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OVERVIEW OF PRIMARY ADULT RETROPERITONEAL TUMOURS

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ABSTRACT

In front of a primary retroperitoneal tumour, it is necessary to have in mind all possible diagnoses in order to specify the diagnostic strategy and the treatment. According to the World Health Organization (WHO) classification of tumours, mesenchymal benign and malignant tumours (including sarcomas and, currently, neurogenic tumours), parasympathetic tumours, extragonadal germ cell tumours, and lymphoid tumours have been identified. By definition, primary retroperitoneal tumours start independently from the retroperitoneal organs. Secondary lesions, carcinoma metastasis, and adenopathy are excluded from this definition, but they can also develop in the retroperitoneal space and lead to misdiagnoses. In the absence of positive tumour markers or an evocative biology, percutaneous biopsy is necessary. Pathological diagnosis is necessary to decide whether surgery must be done, its timing among the other treatments, and its extension. This paper summarizes all the diagnostic possibilities.

1. INTRODUCTION

By definition, primary retroperitoneal tumours start independently from the retroperitoneal organs. Secondary lesions, carcinoma metastasis, and adenopathy are excluded from this definition, but they can also develop in the retroperitoneal space and lead to mistakes during the diagnostic process. According to the World Health Organization (WHO) classification of tumours, mesenchymal tumours (including sarcomas and, currently, neurogenic tumours), parasympathetic tumours, extragonadal germ cell tumours, and lymphoid tumours have been identified. Cystic vestigial tumours are classified according to the embryonal tissue of origin. The retroperitoneal space is limited posteriorly by the vertebral plane and the posterior parietal muscles and anteriorly by the posterior parietal peritoneum; the retroperitoneal space extends from the diaphragm to the pelvic floor. The therapeutic strategies, including both medical and surgical treatment, and even the type and extent of surgery may differ significantly among the different varieties of cancer, and it is therefore crucial to have a reliable diagnosis before considering surgery. Usually, percutaneous biopsy is needed unless tumour markers can provide a diagnosis. Furthermore, even pathologic diagnosis can be challenging. Frozen examinations during surgery, without the contribution of immunohistochemistry and molecular biology, are not recommended and have a risk of detrimental tumour rupture in cases of sarcomas. The aim of this paper is to summarize the principal entities originating from the retroperitoneum that should be considered during the diagnosis process and to describe their principal features, with a special emphasis on mesenchymal tumours – the most frequent tumour in this region. The other histological entities are just briefly mentioned, since they can lead to misdiagnoses, especially when the primary tumour is unknown.

2. MESENCHYMAL TUMOURS

Mesenchymal tumours account for approximately half of all retroperitoneal primary tumours. The WHO 2013 classification is currently used [1], and an update that will be published during 2020. This system is an analogical classification based on the tumour differentiation line. Each subgroup includes varieties that are strictly benign or malignant, or occasionally, those that are intermediate - locally aggressive or with low metastatic potential – such as desmoid tumours or solitary fibrous tumours. Furthermore, this classification also includes a group of tumours with uncertain differentiation, such as

PEComas and deep angiomyxomas (the latter being often infra-peritoneal). Some subtypes that are extremely rare in the retroperitoneum are not currently mentioned. Overall, liposarcomas (LPS) and leiomyosarcomas (LMS) represent approximately 80% of all adult retroperitoneal sarcomas. As nearly half of sarcomas show specific genetic abnormalities, such as translocations, amplifications, and activating or inactivating mutations, molecular biology can help diagnose the tumour or confirm the diagnosis. It is clear that this great variety of different entities, all with specific features and treatments, needs a specialized setting of care. It is therefore recommended that once these tumours are diagnosed, they should be discussed among a dedicated multidisciplinary tumour board to determine a therapeutic strategy [2]. Moreover, the pathologic diagnosis must be validated by an expert pathologist in a reference centre or network. Pathology review can correct the diagnosis in up to 20% of cases.

2.1. Symptoms

There is often a mismatch between tumour volume and the small number of symptoms when the mass is found. The median size at diagnosis is approximately 20 cm [3], and smaller masses are often incidentally found during diagnostic tests for unrelated symptoms. Most frequently, an increasing abdominal circumference and palpable mass are the first signs. It is not uncommon for patients to go on a diet and show biological markers of malnutrition, which need to be investigated before surgery [4]. Weight loss, due to digestive symptoms or diet, can be balanced by the increasing weight of the tumour, and body mass index alone is unreliable and uninformative. Signs of extrinsic compression – such as phlebitis, oedema of the lower limbs, nerve compression, and vague digestive and urinary symptoms – could be found depending on the maximum tumour extent and localization. Often, patients have presented with gastric repletion sensations and difficult feeding due to gastric and digestive compression.

2.2. Imaging

The most frequent mistakes during imaging analyses are to underestimate the extension of the well-differentiated part of a LPS that presents both a well-differentiated and a dedifferentiated component

and to miss extension across abdominal wall openings such as the inguinal ligament [5]. Another mistake is to consider multifocal a unique tumour, by focusing only on the dedifferentiated parts without seeing the well-differentiated part that holds the whole mass together (Fig. 1).

On the CT scan, the mass is located behind the colon, which is displaced forward. However, the tumour can cross the midline and completely shift contralaterally the digestive tract. The kidney can be displaced forward, and it is necessary to systematically compare its position and that of the contralateral kidney.

Fatty tumours may be completely well differentiated, resembling normal fat. In these cases, regardless of its dimensions, the tumours are most frequently well-differentiated LPS, lipomas, extrarenal angioliipomas and myeloliipomas. Quite often, well-differentiated liposarcomas present internal fibrous septa. The kidney may be encased or displaced forward but is rarely infiltrated (unlike with angiomyoliipomas). However, the differential diagnosis of myeloliipomas can be challenging, as these tumours present within the fat marrow that appears as frosted glass after contrast enhancement and can be mistaken as the dedifferentiated part of a LPS (Fig. 2). Angiomyoliipomas show internal vessels. These tumours can be associated with a notch or an infiltration of the renal hilum, which suggests their origin is the kidney and excludes a strictly primary retroperitoneal origin (Suppl Fig. 1).

LPS with both well-differentiated and dedifferentiated components are often poorly evaluated and described as multifocal masses because only the dedifferentiated components are seen, while the well-differentiated part that connects them is not recognized. An incomplete report and analysis describing only the dedifferentiated components can result in incomplete surgery (even without the surgeons' awareness) or fragmentary surgery, which definitively worsens the prognosis (Suppl Fig. 2 and 3).

According to the guidelines of the European Society of Medical Oncology (ESMO), the National Cancer Comprehensive Network (NCCN) and the Trans-Atlantic Retro Peritoneal Sarcoma Working Group (TARPSWG) [7 - 9], if the tumour markers are negative, it is mandatory to perform a preoperative percutaneous biopsy with a coaxial 16G needle to define or confirm the diagnosis and decide among completely different surgical strategies (enucleation for benign tumour, compartmental surgery for sarcomas, lymph node dissection in case of carcinoma) and to discuss, in certain cases, available medical treatments. The biopsy should be performed through a retroperitoneal route with a

coaxial needle to avoid peritoneal seeding. Surgical biopsy, both open and laparoscopic, is contraindicated because it can lead to tumour rupture, which exposes the patient to a risk of peritoneal sarcomatosis and abdominal wall seeding.

The morbidity of percutaneous biopsy is very low. In a recent series [10], of 358 percutaneous CNBs of suspected RPS performed over the study period, 7 (2.0%) resulted in minor bleeding with no transfusion, 3 (0.8%) resulted in significant pain, 1 (0.3%) resulted in unplanned admission to hospital for observation, and 1 (0.3%) resulted in a pneumothorax. There were no infections. In 203 patients who underwent resection of RPS following CNB, there was one case of needle tract seeding at a median follow-up of 44 months. In another recent series [11], no patient developed a biopsy site recurrence.

It is mandatory to evaluate tumour extension across abdominal borders and infiltration into adjacent muscles because these factors can modify the choice of the surgical route: special attention should be paid to the region below the inguinal ligament (where extension into the inguinal channel may be misdiagnosed as inguinal hernia), to the sciatic notch, across the diaphragmatic hiatus and intervertebral foramina. Infiltration into the adjacent muscles (oblique muscle, ilio-psoas, and diaphragm) should be investigated by magnetic resonance imaging (MRI). Bone involvement should be carefully evaluated: it may be limited compared to soft tissue extension, but the presence of bone involvement may change the tumour origin (such as the case of skeletal versus extraskelatal Ewing's sarcoma), resulting in different surgical options. Moreover, MRI is the best technique to define the anatomical extent of infraperitoneal tumours.

As the lungs are the first metastatic site, a preoperative chest CT scan should be performed in all sarcoma patients before deciding on a surgery plan. PET scans may be useful for high-grade sarcomas, but the PET findings can be normal or inconsistent for low-grade tumours. The risk of nodal spread is low except for certain specific subtypes (such as rhabdomyosarcoma and extraskelatal Ewing's sarcoma). Well-differentiated liposarcoma never causes visceral metastasis, but it can cause peritoneal sarcomatosis, especially if tumour rupture occurs during surgery.

2.3. Classification

2.3.1. Fatty tumours

Fatty tumours represent the vast majority of mesenchymal retroperitoneal tumours (approximately 60%) [3]. Among malignant tumours, two subtypes of LPS are described in the retroperitoneum: well-differentiated/dedifferentiated LPS and pleomorphic LPS. Benign tumours are represented by angiomyolipomas, myelolipomas, and lipomas. The denomination “atypical lipomatous tumour”, which refers to well-differentiated LPS, is used only for limb and superficial trunk localizations and should therefore not be used in the retroperitoneum; this term could be misleading since patients may die of iterative local recurrences that may finally become unresectable. Well-differentiated and dedifferentiated LPS present amplification of the MDM2 gene that can be identified by fluorescence in situ hybridization (FISH) and may confirm a challenging diagnosis. Pleomorphic LPS shows complex genetics, without specific abnormalities.

2.3.2. Smooth muscle tumours

Benign retroperitoneal and infraperitoneal leiomyomas are uncommon, occurring mainly in women. LMS account for approximately 20% of retroperitoneal sarcomas [3]. These tumours are sarcomas with complex genetics and high genomic instability. The metastatic risk is remarkable.

In clinical practice, retroperitoneal LMS must be considered if the tumour originates from retroperitoneal vessels. Immunohistochemistry of these tumours shows the expression of smooth muscle actin, desmin and h-caldesmon but is negative for S100. Metastasis from a previously resected uterine sarcoma that was previously wrongly qualified as leiomyoma should be ruled out. A review of the initial specimen can help and positive results from hormone receptor expression can be useful to identifying a gynaecologic origin.

2.3.3. Fibroblastic and myofibroblastic tumours

This group of tumours includes benign tumours often seen in the retroperitoneum as calcifying fibrous tumours, as well as tumours classified as “intermediate” both for their locally aggressive potential, such as desmoid tumours, and for their uncertain metastatic potential, such as solitary fibrous tumours. Malignant fibroblastic tumours and myxofibrosarcomas rarely occur in the retroperitoneum.

It is crucial to obtain a diagnosis of desmoid tumour as the therapeutic strategy has dramatically changed over the last 10 years, with observation or medical treatment being now recommended as the first option (Fig. 3, 4) [12]. CTNNB1 gene (coding beta-catenin) mutations should be investigated routinely to confirm the diagnosis. This mutation is present in up to 95% of cases and may have prognostic value, as the presence of the 45F mutation could be associated with a worse evolution. In nearly 50% of cases, tumours spontaneously stabilize or regress, justifying a primary nonaggressive strategy to avoid over treating the patients. In patients suffering from adenomatous polyposis coli (APC), a mass arising from the root of the mesentery is a highly suspicious sign. The current strategy is active surveillance initially, especially in patients with a history of colectomy and ileal pouch and imaging findings that underestimate the tumour.

Solitary fibrous tumours, previously known as “haemangiopericytomas”, can be identified with imaging because of the large drainage veins surrounding the tumour. SFT account for approximately 6% of retroperitoneal sarcomas [3]. The diagnosis can be confirmed with percutaneous biopsy based on the expression of CD34 and STAT6 in the tumoral cells. This tumour is associated with the specific transcript NAB2-STAT6. The majority of these tumours have a low metastatic risk, which is defined by the age of the patient, tumour size and number of mitoses.

2.3.4. Striated muscle tumours

Adult rhabdomyoma is a benign tumour that occurs on the head and neck and has never been described in the retroperitoneum. Embryonal rhabdomyosarcoma, which is most commonly observed in children and adolescents, may be observed even in young adults in both the retroperitoneal and infraperitoneal space. In children and adolescents, half of the cases occur close to the genitourinary tract. In a minority of cases, these tumours arise because of genetic syndromes, especially when a germline mutation involving RAS or NF1 pathways is present. Multiple chromosomal rearrangements are present in sporadic cases. These tumours typically present lymph node diffusion, and the patients can benefit from induction medical treatment. Alveolar rhabdomyosarcoma is less common than embryonal rhabdomyosarcoma but also affects young adults; however, alveolar rhabdomyosarcomas are rarely described in the retroperitoneum. On the other hand, pleomorphic rhabdomyosarcoma, which affects adults with an average age of 60 years old, can be diagnosed in the retroperitoneum. This sarcoma, which has complex genetics, has a poor prognosis and high metastatic risk.

2.3.5. Vascular tumours

Among benign lesions found in the retroperitoneum, haemangiomas are uncommon, while cystic lymphangiomas are more frequently observed. This second entity presents as a cystic cavity well demarcated by septa. These tumours progressively grow and can reach large volumes that may compress the surrounding structures (Fig. 5). The diagnosis is based on radiological findings, and percutaneous biopsy is usually not recommended. On pathological examination, septa are found consisting of lymphatic vessels of different sizes surrounded by lymphocytes. Large, cystic lymphangiomas can surround and encase other organs, making surgical resection difficult. If tumour rupture occurs, the risk of leaving tumour fragments is high, resulting in the risk of local recurrence. Although there is no risk of malignant transformation, local recurrence could significantly worsen the patient's quality of life.

Malignant vascular tumours are essentially represented by angiosarcoma, with the retroperitoneum as the second most frequent location following the limbs. Clinically, angiosarcomas are much more symptomatic than liposarcomas, with episodes of pain following intratumoral haemorrhage. Subsequent haematomas can cause diagnostic difficulties. In one-third of patients, anaemia or coagulopathy is present. The vast majority of angiosarcomas are high-grade tumours, and a multidisciplinary evaluation is mandatory. In addition to age, tumour size, and Ki67 index, retroperitoneal location is related to worse prognosis.

2.3.6. Nerve tumours

In the retroperitoneum, schwannoma (Suppl Fig. 4) is the most frequent benign nerve tumour, while malignant peripheral nerve sheath tumour (MPNST) is the most frequent malignant entity. There are no radiological features that can clearly distinguish between these two entities. Benign sporadic schwannoma has no specific known aetiology. Multilevel schwannomas may be evocative of type 2 neurofibromatosis. In the retroperitoneum, a mass originating proximally to the spine, in the psoas muscle, or involving the neural foramina (which must be systematically checked) suggests a neural origin, but these findings are not specific. On immunohistochemistry, diffuse expression of S100 and sox10 in the tumoral cells is characteristic of schwannomas. There is no risk of malignant transformation.

MPNST arise from peripheral nerve neurofibromas in patients affected by type 1 neurofibromatosis. A suddenly fast growing previous neurofibroma is highly suspect. Among these patients, those presenting plexiform neurofibromas have a higher risk of malignant transformation. MPNST presents at a younger age in patients suffering from type 1 neurofibromatosis than in sporadic cases. Clinically, the tumour is discovered after the appearance of symptoms such as neurologic pain, motor deficits and paraesthesia and is usually smaller than liposarcomas.

2.3.7. Extraskelatal osseous and cartilaginous tumours

Retroperitoneal localization is extremely rare. Skeletal involvement should be carefully evaluated, as it could be much less visible than extension in the retroperitoneal soft tissues. However, the origin (bone or soft tissue) determines the surgical route.

2.3.8. Uncertain tumour differentiation

Among benign tumours of uncertain differentiation, deep angiomyxoma (Fig. 6) is the most frequent. These tumours are usually localized in the infraperitoneal space and mainly affect women. A main feature is a dissonance of large masses discovered with few symptoms, except perineal masses. These tumours usually arise from the perineum, often inside the labia major, and extend into the ischiorectal space along the pelvic organs to the sacral promontory and higher regions. As these are hypervascular tumours, the skin is erythaematous, often resulting in a misdiagnosis of Bartholin's cyst. On MRI, the characteristic findings include a "combed" appearance on T2 sagittal and coronal images. Although rare, these tumours can infiltrate the pelvic organs, leading to reconstruction issues. These tumours are soft and compressible under pressure, and there is a risk for incomplete gross resection. The surgical objective is conservative surgery in the absence of organ infiltration (vagina, anal sphincter, bladder, etc.) [13]. When facing a residual mass, which occurs frequently after unplanned surgery, observation is an option until significant growth occurs.

Among other rare tumours of uncertain differentiation, PEComas, which are composed of epithelioid perivascular cells, could be both malignant and benign. The cells of these tumours are associated with vessel walls and usually express melanocytic and smooth muscle markers. PEComas include angiomyolipomas and lymphangiomyomatosis, and a group of tumours showing the same histopathologic features can be found throughout the body. Most PEComas other than angiomyolipomas and lymphangiomyomatosis occur sporadically. A small proportion can be

associated with tuberous sclerosis complex (TSC, or Bourneville disease). The metastatic risk ranges from low to moderate. Although there are no established malignancy criteria, tumours with large dimensions, prominent nuclear atypia, high mitotic rate, and necrosis have a higher risk for malignant evolution. The metastatic spread of these tumours significantly differs from that of other sarcomas, with mainly liver, nodes and bone involvement.

2.3.9. Unclassified/undifferentiated sarcomas

These sarcomas without a definite line of differentiation are rare in the retroperitoneum on opposite to extremities.

2.4. Dissemination

Up to 80% of patients die of peritoneal dissemination, which eventually occurs after tumour rupture during inadequate surgery. Leiomyosarcomas and dedifferentiated liposarcomas metastasize more frequently, and the lung is the first site of metastasis. Nodal spread is infrequent, except for certain tumour subtypes, such as rhabdomyosarcoma.

2.5. Treatment of retroperitoneal sarcomas

The cornerstone of treatment for retroperitoneal sarcomas is en bloc surgery with clear margins on the largest surface [8]. It is mandatory to resect the tumour without fragmenting the mass. Indeed, any tumour rupture exposes the patient to peritoneal dissemination, which definitively worsens the prognosis [14]. In most cases, a compartmental surgery is performed, which comprises resection of the tumour with the kidney inside, the colon ahead and the muscular fascia behind [15 – 17]. Adjustments are made according to the histological subtypes. Indeed, the best indications of compartmental surgery are the subtypes with the highest risk for local recurrence in patients who can undergo extended surgery. This surgery must be performed in a referral centre after receiving confirmation of the pathologic diagnosis from a review performed by a sarcoma expert pathologist in reference centres or reference networks. Indeed, in the literature, the data show that centralized surgery could improve the 5-year survival rate by more than 20% [18]. Radiological interpretation mistakes often lead to inadvertent incomplete macroscopic surgery performed by non-specialized

surgeons. Tumour rupture (or macroscopic incomplete resection) definitely worsens the prognosis, exposing the patient to ultimately nonresectable peritoneal dissemination. Salvage surgery never returns the patient to an optimal prognosis.

Preoperative radiation therapy has recently been investigated by an EORTC trial (STRASS 1) and was shown to have significant survival benefits for liposarcomas in a subgroup analysis [19]. Postoperative radiation therapy should not be used because it exposes the patient to digestive morbidity. Chemotherapy is indicated for certain tumour subtypes, such as extraskeletal Ewing sarcoma and rhabdomyosarcoma. For the most common subtypes (liposarcoma and leiomyosarcoma), chemotherapy is currently not a standard of care when the tumour is resectable. An EORTC randomized trial (STRASS 2) is now ongoing to evaluate the role of preoperative chemotherapy [20]. In recurrent disease, the best indications for surgery are low-grade tumours and a large interval time between primary resection and recurrence. The indications for surgery must be discussed by a multidisciplinary tumour board. Anthracycline-based first-line chemotherapy is the standard of care for non-resectable and metastatic patients [21, 22].

2.6. Prognosis of retroperitoneal sarcomas

A correct diagnostic process, a systematic discussion of cases by multidisciplinary tumour boards and surgery in referral centres allow for significantly better survival [2, 18, 23, 24]. For patients treated in referral centres, the 5-year overall survival rate is approximately 70% [3]. However, important differences exist among different subtypes. Well-differentiated liposarcoma has the best prognosis, with a very low metastatic risk. For all subtypes, the 5-year risk of local recurrence is approximately 30%, and the metastatic risk is 20% [3]. Low-grade, well-differentiated liposarcoma mainly leads to local recurrence; while leiomyosarcoma and high-grade dedifferentiated liposarcoma expose the patients to distant metastasis. This must be taken into account while planning the extent of the surgery.

3. EXTRAGONADAL GERM CELL TUMOURS

Germ cell tumours primarily originate in the testicles and ovaries, but metastatic adenopathy is often the first manifestation and is part of the differential diagnosis of a retroperitoneal mass. Nevertheless, in approximately 2% of cases, germ cell tumours originate from an extragonadal site (Suppl Fig. 5), and the retroperitoneum is the second most common location after the mediastinum. Including both primary and secondary tumours, germ cell tumours account for approximately one quarter of all retroperitoneal tumours. For both primary and secondary tumours, serum tumour marker detection could help with the diagnosis (alpha-fetoprotein, human chorionic gonadotropin). On imaging, a polycyclic mass is frequently seen close to the large vessels, but a more well-defined lesion can also be observed. If the tumour markers are negative, a percutaneous biopsy must be performed to obtain a certain diagnosis and determine a therapy plan. The 2016 WHO classification is currently used [25] to identify seminoma and nonseminoma tumours, including embryonal carcinoma, choriocarcinoma and teratomas. Dermoid cysts correspond to mature cystic teratomas. The management of malignant tumours may include neoadjuvant chemotherapy, which should be determined by a multidisciplinary tumour board. As these tumours are considered rare gynaecologic malignancies, women should be treated by dedicated reference networks, undergo a systematic pathologic review and received a multidisciplinary discussion.

4. LYMPHOID TUMOURS

Lymphomas represent approximately one quarter of all retroperitoneal primary tumours. These patients may not have extra-abdominal adenopathy. On imaging, these tumours appear as polycyclic periaortic or pericaval, T1 isointense and T2 iso- or hyperintense lesions. The encasement of major vessels (vena cava and aorta) without occlusion is highly suggestive of these tumours (Suppl Fig. 6) and helps differentiate lymphomas from big vessel malignancies. Serum lactate dehydrogenase (LDH) could be elevated and an evocative sign, but percutaneous biopsy is mandatory to confirm the diagnosis and identify the specific subtype. Chemotherapy is the standard of care.

Castleman disease [26] is a benign lesion characterized by the presence of lymphocytic proliferation. The median age at presentation is 25 years, but these tumours can be observed even in elderly

patients. On imaging, a primary mass with satellite nodules and intense contrast enhancement is characteristic. Surgical resection is the standard of care for localized disease.

5. PARASYMPATHETIC TUMOURS

Mostly comprising paragangliomas, these tumours arise from the parasympathetic ganglion. The secretion of catecholamines can be present and should be investigated on clinical suspicion. Adrenal localization corresponds to pheochromocytoma [27]. Clinically, patients often show symptoms of catecholamine secretion, such as arterial hypertension, diarrhoea, and facial flushing. On imaging, these lesions appear as paraaortic or paracaval masses with large intratumoral vessels and rapid and intense contrast enhancement. The dimensions at diagnosis are often smaller than those of sarcomas since parasympathetic tumours are symptomatic earlier than sarcomas. The main treatment is surgery, with a specific anaesthesiologic preparation in case of hormone secretion. By definition, primary retroperitoneal tumours start independently from the retroperitoneal organs. However, one have to keep in mind adrenocortical carcinoma since the adrenal gland may not be visible.

6. VESTIGIAL CYSTIC TUMOURS

These are retrorectal tumours that are predominantly in females and are discovered incidentally or after the onset of rectal symptoms. Rarely, rectal fistulisation and suppuration are present. The main treatment is surgery. On imaging, a careful evaluation must be performed to exclude bone involvement, as this finding may indicate a sacral chordoma – which requires a completely different treatment and surgical option [28, 29].

7. CONCLUSIONS

During the diagnostic workup of a retroperitoneal mass, a large variety of histological findings in this location must be kept in mind; approximately half of these retroperitoneal masses are mesenchymal tumours. Tumoral markers may provide a diagnosis in some specific cases. In all other cases, percutaneous biopsy is needed to guide therapeutic strategy and define the extent of surgery,

especially in sarcomas, where en bloc surgery with clear margins performed in a reference centre is the standard of care.

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KEY POINTS

- *Retro- and infraperitoneal tumours: Percutaneous biopsy is the standard of care. In fact, the indications for and extent of surgery depends on the diagnosis.*
- *Both the early complication rate and the incidence of needle tract seeding of per cutaneous biopsy of an RP mass are very low.*
- *The well-differentiated components of liposarcomas are often underestimated, leading to incomplete surgery, which definitely worsens prognosis.*
- *The surgical standard of care for sarcoma is a complete, non-fragmented, en bloc resection with clear margins on the larger surface. Typical resection is a compartmental surgery, with en bloc resection of the kidney, colon, and psoas fascia. Adjustments are made according to the histological subtype.*
- *Sarcoma surgery must be performed in a referral sarcoma centre.*

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FIGURES

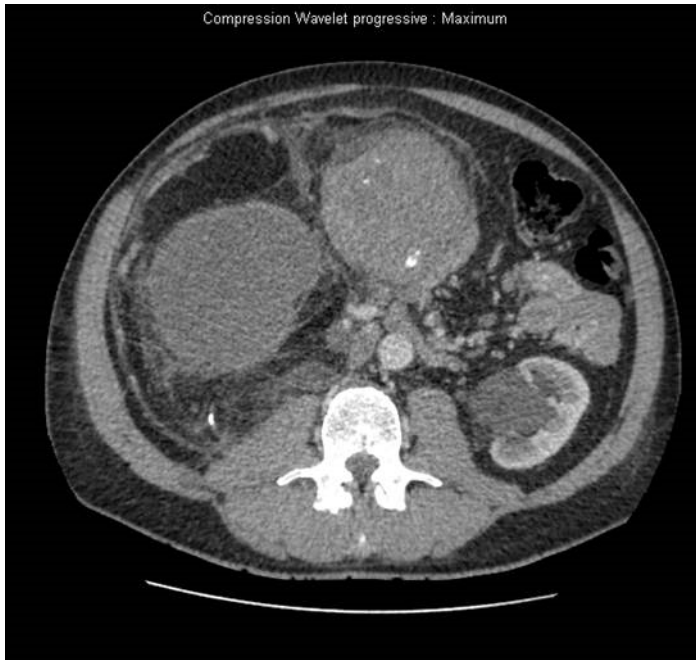


Figure 1: Dedifferentiated liposarcoma. One single mass comprising two dedifferentiated parts surrounded by the well differentiated part. [Single column fitting image]



Figure 2: Myelolipoma. The marrow inside the fat appears like frosted glass after contrast enhancement and can be mistaken as the dedifferentiated part of a liposarcoma. [Single column fitting image]

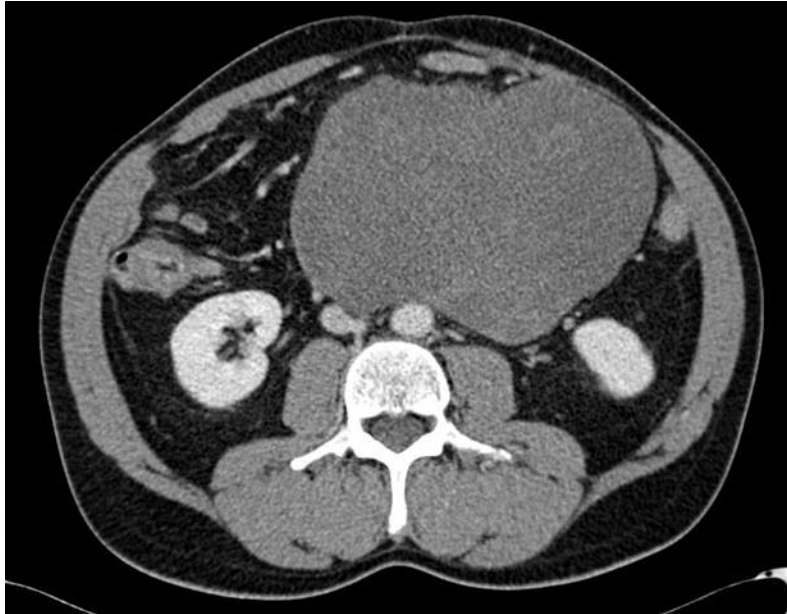


Figure 3: Intra-abdominal mass of 17 cm. Biopsy: Desmoid tumour with beta-catenin mutation Ser45Phe. Few symptoms were present, leading to a decision for active surveillance. [Single column fitting image]



Figure 4: Desmoid tumour. CT scan of the patient in Fig. 3 after 2 years of active surveillance. [Single column fitting image]



Figure 5: Cystic lymphangioma. [Single column fitting image]

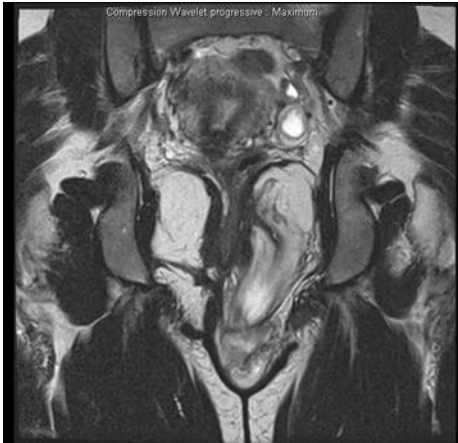


Figure 6: Deep angiomyxoma. [Single column fitting image]