Tumors invading the inferior vena cava with intracardiac extension, imaging features of a series of cases in our institution

Poster No.: C-3219
Congress: ECR 2018
Type: Educational Exhibit
Authors: L. A. Calderón Ramírez, J. P. Garzon, J. C. Aldana, P. Sanchez; Bogota/CO
Keywords: Surgery, Complications, Staging, MR, CT-Angiography, CT, Vascular, Cardiac, Abdomen, Blood, Cancer, Neoplasia
DOI: 10.1594/ecr2018/C-3219

Any information contained in this pdf file is automatically generated from digital material submitted to EPOS by third parties in the form of scientific presentations. References to any names, marks, products, or services of third parties or hypertext links to third-party sites or information are provided solely as a convenience to you and do not in any way constitute or imply ECR's endorsement, sponsorship or recommendation of the third party, information, product or service. ECR is not responsible for the content of these pages and does not make any representations regarding the content or accuracy of material in this file.

As per copyright regulations, any unauthorised use of the material or parts thereof as well as commercial reproduction or multiple distribution by any traditional or electronically based reproduction/publication method is strictly prohibited.

You agree to defend, indemnify, and hold ECR harmless from and against any and all claims, damages, costs, and expenses, including attorneys' fees, arising from or related to your use of these pages.

Please note: Links to movies, ppt slideshows and any other multimedia files are not available in the pdf version of presentations.

www.myESR.org
Learning objectives

96 Normal 0 21 false false false ES-CO JA X-NONE

- Review the main tumoral conditions that compromise the inferior vena cava, some of them with intra cardiac extension and their differential diagnosis.
- Recognize filling defects that affect the inferior vena cava.
- Emphasize the importance of a correct diagnosis for staging and surgical intervention planning.
**Background**

Tumors that arise from the inferior vena cava are extremely rare, with metastatic involvement being the most frequent. When primary, the most common tumors are leiomyosarcoma and leiomyoma (75% and 15% of tumors arising from large veins, respectively). Malignant involvement is more often attributable to direct endovascular extension and/or intraluminal thromboembolization. Cardiac extension is a characteristic that changes staging, treatment and prognosis.

We suggest the following approach to IVC masses:

The **first step** is to differentiate between bland thrombus (clot) and tumor thrombus

- **Tumor thrombus** suggested by:
  1. Expansion of lumen by the thrombus.
  2. Enhancement of filling defect.
  3. Direct continuity between tumor in another organ and thrombus on CT. see **Fig 1**.

It should be remembered that malignancy predisposes to thrombosis due to hypercoagulability, so a bland thrombus may coexist with malignant thrombus into inferior vena cava.

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:**

96 Normal 0 21 false false false ES-CO JA X-NONE

- 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.

**Leiomyosarcoma**

It represents less than 1% of malignant tumors, being the most common primary malignant tumor of the inferior vena cava.

First described in 1871, it mainly affects women between 40-60 years.

This tumor originates from smooth muscle cells and can be classified into 3 groups according to their localization in the inferior vena cava:

- **Upper segment:** From the hepatic vein to the right atrium
- **Middle segment:** From the renal veins to the hepatic vein
- **Lower segment:** Infrarenal area

The majority located in the middle and lower third of the inferior vena cava, those in the upper third are very rare and usually unresectable.

This tumor can exhibit different growth patterns

- Extraluminal (62%)
- Intraluminal (5%)
- Combined (33%)

Clinical manifestation depends on the involved area: (Budd Chiari syndrome, Nephrotic syndrome, Lower extremity edema) or it can present with nonspecific symptoms (abdominal pain, weight loss, mass, nausea, changes in defecation habits, back pain, frequent urination).
The tumor has slow growing, it tends to expand along the least resistant tissue rather than invade other organs. It may extend into the right atrium and, rarely, into the pulmonary artery.

Most patients who survive primary tumor eventually develop metastases, common sites of metastases are the liver, lungs, peritoneum and brain, but can also be found in the skin, bones, omentum and kidneys.

Leiomyosarcomas with extravascular components are difficult to differentiate from retroperitoneal tumors that compress or invade the inferior vena cava. If intraluminal it can be difficult to detect without the use of contrast material. **Fig 2**

**CT findings**

A polypoid lesion or nodular mass firmly attached to the vessel wall can be seen if it is completely intraluminal.

It is usually lobulated, and well defined and can exhibit signs of hemorrhage and necrosis (cystic mass).

It can present as an increased diameter of the inferior vena cava with varying degrees of obstruction and usually seen as an heterogeneous mass with intermediate density and irregular enhancement after administration of contrast media. Calcification is unusual. **Fig 3**

**Intravenous leiomyoma**

It is the most common benign primary tumor of the inferior vena cava

It can originate in the uterine veins or by invasion from uterine fibroid with extension to the inferior vena cava.

Typical of women of reproductive age. There are fewer than 150 cases of intravenous leiomyomatosis described in the literature, with cardiac involvement only seen in 10-40% of cases.

CT may demonstrate continuity in intraluminal tumor growth from the pelvic veins into inferior vena cava or even into the right atrium.
It cannot be differentiated from leiomyosarcoma unless it has progressed and infiltrate or invade abdominal viscera.

Tumors recur in as many as 30% of cases, so it requires long term follow up. Fig 4.

**Renal cell carcinoma**

Renal cell carcinoma is the most common malignancy that extends into the inferior vena cava, it corresponds to 2% of cancers in adults.

It presents expansive growth and can invade the pyelocalicial system and the vessels of the renal hilium, so invasion to venous structures such as the inferior vena cava occurs in 4 to 10% of cases.

Usually it’s an incidental finding, patients are asymptomatic at the time of diagnosis of venous invasion.

CT has a 96% accuracy in determine the extension of renal cell carcinoma into the inferior vena cava. Fig 5.

**Hepatocellular carcinoma**

Usually invades the portal venous system, but it can invade the suprahepatic veins, the inferior vena cava and even the right atrium in up to 5.9% of patients.

It can present with Budd Chiari syndrome.

Characteristic finding on CT is a filling endoluminal defect in the inferior vena cava with dilatation of the involved veins and enhancement after contrast administration. Fig 6.

**Adrenal cortical carcinoma**

Prevalence of 0.5-2 cases per million people with bimodal peak of presentation in the 1st and 4th decades of life.
It is identified early in childhood by being hormonally active (26-94%) (Cushing, Feminization, Virilization, HTA). Its hormonal activity is inversely proportional to its size.

Other symptoms include pain, GI discomfort or palpable mass.

Typically large, most > 6cm (3-25cm), they usually present with necrosis.

They present heterogeneous enhancement (mainly peripheral) with common calcifications (19-33%).

Up to 30% of cases have intravascular extension into the inferior vena cava, being intravascular extension more common in right sided tumors and in those larger than 9cm.

This tumor is more aggressive in adults with approximately 50% with advanced disease at presentation. **Fig 7.**

**Impact of imaging on surgical planning**

- Imaging can play a role in planning surgery for resection of an IVC tumor.
- According to TNM classification of RCC, tumor spread into infradiaphragmatic IVC is T3c stage, while extension in supradiaphragmatic IVC is T4b stage. Also, sub classification into infrahepatic, hepatic and suprahepatic extension can further help the surgeon.
- Surgical resection is the only prospect of cure in leiomyosarcoma, which can be attempted in cases with involvement of the middle and lower inferior vena cava.
- Thrombus extension into the supradiaphragmatic IVC requires cardiopulmonary bypass surgery therefore it is important to identify superior extension of tumor prior to surgery with CT, MRI or intraoperative ultrasound.
- Tumor invasion from renal cell carcinoma of the vena cava does not contraindicate surgical resection.
- Invasion of the inferior vena cava can be considered an exclusion criterion for surgical resection of hepatocellular carcinoma.
- If tumor invades vessel wall, segmental resection of IVC is necessary.
- Involvement of IVC at confluence of hepatic veins would likely necessitate partial liver resection.
Fig. 1

Fig. 2: Coronal CT image shows extraluminal extension of a heterogeneous mass (arrow) and intraluminal tumor (arrowhead).

**Fig. 3:** Axial CT image shows expansion of the IVC lumen by a heterogeneously contrast enhancing partially necrotic mass (arrow).

**Fig. 4:** Coronal reconstructed CT image shows an hypoattenuating tubular-shaped mass (curved arrow) originating in the right side of the pelvis within the lumen of the ovarian vein and demonstrating a 30-cm-long contiguous extension into the proximal IVC and right atrium (arrowhead). The distal IVC (straight arrow) and iliac veins are not involved. An incidental upper pole left renal cyst (star) is seen.

© From Low Gavin, Rouget Adrien, Crawley Cinzia. Intravenous Leiomyomatosis with Intracaval and Intracardiac Involvement. Radiology 2012; 265:971-975
**Fig. 5:** Coronal CT image show a right kidney mass with invasion to the inferior vena cava and extension to the right atrium


---

**Fig. 6:** Axial and coronal CT shows an heterogeneous hepatic mass that invades the hepatic vein and the IVC

**Fig. 7:** Coronal contrast-enhanced CT image show a large left-sided adrenal cortical carcinoma with extension of the tumor thrombus into the IVC.

Findings and procedure details

We described four Fundación Clínica Shaio cases, all evaluated with volumetric acquisitions in multiplanar images, using a MDCT (320 channels) after the injection of 75 cc of iodinated nonionic contrast media.

We found two cases of clear cell renal cell carcinoma, a leiomyosarcoma arising from the abdomen and a leiomyoma arising from the vessel wall, all cases were biopsy proven.

**CASE 1: Leiomyosarcoma**

47 years old male with two month history of epigastric pain and weight loss. **Fig 8 and 9.**

**CASE 2: Leiomyoma**

96 Normal 0 21 false false false ES-CO JA X-NONE

44 years old female with one month of worsening chest pain and dyspnea. **Fig 10 and 11.**

**CASE 3: Clear cell renal cell carcinoma**

96 Normal 0 21 false false false ES-CO JA X-NONE

48 years old male with five days of epigastric pain irradiated to the thorax and upper limbs. **Fig 12 and 13.**

**CASE 4: Clear cell renal cell carcinoma**

56 years old male with four-month history of worsening thoracic pain, dyspnea, nausea and vomiting. **Fig 14.**

96 Normal 0 21 false false false ES-CO JA X-NONE
**Fig. 8:** Leiomyosarcoma. CT performed in Toshiba Aquilion One (320 channels multislice CT), A, B & C. Contrast enhanced CT in coronal, sagittal and axial planes. arising anterior to the left kidney there is a soft tissue density mass with mixed enhancement invading the left renal vein and extending into the inferior vena cava all the way up into the right atrium.
Fig. 9: Leiomyosarcoma. CT performed in Toshiba Aquilion One (320 channels multislice CT), A, B & C. Contrast enhanced CT in coronal, sagittal and axial planes. arising anterior to the left kidney there is a soft tissue density mass with mixed enhancement invading the left renal vein and extending into the inferior vena cava all the way up into the right atrium.

© Universidad de la Sabana, Clínica Abood Shaio - Bogota/CO
**Fig. 10:** Leiomyoma. CT performed in Toshiba Aquilion One (320 channels multislice CT), A, B & C. Contrast enhanced CT in coronal, sagittal and axial planes, arising from the inferior vena cava there is a soft tissue density mass with mixed enhancement extending into the right atrium and right ventricle. Cardiomegaly is also noted. D. Macroscopic specimen obtained after surgery.

© Universidad de la Sabana, Clínica Abood Shaio - Bogota/CO

**Fig. 11:** Leiomyoma. CT performed in Toshiba Aquilion One (320 channels multislice CT), A, B & C. Contrast enhanced CT in coronal, sagittal and axial planes, arising from the inferior vena cava there is a soft tissue density mass with mixed enhancement extending into the right atrium and right ventricle. Cardiomegaly is also noted. D. Macroscopic specimen obtained after surgery.

© Universidad de la Sabana, Clínica Abood Shaio - Bogota/CO
**Fig. 12:** Renal cell carcinoma. CT performed in Toshiba Aquilion One (320 channels multislice CT), A, B & C. Contrast enhanced CT of the upper abdomen in coronal, sagittal and axial planes showing heterogeneous enhancement and enlargement of the left kidney by a diffuse infiltrating mass that extends into the left renal vein and the inferior vena cava, note the expansion of the vessel lumen.

© Universidad de la Sabana, Clínica Abood Shaio - Bogota/CO
Fig. 13: Renal cell carcinoma. CT performed in Toshiba Aquilion One (320 channels multislice CT), A, B & C. Contrast enhanced CT of the upper abdomen in coronal, sagittal and axial planes showing heterogeneous enhancement and enlargement of the left kidney by a diffuse infiltrating mass that extends into the left renal vein and the inferior vena cava, note the expansion of the vessel lumen.

© Universidad de la Sabana, Clínica Abood Shaio - Bogota/CO

Fig. 14: Renal cell carcinoma. CT performed in Toshiba Aquilion One (320 channels multislice CT), A & B. Contrast enhanced CT of the upper abdomen in coronal and axial planes showing enlargement and loss normal anatomy of the right kidney by a large diffuse infiltrating mass with heterogeneous enhancement that extends into the right renal vein and the inferior vena cava.

© Universidad de la Sabana, Clínica Abood Shaio - Bogota/CO
Conclusion

Based on imaging features, the radiologist should be able to differentiate between a tumor thrombus and a bland thrombus, suggest the extension, and determinate the primary or secondary origin of the lesion if it is possible as this information has a direct impact on staging and management.


