Retroperitoneal tumors: Computed Tomography (CT) and Magnetic Resonance (MR) patterns

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Learning objectives

1) To illustrate the most frequent CT and MR imaging appearance of the retroperitoneal tumors.

2) To review the most frequent recurrence patterns after retroperitoneal tumors surgery.

3) To evaluate the most effective CT and MR imaging examination techniques.

Background

Primary retroperitoneal neoplasms are a rare but diverse group of benign and malignant tumors that arise within the retroperitoneal space but outside the major organs in this space (1).

WHO 2002 classification of retroperitoneal tumors divided this group of diseases by means of the origin tissue (Tab.1) (2).

Images for this section:

**Fig. 1:** Tab.1 WHO RETROPERITONEAL TUMORS CLASSIFICATION
Imaging findings OR Procedure details

MR-CT IMAGING TECHNIQUE

We retrospectively evaluated 349 cases of CT and MR examinations of patients with retroperitoneal tumors.

MR examinations were performed with a 1.5T MR system (Philips Gyroscan Intera Power) using multiplanar Gradient Echo (GRE) T1w, Turbo Spin Echo (TSE) T2w sequences with and without fat suppression (FS) and contrast-enhanced (ce) dynamic GRE FS-T1w sequence. Post contrast GRE FS-T1w images were also evaluated before and after digital subtraction procedure.

CT examinations were performed with a MDCT system (Toshiba Aquilion Multi 4), using unenhanced and multiphasic post-contrast axial scans, associated to multiplanar reconstructions.

CT AND MR IMAGING PATTERNS OF THE RETROPERITONEAL TUMORS

Most important parameters in retroperitoneal tumors evaluation are: (A) determining tumor location into the retroperitoneal space; (B) identifying the organ of origin; (C) recognizing specific features of various retroperitoneal tumors (evaluating tumor components and vascularity)

(A) determining tumor location into the retroperitoneal spaces

Anterior displacement of retroperitoneal structures suggests that a mass arises in the superior part of the retroperitoneum. (Anterior displacement of pancreas and portal vein for masses arising from the central area of the retroperitoneum (Fig. 1A); anterior displacement of the kidney for lateral masses (Fig. 1B)).

In the inferior part of the retroperitoneum, typical signs of retroperitoneal masses are displacement of major vessels and infiltration of psoas muscles (Fig. 2).

Nevertheless, determining retroperitoneal tumor location could be difficult in case of big masses or mesenterial lesions (1).

(B) Identification of 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. Some radiologic signs are helpful in determining tumor origin (1):
**Positive Beak Sign:** When a mass deforms the edge of an adjacent organ into a "beak" shape, it is likely that the mass arises from that organ (Fig. 3).

**Negative Beak Sign:** If a mass deforms the edge of an adjacent organ with dull edges, it suggests that the tumor compresses the organ but does not arise from it (Fig. 4).

**Positive Embedded Organ Sign:** When a tumor compresses an adjacent plastic organ that is not the organ of origin, the organ is deformed into a crescent shape (Fig. 5).

**Negative Embedded Organ Sign:** when part of an organ appears to be embedded in the mass, the tumor is in close contact with the organ and the contact surface is typically sclerotic with desmoplastic reaction (Fig. 6).

**Phantom (Invisible) Organ Sign:** When a large mass arises from a small organ, the organ sometimes becomes undetectable (Fig. 7).

(C) Recognizing specific features of various retroperitoneal tumors

Some tumor contents and vascularization can be clearly demonstrated at CT and MR imaging and it can help in the differential diagnosis.

**FAT**

The presence of fat is easily recognized due to its low attenuation at CT or its high signal intensity at T1w MR imaging, with loss of signal intensity on fat-suppressed MR images.

A mass that is homogeneous and well defined and consists almost entirely of fat represents lipoma. A mass that is irregular and ill defined but contains fat, represents liposarcoma.

**Liposarcomas** are the most common sarcomas of the retroperitoneum (57% of the cases). They are classified at pathologic analysis into well-differentiated, pleomorphic, myxoid and dedifferentiated subtypes. Well-differentiated liposarcomas usually contain an appreciable amount of fat, whereas high-grade liposarcomas may not demonstrate appreciable fat. Most frequent site in the retroperitoneum is the renal fossa. At imaging liposarcomas appear well-circumscribed, encapsulated and multilobulated fatty mass, with internal fibrous septa. At CT liposarcomas show negative attenuation coefficient and can have large internal enhancing fibrous septa (Fig. 8). At MR imaging the mass shows signal intensity identical to that of subcutaneous adipose tissue, with thick peripheral ring and fibrous septa in the well-differentiated variant (Fig. 9). Pleomorphic subtype has a poor adipose content, with inhomogeneous signal intensity on T1w and T2w images. The myxoid variant is isointense compared to the muscle, with cystic appearance on unenhanced images but solid aspect after CM administration. Dedifferentiated subtypes show heterogeneous aspect, with nodular areas of contrast enhancement. CT and MR imaging features that suggest malignancy include diameter larger than 10 cm, presence
of thick septa, presence of globular and/or nodular nonadipose areas, and percentage of fat composition less than 75% (4). Only 50% of the tumors can be completely removed at the moment of diagnosis. The best treatment is surgery, but with great incidence of local recurrence. Retroperitoneal liposarcoma frequently recurs asymptomatically within 2 years of the initial surgical resection (3,5) (Fig. 10).

HYPERVASCULAR LESIONS WITH NECROSIS

Necrosis (low attenuation areas without contrast enhancement on CT imaging and hyperintense areas on T2w MR imaging) is usually seen in tumors of high-grade malignancy such as leiomyosarcomas.

Leiomyosarcomas are the second sarcomas of the retroperitoneum (16% of the cases). These tumors originate from smooth muscle, particularly that of the retroperitoneal vessel walls. They appear at imaging as a voluminous solid lobular retroperitoneal mass with cystic areas caused by necrosis. Lesions take up the CM both on CT and MR imaging (Fig. 11). Lesion enhancement involves the muscular or fibrous content of the lesion, but it is usually delayed (dd with other hypervascularized tumors such as the hemangiopericytoma). The amount of necrosis represents a pattern of aggressiveness. Leiomyosarcomas may also spread to lung, muscles, liver and kidney by means of hematogenous dissemination (Fig. 12). (3,5).

LESIONS WITH MYXOID STROMA

Few soft-tissue masses contain a myxoid stroma, characterized by a mucoid matrix rich in acid mucopolysaccharides (Malignant fibrous histiocytomas, Myxoid liposarcomas, Schwannomas). Lesions with myxoid stroma showed hypodensity on CT images and high signal intensity on T2-weighted MR images. After contrast medium injection the degree of enhancement depends on the extent of the vascular network within the myxoid stroma (3,5).

Malignant fibrous histiocytomas are the third sarcomas of the retroperitoneum (9% of the cases). At imaging these tumors appear as large retroperitoneal tumor with heterogeneous, predominantly peripheral enhancement. CT images show liqid-like appearance, with hypodensity on unenhanced images and contrast enhancement after CM injection (Fig. 13). MR images show low signal intensity on T1w and mixed low, intermediate, and markedly high signal intensity (the so-called "bowl of fruit sign") as a result of admixture of solid components, cystic degeneration, hemorrhage, myxoid stroma, and #brous tissue on T2w images. Associated lymphadenopathy is commonly observed in malignant #brous histiocytoma (3,5).

VASCULAR LESIONS
Extremely hypervascular tumors include paragangliomas and hemangiopericytomas. Other hypervascular retroperitoneal tumors are leiomyosarcomas and dedifferentiated liposarcomas.

**Hemangiopericytomas** represents 3% of the retroperitoneal tumors. These rare tumors apparently arising from pericytes. Most frequent sites are the muscles of the lower extremities and subcutaneous tissue. At imaging these lesions appear as large hypervascularized mass with polylobular contour, with enhancement patterns and MR signal intensity similar to hemangiomas (Fig. 14). Rapid enhancement after CM administration and signal void artifacts on MR images are useful in the differential diagnosis respect to other vascularized retroperitoneal neoplasms (leiomyosarcomas, dedifferentiated liposarcomas).

**CYSTIC LESIONS**

Some tumors are completely cystic in appearance. These include lymphangiomas and mucinous cystic tumors.

**Cystic Lymphangiomas** are uncommon, congenital benign tumors and occur due to failure of the developing lymphatic tissue to establish normal communication with the remainder of the lymphatic system. Cystic lymphangiomas can occur anywhere in the perirenal, pararenal or pelvic extraperitoneal spaces. At imaging, cystic lymphangioma typically appears as a large, thin-walled, multiseptate cystic mass. Its attenuation values on CT images vary from that of fluid to that of fat. MR allows a better evaluation of the multiloculated cystic pattern on T2w and on fat suppressed enhanced T1w images (Fig. 15).

**Images for this section:**
Fig. 1: Tab.1 WHO RETROPERITONEAL TUMORS CLASSIFICATION

<table>
<thead>
<tr>
<th>TISSUE OF ORIGIN</th>
<th>NEOPLASMS</th>
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<tbody>
<tr>
<td>Fat tissue</td>
<td>Lipoma</td>
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<tr>
<td></td>
<td>Liposarcoma</td>
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<tr>
<td>Connective tissue</td>
<td>Fibroma</td>
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<tr>
<td></td>
<td>Malignant fibrous histiocytoma</td>
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<tr>
<td>Muscle tissue</td>
<td>Leiomyoma</td>
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<td></td>
<td>Leiomyosarcoma</td>
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<tr>
<td>Vascular tissue</td>
<td>Lymphangioma</td>
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<td></td>
<td>Malignant hemangiopericytoma</td>
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<tr>
<td>Neurogenic tissue</td>
<td>Neurofibroma</td>
</tr>
<tr>
<td>Miscellaneous</td>
<td>Schwannoma</td>
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<td></td>
<td>Retroperitoneal fibrosis</td>
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</table>

Fig. 2: (FIG 1A-B) DETERMINING TUMOR LOCATION INTO THE RETROPERITONEAL SPACES Anterior displacement of retroperitoneal structures suggests that a mass arises in the superior part of the retroperitoneum, with anterior displacement of pancreas and portal vein for masses arising from the central area of the retroperitoneum (recurrence of liposarcoma, A) and anterior displacement of the kidney for lateral masses (Malignant fibrous histiocytoma, B).
Fig. 3: (FIG 2) DETERMINING TUMOR LOCATION INTO THE RETROPERITONEAL SPACES Displacement of major vessels and infiltration of psoas muscles are typical signs of retroperitoneal masses in the inferior part of the retroperitoneum (Malignant \#brous histiocytooma).
**Fig. 4:** (FIG. 3) IDENTIFICATION OF THE ORGAN OF ORIGIN Positive Beak Sign (pancreatic cystic tumor)

**Fig. 5:** (FIG. 4) IDENTIFICATION OF THE ORGAN OF ORIGIN Negative Beak Sign (retroperitoneal leiomyosarcoma)
Fig. 6: (FIG. 11) LEIOMYOSARCOMA CT of leiomyosarcoma of the upper left retroperitoneum (a,b,c,d). Voluminous solid lobular mass with hypodense areas caused by necrosis.
**Fig. 7:** (FIG. 12) LEIOMYOSARCOMA Secondary lesions from leiomyosarcoma on MR and CT images (lung, liver, kidney, peritoneum, muscles)
**Fig. 8:** (FIG. 13) MALIGNANT FIBROUS HISTIOCYTOMA CT images show large retroperitoneal tumor with heterogeneous, predominantly peripheral enhancement.
Fig. 9: (FIG. 14) MALIGNANT HEMANGIOPERICYTOMA Large hypervascularized mass with polylobular contour, with enhancement patterns similar to hemangiomas on CT (a,b) and MR (c,d) images.
**Fig. 10:** (FIG. 10) LIPOSARCOMA CT of recurrent liposarcoma of the upper right retroperitoneum (a,b,c). MR of recurrent liposarcoma of the pelvis (d,e,f)
**Fig. 11:** (FIG. 9) LIPOSARCOMA MR of well differentiated liposarcoma (multilobulated fatty mass, with internal fibrous septa (arrows)).

**Fig. 12:** (FIG. 8) LIPOSARCOMA CT of dedifferentiated liposarcoma (mass with poor adipose content and inhomogeneous contrast enhancement)
Fig. 13: (FIG. 7) IDENTIFICATION OF THE ORGAN OF ORIGIN Phantom (Invisible) Organ Sign (adrenal metastasis)

Fig. 14: (FIG. 6) IDENTIFICATION OF THE ORGAN OF ORIGIN Negative Embedded Organ Sign (GIST arising from duodenum)
Fig. 15: (FIG. 5) IDENTIFICATION OF THE ORGAN OF ORIGIN Positive Embedded Organ Sign (retroperitoneal malignant fibrous histiocytoma)

Fig. 16: (FIG. 15) CYSTIC LYMPHANGIOMA Large, thin-walled, multiseptate cystic mass (arrows). CT images show a hypodense mass with thin septa (a,b,c). MR images allow a better evaluation of the multiloculated cystic pattern on T2w (d,f) and on fat suppressed enhanced T1w images (e).
Conclusion

CT and MR are effective imaging techniques in the evaluation of retroperitoneal tumors. The most effective techniques are multiphasic acquisition (CT) and TSE T2w, conventional dynamic ce-FS-GRE T1w and subtracted dynamic ce-FS-GRE T1w sequences (MR). The most important parameters in the study of the retroperitoneal tumors were the anatomy and the topography of the lesions for the surgical planning and the vascularization to evaluate the aggressiveness of the lesions.

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