitted to an outside hospital with chest pain, and was found to have a pulmonary embolism (PE).

- His INR at the time was therapeutic, between 2 and 2.5. He was therefore begun on enoxaparin for anti-coagulation.
Pulmonary Embolism on CTA

This is the patient’s CTA image showing a pulmonary embolism.

- Findings: **Filling defect** in right pulmonary artery

Other structures:
- Aortic outflow tract
- Right ventricle
- Descending aorta

Outside hospital records
Abnormal mass on CTA

- This is the most caudal image from the CTA performed at the outside hospital when the patient presented with a PE.
- This **abnormal structure** was not mentioned on the radiologic report.
- This reminds us to always remember to inspect all images!
Our patient: History continued

- The patient had an outpatient hematology appointment in February 2012
- Because the patient suffered a pulmonary embolism despite being on warfarin with a therapeutic INR, a CT abdomen and pelvis was performed to rule out occult malignancy.
Mass on CT abdomen and pelvis

- A **large mass** with areas of increased density can be seen in the retroperitoneum
  - Measured 7.4 x 7.4 x 9.7 cm
- The inferior vena cava (IVC) is not visualized
- Multiple abnormal **subcutaneous collateral vessels** have formed to bypass the obstructed IVC
Differential diagnosis for retroperitoneal mass

- Soft tissue sarcoma
- Lymphoma
- Primary germ cell tumor
  - Check AFP and HCG levels to assess likelihood (both were normal in this patient)
- Metastatic testicular cancer
  - Perform scrotal ultrasound to assess likelihood (was normal in this patient)
- Neoplasms from duodenum, pancreas, adrenal glands, or kidneys
- Schwannomas, paragangliomas
- Benign processes: Castleman’s disease (angiofollicular lymph node hyperplasia), retroperitoneal fibrosis
ICV mass on CT abdomen & pelvis

- Imaging suggests the mass is within the IVC itself (and not simply retroperitoneal and externally compressing the IVC).
- Continue to view the menu of tests for imaging the IVC.
Imaging the IVC: Menu of Tests

- **Conventional venography**
  - Historical gold standard; now rarely used

- **Ultrasonography**
  - Color Doppler flow imaging is used to assess for blood flow

- **MDCT**
  - Different phases of IV contrast administration are used
    - **Arterial phase:** Identify hypervascular tumors. Assess for pulmonary emboli or lung metastases.
    - **Portal venous phase:** 60-70 seconds after contrast injection. Typically used to evaluate IVC. Can also evaluate liver parenchyma.
      - IVC heterogeneously enhances in this phase. Non-opacified blood from lower extremities mixes with opacified blood from renal veins. Can make IVC thrombus assessment difficult.
    - **Three-minute delay phase:** Provides more homogenous enhancement of IVC lumen; allows better assessment of superior and inferior extension of tumor.

- **MR**
  - T2-weighted images as well as pre- and post-IV contrast injection T1-weighted images
The menu of tests for imaging the IVC has been presented.

Please proceed to view a general approach to IVC masses.
Approach to IVC masses: Step 1

- The **first step** is to differentiate between bland thrombus (clot) and tumor thrombus
  - Tumor thrombus suggested by:
    - Expansion of lumen by the thrombus
    - Enhancement of filling defect
    - Direct continuity between tumor in another organ and thrombus

- Please proceed to the next slide to view an image of a companion patient which illustrates the characteristic features of tumor thrombus
Companion patient: tumor thrombus on CT

- This companion patient was found to have tumor thrombus in the IVC secondary to renal cell carcinoma
- The thrombus obstructs and expands the IVC
- There is direct continuity between the renal mass and the IVC thrombus
- Neovascularity in the main thrombus confirms the finding is tumor thrombus and not bland thrombus
  - This image was obtained in the arterial phase.

Approach to IVC masses: Step 2

- Once a mass is identified as tumor thrombus and not clot, the **second step** is to differentiate between primary and secondary IVC tumors

- Primary IVC Tumors
  - Leiomyosarcoma is the most common malignant primary tumor of the IVC

- Secondary IVC Tumors
  - Please proceed to the next slide to view a list of secondary IVC tumors
Approach to IVC masses: Secondary IVC tumors

- Secondary IVC Tumors
  - Tumors extending contiguously from a primary tumor
    - Renal cell carcinoma (most common)
    - Hepatocellular carcinoma
    - Adrenocortical carcinoma
    - Wilms’ tumor (children)
  - Leiomyosarcoma arising in retroperitoneum can secondarily invade IVC
  - Rarely, renal angiomyolipoma and pheochromocytoma involve the IVC. Metastatic disease in retroperitoneal lymph nodes can also extend into the IVC.
  - Females: Intravenous leiomyomatosis
    - Smooth muscle tumor; either arises in uterine veins or represents extension of uterine fibroma into the IVC
● You have now seen a general approach to IVC masses

● In this patient, imaging helped to narrow the differential diagnosis for IVC masses

● Please proceed to see images which caused the radiologist to favor a primary IVC tumor over a secondary tumor
Narrowing the differential with imaging: Ruling out renal or adrenal tumor

- **Left adrenal gland** (y-shaped) appears normal
- **Right adrenal gland** is difficult to visualize, but appears normal
- Both **kidneys** are unremarkable
Primary IVC tumor seemed most likely based on CT findings

- The liver, spleen, and bowel appeared normal. The pancreas was deviated anteriorly by the mass, but was otherwise unremarkable. Lymph nodes were not enlarged.
- It seems the mass is arising from the IVC itself, and not from another retroperitoneal organ or structure.
  - This suggests primary IVC tumor, with leiomyosarcoma being most common.
Pathologic diagnosis: Leiomyosarcoma

- CT-guided core needle biopsy was performed to confirm the diagnosis
  - Pathologic diagnosis: Leiomyosarcoma
- IVC Leiomyosarcoma
  - Arises from smooth muscle cells in vessel wall
  - Often presents late; occasionally with leg swelling, ascites, or Budd-Chiari syndrome
  - Most often affects lower 2/3 of IVC. Can be entirely intraluminal or extend outside lumen
  - Usually seen in women in 5th and 6th decades.
  - Poor prognosis: 14% survival at 10 years
- Treatment of IVC Leiomyosarcoma
  - Surgery. Complete resection with microscopically negative margins offers best hope for cure.
  - Role for radiation and chemotherapy is debated.
Impact of imaging on surgical planning

- Imaging can play a role in planning surgery for resection of an IVC tumor.

- Thrombus extension into the supradiaphragmatic IVC requires cardiopulmonary bypass surgery
  - It is important to identify superior extension of tumor prior to surgery with CT, MRI or intraoperative ultrasound

- If tumor invades vessel wall, segmental resection of IVC is necessary

- In this patient, involvement of IVC at confluence of hepatic veins would likely necessitate partial liver resection
The patient underwent surgery in February 2012.

The surgeon wanted to determine whether the IVC was involved at the level of the hepatic veins. Involvement of the hepatic veins would necessitate partial liver resection.

The next slide will review relevant anatomy.

Following the anatomy review, please proceed to view intraoperative ultrasound images.
The surgeon wished to determine whether the IVC was involved at the level of the hepatic veins.
Intraoperative ultrasound: Locating the mass

The IVC mass was located on intraoperative ultrasound

Findings:

- **IVC** superior to mass
- **Superior aspect of IVC mass**
- **Liver**
Mass on intraoperative ultrasound

- The mass shows mixed echogenicity
- Doppler reveals blood flow to the mass
IVC is patent at level of hepatic veins on intraoperative ultrasound

- **Image 1** shows hepatic veins draining into a patent IVC

- **Image 2**, taken from a more inferior level, shows increased echogenicity within the IVC due to the mass

  - Ultrasound demonstrated 3.5 cm between the superior tumor edge and the confluence of the hepatic veins. Therefore, resection of the liver was not necessary.
Our patient: History continued

- The mass was removed, including a portion of the IVC
  - Possible because of extensive collateral circulation
- Pathology showed negative margins, and confirmed diagnosis of leiomyosarcoma (high-grade)

Example of collateral circulation that allows blood to bypass the IVC

Follow-up for patients with leiomyosarcoma

- Leiomyosarcoma most commonly metastasizes to lung and liver
  - Follow-up after resection should involve imaging of chest, abdomen, and pelvis on a regular schedule
  - Imaging is recommended every 3-6 months for 2-3 years; however, there have been no randomized trials comparing different surveillance strategies
Lung nodules on follow-up CT

**August 2012**
- Left lower lobe **nodules** noted on follow-up CT
- Underwent VATS left lower lobe wedge resection in August 2012. Nodules proved to be leiomyosarcoma on pathology

**December 2012**
- New bilateral pulmonary **nodules** noted on follow-up CT, very concerning for progression of metastatic disease
- **Evidence of prior resection** can be seen
Staging of Leiomyosarcoma

- Unfortunately, this patient has stage IV disease
- Five-year survival estimates for stage IV disease range from 0-17%

References

Acknowledgements

- Dr. Gunjan Senapati
- Dr. Gillian Lieberman