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Retroperitoneal masses:

A diagnostic approach

5-8 June 2019, Rome, Italy

Learning Objectives

The purpose of this poster is:

- ✓ Review the anatomy of the retroperitoneal space;
- ✓ Review the imaging features of the main groups of retroperitoneal masses, with emphasis on computed tomography (CT) and magnetic resonance (MR) findings;
- ✓ To give diagnostic clues for the correct differential diagnosis of these entities.

Background

The retroperitoneal space

- Part of the abdominal cavity that lies between the posterior parietal peritoneum, anterior to the transversalis fascia and extends from the diaphragm superiorly to continue into the extraperitoneal space in the pelvis inferiorly.

- Divided into three spaces by the **perirenal fascia:**

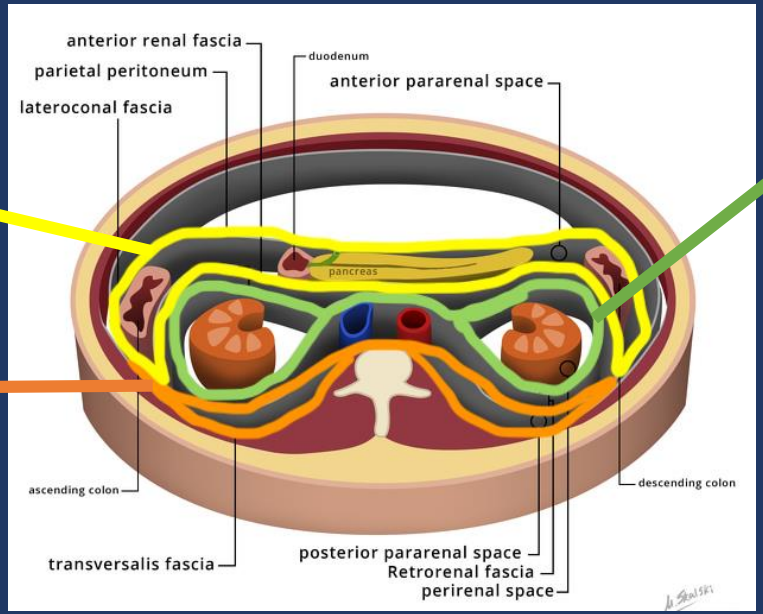
1. **Anterior pararenal space – p**ancreas, ascending and descending colon, duodenum
2. **Perirenal space - a**drenal glands, kidneys, renal vessels.
3. **Posterior pararenal space -** fat

Background

The retroperitoneal space

Anterior pararenal space

Posterior pararenal space



Perirenal space

The anterior and posterior pararenal spaces merge inferior to the level of the kidneys, which communicates inferiorly with the prevesical space and extraperitoneal compartments of the pelvis.

Fig. 1 – The retroperitoneal compartments.
Source: Adapted from www.radiopaedia.com (on 30/03/2019)

Background

Retroperitoneal masses – Diagnostic approach

STEP 1: **D**etermine if the tumor is located within the retroperitoneal space.

STEP 2: Identify the organ of origin.

STEP 3: Specific Patterns of Spread.

STEP 4: Characteristic Tumor Components.

STEP 5: Vascularity.

Background

Retroperitoneal masses – Diagnostic approach

STEP 1: Determine if the tumor is located within the retroperitoneal space.

Displacement of normal anatomic structures of the retroperitoneum

Anterior displacement of retroperitoneal organs as well as the great vessels strongly suggests that the tumor arises in the retroperitoneum.

Background

Retroperitoneal masses – Diagnostic approach

STEP 2: Identify the organ of origin.

Before a tumor can be described as primarily retroperitoneal, the possibility that the tumor originates from a retroperitoneal organ must be excluded!

Primary

Arise within the **retroperitoneal space** but **outside the major organs in this space**

vs.

Secondary

Origin in **retroperitoneal major organ** or **metastatic disease from a extraperitoneal primary tumor**

STEP 3: Specific Patterns of Spread

STEP 4: Characteristic Tumor Components

STEP 5: Vascularity

Background

Primary Retroperitoneal masses

- 75% of primary retroperitoneal tumors are histologically confirmed malignancies.
- Malignant tumours of the retroperitoneum occur four times more frequently than benign lesions.
- Sarcomas comprise 1/3 of this lesions.

Malign

1. **Lymphoma – 33%**
2. **Mesodermal system – 30%:**
   - Lipossarcoma
   - Leiomyosarcoma
   - Malignant fibrous histiocytoma
3. **Nervous system**
4. **Germ cell tumors**

80% of primary retroperitoneal sarcomas

Benign

1. **Neurogenic tumours:**
   - Schwannomas
   - Neurofibromas
2. **Paragangliomas**
3. **Retroperitoneal fibrosis**
4. **Retroperitoneal lymphangioma**
5. **Castleman Disease**
6. **Lipomas and angiomyolipomas**

Imaging Findings

Primary Retroperitoneal masses - Malignant LYMPHOMA

- Lymphoma is the **most common retroperitoneal malignancy**, accounting for 33% of all primary retroperitoneal masses.
- Hodgkin lymphoma involves the spleen and mediastinum, with involvement of para-aortic nodal groups in 25% of cases.
- Non -Hodgkin lymphoma (NHL) involves extranodal sites as well as discontinuous nodal groups, with para-aortic nodes affected in 55% of cases.

Characteristic Imaging Findings:

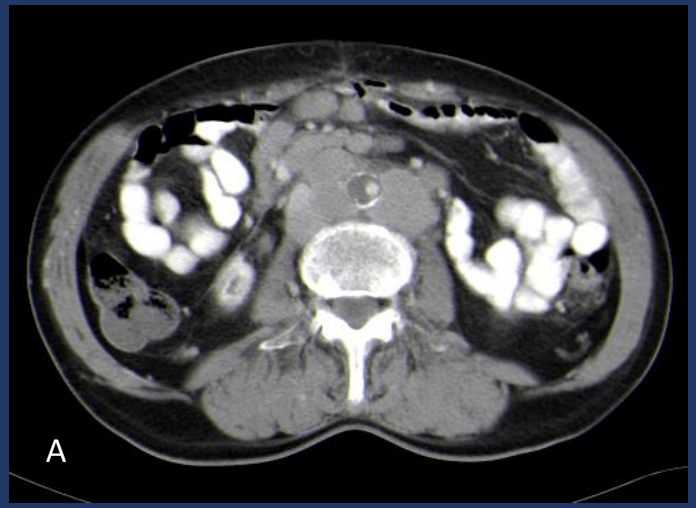
CT:
- Well- defined para-aortic or pelvic masses, which demonstrate homogeneous attenuation on non-contrast CT
- Tend to be hypovascular and demonstrate mild **homogeneous enhancement** on contrast-enhanced CT
- 23% of NHL enhance heterogeneously
- Infiltrative aspect and often appear as a mantle-like mass around the aorta or IVC, **extending between and encasing structures without compressing them**
- Vascular invasion and thrombosis are rare

MRI:
- **T1:** isointense to muscle
- **T2:** iso- or hyperintense
- **T1+Gd:** moderately homogeneous or patchy enhancement

Imaging Findings

Primary Retroperitoneal masses - Malignant

LYMPHOMA



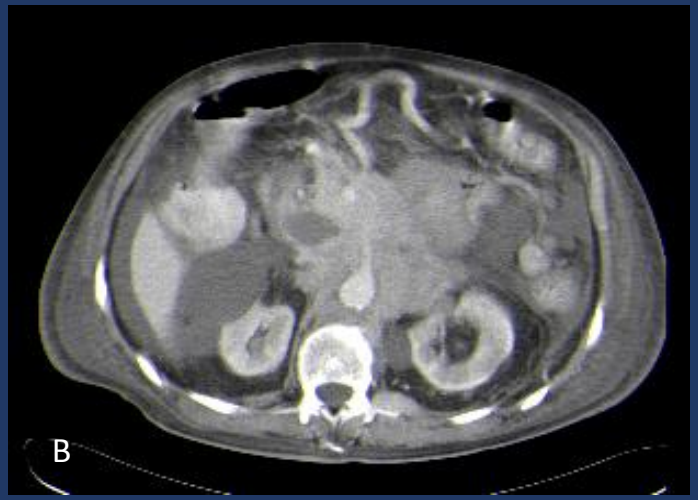


Fig. 2 – Lymphoma – Axial contrast-enhanced CT reconstrutions (A and B) demonstrates enlarged lymph nodes involving the aorta, IVC (A) and the mesenteric vessels (B) in two diferent patients with lymphoma.
Source: Radiology department, Hospital do Divino Espirito Santo de Ponta Delgada, EPE

Imaging Findings

Primary Retroperitoneal masses - Malignant

MESODERMAL ORIGIN - LIPOSARCOMA

- Most common primary retroperitoneal sarcoma - 40% of cases.
- 4 histologic subtypes with diferente imaging features.

Characteristic Imaging Findings:

- **Well-differentiated (low-grade tumors)** - macroscopic fat comprising >75% of the tumor; smooth margins and lobular contours; septae >3 mm in thickness and mild to moderate contrast enhancement.
- **De-differentiated (high-grade tumors) -** non-lipomatous mass within, adjacent to, or surrounding a fatty mass, enhancing septa, calcifications (30%). No evidence of fat in up to 20%.
- **Myxoid (intermediate grade tumor)** - heterogeneous, lobular contours, internal septations; near water attenuation of the myxoid component that gradually enhance on delayed post-contrast images. No macroscopic fat in 50% of the cases.
- **Pleiomorphic (high-grade tumors) -** little or no fat heterogeneous masses, isodense to muscle on CT; commonly have areas of low attenuation representing necrosis. Calcifications are rare.

Imaging Findings

Primary Retroperitoneal masses - Malignant

MESODERMAL ORIGIN - **LIPOSARCOMA**

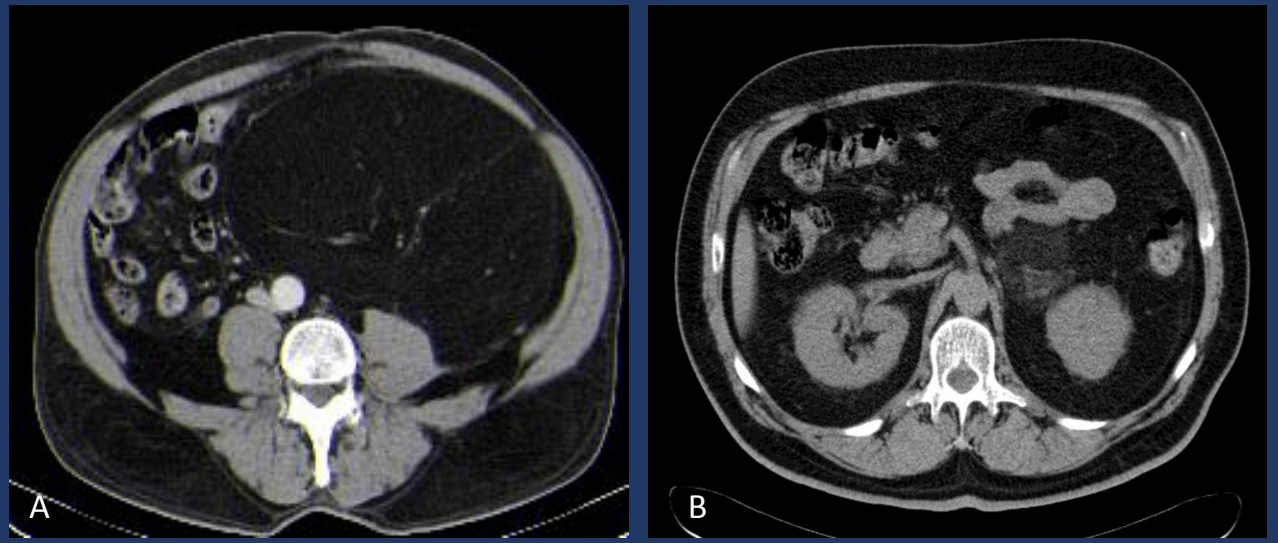


Fig. 3 - Well-differentiated liposarcoma – Axial contrast-enhanced CT reconstrutions demonstrates a well-circumscribed mass, predominantly fatty (>75%), with enhancing septations > 3mm in thickness in two diferente pacientes (A and B).

Source: Radiology department, Hospital do Divino Espirito Santo de Ponta Delgada, EPE

Imaging Findings

Primary Retroperitoneal masses - Malignant

MESODERMAL ORIGIN - **LIPOSARCOMA**

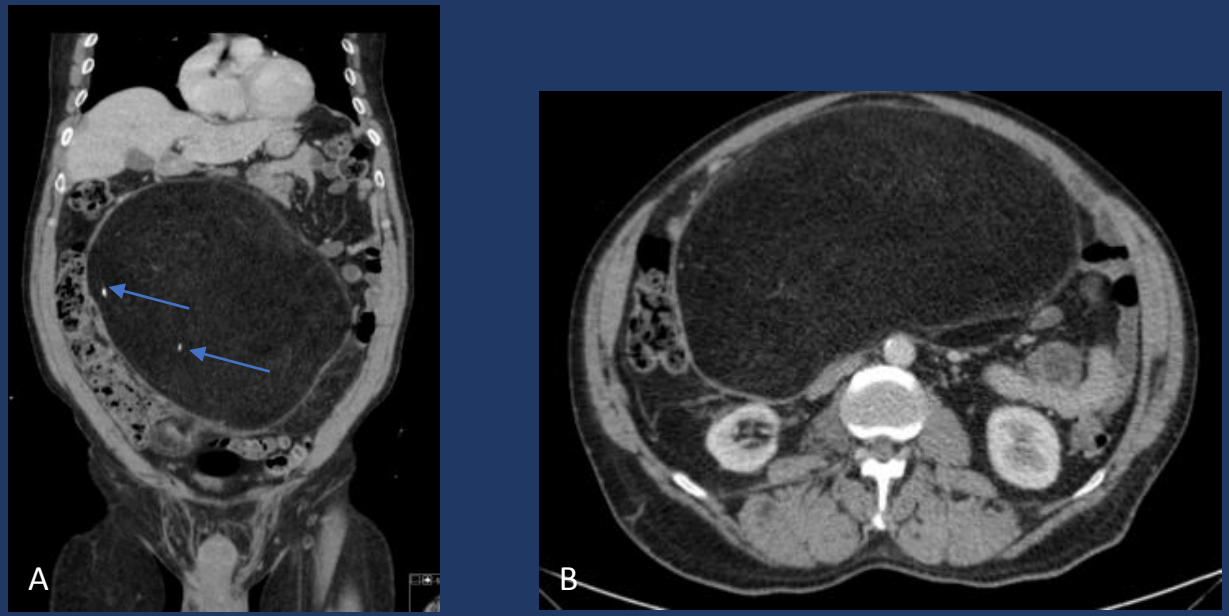


Fig. 4 - Well-differentiated liposarcoma – Coronal (A) and axial (B) contrast-enhanced CT reconstrutions showing a well-circumscribed mass, predominantly fatty (>75%), with some calcifications (arows).
Source: Radiology department, Centro Hospitalar de São João do Porto, EPE

Imaging Findings

Primary Retroperitoneal masses - Malignant

MESODERMAL ORIGIN - LEIOMYOSARCOMA

- Second most common primary retroperitoneal tumor in adults.
- Divided in three main categories: extravascular (62 %), intravascular (5 %) and a combination of both (33 %).
- Believed to arise from blood vessels, spermatic cord or wolfian and mullerian duct remnants within the retroperitoneum.

Characteristic Imaging Findings:

CT:

• Large masses with attenuation equal to or less to that of muscle
• Internal regions of low attenuation corresponding to areas of necrosis
• Areas of haemorrhage may occasionally be seen within.
• Tumors involving blood vessels such as the IVC may show polypoid intra-luminal extension with expansion of the blood vessel - the tumor thrombus typically enhances.
• **Fat and calcification are not typically present.**

MRI:

- **T1:** intermediate to low signal intensity
- **T2:** intermediate to high signal intensity
- **T1+Gd :** heterogeneous enhacement

Heterogeneous large solid mass in the retroperitoneum with necrotic areas and contiguous involvement of a vessel, is **highly suggestive of a leiomyosarcoma!**

Imaging Findings

Primary Retroperitoneal masses - Malignant

MESODERMAL ORIGIN - MALIGNANT FIBROUS HISTIOCYTOMA

- Third most common retroperitoneal sarcoma (19%) and the most common soft-tissue sarcoma in the body.
- +++ males / +++ 50–60-years.

Characteristic Imaging Findings:

CT:
- Large, infiltrating soft-tissue mass with areas of necrosis and hemorrhage and with invasion of adjacent organs
- Density typically similar to adjacent muscle
- Heterogeneous lower density areas if hemorrhage, necrosis or myxoid material is abundant
- Peripheric calcification can be seen (7%–20%)

Presence of calcification may help to distinguish malignant fibrous histiocytoma from leiomyosarcoma!

MRI:
- **T1:** intermediate signal intensity, similar to adjacent muscle; heterogeneity if hemorrhage, calcification, necrosis or myxoid material present; prominent enhancement of solid components
- **T2:** intermediate to high signal intensity; heterogeneity if hemorrhage, calcification, necrosis or myxoid material present
- **T1+Gd:** heterogeneous enhacement

Imaging Findings

Primary Retroperitoneal masses - Malignant

NEUROGENIC ORIGIN - NEUROBLASTOMA

- The vast majority arise from the adrenal gland and from the retroperitoneum in 30-35% of cases.
- Most common extracranial solid childhood malignancy.
- +++ males / +++ 1st decade of life.

Characteristic Imaging Findings:

CT:
- Heterogeneous masses with areas of necrosis witch are of low attenuation
- Calcifications in 80-90% of cases
- The mass usually insinuate itself beneath the aorta and lift it off the vertebral column
- Invasion of adjacent organs, displacing them and encasement of vessels with luminal compression
- Lymph node enlargement is often present

MRI:
- **T1:** heterogeneous and iso to hypointense
- **T2:** heterogeneous and hyperintense; cystic/necrotic areas very high intensity; signal voids may be evident due to calcifications
- **T1+Gd:** variable and heterogeneous enhancement

Imaging Findings

Primary Retroperitoneal masses - Malignant

NEUROGENIC ORIGIN - **NEUROBLASTOMA**

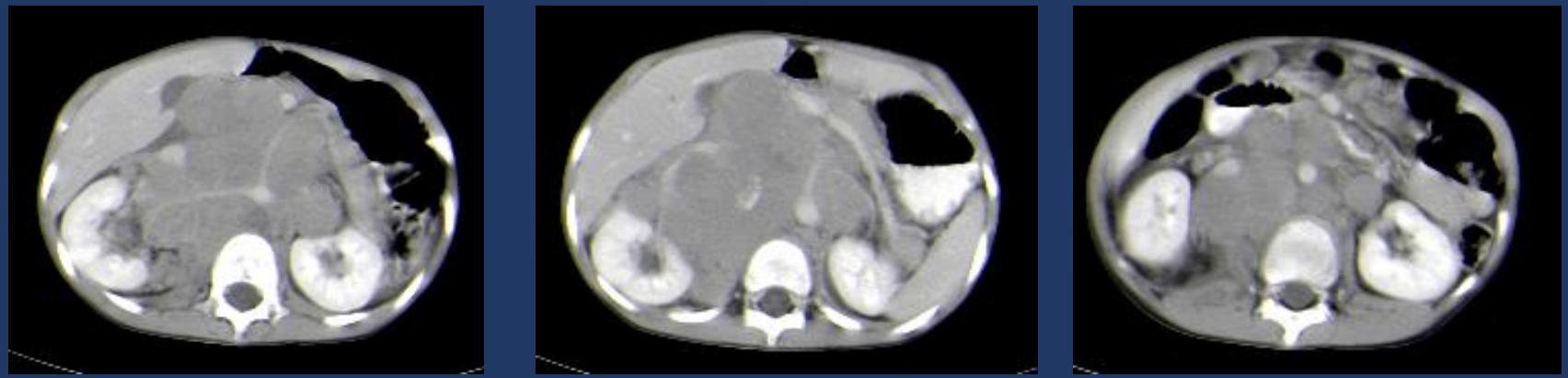


Fig. 5 – Retroperitoneal neuroblastoma – Axial contrast-enhanced CT reconstrutions demonstrates a large, lobulated, heterogeneous retroperitoneal mass that causes invasion of adjacent organs, displacing them and encasement of vessels with luminal compression.
Source: Radiology department, Hospital do Divino Espirito Santo de Ponta Delgada, EPE

Imaging Findings

Primary Retroperitoneal masses - Malignant

- Germ cell tumour.
- +++ female / +++ in children.

Characteristic Imaging Findings:

CT
- Variable appearance: solid, cystic or mixed
- Fat and calcification may be present
- The sacral or coccyx bone may be involved

MRI:
- **T1:** fat components - high signal; calcific/bony components - low signal
- **T2:** fluid components - high signal; calcific bony componentes - low signal
- **T2* GRE:** magnetic susceptibility artefact because of calcifications
- **T1 + Gd:** enhancing solid components

GERM CELL TUMORS - **SACROCOCCYGEAL TERATOMA**

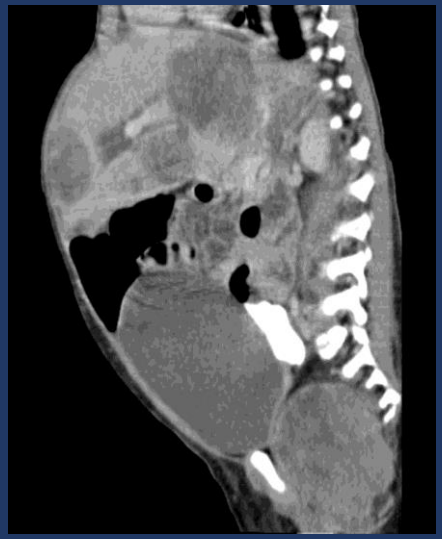


Fig. 6 – Sacrococcygeal teratoma – Sagital contrast-enhanced CT reconstrutions showing a well delimited pre-sacral heterogeneous mass
Source: Radiology department, Hospital do Divino Espirito Santo de Ponta Delgada, EPE

Imaging Findings

Primary Retroperitoneal masses - Benign

NEUROGENIC ORIGIN - SCHWANNOMA

- Benign tumor that arises from the perineural sheath of Schwann.
- 6% of retroperitoneal neoplasms.
- +++ females / +++ 20–50-years.

Characteristic Imaging Findings:

CT:
- Small schwannomas –r ound, well defined, homogeneous
- Large schwannomas - may be heterogeneous
- Calcifications may be present
- Variable homogeneous or heterogeneous enhancement

MRI:
- **T1**: cellular areas are hypointense
- **T2:** cellular areas are hypointense while cystic areas appear hyperintense
- **T1 +Gd:** Contrast enhancement is heterogeneous, with nonenhancing cystic components and enhancing solid components

Imaging Findings

Primary Retroperitoneal masses - Benign

PARAGANGLIOMA

- Neuroendocrine tumors arising from paraganglia.
- 10 % of all pheochromocytomas arising from the neural crest cells along the sympathetic chain.
- May be associated with **von Hippel Lindau syndrome, MEN Type II and NF1.**
- Generally more malignant with higher metastatic rate than their adrenal counterparts - Malignancy is defined as evidence of metastases.

Characteristic Imaging Findings:

CT:

- Density greater than 10Hu on non-contrast imaging (differentiates from adenoma)
- Large, lobulated and heterogeneous masses due to presence of necrosis, blood and calcification
- Mostly hypervascular after contrast administration - avidly enhances with contrast with delayed washout

MRI:

- **T1**: hypointense to liver and adrenal; salt and pepper appearance due to enhancing parenchyma and signal flow voids of vessels
- **T2**: hyperintense - 'light bulb' appearance
- **T1+Gd:** heterogenous prolonged enhancement

Imaging Findings

Primary Retroperitoneal masses - Benign

- Collagen vascular disease.
- +++ males / +++ 40-60 years .

Characteristic Imaging Findings:

- Ill-defined soft tissue mass most often centered on the aorta, IVC, iliac vessels, and ureters (may produce hydronephrosis by extrinsic compression)
- **Does not displace the involved vessels/organs**
- Early enhancement is commonly seen in the initial inflammatory stage; delayed enhancement occurs when the disease progresses to fibrosis
- **MRI** - fibrotic component typically demonstrates diffuse low T1 signal. The T2 signal varies according to the degree of active inflammation and edema - initially, high T2 signal is present and progressively decreases over time. Delayed enhancement is typical.

RETROPERITONEAL FIBROSIS

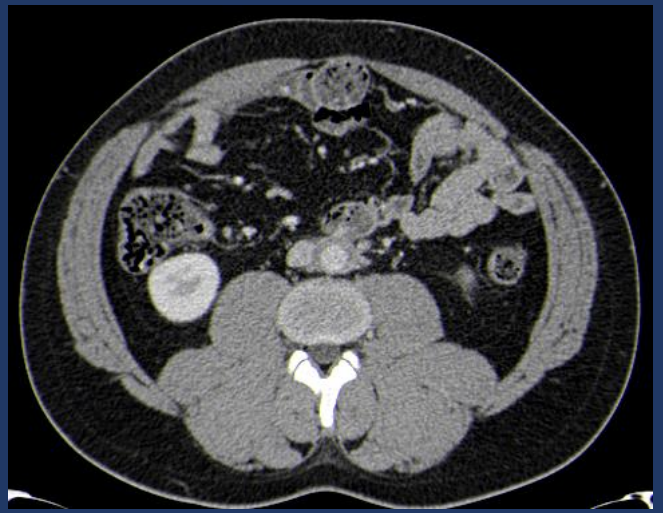


Fig. 7– Retroperitoneal Fibrosis – Axial contrast-enhanced CT reconstrutions demonstrates an ill-defined soft tissue mass around the aorta and IVC, not displacing them.
Source: *Radiology department, Hospital do Divino Espirito Santo de Ponta Delgada, EPE*

Imaging Findings

Primary Retroperitoneal masses - Benign

- Congenital malformation.
- Rare (<1% of cases) / +++ males.

Characteristic Imaging Findings:

- Involve more than one retroperitoneal compartment
- Invaginate between structures without displacing them
- Large, thin-walled, multiseptated cysts
- Calcifications are rare

- Low T1- and high T2-signal intensity, with no significant enhancement on post-contrast images

RETROPERITONEAL LYMPHANGIOMA

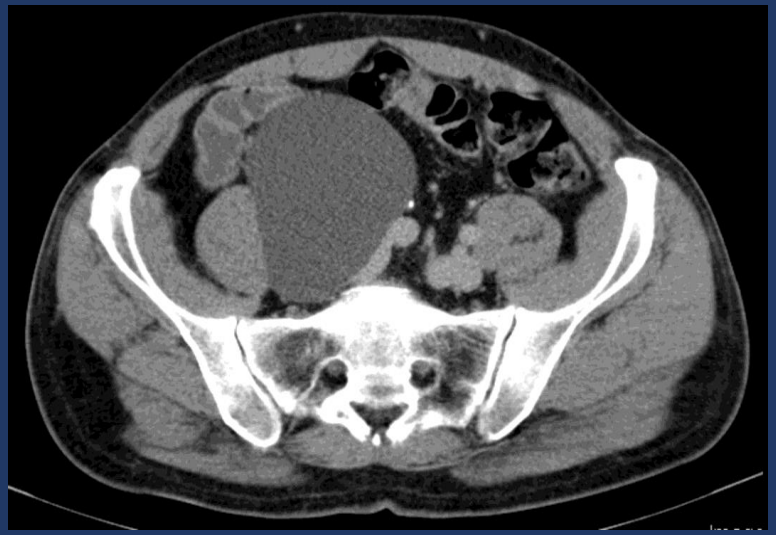


Fig. 8 – Retroperitoneal lymphangioma – Axial contrast-enhanced CT reconstrutions demonstrates a large cystic, thin-walled lesion that invaginates between the right iliac vessels and the homolateral psoas muscle without displacing them.
Source: *Radiology department, Hospital do Divino Espirito Santo de Ponta Delgada, EPE*

Imaging Findings

Primary Retroperitoneal masses - Benign

FLUID COLECTIONS AND INFECTION

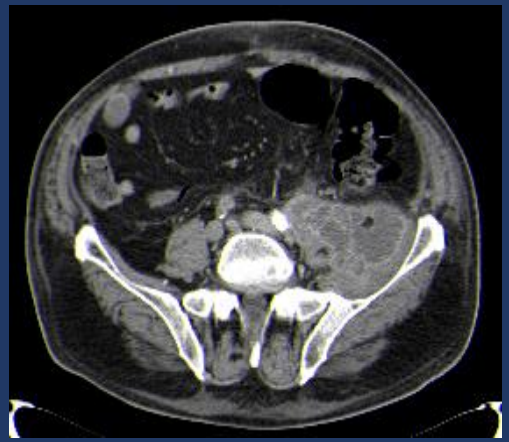


Fig. 9– Abcess of the left psoas muscle. Axial contrast-enhanced CT reconstrutions demonstrates Mutiloculated fluid collection with an air bubble inside and enhancing wall.
Source: *Radiology department, Hospital do Divino Espirito Santo de Ponta Delgada, EPE*

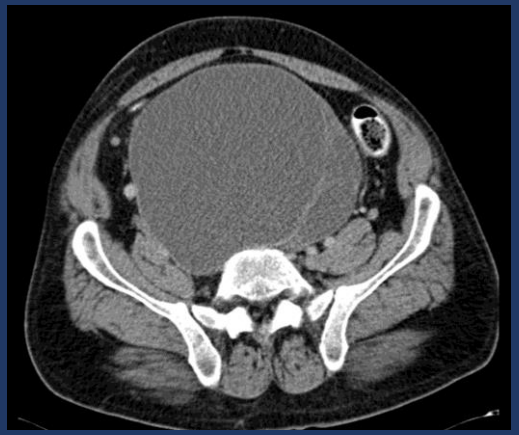


Fig. 10– Retroperitoneal cyst. Axial contrast-enhanced CT reconstrutions demonstrates a large, round, thin-walled cystic lesion with a thin enhancing septae.
Source: *Radiology department, Hospital do Divino Espirito Santo de Ponta Delgada, EPE*

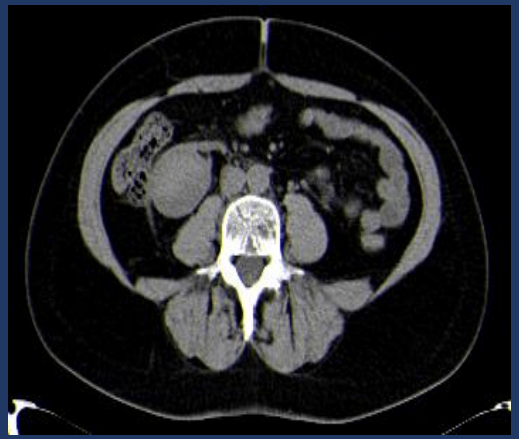


Fig. 11– Retroperitoneal haematoma. Axial non-enhanced CT reconstrutions demonstrates Collection with spontaneously hyperdense content suggesting blood.
Source: *Radiology department, Hospital do Divino Espirito Santo de Ponta Delgada, EPE*

Imaging Findings

Secondary Retroperitoneal masses - Malignant

METASTATIC DISEASE

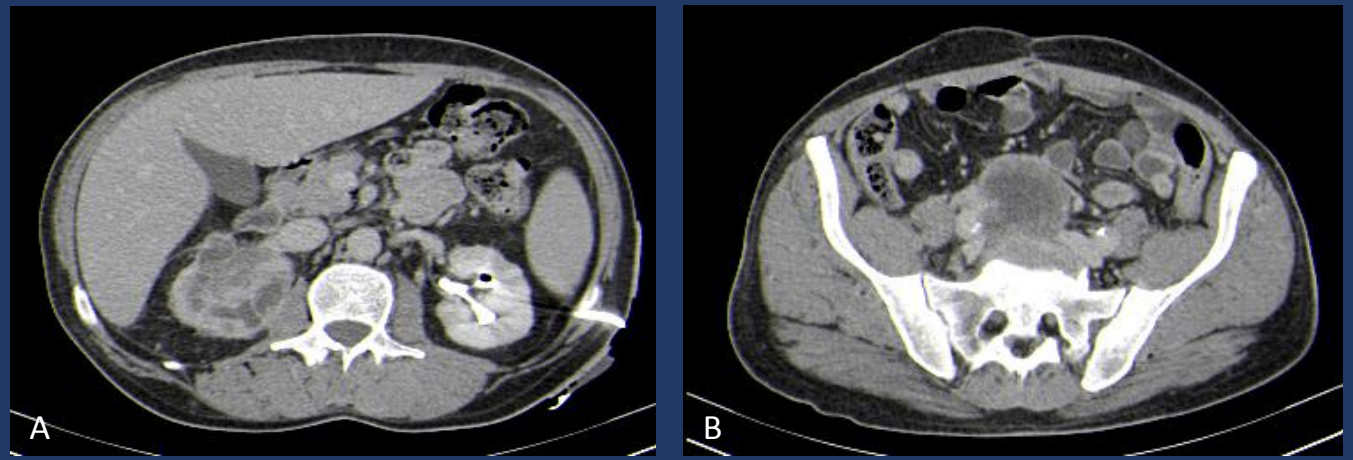


Fig. 12 – Metastatic bladder carcinoma – Axial contrast-enhanced CT reconstrutions demonstrates neoplasic involvement of the right kidney (A) and a retroperitoneal mass (B), secondary to a bladder carcinoma.
Source: *Radiology department, Hospital do Divino Espirito Santo de Ponta Delgada, EPE*

Imaging Findings

Secondary Retroperitoneal masses - Malignant

PRIMARY TUMORS OF THE RETROPERITONEAL MAJOR ORGANS

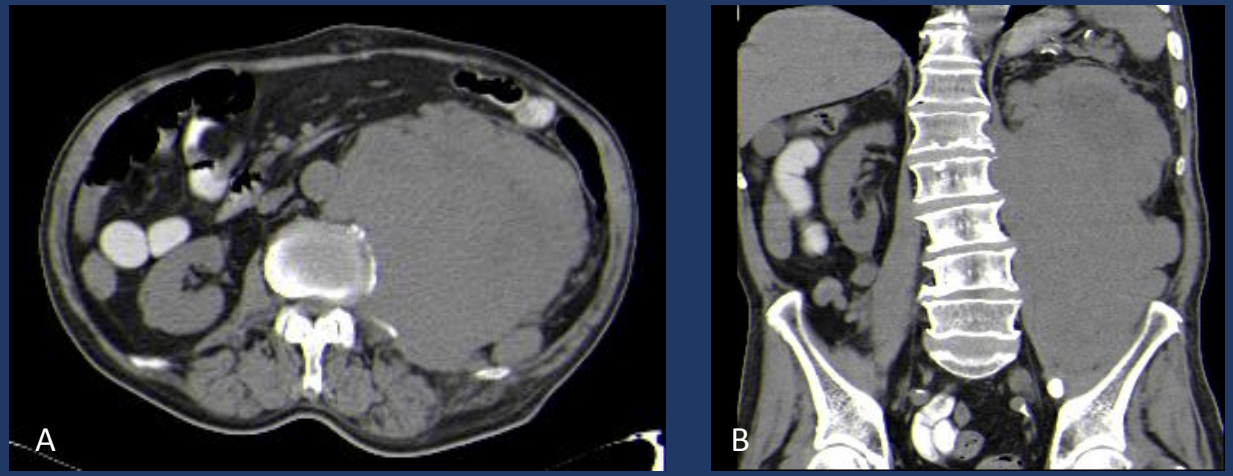


Fig. 13 – Carcinoma of the left kidney. Axial (A) and coronal (B) non-enhanced CT reconstrutions showing a large solid mass occupying the left retroperitoneum, that originates from the left kidney.
Source: *Radiology department, Hospital do Divino Espirito Santo de Ponta Delgada, EPE*

Imaging Findings

Secondary Retroperitoneal masses - Benign

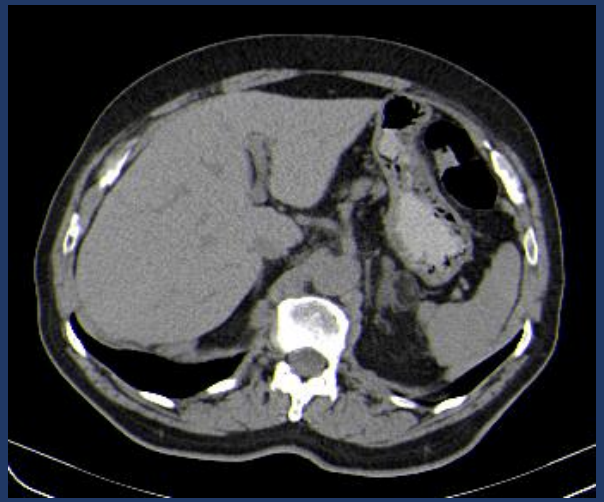


Fig. 14 – Left adrenal myelolipoma. Axial non-enhanced CT reconstrutions showing a well circumscribed fatty lesions on the left adrenal gland.
Source: *Radiology department, Hospital do Divino Espirito Santo de Ponta Delgada, EPE*

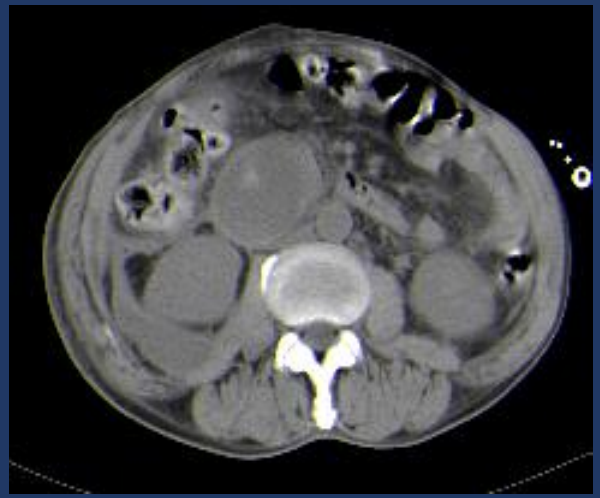


Fig. 15 – Pancreatic pseudocyst. Axial non-enhanced CT reconstrutions showing a well circumscribed fluid collection inferior to the pancreatic head in a pacient with history of previous acute pancreatitis.
Source: *Radiology department, Hospital do Divino Espirito Santo de Ponta Delgada, EPE*

Conclusions

- Familiarity with retroperitoneal anatomy and the radiographic signs to identify an intra-abdominal mass as primary retroperitoneal enable differentiation between primary and secondary masses.
- The differential diagnosis of primary retroperitoneal masses may be based on the predominant cross-sectional imaging appearance as either cystic or solid and neoplastic and non-neoplastic.
- The recognition of the imaging features of various retroperitoneal masses (composition, enhancement pattern, location, relationship to adjacent structures) together with relevant clinical information and laboratorial findings helps to narrow the differential diagnosis.
- Imaging plays an essential rule on the early diagnosis of retroperitoneal masses, allowing a suitable decision of treatment for the patient, in order of the best prognosis possible.

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