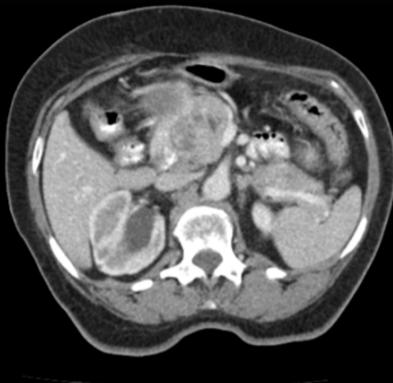


Radiographic QUIZ

Julio Manuel Díaz Riverol¹

A 78-year-old woman presented with intermittent abdominal pain that she had been experiencing for several weeks.

Contrast enhancement abdominal CT images are shown below:



Where is the salient abnormality located?

- Retroperitoneal space
- Intraperitoneal space

Respond True or False

- The mass is involving the interior vena cava.
- The mass is displacing the abdominal aorta.
- The mass is causing right hydronephrosis.
- The mass is causing dilatation of the CBD and pancreatic duct.
- The mass is in contact with the right psoas muscle.

In which of the retroperitoneal compartment the mass is extended?

- Anterior perirenal space
- Posterior perirenal space
- Perirenal space
- Central retroperitoneal space
- All of the above

Which of the following statements is incorrect?

- The mass reveals necrotic areas.
- There are calcifications within the mass.
- The mass shows heterogeneous enhancement.
- Retroperitoneal masses constitute a heterogeneous group of lesions, originating in the retroperitoneal space, which pose a diagnostic challenge for radiologists.
- Retroperitoneal masses are classified as primary when they do not originate from organs such as the kidneys, adrenal glands, pancreas, or bowel loops.

DESCRIPTION OF THE EXAM

CT scan showing a bulky retroperitoneal lesion with heterogeneous enhancement and foci of necrosis involving the inferior vena cava, displacing the abdominal aorta, causing dilatation of the CBD and pancreatic duct by external compression and right hydronephrosis.

Biopsy was performed indicating Mesenchymal Fusocellular Neoplasia compatible with leiomyosarcoma.

Surgical excision of the mass was performed confirming the retroperitoneal origin arising from the inferior vena cava wall.

BRIEF REVIEW OF THE PATHOLOGY

Tumor location

Characterization of the retroperitoneal space

The first step is to decide whether the tumor is located within the retroperitoneal space. It is useful to observe the displacement of normal anatomic structures. Anterior displacement of retroperitoneal organs (eg, kidneys, adrenal glands, ureters, ascending and descending colon, pancreas, portions of the duodenum) strongly suggests that the tumor arises in the retroperitoneum. Major vessels and some of their branches are also found in the retroperitoneal cavity, so that displacement of these vessels can be helpful as well.

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 that are helpful in determining tumor origin include the “beak sign,” the “phantom (invisible) organ sign,” the “embedded organ sign,” and the “prominent feeding artery sign”. When there is no definite sign that suggests an organ of origin, the diagnosis of primary retroperitoneal tumor becomes likely.

Leiomyosarcomas usually manifest as large soft-tissue masses, with internal heterogeneity and heterogeneous enhancement usually secondary to necrosis and hemorrhage. The mean tumor size for lesions with and without vascular involvement has been reported as 10.4 cm and 11.3 cm, respectively. Calcifications are not commonly found, and adipose tissue is absent. Purely intravascular lesions appear as heterogeneously enhancing expansile masses. They commonly exhibit low to intermediate signal intensity at T1-weighted MRI and intermediate to high signal intensity at T2-weighted MRI. They also occasionally demonstrate fluid-fluid levels secondary to hemorrhage.

A large non-fat-containing retroperitoneal mass with involvement of a contiguous vessel and varying internal necrosis should raise the possibility of a leiomyosarcoma.

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 (beak sign). On the other hand, an adjacent organ with dull edges suggests that the tumor compresses the organ but does not arise from it.

Phantom (invisible) organ sign

When a large mass arises from a small organ, the organ sometimes becomes undetectable. This is known as the phantom organ sign. However, false-positive findings do exist, as in cases of huge retroperitoneal sarcomas that involve other small organs such as the adrenal gland.

Embedded organ sign

When a tumor compresses an adjacent plastic organ (eg, gastrointestinal tract, inferior vena cava) that is not the organ of origin, the organ is deformed into a crescent shape. In contrast, when part of an organ appears to be embedded in the tumor (negative embedded organ sign), the tumor is in close contact with the organ and the contact surface is typically sclerotic with desmoplastic reaction. Occasionally, the contact surface becomes ulcerative. When the embedded organ sign is present, it is likely that the tumor originates from the involved organ.

Prominent feeding artery sign

Hypervascular masses are often supplied by feeding arteries that are prominent enough to be visualized at CT or MR imaging, a finding that provides an important key to understanding the origin of the mass.

Retroperitoneal masses

Retroperitoneal masses constitute a heterogeneous group of lesions, originating in the retroperitoneal space, which pose a diagnostic challenge for radiologists. The majority of cases are malignant tumors, of which approximately 75% are mesenchymal in origin. Although such tumors are more prevalent in adults, they can occur at any age. Primary malignant tumors that originate in the soft tissue of the retroperitoneum (ie, outside the solid retroperitoneal organs such as the kidneys, adrenal glands, and portions of the small and large bowel) are rare. In one review of 25 647 malignant neoplasms from a tumor registry, retroperitoneal

tumors accounted for 0.16% of all tumors. Nevertheless, 79%-90% of all retroperitoneal tumors are malignant, and they are associated with relatively high mortality rates, owing partly to the nonspecific symptoms, or lack thereof, in patients at presentation. When they do not originate from organs such as the kidneys, adrenal glands, pancreas, or bowel loops, retroperitoneal masses are classified as primary and are categorized as solid or cystic, depending on their appearance on imaging. Solid lesions can be divided into four groups, by origin: mesenchymal, neural, germ-cell, and lymphoproliferative. Among the cystic lesions, the most common are lymphangioma and cystic mesothelioma. There are also non-neoplastic processes, primarily retroperitoneal fibrosis, non-Langerhans histiocytosis (Erdheim-Chester disease), and extramedullary hematopoiesis.

Leiomyosarcoma

Leiomyosarcoma is a malignant tumor of smooth muscle cells, with a retroperitoneal location in 12%-69% of cases. It is the second most common primary malignant retroperitoneal sarcoma, accounting for 28% of cases. Leiomyosarcomas are typically diagnosed during the fifth to sixth decades of life, more often in women than in men. Leiomyosarcoma is believed to originate from the large blood vessels in the retroperitoneum (eg, the inferior vena cava) and/or their tributaries. Nevertheless, leiomyosarcomas most commonly manifest as extravascular tumors (62% of cases) and are rarely completely intravascular (5%). In 33% of cases, leiomyosarcomas have both an intravascular and an extravascular component. Similar to liposarcomas, leiomyosarcomas are often large when patients present, and symptoms are usually related to compression of adjacent structures. Metastases are seen at the time of diagnosis in 9% of patients with extravascular tumors and 23% of patients with intravascular tumors, with the lungs, liver, and peritoneum being the most common sites. Prognosis is typically dependent on achieving complete excision with wide negative margins.

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