11
Imaging of Primary Malignant Tumors of Peritoneal and Retroperitoneal Origin

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Key Points

- Primary malignancies arising from the peritoneal, subperitoneal and retroperitoneal spaces occur much less frequently than metastatic involvement from primary organ-based tumors or lymphoproliferative diseases.
- Nonetheless, these rare primary lesions should be considered in the absence of a known or suspected organ-based malignancy.
- Cross-sectional imaging can be useful for detection, characterization, staging, guiding biopsy for tissue diagnosis and evaluating response to therapy.

Abstract Peritoneal carcinomatosis and metastatic involvement of the retroperitoneum are relatively common manifestations of many organ-based malignancies and lymphoproliferative disorders. *Primary* malignancies of peritoneal and retroperitoneal origin occur much less frequently, and can be difficult to distinguish from metastatic disease. In many cases, a precise diagnosis based on imaging findings alone is not possible. However, the imaging features of these primary tumors, in combination with the clinical and demographic data, can be utilized to narrow the scope of the differential diagnosis. This chapter will present the clinical and imaging features of primary peritoneal and retroperitoneal tumors arising from the various tissue components that comprise the ligaments, mesenteries and connective tissues of these anatomic spaces.

1 Introduction

Peritoneal carcinomatosis is a relatively common manifestation of many organ-based malignancies, particularly of the GI tract and ovaries. Likewise, metastatic retroperitoneal lymphadenopathy and direct extension from an organ-based primary

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M.A. Blake and M.K. Kalra (eds.), *Imaging in Oncology.*

tumor are also common findings at imaging evaluation. Primary tumors of peritoneal and retroperitoneal origin occur much less frequently, but are often first identified on cross-sectional radiologic imaging studies, such as computed tomography (CT), ultrasound (US) or magnetic resonance imaging (MRI).

Neoplastic involvement of the peritoneum and retroperitoneum generally manifest with an abnormal increase in soft tissue, which can appear infiltrative or tumerous and be associated with variable amounts of cystic change, calcification, fatty composition, intravenous contrast enhancement and surrounding fluid. However, since many non-neoplastic and metastatic processes demonstrate similar imaging findings, the appearance of many primary malignancies of the peritoneum and retroperitoneum is nonspecific [1]. As a result, even in the absence of a known organ-based primary malignancy, metastatic disease is often the first consideration when confronted with an abnormal soft tissue process arising within the peritoneal or retroperitoneal space. However, primary malignancies should also be considered in this setting.

This chapter will present the salient clinical and imaging features of the majority of the primary neoplasms (Table 11.1) arising from the various tissue components that comprise the ligaments, mesenteries and connective tissues of the peritoneal and retroperitoneal spaces. The differential diagnosis for peritoneal-based and retroperitoneal-based neoplasms can often be refined by combining the imaging features with the patient's relevant clinical and demographic information. In addition to detection and characterization, cross-sectional imaging is useful for directing biopsy for tissue diagnosis.

2 Anatomic Considerations

The visceral and parietal peritoneum enclose the large potential space referred to as the peritoneal cavity. Pathologic processes that gain access to the peritoneal cavity can disseminate throughout this space via the relatively unrestricted movement of fluid and cells. Pathologic processes can also be disseminated within the subperitoneal space, which lies deep to the surface lining of the visceral and parietal peritoneum, omentum

Table 11.1 Primary Peritoneal and Retroperitoneal Malignancies

Primary Peritoneal Malignancies
Mesothelioma
Papillary serous carcinoma
Desmoplastic small round cell tumor
Malignant fibrous histiocytoma
Liposarcoma
Other mesenchymal tumors
Primary Retroperitoneal Malignancies
Liposarcoma
Leiomyosarcoma
Malignant fibrous histiocytoma
Other mesenchymal tumors
Paraganglioma
Extragonadal germ cell tumors

and the various peritoneal ligaments and mesenteries [2]. The subperitoneal space has both intraperitoneal and extraperitoneal components that bridge the peritoneum and retroperitoneum, which can result in bi-directional spread of disease processes. This concept helps to explain the involvement of both the peritoneal and retroperitoneal space that is sometimes encountered.

The retroperitoneal space is not defined by specific anatomic structures delineating its borders, but rather as the space posterior to the peritoneal cavity. Retroperitoneal structures may be defined as primary (e.g., retroperitoneal from the beginning of embryogenesis) or secondary (e.g., an area initially suspended by a mesentery during early embryogenesis that subsequently migrated and fused to become retroperitonealized). The extraperitoneal pelvis essentially represents the inferior continuation of the retroperitoneal space. The retroperitoneum and extraperitoneal pelvis represent a crossroads for a number of organ systems containing portions of the gastrointestinal and genitourinary tracts, as well as major vascular structures. The retroperitoneum, however, also contains intrinsic connective tissues, fat and neural elements.

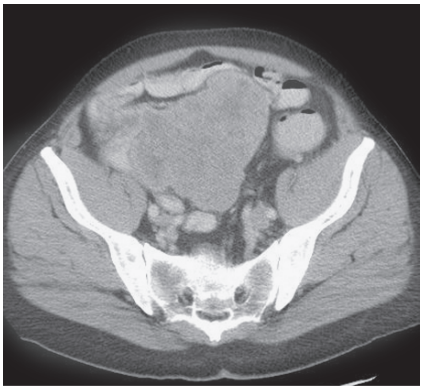
This chapter will focus on primary malignancies arising directly from the supporting tissues of the peritoneal, subperitoneal and retroperitoneal spaces, rather than tumors that arise from the organs contained within these spaces.

3 Primary Peritoneal Malignancies

3.1 Mesothelioma

Clinical Features. Mesothelial cells line the internal body cavities, including the pleura, peritoneum, pericardium and paratesticular space. Mesothelioma is a rare tumor which arises from these cells and most frequently involves the pleural space. However, approximately 30 percent arise solely from the peritoneum [3]. There are benign, borderline and malignant variants, but benign cystic mesothelioma is not related to malignant mesothelioma. Compared with the pleural form, malignant mesothelioma of the peritoneum is less often associated with asbestos exposure [3, 4]. However, cases with both pleural and peritoneal involvement are usually asbestos-related. In general, malignant mesothelioma of the peritoneum is an aggressive tumor with a rapidly progressive clinical course and a universally poor prognosis.

Imaging Features. The imaging features of peritoneal mesothelioma are variable [4]. The “dry” appearance consists of single or multiple peritoneal-based soft tissue masses that may be large or confluent (Fig. 11.1). The “wet” appearance consists of peritoneal thickening that may be nodular and/or diffuse and is associated with peritoneal fluid (ascites) (Fig. 11.1). Scalloping and mass effect upon adjacent abdominal organs can be seen. Calcification, either within the mass or associated with peritoneal plaques, is uncommon and one should consider other etiologies in the setting of extensive calcification in a peritoneal-based tumor.



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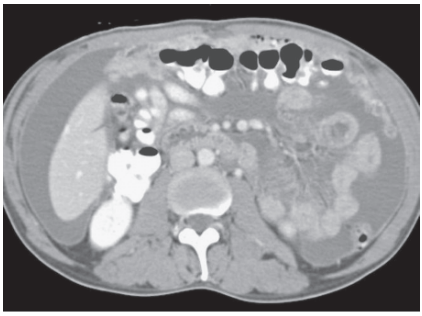


(b)

Fig. 11.1 Malignant peritoneal mesothelioma. **(a)** Contrast-enhanced CT image shows a large, confluent peritoneal-based mass with heterogeneous attenuation, but no calcification. There is no associated ascites present, reflecting the "dry form" of peritoneal mesothelioma. **(b)** Contrast-enhanced CT image from a different patient shows ascites with diffuse thickening of both the visceral and parietal peritoneum, as well as omental and mesenteric soft tissue infiltration. This appearance reflects the so-called "wet form"

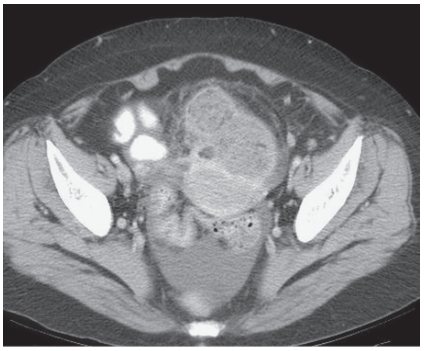
4 Papillary Serous Carcinoma

Clinical Features. Primary papillary serous carcinoma of the peritoneum is a rare malignancy that predominately affects post-menopausal women [5]. This tumor is histologically identical to serous ovarian papillary carcinoma and is clearly distinguishable when the ovaries are either not involved or only superficially involved [6]. Treatment generally consists of an abdominal hysterectomy, bilateral salpingo-oophorectomy and debulking surgery, which are followed by combination chemotherapy. Despite these interventions, the prognosis is dismal.

Imaging Features. Cross-sectional imaging often shows extensive, multifocal involvement of the peritoneum, with omental caking, ascites and, importantly, no primary ovarian mass (Fig. 11.2). There is often extensive calcification within the omental masses, which can be a useful CT finding for differentiating this tumor from peritoneal mesothelioma [6].

5 Desmoplastic Small Round Cell Tumor

Clinical Features. Desmoplastic small round cell tumor is a rare, highly aggressive malignancy that was first described relatively recently [7]. It generally behaves like a high-grade soft tissue sarcoma, but has a predilection for primary peritoneal involvement. The disease tends to progress rapidly and metastatic disease to the liver, lungs, lymph nodes and bones are often present at diagnosis. Unlike most



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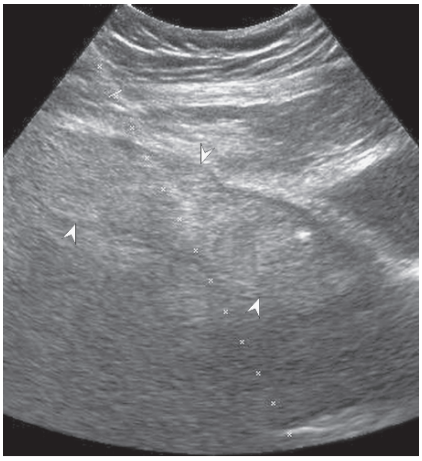


(b)

Fig. 11.2 Primary peritoneal papillary serous carcinoma. **(a)** Contrast-enhanced CT image from a middle-aged female shows a heterogeneous infiltrative soft tissue mass involving the omentum; the ovaries were normal in appearance (not shown). **(b)** Diagnosis was confirmed by ultrasound-guided core biopsy of the thickened omentum (arrowheads)

other primary peritoneal neoplasms discussed herein, this tumor most often affects adolescents and young adults, particularly males. Treatment is relatively ineffective, but attempted therapy often includes surgical debulking, chemotherapy and radiation therapy.

Imaging Features. The most common imaging appearance is that of multiple, bulky rounded peritoneal-based masses [7] (Fig. 11.3). There can be associated ascites, and heterogeneous enhancement of the masses with areas of central necrosis is common. The omentum and paravesical regions are often involved. Although the lesions are usually discrete, an infiltrative appearance is sometimes seen. Calcifications and lymphadenopathy are not usually present.

6 Malignant Fibrous Histiocytoma

Clinical Features. Primary sarcomas of the peritoneal/subperitoneal space, such as malignant fibrous histiocytoma (MFH) and liposarcoma, occur less frequently than their retroperitoneal counterparts [4, 8]. These tumors are most frequently seen in adults and are typically quite large at diagnosis. MFH accounts for approximately 20 percent of all soft tissue sarcomas, most commonly arising in the extremities and retroperitoneum [9]. However, MFH is also reported by some sources to be the single most common primary peritoneal sarcoma [8]. It occurs more frequently in males and has a peak incidence in the fifth and sixth decades of life. The mass is often clinically silent until it is quite large. Constitutional symptoms such as fever,

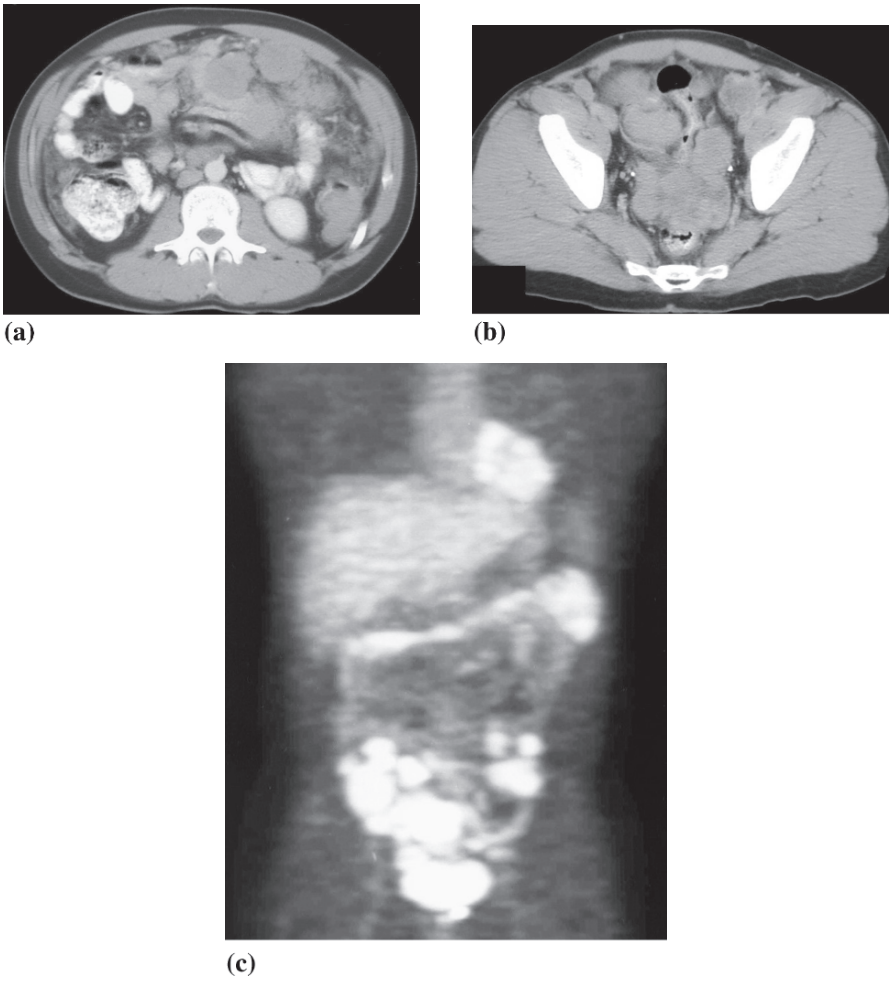


Fig. 11.3 Desmoplastic small round cell tumor. **(a** and **b)** Contrast-enhanced CT images show multiple, rounded, enhancing peritoneal-based masses, which is the most common appearance of this disease. **(c)** Coronal image from FDG-PET study shows multiple hypermetabolic foci corresponding to the rounded peritoneal-based masses seen on CT

malaise and weight loss can occur, but are nonspecific. The only treatment, if possible, is complete surgical resection. Metastatic disease most often involves the lungs, bone and liver. Prognosis is related to tumor grade, size and the presence or absence of metastatic disease. Specifically, high-grade tumors and tumors larger than 10 cm in size have a poor outcome with 10-year survival rates of less than 50 percent [9].

Imaging Features. Radiographically, MFH typically manifests as a large heterogeneous soft tissue mass (Fig. 11.4), as do most sarcomas. Biopsy is required to make a specific diagnosis. The mass is frequently lobulated with peripheral nodular

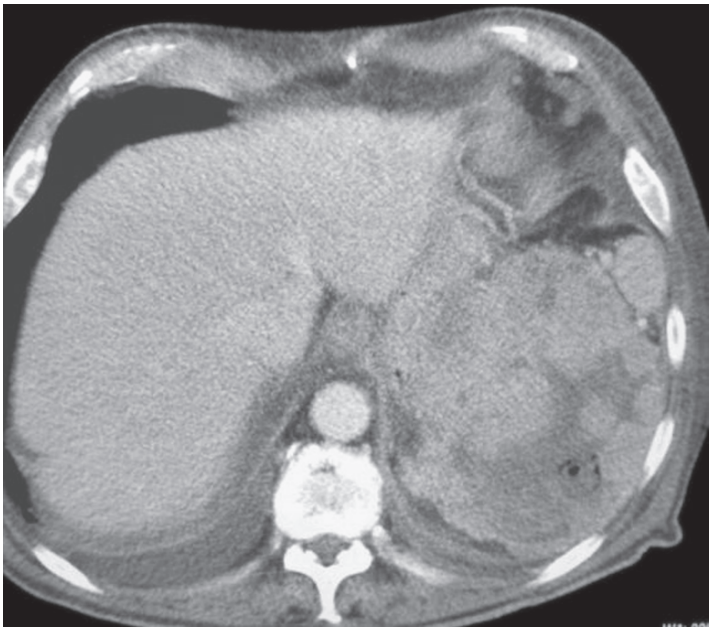


Fig. 11.4 Peritoneal malignant fibrous histiocytoma (MFH). Contrast-enhanced CT image shows a large lobulated heterogeneous peritoneal-based mass occupying the left subphrenic space. Ascites is also present

enhancement, can have associated calcifications (in approximately 10 percent), and may demonstrate heterogeneity from central necrosis, hemorrhage or myxoid degeneration. Fatty components are not seen in MFH [10]. The tumor may directly invade the abdominal musculature, but vascular invasion is rare.

7 Liposarcoma

Clinical Features. Fat-containing tumors are very common in general and account for approximately half of all soft tissue tumors in most surgical series [11, 12]. However, the vast majority of these represent benign lipomas, and differentiating these tumors from liposarcoma is not a trivial matter. Although liposarcoma is one of the most common primary retroperitoneal malignancies, primary peritoneal liposarcoma is relatively rare [13]. The clinical presentation is usually delayed due to the lack of associated symptoms. Ultimately, the mass may become palpable, create symptoms related to mass effect on adjacent structures, or may be incidentally identified at the time of imaging. Treatment is surgical resection, with or without chemotherapy and radiotherapy. Prognosis is inversely related to cellular differentiation of the tumor and directly related to completeness of resection.

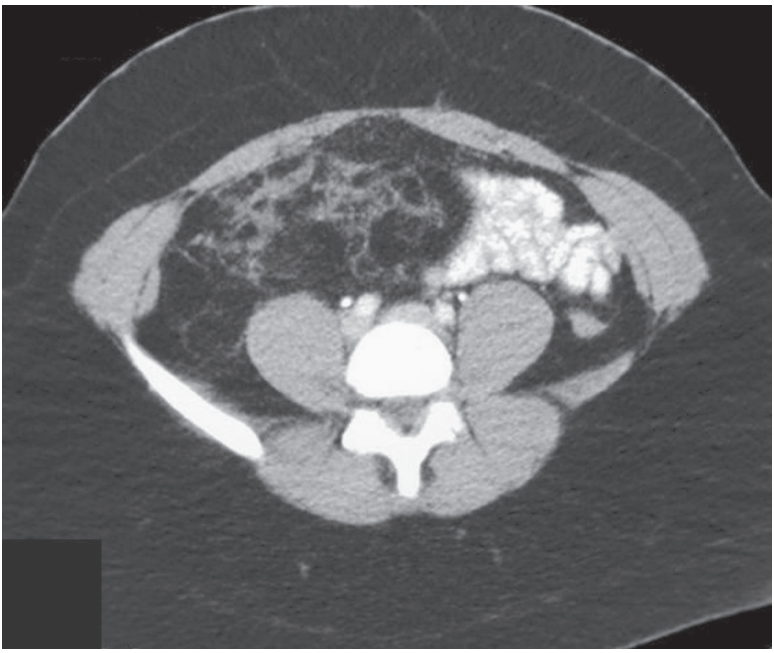


Fig. 11.5 Peritoneal liposarcoma. Contrast-enhanced CT image shows a heterogeneous fatty mass with enhancing soft tissue elements. Note that it is displacing the adjacent small bowel

Imaging Features. Fat-containing tumors are readily and confidently recognized on CT and MRI when demonstrable macroscopic fat is present, which significantly limits the differential diagnosis. If the mass is homogeneous, well-defined and consists almost entirely of fat with only minimal if any soft tissue component, the diagnosis of a benign lipoma is almost certain. Liposarcomas are typically less well-defined, have indistinct borders and contain variable but increased amounts of soft tissue [14] (Fig. 11.5). In fact, some poorly differentiated liposarcomas have no demonstrable fat on cross-sectional imaging and are, therefore, indistinguishable from other sarcomas.

8 Other Malignant Mesenchymal Tumors

Clinical Features. The remaining malignant mesenchymal tumors beyond MFH and liposarcoma essentially lack any distinguishing clinical or radiographic features. As a result, tissue biopsy or surgical resection is required for definitive diagnosis. Malignant nerve sheath tumors and gastrointestinal stromal tumors (GIST) in the setting of neurofibromatosis type 1 (NF-1), however, can be an exception since the patient will often have clinical stigmata of NF-1 (e.g., café au lait spots and cutaneous neurofibromas), or will already carry the diagnosis of NF-1.

Peritoneal involvement by leiomyosarcoma and malignant GIST are most frequently due to metastatic spread from a primary gastrointestinal site, but primary peritoneal tumors can and do occur [15]. In the past malignant GISTs were incorrectly classified as leiomyosarcomas (see section on retroperitoneal leiomyosarcomas). Fibrosarcoma of the mesentery and omentum in young patients can be difficult to differentiate from inflammatory pseudotumor, both at imaging and at pathologic evaluation [16]. Angiosarcoma can develop from the vascular elements of the subperitoneal space. Even synovial sarcomas can arise within the peritoneum, and these tumors can have associated dystrophic calcifications [17].

Imaging Features. Malignant nerve sheath tumors are often multifocal and have a branching or coalescent appearance. They are frequently of low attenuation on CT and have high signal on T2-weighted MR images. As a result they are sometimes mistaken for cystic lesions. Frequently, there are associated nerve root lesions or other findings of NF-1. GIST should also be considered for peritoneal or retroperitoneal tumors in the setting of NF-1 (Fig. 11.6).

The remaining sarcomas are usually indistinguishable from each other on cross-sectional imaging, usually presenting as large soft tissue masses. Synovial sarcomas may have associated dystrophic calcifications (Fig. 11.7), and angiosarcomas are typically hypervascular, but these features are not always present and significant overlap in imaging features exists.

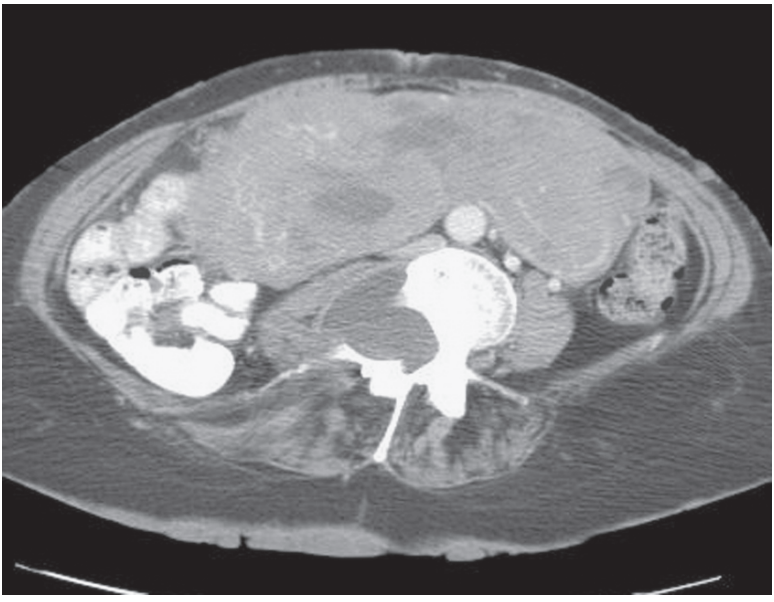


Fig. 11.6 Peritoneal GIST in the setting of NF-1. Contrast-enhanced CT image shows large heterogeneously enhancing mesenteric soft tissue mass with areas of necrosis or cystic change. Note the mesenchymal dysplasia involving the lumbar spine with associated lateral meningocele, as well as numerous cutaneous neurofibromas. This combination of findings is essentially diagnostic for NF-1. The two most likely considerations for the complication tumor would be malignant nerve sheath tumor and GIST. This lesion proved to be a large mesenteric GIST

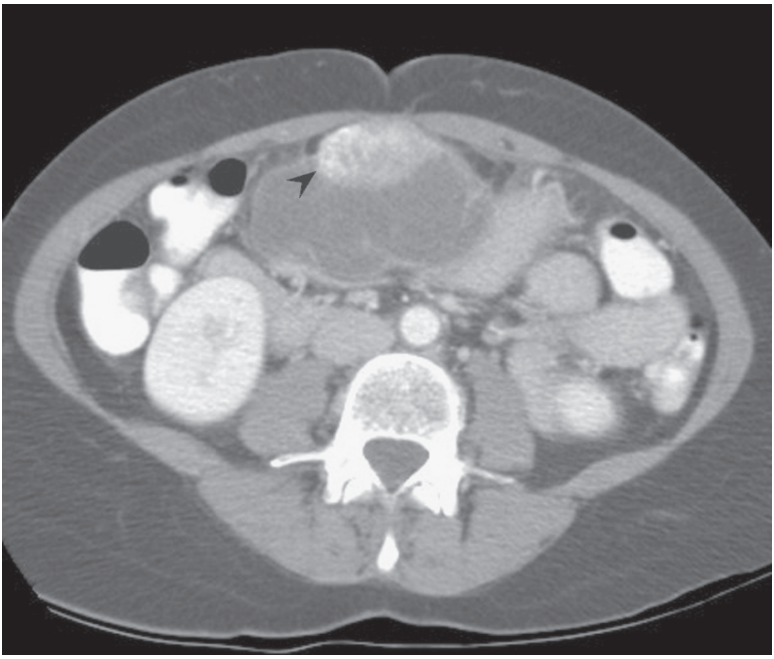


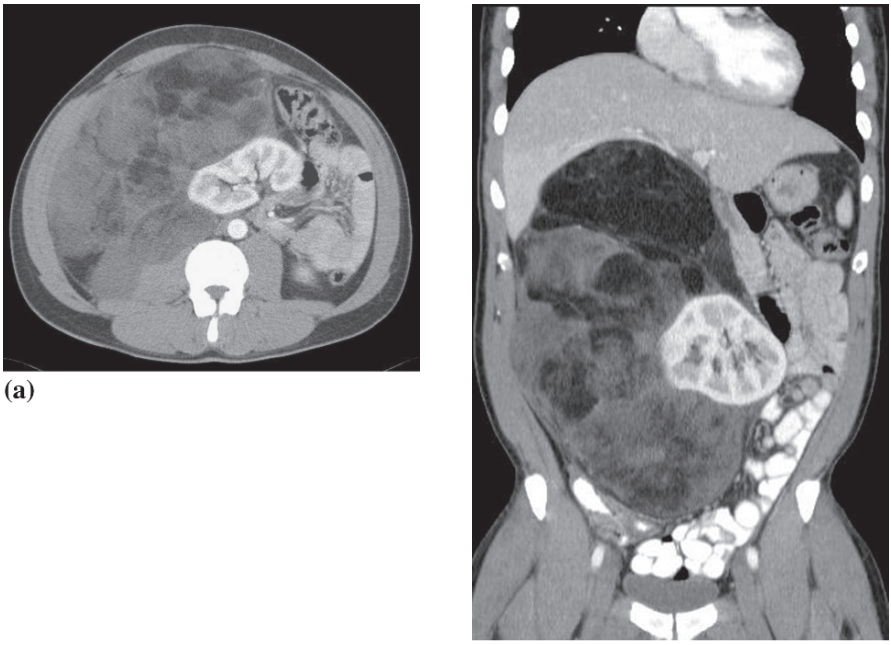
Fig. 11.7 Primary peritoneal synovial sarcoma. Contrast-enhanced CT image shows a mixed cystic and solid mass arising within the gastrocolic ligament. Dystrophic calcification is present within the anterior soft tissue component of the mass (arrowhead). Primary peritoneal origin is a rare extra-articular location for this tumor

9 Primary Retroperitoneal Neoplasms

9.1 Liposarcoma

Clinical Features. As a group, sarcomas are the most common primary malignancies of the retroperitoneum. The three most common cell types include liposarcoma, leiomyosarcoma and malignant fibrous histiocytoma (MFH). Beyond liposarcoma where the presence of fat usually provides a specific clue [18], most of these malignant mesenchymal tumors arising within the retroperitoneum are difficult to differentiate on imaging or clinical grounds.

As with peritoneal fat-containing tumors, differentiating benign lipomas from liposarcomas can sometimes be extremely difficult. Liposarcoma is one of the most common primary retroperitoneal malignancies [19]. The mass is often large at the time of diagnosis due to the lack of associated clinical manifestations. It is treated by surgical resection and the decision to administer additional chemotherapy and radiotherapy is made on a case-by-case basis. Prognosis is related to the grade of the tumor and completeness of resection.



(a) **(b)**

Fig. 11.8 Retroperitoneal liposarcoma. **(a** and **b)** Contrast-enhanced transverse and coronal CT images show a large retroperitoneal mass that contains both fatty and soft tissue components. Note that the tumor displaces the right kidney anteromedially, which confirms its retroperitoneal origin

Imaging Features. A well-defined, homogeneous fatty mass is likely to represent a benign lipoma. However, characteristics that are associated with a higher risk of liposarcoma include a lesion size > 10 cm, thick septations, globular and/or nodular nonadipose regions and a relative proportion of fat < 75 percent [14] (Fig. 11.8). Note that thin septations are seen in both benign and malignant lesions and are not predictive. There is also significant overlap in both the imaging findings and histologic findings of lipomas and well-differentiated liposarcomas.

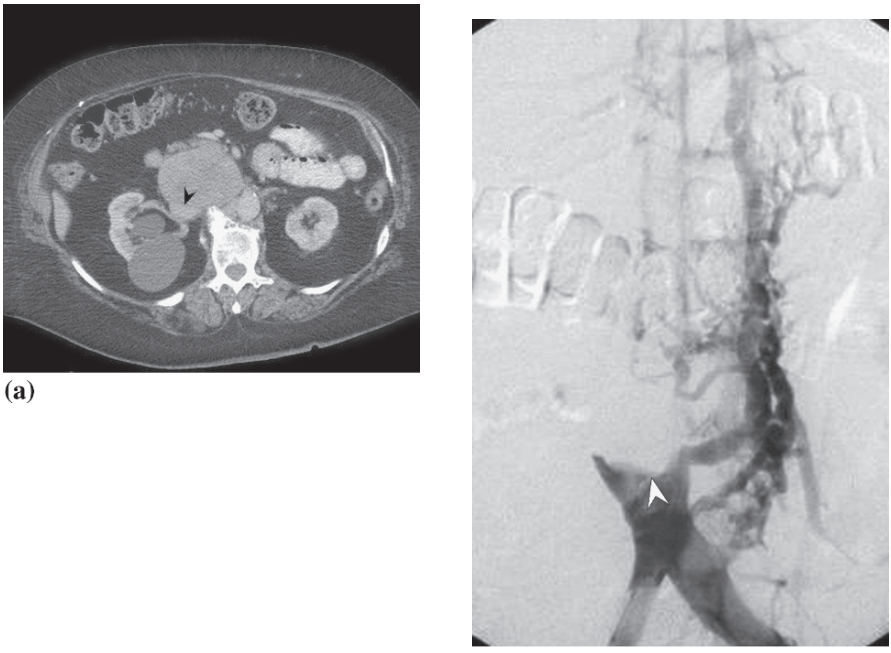
10 Leiomyosarcoma

Clinical Features. The majority of retroperitoneal leiomyosarcomas occur in women, usually in the fifth or sixth decade of life. The retroperitoneum represents the most common primary site of origin, followed by the uterus [20]. A significant fraction of these tumors arise from the inferior vena cava [21]. Clinical presentation largely relates to whether or not an intravascular component is present; most cases are large heterogeneous tumors demonstrating an extraluminal growth pattern. Tumors with an intravascular component may present with symptoms relating to

venous compromise or thrombosis. The lungs are the most frequent site of metastatic involvement [22]. Treatment is difficult because surgical resection is often limited by the size and extent of the mass, while adjuvant chemotherapy and radiation therapy are relatively ineffective.

Until recently many GISTs were incorrectly classified as smooth muscle tumors (leiomyomas and leiomyosarcomas), but recent advances in immunohistochemistry and electron microscopy have shown that these tumors are indeed unique [23]. In comparison, true retroperitoneal GISTs are extremely rare and although primary peritoneal origin is more common, it is still quite rare compared with a primary gastrointestinal tract origin.

Imaging Features. Three growth patterns can be seen at imaging: extravascular (most common), completely intravascular (least common) and combined extra- and intravascular [21] (Fig. 11.9). US and angiography (by CT, MRI or conventional means) may be useful in cases with an intravascular component (Fig. 11.9). Although involvement of the inferior vena cava is suggestive of leiomyosarcoma, other sarcomas can secondarily invade this structure, reducing the specificity of this finding somewhat.



(a) **(b)**

Fig. 11.9 Retroperitoneal leiomyosarcoma. **(a)** Contrast-enhanced CT image shows an enhancing retroperitoneal mass, which appears to arise from the adjacent inferior vena cava (IVC, arrowhead). **(b)** Image from direct venography from a different patient shows obstruction of the IVC by an intravascular leiomyosarcoma, which gives rise to a large filling defect (arrowhead). There is associated collateralization into the azygos system. Although this appearance is suggestive of leiomyosarcoma, other malignancies can secondarily invade the IVC

11 Malignant Fibrous Histiocytoma

Clinical Features. MFH contains both fibroblastic and histiocytic cells in various proportions. Pleomorphic is the most common histiologic subtype, but there are also myxoid, giant cell, inflammatory and angiomatoid subtypes [9]. MFH generally presents in the fifth and sixth decades and the most common symptoms are fever, malaise and weight loss. Metastatic disease most frequently involves the lungs, but osseous and hepatic metastases are sometimes seen. Treatment is surgical resection and although the risk of local recurrence is directly related to completeness of resection, the overall prognosis is more closely related to tumor grade (low, intermediate or high), tumor size and the presence of metastases [9].

Imaging Features. MFH usually manifests as a large heterogeneous soft tissue mass on cross-sectional imaging. The tumor heterogeneity is related to a variable combination of central necrosis, hemorrhage and myxoid degeneration. Enhancement is variable, but often nodular and peripheral. Calcifications are seen in approximately 10 percent of patients, but no fatty component or vascular invasion should be present [10]. MFH is often locally aggressive and invades adjacent structures.

12 Other Malignant Mesenchymal Tumors

Clinical Features. Similar to peritoneal sarcomas, other retroperitoneal sarcomas generally lack distinguishing features. However, there are some characteristics that may be helpful in differentiating the various subtypes. As previously discussed, malignant nerve sheath tumors and GISTs are associated with NF-1, although primary retroperitoneal GISTs are rare. Fibrosarcoma is another rare retroperitoneal tumor, which can have variable biologic behavior. Inflammatory fibrosarcoma in children can be difficult to distinguish from benign myofibroblastic tumor (inflammatory pseudotumor); this malignancy can be locally aggressive and has the potential for metastasis [16]. Angiosarcoma, rhabdomyosarcoma and hemangiopericytoma are rare, aggressive neoplasms that have an extremely poor prognosis.

Imaging Features. Most of these sarcomas manifest as large, heterogeneous, locally invasive masses and the imaging findings are generally nonspecific. The role of imaging is more to evaluate for the extent of disease, to guide biopsy for tissue diagnosis and to assess response to therapy. Malignant nerve sheath tumors in the setting of NF-1 often have a branching, plexiform morphology, with low attenuation on CT and high signal on T2-weighted MRI that can mimic a cystic appearance [24].

13 Malignant Paraganglioma

Clinical Features. Paragangliomas arise from neuroendocrine cells derived from the embryologic neural crest. These tumors can occur anywhere along the sympathetic chain, including both adrenal (e.g., pheochromocytoma) and extra-adrenal

(e.g., paraganglioma) origin [25, 26]. Paragangliomas can be hormonally active, secreting catecholamines which can result in labile hypertension, palpitations, sweating and headaches. Most paragangliomas are benign, but up to 10 percent metastasize and display malignant behavior. They are most likely to occur between the ages of 30 and 45.

Imaging Features. Other than a characteristic location adjacent to the aorta (including the organ of Zuckerkandl) (Fig. 11.10), there are no imaging-specific features for extra-adrenal paragangliomas. These tumors are generally hypervascular, which can be a suggestive imaging feature. The tumor can be solitary or multifocal, and

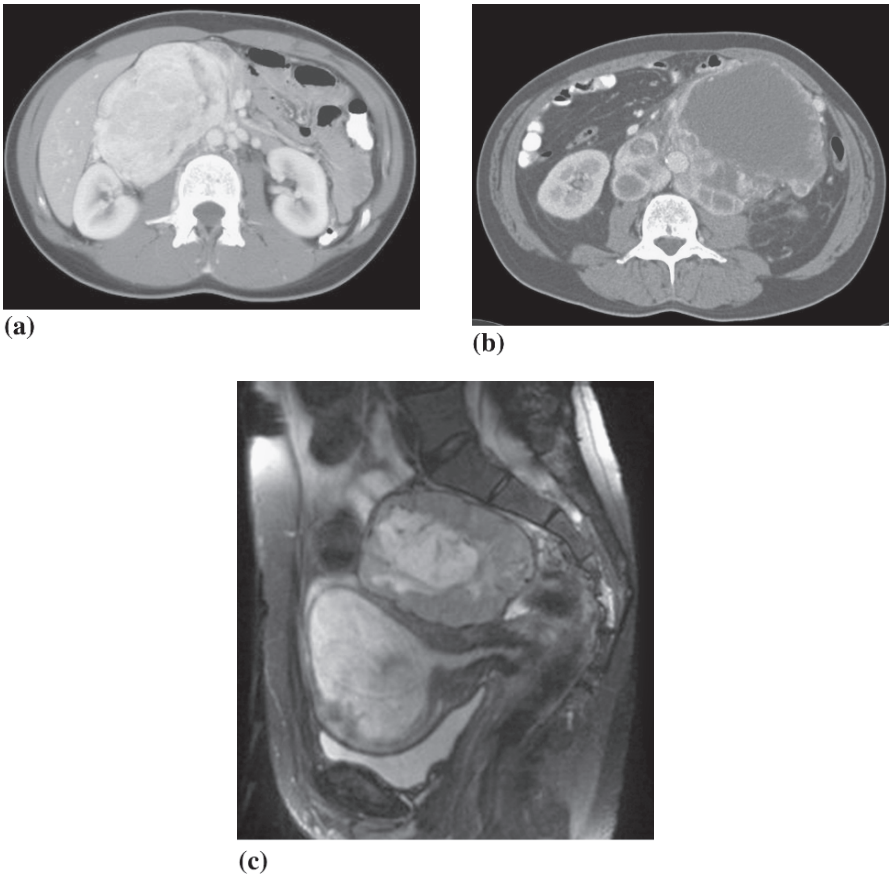


Fig. 11.10 Malignant retroperitoneal paraganglioma. **(a)** Contrast-enhanced CT image shows a large hypervascular retroperitoneal mass, which is relatively homogeneous in appearance considering its large size. **(b)** Contrast-enhanced CT image from a second patient shows a large paraganglioma with a predominately cystic appearance, likely from central necrosis. Adjacent low-attenuation retroperitoneal lymphadenopathy is present. **(c)** Sagittal T2-weighted MRI from a pregnant female with hypertension and palpitations shows a large mass with central high signal from cystic change arising from the organ of Zuckerkandl. Note the gravid uterus inferior and adjacent to the mass

is often relatively homogeneous in appearance, although malignant lesions tend to be larger and demonstrate areas of central necrosis [27]. Associated calcifications are present in about 15 percent of cases. Local invasion or distant metastases are diagnostic of malignancy.

14 Extragonadal Germ Cell Tumors

Clinical Features. Extragonadal germ cell tumors (EGGCT) represent 5 percent to 10 percent of all germ cell tumors, and are characterized by a midline location extending from the pineal gland to the coccyx [28, 29]. Approximately 20 percent to 40 percent of EGGCTs are seminomas, with nonseminomatous germ cell tumors (e.g., embryonal carcinoma, yolk-sac tumor, choriocarcinoma, teratoma or combined) representing the remaining 60 percent to 80 percent. The majority of these tumors occur in the mediasteinum, but the second most common site of involvement is the retroperitoneum (30 percent to 40 percent). Because metastatic retroperitoneal involvement from a testicular primary germ cell tumor is much more common than a primary EGGCT, males should undergo testicular US to exclude this possibility [28].

The most common clinical symptoms from retroperitoneal EGGCT include a palpable abdominal mass, abdominal or back pain and weight loss. Treatment generally includes primary chemotherapy, followed by surgical resection of any significant residual mass. Although controversial, any residual mass measuring > 3 cm is usually resected and, if residual disease is identified in the pathologic specimen, then further chemotherapy is given [28]. Although generally treated like a metastatic gonadal germ cell tumor, the prognosis of primary retroperitoneal EGGCT is somewhat worse, but still rather favorable overall. Negative prognostic factors include nonseminomatous histology, elevated tumor markers at the time of diagnosis and the presence of metastatic disease.

Imaging Features. The cross-sectional imaging appearance of these tumors varies according to the underlying tissue type. Teratomas often have a markedly heterogeneous appearance due to varying combinations of soft tissue, calcification, fat and fluid. In contrast, seminomas tend to be large, homogeneous lobulated soft tissue masses. Nonseminomatous germ cell tumors are often very irregular in morphology and heterogeneous in appearance, with variable amounts of necrosis and hemorrhage (Fig. 11.11).

Conclusion

Primary malignancies of the peritoneal, subperitoneal and retroperitoneal spaces occur much less frequently than metastatic involvement from primary organ-based tumors or lymphoproliferative diseases. Cross-sectional imaging techniques, such as CT and PET/CT, can be useful for detection, characterization, staging, guiding biopsy for tissue diagnosis and evaluating response to therapy.

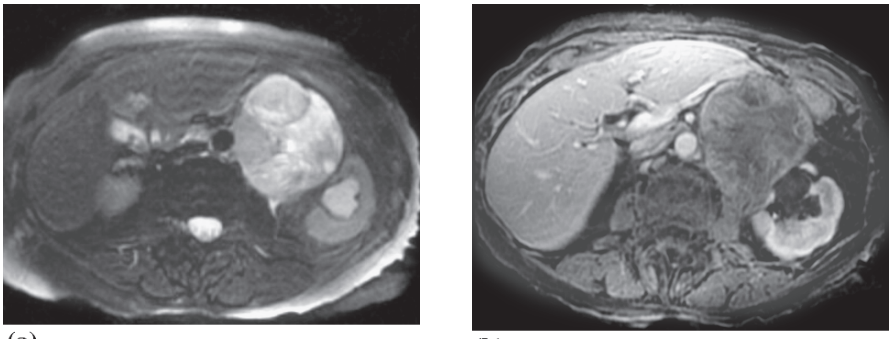


Fig. 11.11 Extragonadal primary retroperitoneal germ cell tumor. **(a** and **b)** T2-weighted and contrast-enhanced T1-weighted MR images show a complex T2 hyperintense retroperitoneal mass that causes left-sided obstructive hydronephrosis. At biopsy, this proved to be a malignant mixed mullerian tumor (MMMT) arising within the retroperitoneum

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