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Giant Retroperitoneal Myxoid Liposarcoma: Imaging Diagnosis of a Rare Tumor and Histopathological Classification of Liposarcomas: A Case Report and Literature Review

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Abstract **Case Report**

Sarcomas are rare malignant tumors. Within this category, among liposarcomas, the myxoid type is the most frequent subtype. Abdominal liposarcomas are most often retroperitoneal and can reach considerable sizes before becoming clinically detectable due to their slow and insidious progression, which frequently leads to delayed diagnosis. Clinical presentation is variable and depends on tumor size and location. It most commonly manifests as a large abdominal mass responsible for abdominal distension, with or without abdominal pain or compressive symptoms. On imaging, myxoid liposarcoma typically appears as a large solid-cystic mass exerting compressive effects, particularly on digestive structures. Histologically, it is distinguished from other liposarcomas by its characteristic myxoid component. Treatment consists of complete surgical resection of the tumor. We report a case of retroperitoneal myxoid liposarcoma in a 50-year-old patient revealed by a progressively enlarging painful abdominal mass. Management consisted of complete tumor excision. Histological examination confirmed a low-grade myxoid liposarcoma. No adjuvant therapy was deemed necessary, with favorable postoperative outcome. The aim of this work is to report, through this case, the clinical and radiological features of myxoid liposarcoma, to detail the elements of positive and topographic diagnosis, and to highlight the differential diagnostic challenges encountered in imaging. The histopathological section addresses the current classification of liposarcomas, which has undergone modifications compared to previous versions, the understanding of which allows a better imaging approach to these tumors.

Keywords: Myxoid liposarcoma, Retroperitoneum, Soft tissue sarcoma, Computed tomography, Surgical resection, Histopathology.

INTRODUCTION

Soft tissue sarcomas are rare and account for less than 1% of all malignant tumors. Liposarcoma is a common type within this category. The myxoid subtype is frequently found in the extremities and it is the most common subtype among retroperitoneal liposarcomas. [1] In the most recent version of the WHO classification of liposarcomas, myxoid liposarcoma also includes round cell liposarcoma, which previously represented a separate entity. These two types actually represent a continuum in which round cell liposarcoma corresponds to the poorly differentiated and higher-grade form. [2] Imaging plays a crucial role in the positive diagnosis of these slowly evolving and often clinically silent tumors, and helps guide subsequent management. [1,3,4]

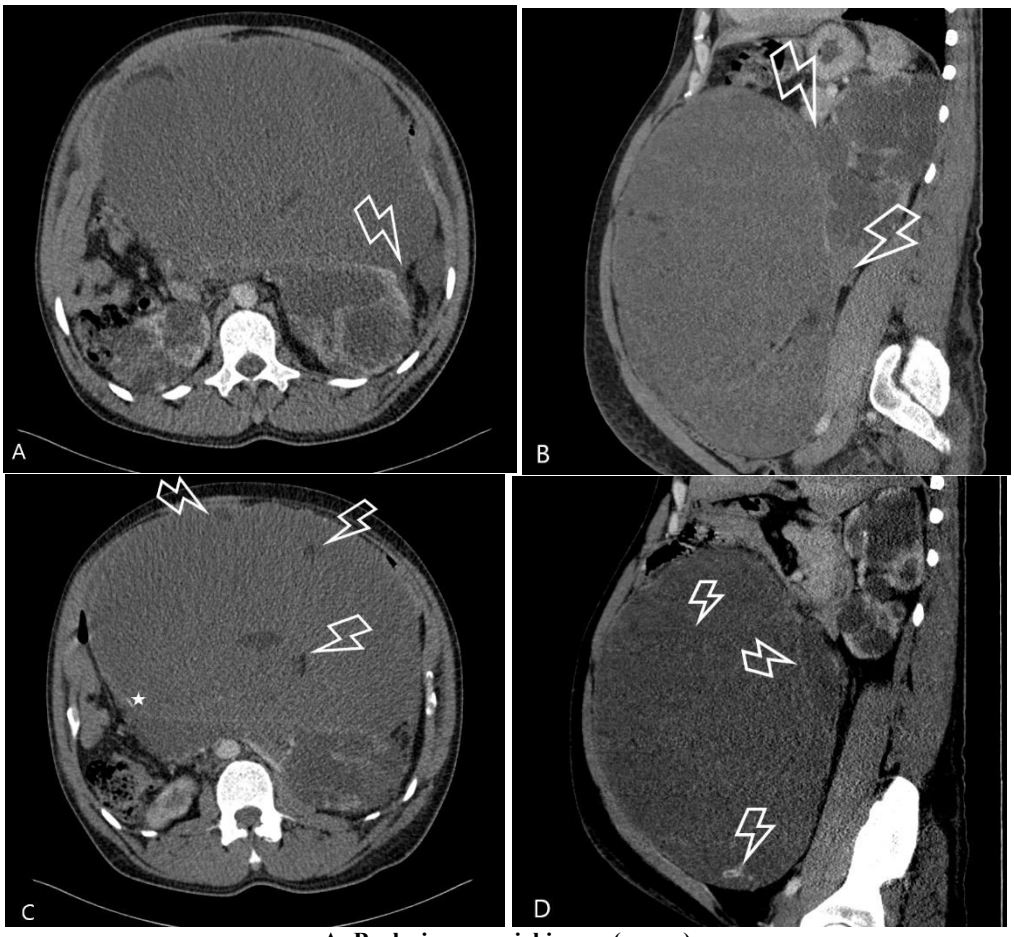
CASE REPORT

A 50-year-old patient, with no significant past medical history, presented with marked abdominal distension that had been progressing for 20 days. Initial standard biological workup was unremarkable, apart from moderately impaired renal function, and infectious screening was negative. An abdominal CT scan was performed. It revealed a voluminous abdominopelvic and retroperitoneal cystic mass measuring 32x20x34 cm (thickness, width, height, respectively). It was multi-loculated, well-circumscribed, bordered by a thin, iso-

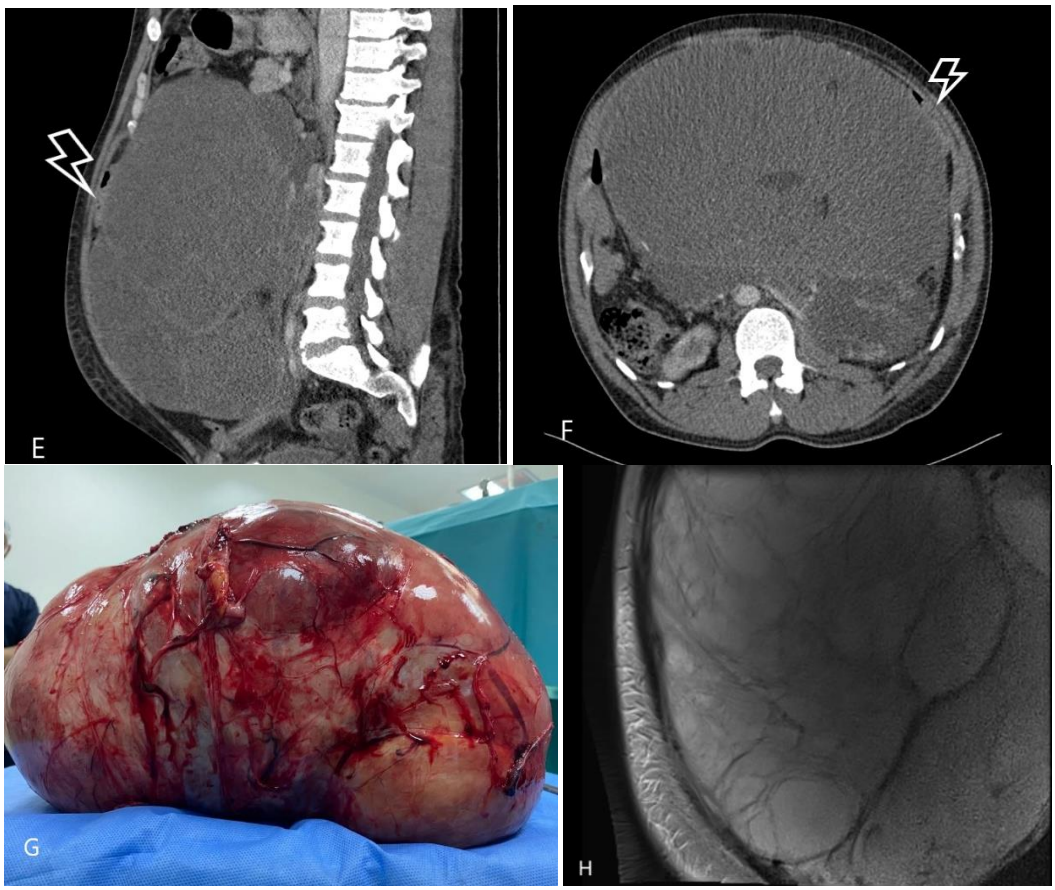
dense capsule, and displaced the intra-abdominal viscera anteriorly and laterally against the anterolateral abdominal wall, particularly the left and transverse colon. Posteriorly, it displaced the retroperitoneal vessels. The mass showed low attenuation on non-contrast images with an average attenuation between 0 and 25 Hounsfield Units (HU). It contained fatty areas, some calcifications, and multiple thin septations, with no significant contrast enhancement and no detectable opacification within it on delayed phases. The kidneys were enlarged with bosselated contours, containing multiple confluent simple cystic formations that thinned the cortex and caused scalloping of the excretory tracts, associated with multiple simple hepatic cysts (hepatorenal polycystic disease). A small amount of fluid was also noted in the left parietocolic gutter with thickening of the left perirenal fascia. The mass was adherent to the left kidney, on which it exerted a scalloping effect, to the anterior pararenal fascia, and to the posterior abdominal wall. The beak sign was deemed negative. The adrenal glands were well visualized and appeared normal. No secondary lesions were found on bone windows. Complementary MRI images were acquired, though exploration was hampered by the tumor's size and abdominal distension. The mass predominantly showed intermediate T2 hyperintensity with T2 hypointense septations.

Surgical exploration revealed a voluminous solid-cystic mass displacing the transverse colon against the anterior abdominal wall, adherent posteriorly to the left kidney and the posterior abdominal wall. Excision was performed with laborious opening of Told's fascia and dissection of the left colic flexure, the transverse colon, and further liberation of the mass from its posterior and lateral aspects. An accidental breach occurred in the upper third of the left ureter, necessitating the placement of a left double-J stent and repair of the breach by a termino-terminal ureteral anastomosis. Histopathological examination confirmed a low-grade (grade 1) myxoid liposarcoma.

Figures A-F: Abdominal CT scan at portal venous phase: axial and sagittal views: Voluminous, multi-septated, compressive abdominopelvic cystic mass adherent to the retroperitoneum, containing fatty areas, some parietal and internal calcifications, with no enhancement after contrast injection, consistent with a low-grade myxoid liposarcoma.



A: Beak sign on axial image (arrow)
B: Beak sign on sagittal image (arrow)
C: Fatty areas (arrows) and fine parietal calcification (asterisk).
D: Thin internal septa and calcification (arrows)



E and F: Anterior displacement and compression of the left colon against the anterior abdominal wall (arrows).
G: Surgical specimen.
H: Tumor showing intermediate to high signal intensity on sagittal T2-weighted BLADE sequence, with septa appearing as low signal intensity on T2 (arrows)

CASE DISCUSSION

The primary difficulty in the CT-based diagnosis of this case was determining the site of origin of the tumor. Indeed, the mass was very large and involved multiple abdominal compartments. Assessment of the beak sign with the left kidney was relatively challenging, particularly on sagittal reconstructions. This was due to the fact that the tumor was closely adherent to the left kidney and to its exophytic cortical cystic lesions (which had nearly identical attenuation), as well as to the posterior parietal peritoneum.

On axial and oblique planes, the beak sign was negative. Identification of normal adrenal glands excluded an adrenal origin. The absence of opacification of the tumor's cystic locules on delayed-phase imaging further supported the hypothesis of an extra-renal origin.

Furthermore, the adherence of the tumor to the posterior abdominal wall, its relationship with the retroperitoneal vessels, and the mass effect exerted on the colon particularly the left colon, which was compressed against the anterior abdominal wall along almost its entire course favored a retroperitoneal origin. Regarding the histological type, one might be tempted, somewhat prematurely, to consider a cystic lymphangioma given the context of hepatorenal polycystic disease (in the context of Von Hippel–Lindau disease) and the predominantly cystic attenuation of the mass. However, cystic lymphangiomas are generally less compressive. In addition, the presence of a fatty component and the common retroperitoneal location are more suggestive of a liposarcoma.

DISCUSSION

Sarcomas are rare, heterogeneous malignant mesenchymal tumors with ubiquitous distribution, accounting for less than 1% of all malignant tumors. They are classified into soft tissue sarcomas (60%), gastrointestinal stromal tumors (GIST) (30%), and bone sarcomas (10%). Retroperitoneal localization accounts for 10–15% of all sarcomas, where they are predominantly represented by liposarcomas (40–50% of retroperitoneal sarcomas). Furthermore, abdominal sarcomas can be categorized according to their location into retro- or subperitoneal, parietal, and visceral sarcomas. Mesenteric sarcomas are rare, with no standardized definition of their anatomical location. [1,3-6]

On imaging, retroperitoneal sarcomas typically present as large heterogeneous masses with fatty and soft-tissue attenuation, displacing adjacent retroperitoneal structures or viscera. Tumor size at diagnosis is usually greater than 10 cm. Among these retroperitoneal sarcomas, liposarcoma and leiomyosarcoma alone account for more than 90% of cases. They are often asymptomatic until they reach a sufficiently large size to produce clinical manifestations related to a mass effect. [5-9]

Regarding liposarcomas, they most commonly affect adults over 40 years of age, with no clear sex predilection. They represent the most frequent soft tissue sarcoma in adults and the most common retroperitoneal tumor. According to the latest WHO classification of soft tissue tumors (5th edition, 2020) [2], five histological subtypes of liposarcomas are recognized: well-differentiated liposarcoma (or atypical lipomatous tumor), dedifferentiated liposarcoma, myxoid liposarcoma, pleomorphic liposarcoma, and myxoid pleomorphic liposarcoma.

From a macroscopic standpoint, these tumors are generally relatively well-defined and surrounded by a capsule or pseudocapsule. The proportion of fat and soft tissue varies depending on the histological subtype and the fibrous septa separating lobules. Areas of degeneration and necrosis are frequently observed in large tumors. Calcifications may be present inconsistently, and vascularization is usually poor. Histopathological–radiological correlation remains challenging. However, in general, well-differentiated liposarcoma, composed predominantly of mature fat, shows markedly negative attenuation values; a more compact and soft-tissue attenuation suggests a high-grade liposarcoma; whereas low attenuation with fluid-like density, rich in edematous connective tissue and gelatinous myxoid stroma, is suggestive of myxoid liposarcoma. [2]

On CT, liposarcoma demonstrates variable appearances depending on the amount of intratumoral fat. In the fatty (well-differentiated) form, the lesion is composed almost entirely of fat. In the mixed form, discrete fatty areas (< –20 HU) are associated with soft-tissue density components (> +20 HU). In the pseudocystic form, the lesion exhibits homogeneous fluid attenuation between +20 and –20 HU and may therefore mimic a cystic tumor. Finally, the solid form shows attenuation greater than 20 HU with no demonstrable fat on cross-sectional imaging, making it indistinguishable from other sarcomas; this corresponds to poorly differentiated liposarcoma. [3,7]

On CT, these tumors most often appear as encapsulated masses with variable parietal calcifications, causing displacement, compression, or distortion of adjacent structures (kidneys, gastrointestinal tract), with possible invasion of surrounding tissues. Contrast enhancement may be present and can be either homogeneous or heterogeneous, and prominent intratumoral vessels are generally absent. On MRI, signal characteristics and enhancement patterns vary depending on the proportions of fat, solid, cystic, necrotic, or hemorrhagic components. [3,7,8] Myxoid liposarcoma (MLS) is the second most frequent subtype of liposarcoma (approximately 15%) and typically arises in the deep soft tissues of the extremities (thigh and popliteal region) in young adults. It is also the most common subtype among retroperitoneal liposarcomas. On CT, it appears as a large, well-defined mass with fluid-like attenuation on non-contrast images (lower than muscle), reflecting its high-water content and myxoid component. It contains a low proportion of fat (<10%), which may be visible or sometimes absent. It is usually associated with septa, lobulations, or a soft-tissue component, while calcifications are rare and less frequent than in other liposarcoma subtypes. [1,2,3,7,9-11]

On MRI, myxoid liposarcoma is typically hypointense on T1-weighted images, although small hyperintense foci may be present, reflecting intratumoral fat. It demonstrates high signal intensity on T2-weighted images due to its abundant myxoid (gelatinous) stroma, and usually shows delayed enhancement after gadolinium administration on T1-weighted sequences. Ultrasound may demonstrate a complex hypoechoic mass with posterior acoustic enhancement, which can help differentiate myxoid liposarcoma from purely cystic lesions.

On imaging- particularly CT-myxoid liposarcoma, like other liposarcoma subtypes, often raises differential diagnostic challenges. These difficulties arise at two levels: first, determining the site of origin of the tumor may be challenging and requires careful analysis of imaging features, particularly on CT, to establish an accurate topographic diagnosis; second, the tumor's appearance may overlap with several other entities. In this context, we detail several major differential diagnoses that may closely mimic myxoid liposarcoma on imaging. [1,3,7-10]

For topographic diagnosis, several imaging signs are useful in clinical practice. The first step is to determine whether the mass is retroperitoneal or intraperitoneal. This requires careful assessment of the displacement of normal anatomical structures. Anterior displacement of retroperitoneal organs (e.g., kidneys, adrenal glands, ureters, aorta) strongly suggests a retroperitoneal origin. A thorough understanding of peritoneal anatomy is essential. The boundary between the two compartments is defined by the posterior layer of the parietal peritoneum; therefore, anterior displacement of organs adherent to or contained within this layer (such as the colon, pancreas, and duodenum) also supports a retroperitoneal origin. The second step is to exclude the possibility that the tumor arises from a retroperitoneal

organ. Several radiological signs are useful in determining tumor origin, including the beak sign, the phantom (invisible) organ sign, the embedded organ sign, and the prominent feeding artery sign. The beak sign (also referred to as the "claw sign") corresponds to a tapering extension of normal parenchyma from an involved organ that appears to embrace the mass, indicating that the tumor arises from that organ. This sign may be difficult to assess and should be evaluated in multiple orthogonal planes using multiplanar reconstructions (MPR). In contrast, smooth indentation or deformation of an adjacent organ suggests extrinsic compression rather than tumor origin. When a large mass arises from a small organ, the latter may become undetectable, resulting in the so-called phantom organ sign. However, false positives may occur, particularly in large retroperitoneal sarcomas involving small organs such as the adrenal gland. The embedded organ sign is observed when a tumor compresses a deformable adjacent organ (e.g., bowel loops or the inferior vena cava), which then assumes a crescent shape molded by the mass, indicating that the tumor does not arise from that structure. Hypervascular masses may be supplied by prominent feeding arteries visible on CT or MRI, providing an important clue to their origin (prominent feeding artery sign). Moreover, a malignant tumor that abuts a neighboring organ without invading it is unlikely to arise from that organ. Finally, in the absence of imaging features suggesting a specific organ of origin, the diagnosis of a primary retroperitoneal tumor becomes likely. [3,4,6,9-12]

Regarding differential diagnosis, the main consideration is with other subtypes of liposarcoma, which share overlapping imaging features, as discussed above. Other soft tissue sarcomas, particularly leiomyosarcoma, should also be considered. Leiomyosarcoma is the second most common primary retroperitoneal sarcoma after liposarcoma. It typically presents as a large heterogeneous retroperitoneal mass, often with areas of necrosis or cystic degeneration, but generally lacking a fat component. Hepatic metastases (often necrotic or cystic) may also be present. Another important differential diagnosis is cystic lymphangioma with chylous content. It is a polymorphic tumor that predominantly involves the neck (approximately 75%) and more rarely the mesentery or retroperitoneum (<1%). It is more common in children (malformative origin) but may also occur in adults (approximately 40%), either congenital or secondary to lymphatic obstruction. It typically appears as a multiloculated cystic mass surrounded by a fibrous capsule and containing serous lymphatic fluid, chylous (fat-density) fluid, hemorrhagic content, or a mixture of these components. Due to anatomical continuity between the mesentery, its root, and the retroperitoneum, distinguishing mesenteric from retroperitoneal lymphangiomas may be difficult. Clinical presentation is often similar (asymptomatic or mass effect with possible compressive complications). [3,11-14]

On CT, cystic lymphangiomas appear as uni- or multiloculated masses with homogeneous content, no internal enhancement, and thin walls and septa that may enhance slightly. Parietal calcifications are possible but rare. Involvement of multiple retroperitoneal compartments is common. Fat content in chylous lymphangiomas is better detected on MRI than CT, showing signal drop on out-of-phase sequences compared with in-phase sequences. Another differential diagnosis is a large exophytic renal angiomyolipoma (AML), which contains fat and may mimic retroperitoneal liposarcoma. Features favoring AML include a defect in the renal parenchyma, enlarged intralesional vessels, and clear renal origin, which can be assessed using MPR and the beak sign. In contrast, smooth compression of the kidney and extension beyond the perirenal space favor liposarcoma. Clear cell renal cell carcinoma (RCC), which accounts for approximately 90% of malignant renal tumors in adults, should also be considered. It typically appears as a heterogeneous mass with variable cystic, hemorrhagic, or necrotic components. Calcifications are uncommon, and small fat foci may rarely be present. The combination of fat and calcifications favors RCC over angiomyolipoma. The cystic form of RCC appears as a uni- or multiloculated cystic mass with thick walls, septal or capsular calcifications, and contrast enhancement (≥20 HU). [1,3,11-15]

Another interesting differential diagnosis is giant retroperitoneal myxoma, although this location is extremely rare. This benign mesenchymal tumor is composed of stellate undifferentiated cells within a myxoid stroma. It most commonly arises in the heart, followed by bone, muscle, skin, and subcutaneous soft tissues, and typically affects adults aged 40–50 years. Tumor size is variable, and clinical presentation is often similar. On CT, it appears as a large, well-circumscribed hypodense multiloculated mass with internal septa and occasional calcifications. On MRI, it is hypointense on T1-weighted images and hyperintense on T2-weighted images, with enhancement of septa after gadolinium administration.

Other possible differential diagnoses include peritoneal inclusion cysts (typically pelvic, fluid attenuation, sometimes with thin wall calcifications), dermoid cysts (benign tumors with fatty content, often unilocular, with more frequent calcifications), and lymphoceles (which should be excluded in the absence of prior surgery). [1,3,16-20] Current histopathological classification of liposarcomas: [2,21-24]

According to the latest edition of the WHO histopathological classification of soft tissue tumors (2020) [2], liposarcomas are malignant adipocytic tumors and are classified into five histological types:

1- Atypical lipomatous tumor / well-differentiated liposarcoma (ALT/WDLPS):

This is a locally aggressive mesenchymal neoplasm composed, in whole or in part, of an adipocytic proliferation showing at least focal nuclear atypia in adipocytes and stromal cells. The terms ALT and WDLPS are synonymous, describing morphologically and genetically identical lesions. Amplification of MDM2 and/or CDK4 is almost constant. Macroscopically, it is usually a large, well-defined, and lobulated mass. The consistency is variable, ranging from firm grayish areas to gelatinous areas depending on the proportion of fibrous and myxoid components. Large retroperitoneal tumors are often heterogeneous, with fat necrosis and punctate hemorrhages. From a histopathological point of view, three main morphological subtypes are described: adipocytic (lipoma-like), sclerosing, and inflammatory. The coexistence of several patterns is frequent, especially in retroperitoneal locations.

The lipoma-like type is composed of mature adipocytes in which, unlike benign lipoma, a substantial variation in cell size is observed, as well as nuclear atypia in adipocytes or stromal spindle cells. Hyperchromatic stromal spindle cells scattered are easily identified in fibrous septa or vessel walls. Occasionally, adipocytes may acquire hibernoma-like features. A variable number of lipoblasts (from numerous to none) may be found. Importantly, the mere presence of lipoblasts neither establishes nor is required for a diagnosis of liposarcoma. The sclerosing type is the second most frequent. This pattern is most often observed in the retroperitoneum or the spermatic cord. The main histological finding is the presence of scattered bizarre stromal cells showing marked nuclear hyperchromasia and located within an extensive fibrillary collagenous stroma. Multivacuolated lipoblasts may be observed. The fibrous component may overshadow the lipogenic areas, which may therefore be easily missed in small samples.

Inflammatory ALT/WDLPS represent the rarest subtype, most often occurring in the retroperitoneum. A chronic inflammatory infiltrate predominates to such an extent that the adipocytic nature of the neoplasm may be masked. When the adipocytic component is scarce, the presence of bizarre multinucleated stromal cells represents a useful diagnostic clue. Rarely, ALT/WDLPS may show mature heterologous differentiation, which may be osseous or myogenic, but does not in itself imply dedifferentiation. Nuclear immunopositivity for MDM2 and/or CDK4 is present in most cases. In the lipoma-like type, the expression of MDM2 and CDK4 may be difficult to assess, making FISH a valuable alternative. A major pitfall is represented by nuclear positivity of MDM2 in histiocytes within areas of fat necrosis. ALT/WDLPS associated with Li-Fraumeni syndrome are negative for MDM2; however, they express p53.

2- Dedifferentiated liposarcoma (DDLPS):

DDLPS is an atypical lipomatous tumor or a well-differentiated liposarcoma (ALT/WDLPS) showing progression, either in the primary tumor or in a recurrence, toward a sarcoma (generally non-lipogenic) of variable histological grade. In most cases, there is amplification of MDM2 and CDK4. A well-differentiated component may not be identifiable. From a macroscopic point of view, DDLPS usually consists of large multinodular yellow masses containing discrete, solid, often non-lipomatous (dedifferentiated) areas of gray-brown color. The dedifferentiated areas may show necrosis. The transition between lipomatous and dedifferentiated areas may sometimes be progressive. From a histopathological point of view, the hallmark of DDLPS is the transition from ALT/WDLPS to a non-lipogenic sarcoma, which in most cases is of high grade. The extent of dedifferentiation is variable. The transition is generally abrupt; however, in some cases, it may be more progressive and, exceptionally, low- and high-grade areas appear to be intermixed. In some cases, a well-differentiated lipomatous component is difficult to identify.

The dedifferentiated areas show variable histological features but most often resemble an undifferentiated pleomorphic sarcoma or a myxofibrosarcoma of intermediate to high grade. Although dedifferentiation was initially defined by high-grade morphology, cases with low-grade dedifferentiation have increasingly been recognized. Low-grade dedifferentiation is most often characterized by the presence of uniform fibroblastic spindle cells with mild nuclear atypia, often arranged in a fascicular pattern and showing cellularity intermediate between well-differentiated sclerosing liposarcoma and the usual high-grade areas.

Low-grade DDLPS should not be confused with atypical spindle cell lipomatous tumors; the latter contain atypical adipocytes or lipoblasts, whereas dedifferentiated areas, both low and high grade, are generally non-lipogenic. Low-grade DDLPS is practically impossible to distinguish from cellular WDLPS.

Occasionally, the high-grade component may show overt lipoblastic differentiation, either in the form of scattered lipoblasts within the high-grade component or in the form of sheets of pleomorphic atypical adipocytic cells resulting in areas morphologically indistinguishable from pleomorphic liposarcoma. This phenomenon has been referred to as homologous lipoblastic differentiation or pleomorphic liposarcoma-like features. A solitary fibrous tumor-like morphology and an inflammatory myofibroblastic tumor-like morphology may rarely be observed. The main role of immunohistochemistry is to confirm divergent differentiation and to exclude other tumor types. Diffuse nuclear expression of MDM2 and/or CDK4 is almost

invariably observed and also allows distinction of homologous DDLPS from pleomorphic liposarcoma.

3- Myxoid liposarcoma (MLPS):

This is a malignant tumor composed of uniform round to ovoid cells, with a variable number of small lipoblasts, within a myxoid stroma with a branching capillary vascular network. Translocations producing FUS-DDIT3 or, more rarely, EWSR1-DDIT3 fusion transcripts are pathognomonic. Included in this category are more cellular and higher-grade tumors, formerly known as round cell liposarcoma.

From a macroscopic point of view: MLPS are generally large (>10 cm), circumscribed, multinodular intramuscular neoplasms. The cut surface is smooth, gelatinous, and glistening. Higher-grade tumors show a firmer, fleshy, beige appearance. Macroscopic necrosis is rare. Adequate sampling to estimate the degree of hypercellularity is essential, as this represents a major prognostic determinant.

From a histopathological point of view: at low magnification, MLPS are moderately cellular and lobulated tumors with increased peripheral cellularity, composed of patternless arrangements of small uniform ovoid cells without morphological adipocytic differentiation, with a variable number of small lipoblasts.

The tumors contain abundant, slightly basophilic myxoid stroma with a delicate plexiform and arborizing capillary network ("chicken-wire" pattern), around which neoplastic cells often cluster. Areas of paucicellular extracellular mucin may be present, conferring a microcystic or pulmonary edema-like appearance.

MLPS is generally devoid of significant atypia, substantial mitotic activity, or spindle cells. Lipoblasts are smaller than in other liposarcomas and are mainly univacuolated or bivacuolated. Lipoblasts may be rare or even absent. A variable proportion of mature fat may be present. Chondroid and osseous elements are rare and are considered to be of metaplastic nature.

High-grade MLPS shows >5% of the tumor with cellular overlap, decreased myxoid matrix, less apparent capillary vascularization, high nuclear grade, and increased mitotic activity. A cord-like or trabecular arrangement is often present.

Pure high-grade MLPS may be impossible to distinguish from other round cell sarcomas, requiring molecular genetic studies for diagnosis. The presence of >5% hypercellularity is associated with a significantly worse prognosis (differentiation score of 3 in the FNCLCC grading system – Fédération Nationale Française des Centres de Lutte Contre le Cancer). The presence and percentage of hypercellular areas must be reported.

Some cases show so-called transition zones with modestly increased cellularity without high nuclear grade or increased mitotic activity. Immunohistochemistry plays a minor role in the diagnosis of MLPS but may have some value in distinguishing high-grade tumors from other round cell sarcomas.

4- Pleomorphic liposarcoma:

This is a high-grade pleomorphic sarcoma containing a variable number of pleomorphic lipoblasts. No areas of ALT/WDLPS or other lines of differentiation are present. Macroscopically: most tumors are large, with a median size of 8 to 10 cm. They are well delineated but non-encapsulated, or poorly defined and infiltrative, and sometimes multinodular. On cut section, most tumors are white to yellow. Myxoid changes and foci of necrosis are often observed. From a histopathological point of view, most cases have infiltrative margins, and all tumors contain a variable proportion of pleomorphic lipoblasts on a background of high-grade undifferentiated sarcoma, generally pleomorphic. The presence of lipoblasts is required for diagnosis, but their number varies considerably from case to case and from one area to another within the same tumor, highlighting the importance of adequate sampling. In most cases, the non-lipogenic component resembles an undifferentiated pleomorphic sarcoma with spindle cells and multinucleated giant cells arranged in short fascicles, with some notable features: namely, the presence of extremely large tumor cells often showing clear or vacuolated cytoplasm, and the presence of extracellular and sometimes intracellular eosinophilic hyaline droplets. Nearly half of cases contain at least one focal area similar to intermediate- to high-grade myxofibrosarcoma-like areas associated with pleomorphic lipoblasts. This myxofibrosarcoma-like component is predominant in some cases. An epithelioid morphology is observed in approximately one quarter of cases, with areas resembling poorly differentiated carcinoma, clear cell renal cell carcinoma, adrenocortical carcinoma, or melanoma. Necrosis is present in more than half of cases. Unlike dedifferentiated liposarcoma with homologous differentiation, staining for MDM2 and CDK4 is generally negative in pleomorphic liposarcoma. The epithelioid subtype may be positive for keratins and melan-A.

5- Myxoid pleomorphic liposarcoma:

Myxoid pleomorphic liposarcoma is an exceptionally rare, aggressive adipocytic neoplasm, usually occurring in children and adolescents. Myxoid pleomorphic liposarcoma shows mixed histological features of conventional myxoid liposarcoma and pleomorphic liposarcoma and lacks the gene fusions and amplifications seen in myxoid liposarcoma, atypical

lipomatous tumor (ALT), and dedifferentiated liposarcoma (DDLPS). Macroscopically, myxoid pleomorphic liposarcomas are non-encapsulated tumors with poorly defined margins. Histopathologically, the tumors show variable proportions of myxoid liposarcoma-like areas, characterized by abundant myxoid matrix, scattered lipoblasts, relatively bland primitive round to oval cells, and a delicate curvilinear to plexiform capillary network. Lymphangioma-like myxoid areas may be observed. Pleomorphic spindle or ovoid cells with hyperchromatic nuclei may be scattered within the myxoid component, with a progressive transition to more cellular, high-grade pleomorphic liposarcoma-like areas, showing marked cytological atypia, increased mitotic activity, atypical mitoses, pleomorphic lipoblasts, and occasional necrosis. Myxoid pleomorphic liposarcomas have a non-specific immunophenotype. [2,21-24]

Main advances in this edition:

This classification introduces important modifications compared with previous versions, with the first major modification being that "round cell liposarcoma" is no longer considered a distinct entity, but rather as the high-grade form of myxoid liposarcoma. It also allowed the introduction of new diagnostic entities, among which the recognition of myxoid pleomorphic liposarcoma is cited first. This entity shows mixed histological features between myxoid liposarcoma and pleomorphic liposarcoma, but lacks the typical genetic abnormalities of these entities (absence of FUS-DDIT3 fusion, absence of MDM2 amplification) Another entity has been recognized and remains an important differential diagnosis, namely the atypical spindle cell/pleomorphic lipomatous tumor (Atypical Spindle Cell/Pleomorphic Lipomatous Tumor). This represents one of the most notable additions. This new category allows classification of adipocytic tumors that present intermediate features, not fitting perfectly with either classical atypical lipomatous tumor (ALT) or pleomorphic or dedifferentiated liposarcoma. In addition, this classification has enabled refinement of prognostic criteria. Thus, for dedifferentiated liposarcoma (DDLPS), it provides new clinically significant prognostic information and describes the recognition of low-grade dedifferentiation (not to be confused with sclerosing-type ALT) and homologous lipoblastic differentiation. The distinction between these different aspects has both prognostic and therapeutic importance. Furthermore, it reinforces the role of molecular pathology, as it is increasingly based on molecular characteristics, thereby consolidating its central role in the diagnosis of liposarcomas. For example, amplification of MDM2 and CDK4 is confirmed as the nearly constant genetic abnormality defining the ALT/WDLPS and DDLPS spectrum. Detection of this amplification by immunohistochemistry (MDM2/CDK4) or by FISH is an essential diagnostic tool for distinguishing these entities from other liposarcomas and from benign lesions.

On the other hand, FUS-DDIT3 / EWSR1-DDIT3 translocations remain the pathognomonic feature of myxoid liposarcoma. [2,21-24]

CONCLUSION

Liposarcoma is the most common soft tissue sarcoma in adults and represents the most frequent primary retroperitoneal tumor. Myxoid liposarcoma is the second most frequent subtype among all liposarcomas (all locations included) and is the most frequent among retroperitoneal liposarcomas. On imaging, the diagnosis of myxoid liposarcoma is associated with numerous difficulties, with sometimes problems of topographic diagnosis and similarities with numerous radiological differential diagnoses. It presents as a large, well-defined mass, slowly evolving in an insidious manner. Visualization of a fatty component suggests the diagnosis of liposarcoma; the myxoid component is characteristic, with an intermediate fluid-like attenuation, and the solid or vascularized component has a poorer prognostic value. Finally, understanding the histological subtypes is crucial in the approach to these tumors. In this regard, the 2020 WHO classification represents a major evolution, allowing refinement of the classification of liposarcomas by relying on advances in molecular genetics for improved diagnostic accuracy. Former round cell liposarcomas are now included within the myxoid liposarcoma type, and new entities have been formally introduced, such as the recognition of myxoid pleomorphic liposarcoma, which presents mixed histological features between myxoid liposarcoma and pleomorphic liposarcoma but lacks the typical genetic abnormalities of these entities.

REFERENCES

1. Terrier, P. (2012). Les liposarcomes. Annales de Pathologie, 32(5), e59–e68. https://doi.org/10.1016/j.annpat.2012.07.004
2. WHO Classification of Tumors 5th Edition, Soft tissus and Bone Fifth edition. International Agency for Research on Cancer, 2020. ISBN 9789283245025 (pbk.) | ISBN 9789283245032 (ebook)
3. Hinshaw, J. L., & Pickhardt, P. J. (2008). Imaging of primary malignant tumors of peritoneal and retroperitoneal origin. In Cancer Treatment and Research (pp. 209–230). Springer US. https://doi.org/10.1007/978-0-387-75587-8_11
4. Dariane, C *et al.*,Le point sur les masses rétropéritonéales : pièges à éviter et recommandations de prise en charge. Progrès en Urologie - FMC, 28(15), 826–833. https://doi.org/10.1016/j.fpurol.2018.08.001
5. Penel, N., & Cazin, J. (2020). Sarcomes. In Pharmacie Clinique Pratique en Oncologie (pp. 381–391). Elsevier. https://doi.org/10.1016/b978-2-294-76375-5.00022-1

6. Vilcot, L *et al.,*Comment je raisonne devant une masse rétropéritonéale. Medecine Nucleaire-imagerie Fonctionnelle Et Metabolique, 41(4), 241–243. https://doi.org/10.1016/j.mednuc.2017.06.001
7. Mujtaba, 2019 Mujtaba, B., Patel, D., & Speth, J. (2019). Myxoid Liposarcoma With Skeletal Metastases: Pathophysiology and Imaging Characteristics. Current problems in diagnostic radiology, 48(6), 596-600.
8. Imagerie abdominale, 1re édition, de Michael P. Federle, Siva P. Raman, traduit par Nicolas Badet. © 2020 Elsevier Masson SAS. ISBN: 978-2-294-75344-2 e-ISBN: 978-2-294-75640-5.
9. Chouli, M., M. T. Cherif, A. A. Y. Abdou, A. Taleb, & A. Abidia. (2006). Liposarcome du rétropéritoine. Feuillets De Radiologie, 46(1), 39–46. https://doi.org/10.1016/S0181-9801(06)70518-8
10. Tattersall, H., *et al.,* (2020). Primary mesenteric sarcomas: Collaborative experience from the Trans-Atlantic Australasian Retroperitoneal Sarcoma Working Group (TARPSWG). *Journal of Surgical Oncology*. https://doi.org/10.1002/jso.26353
11. Xiao, J., Zhu, Y., Niu, Z., Chen, Y., Guan, W., & Song, Y. (2020). Diagnosis and Prognosis of retroperitoneal Liposarcoma: A Single Asian Center Cohort of 57 Cases. *Journal of Oncology*, 2021, 7594027.
12. Zhang, Z., Li, Y., Gao, Y., & Wang, J. (2021). Giant retroperitoneal myxoma: A case report and literature review. International Journal of Surgery Case Reports, 82, 106055.
13. Nishino, M., Hayakawa, K., Minami, M., Yamamoto, A., Ueda, H., & Takasu, K. (2003). Primary retroperitoneal neoplasms: CT and MR imaging findings with anatomic and pathologic diagnostic clues. Radiographics, 23(1), 45-57.
14. Sung, M. S., *et al.,*(2000). Myxoid liposarcoma: appearance at MR imaging with histologic correlation. *Radiographics*, 20(4), 1007-1019.
15. Garg PK, Jain BK, Dahiya D, Bhatt S, Arora VK, Liposarcome M. Rapport de deux cas avec revue de la littérature. J Gastro Intest Cancer 2014 ;45(1):170–4.
16. Soufi M, Mdaghri J, Benamr S, Lahlou MK, Massrouri R, Essadel A, *et al.,*Liposarcome géant de l'épiploon mimant une tumeur ovarienne. Un rapport de cas. Indian J Surg 2012 ;74(5):425, 10.1007/s12262-012-0718-y.
17. Benjaminov O, Gutman H, Nyabanda R, Keinan R, Levavi GSH. Liposarcome myxoïde : une présentation inhabituelle. Am J Roentgenol 2007 ;188 :817–21. Doi :10.2214/AJR.05.0246.
18. Hashimoto S, Arai J, Nishimuta M, Matsumoto H, Fukuoka H, Muraoka M, *et al.,*Résection d'un liposarcome du grand épiploon : à propos d'un cas et revue de la littérature. Int J Surg Case Rep 2019 ; 61 : 20–5. Doi : 10.1016/j.ijscr.2019.06.067.
19. Mulita F, Verras GI, Liolis E, Tchabashvili L, Kehagias D, Kaplanis C, *et al.,*Liposarcome rétropéritonéal récurrent : à propos d'un cas et revue de la littérature. Clin Case Rep 2021 ;00:04717e. Doi :10.1002/ ccr3.4717.
20. Harhar M, Ramdani A, Bouhout T, Serji B, Harroudi Tél. Liposarcome myxoïde : à propos d'un cas d'une localisation rare de la paroi abdominale. Cureus 2020 ;12(6):1–5. Doi : 10.7759/cureus.8715.
21. Sachidananda S, Kumar Krishnan A, Ramesh R, Kuppurao S. Liposarcome mésentérique multiple primaire du mésocôlon transverse. Ann Coloproctol 2013 ;29(3):123–5.
22. Karanian M, Coindre J-M. Quatrième édition de la classification OMS des tumeurs des tissus mous. Annales de pathologie (2014), http://dx.doi.org/10.1016/j.annpat.2014.11.003
23. Choi, J. H., & Ro, J. Y. (2021). The 2020 WHO Classification of Tumors of Soft Tissue: What You Need to Know. Korean Journal of Pathology, 55(1), 1–15. https://doi.org/10.4132/jptm.2020.12.28
24. Kallen, M. E., & Hornick, J. L. (2021). The 2020 WHO Classification of Tumors of Soft Tissue: An Update and Emphasis on the New Entities. The American Journal of Surgical Pathology, 45(5), 563–577. https://doi.org/10.1097/PAS.0000000000001663
25. Lam, S. W., Briaire-de Bruijn, I. H., Cleven, A. H. G., *et al.,*(2022). Myxoid pleomorphic liposarcoma: a clinicopathological, immunohistochemical and molecular genetic study of 11 cases. Histopathology, 80(3), 479–490. https://doi.org/10.1111/his.14574