

# LEIOMYOSARCOMA OF INFERIOR VENA CAVA

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# LEARNING OBJECTIVES

- Describe the epidemiology and clinical manifestations of this pathology.
- Describe the imaging findings, focusing on CT.
- Explain the principle entities that can invade or infiltrate the inferior vena cava (IVC).

# BACKGROUND

- Affection of IVC can be due to both benign and malignant conditions.
- Tumours of the IVC can be either primary or secondary in origin.
- Primary tumours originate from within the vessel wall, while secondary tumours originate in the adjacent tissue and surround, compress or invade the IVC.
- Primary vascular leiomyosarcoma (LMS) is a rare tumour that originates from the smooth muscle cells of the tunica media and accounts for 2% of all LMS, the most common site being the inferior vena cava. It is more common during the sixth decade of life, more frequently in women. Although rare, LMS is the most common primary malignancy that affects the IVC.

# BACKGROUND: Classification

- Keiffer et al. classified IVC LMS into three groups according to its location:
  - Infrarenal (between the confluence of the common iliac and renal veins).
  - Suprarenal (between the renal and hepatic veins).
  - Suprahepatic (between the hepatic veins and the right atrium).
  - The most common site is the middle section, followed by the lower and the upper section.
- Three main growth patterns can be identified:
  - Extraluminal: in two thirds of the cases, simulating the invasion of the IVC from a retroperitoneal mass.
  - Intraluminal: the least frequent growth pattern.
  - Both extra and intra luminal.

# BACKGROUND: Symptoms

- The location and size of the tumour determines the symptoms of the patient and its posterior management.
- As LMS is a slow-growing progressive tumour, many individuals are asymptomatic at first, and up to 10.5% of patients are diagnosed incidentally.
- Patients with tumours in the lower portion of the inferior vena cava may have lower back pain and generalized swelling.
- If the tumour is located in the middle portion of the vein, patients may have a nephrotic syndrome due to the occlusion of the renal vein.
- In cases with upper segment affection, the patients may develop a Budd Chiari Syndrome.
- Non-specific symptoms due to compression, such as palpable abdominal mass, abdominal pain, lower limb edema, thrombosis and venous stasis, may be seen.

# BACKGROUND: Treatment

- LMS has an extremely poor prognosis due to its late presentation.
- When diagnosed early, it has a better prognosis than other tumours, with surgical treatment and complete resection of the vein being the treatment of choice in these patients, as opposed to the radiotherapy and chemotherapy given to other tumours.
- Tumours affecting the upper segment are unresectable and patients often undergo bypass surgery to preserve circulation.
- Adjuvant treatments are reserved for patients with locally advanced, metastatic or recurrent disease.

# IMAGING FINDINGS

- LMS is often found incidentally on routine imaging exams.
- The most common sign is the lack of visibility of the venous structure, followed by the presence of an intraluminal thrombus inside a dilated vessel.
- Ultrasound can demonstrate the dilation of the IVC (Fig.1) as well as the vascularity of the tumour when using Doppler ultrasonography. (Fig.2).
- CT plays an important role in the diagnosis and follow-up of these patients (Fig.3 and Fig.4). It is the imaging modality of choice for detecting the origin and extension of the tumour, evaluating the presence of metastases and in helping the preoperative surgical planning. Metastases occur at late stages of the disease, most frequently in the liver, lungs and lymph nodes.
- MRI can also be practiced, especially in cases where CT is contraindicated and above all in patients with iodine contrast allergy.

Fig.1. 54 year old man with mental retardation arrives in the emergency room with severe limb pain. Doppler US discards deep venous thrombosis. After 24h under antibiotic treatment for cellulitis, right flank pain starts and US is performed to discard nephritic colic. US demonstrates right hydronephrosis. Dilated IVC and an heterogeneous mass with intraluminal and extraluminal component is seen.

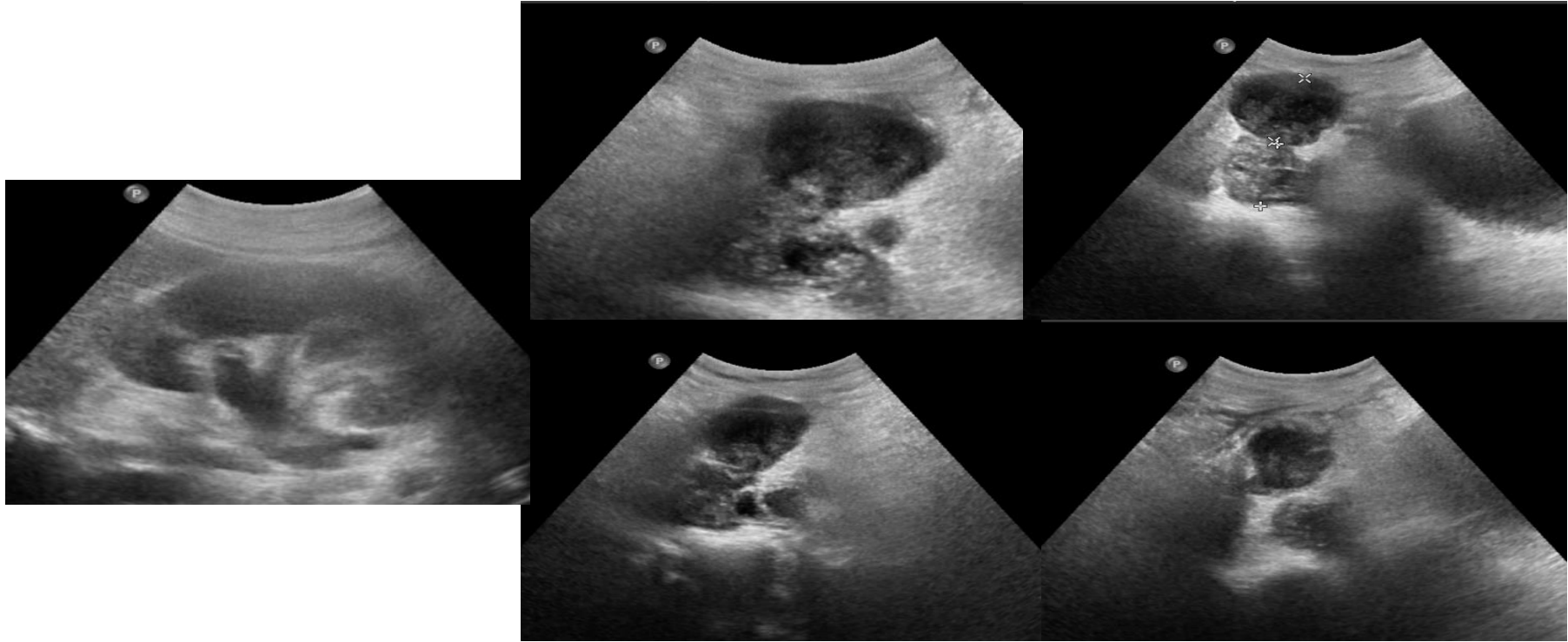




Fig.2. 45 year old woman describes a 3 month history of dyspnea and oppressive sensation which she associated with stress. On physical examination, a mass was palpable in the right flank. Abdominal US demonstrates a heterogeneous mass in the right flank, with increased Doppler signal in the IVC location and lack of visibility of the vascular structure.



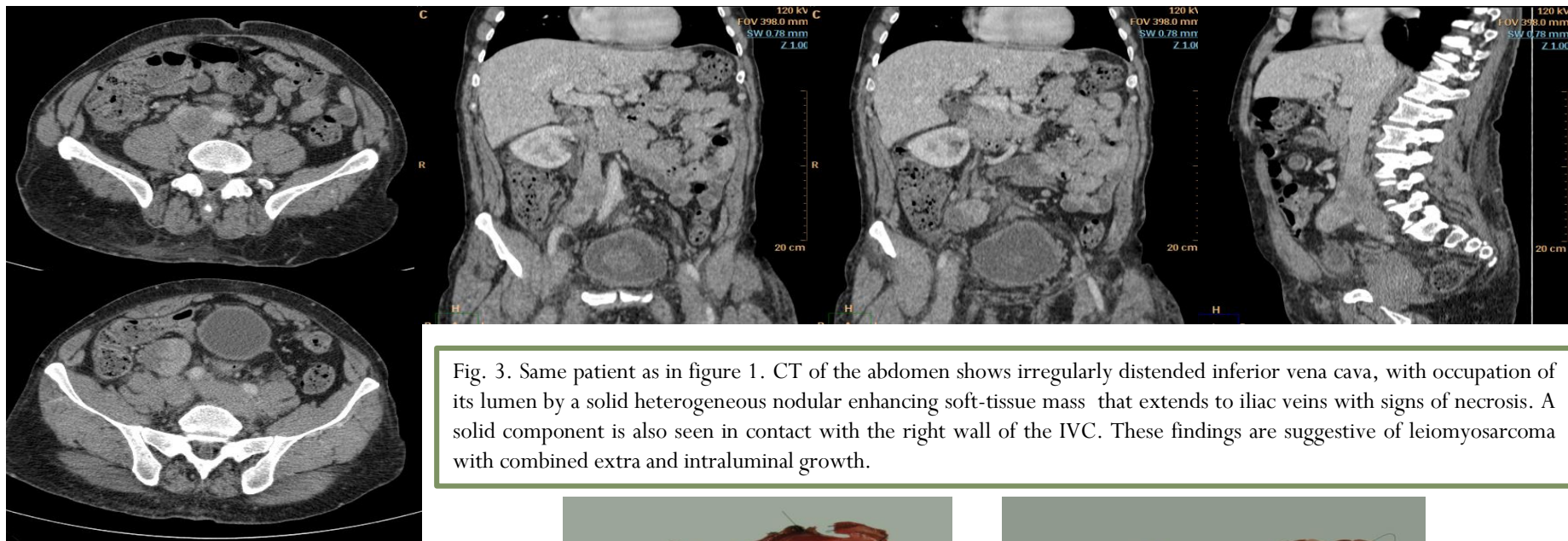
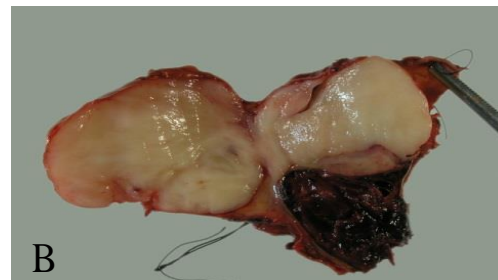
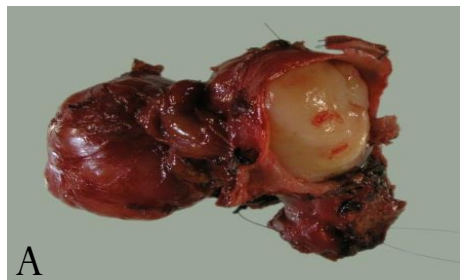
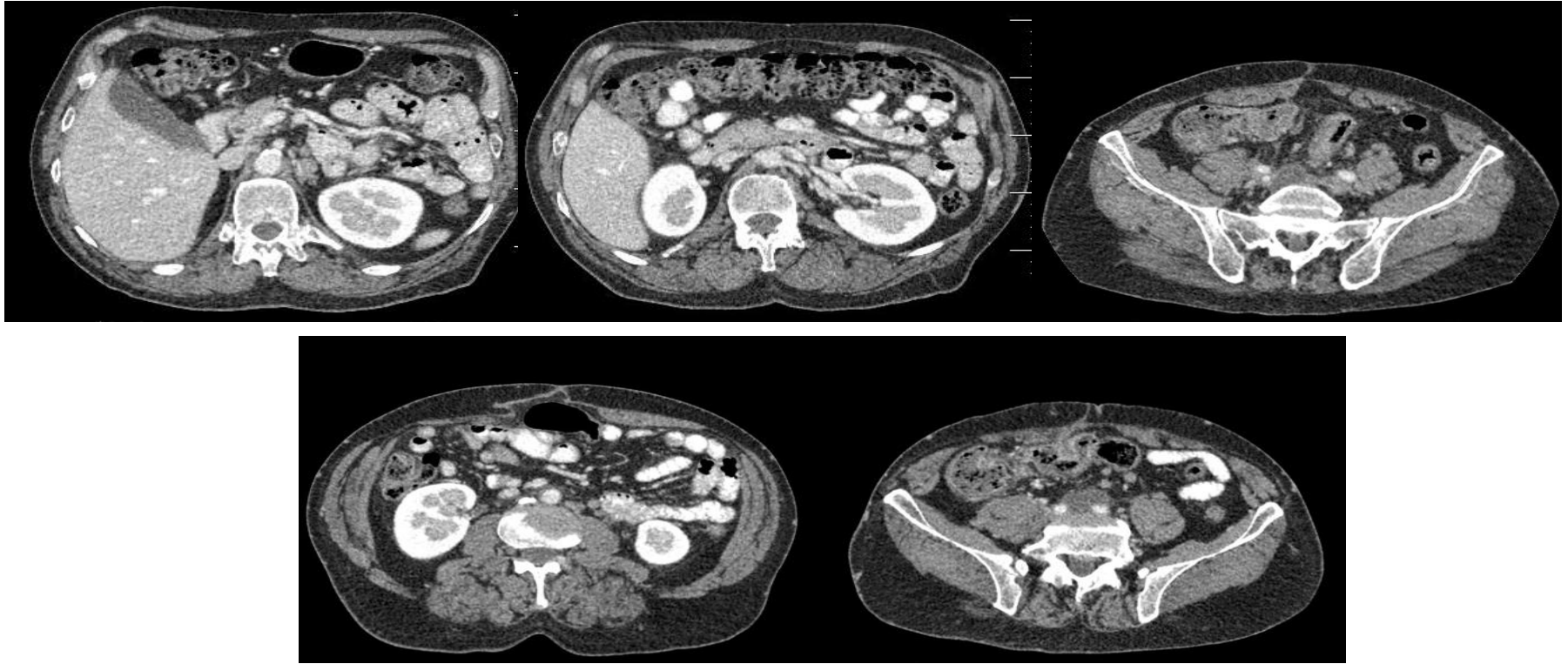


Fig. 3. Same patient as in figure 1. CT of the abdomen shows irregularly distended inferior vena cava, with occupation of its lumen by a solid heterogeneous nodule enhancing soft-tissue mass that extends to iliac veins with signs of necrosis. A solid component is also seen in contact with the right wall of the IVC. These findings are suggestive of leiomyosarcoma with combined extra and intraluminal growth.



Macroscopic appearance of the tumour after resection. Picture A shows the tumour growing inside the IVC. Picture B corresponds to a cross section through the tumour, showing the intraluminal component as well as the extraluminal component.

Fig.4. CT scan 1 year after surgery (infrarenal cavectomy) in previous patient (Fig.1 and 3). Follow-up CT demonstrates postoperative small fluid collection in the surgical bed, without tumoral component. Hydronephrosis resolution is also seen.



# IMAGING FINDINGS

- CT normally demonstrates an irregular distended inferior vena cava, with an occupation of its lumen by a solid heterogeneous nodular enhancing soft-tissue mass. (Fig.5).
- A central low attenuation representing necrosis is a typical finding and haemorrhage is common.
- Calcifications are extremely rare.
- LMS can have a retroperitoneal origin and affect the inferior vena cava secondarily. (Fig.6).

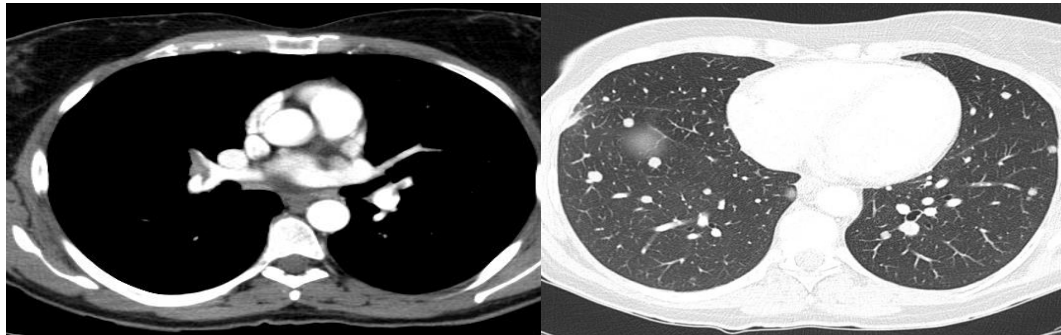


Fig.5. Same patient as in Figure 2.

CT of the abdomen shows the entire inferior vena cava distended and occupied by a heterogenous enhancing soft tissue mass with extraluminal growth.

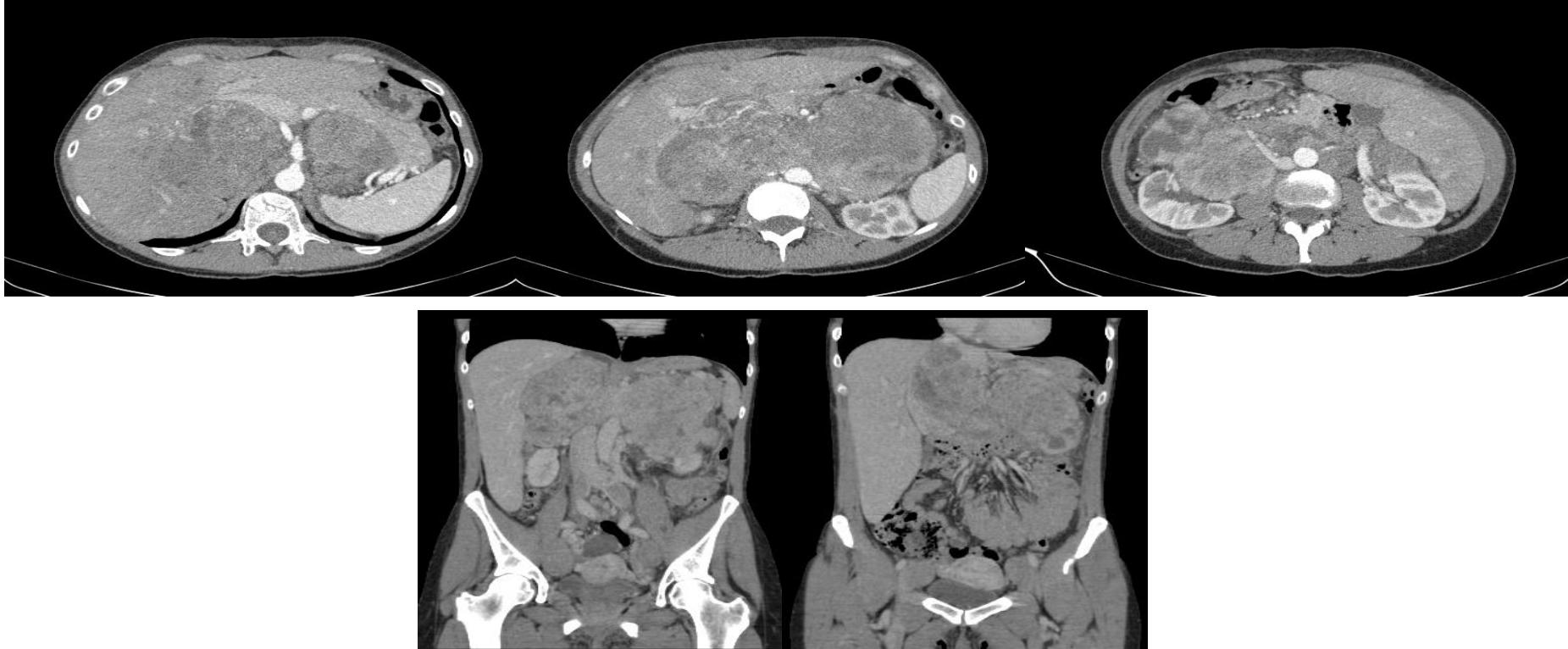
CT of the chest shows intraluminal filling defects in relation to pulmonary embolism. Multiple bilateral pulmonary nodules are seen, due to pulmonary metastases.

Biopsy confirmed leiomyosarcoma.



Fig.6. 45 year old woman with epigastric pain and solid hypoechoic mass seen in US study.

CT scan shows a great solid heterogeneous lobulated mass in the retroperitoneal space. The differential diagnosis should include: sarcoma (leiomyosarcoma with both intra and extravascular involvement of the IVC, angiosarcoma), desmoid tumour, lymphoma and perivascular epithelioid cell tumours (PEComa). Biopsy of the mass confirmed retroperitoneal leiomyosarcoma.



# IMAGING FINDINGS:

## Differential Diagnosis

- The differential diagnosis is wide and must include:
  - benign diseases such as venous thrombosis and flux artefacts.
  - neoplastic invasion from nearby organs: pheochromocytoma, renal cell carcinoma, adrenal carcinoma and hepatocellular carcinoma.(Fig. 7,8,9 and 10).

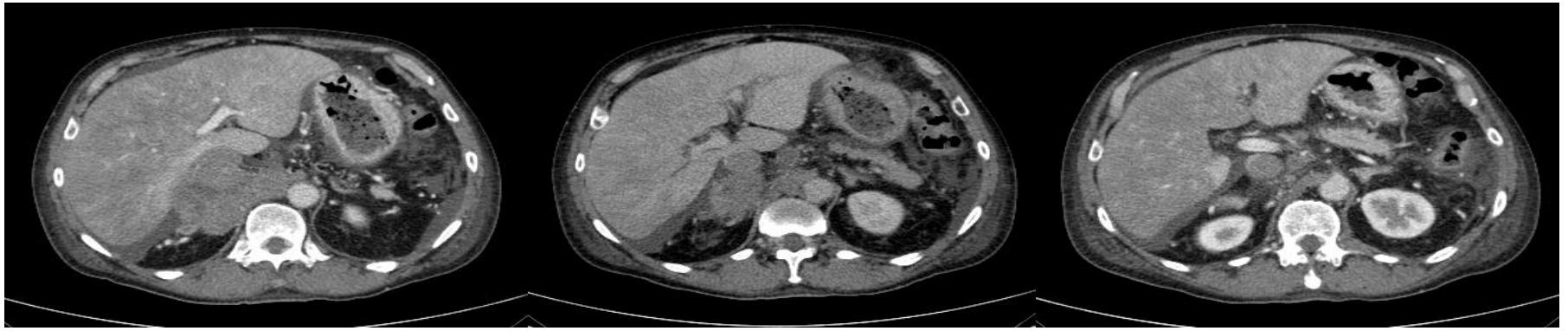


Fig.7. 74 year old male with dyspnea and pulmonary nodules in chest radiograph. Body CT showed a voluminous mass in the right adrenal fossa, with liver and IVC infiltration. Autopsy confirmed adrenal carcinoma.



Fig.8. 59 year old man with squamous cell lung cancer. CT shows heterogeneous retroperitoneal mass affecting the right adrenal, the hepatic segment VI, the IVC and the upper renal pole in relation to adrenal metastases.



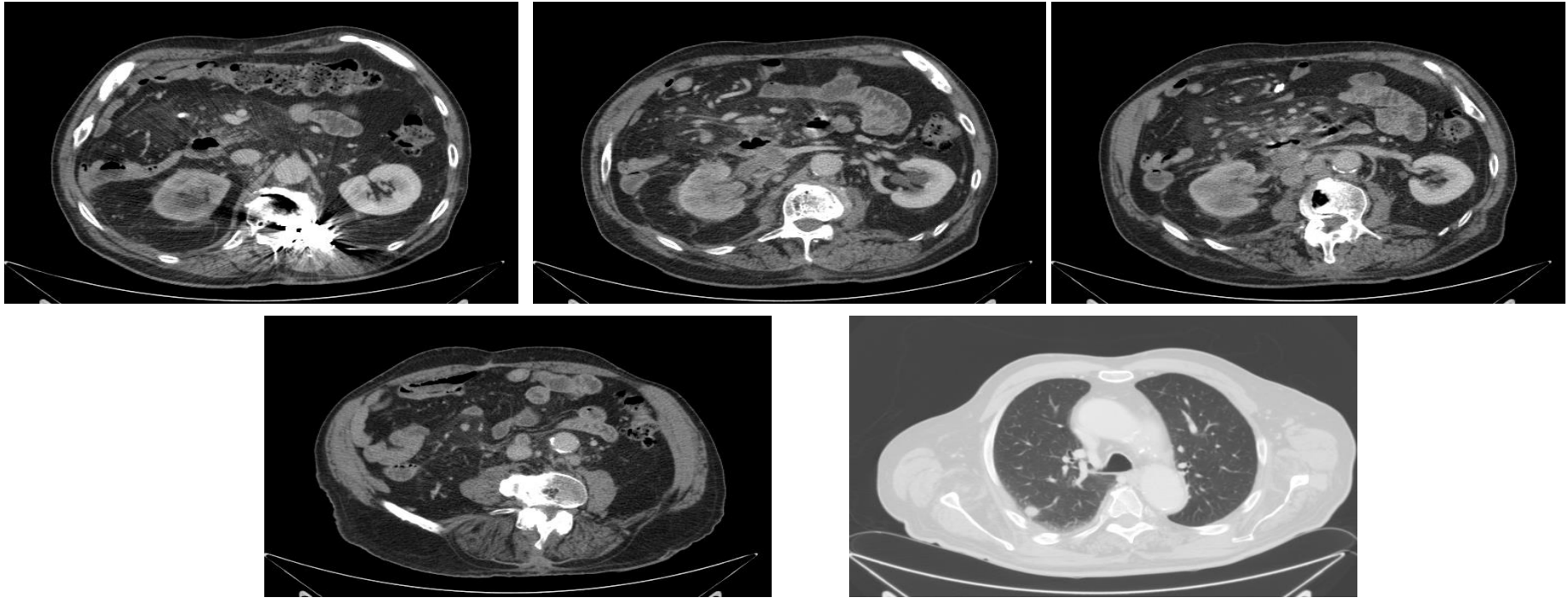


Fig.9. 84 year old man with right colon cancer and hematuria. CT shows an infiltrative lesion in the right kidney extending into the renal hilum with affection of the excretory urinary tract and the renal vein reaching the IVC. Multiple enlarged lymph nodes and pulmonary metastases are also seen.

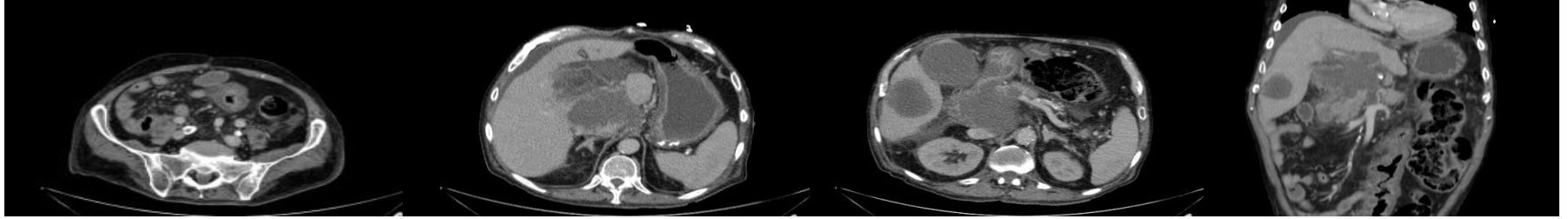


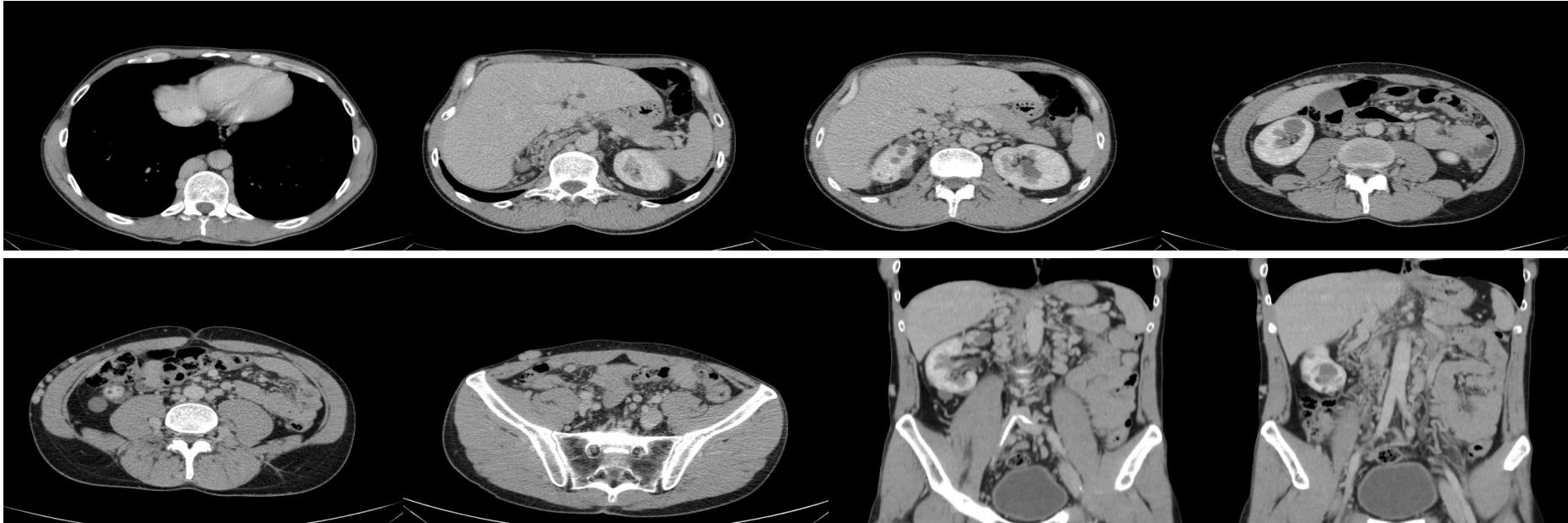
Fig.10. Hepatic hilar hypodense mass that infiltrates liver, pancreatic head and duodenum, with severe thrombosis of portal vein, superior mesenteric vein and partial inferior vena cava thrombosis. Intestinal malrotation with a concentric wall thickening of the cecum and ascendent colon is also seen. Colon neoplasm and hepatic hilar M1 was confirmed.

# IMAGING FINDINGS:

## Differential Diagnosis: Benign thrombosis

- Benign thrombosis is the principle cause of IVC obstruction.
- Frequently, it originates in the lower extremities or pelvis in association with dehydration, immobilization, coagulopathy, pelvis inflammatory disease or congestive cardiac failure.
- US is normally enough to depict lower limb thromboses, but CT is necessary to study abdominal and pelvis vessels.
- CT will demonstrate occupation of the vessel's lumen by a soft tissue mass with regular margins and no enhancement after contrast administration, while tumoral thrombosis will have irregular margins and enhance after contrast administration, behaving as the primary tumour.
- Sometimes chronic thrombi may calcify and can be seen as hyperdense linear structures inside the vein on non contrast CT scans.(Fig.11).

Fig.11. CT scan practised on a 36 year old man with trombophilia. Reduced diameter of the IVC was seen, with correct permeability of both iliac veins which present a “Y” shape hyperdense structure in their proximal segment suggestive of calcification of a laminate chronic thrombosis. Multiple collateral vessels and dilated azygos and hemiazygos veins are also seen.



# IMAGING FINDINGS:

## Differential Diagnosis: Flux Artefacts

- It's important to bear in mind flux artefacts in order to avoid more invasive procedures. (Fig.12).
- On early CT contrast scans, a hypodense intraluminal defect is usually seen in the IVC where the renal veins drain, disappearing in venous acquisition scans due to contrast homogenization.
- This phenomenon is due to mixed contrast opacified and non-opacified blood and can be confused with thrombi.
- It is a feature of early phase dynamic scanning.
- Later scan times allow contrast enhanced blood to enter the lower limbus, mixing with the renal vein blood.

A



B



Fig.12. Intraluminal filling defect of IVC on “A” images (CT in pancreatographic phase) with complete resolution on “B” images (CT in portal phase) in a study for pancreatitis. This phenomenon is due to mixed contrast opacified and non opacified blood and can be confused with thrombus. It is a feature of early phase dynamic scanning. Later scan times allow contrast enhanced blood to enter the lower limb mixing with the renal vein blood.

# IMAGING FINDINGS:

## Differential Diagnosis: Leiomyoma

- Leiomyoma is the most frequent benign tumour affecting the IVC.
- Normally in women of reproductive age.
- It can originate from the uterine veins or be due to invasion from a uterine fibroid extending to the IVC.

# CONCLUSION

- Radiologists must be familiar with the different entities that can affect the IVC.
- CT is the imaging modality of choice and allows a correct interpretation, facilitating the choice of treatment.