

Primary Neoplasms of Peritoneal and Subperitoneal Origin: CT Findings¹

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CME FEATURE

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LEARNING OBJECTIVES FOR TEST 3

After reading this article and taking the test, the reader will be able to:

- Describe the CT features of a variety of primary peritoneal neoplasms.
- Identify the pertinent clinical and demographic features associated with these neoplasms.
- Discuss peritoneal anatomy as it relates to neoplastic disease.

Peritoneal carcinomatosis is a common metastatic manifestation of many organ-based malignancies, particularly carcinomas of the gastrointestinal tract and ovaries. Primary neoplasms of peritoneal and subperitoneal origin occur much less frequently than metastatic peritoneal involvement from a known or occult primary tumor; however, these rare primary lesions (peritoneal mesothelioma, papillary serous carcinoma, desmoplastic small round cell tumor, benign and malignant mesenchymal tumors, lymphoproliferative disorders) are often first detected at computed tomography (CT) and should be considered in the absence of a known or suspected organ-based malignancy. A precise diagnosis based on imaging findings alone is often not possible. Furthermore, distinguishing a benign from a malignant process and a primary from a metastatic process is also challenging. Nevertheless, CT features combined with the patient's relevant clinical and demographic data can help narrow the differential diagnosis for a peritoneum-based neoplasm in many cases. CT is useful not only for the detection, characterization, and staging of primary neoplasms of peritoneal and subperitoneal origin, but also for guiding biopsy for tissue diagnosis.

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Abbreviations: GIST = gastrointestinal stromal tumor, NF-1 = neurofibromatosis type 1

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Introduction

Peritoneal carcinomatosis is a relatively common metastatic manifestation of a variety of organ-based malignancies, particularly of the gastrointestinal tract and ovaries. Primary neoplasms of peritoneal and subperitoneal origin occur much less frequently than metastatic disease in these locations; however, they are often first detected at computed tomography (CT).

In general, neoplastic involvement of the peritoneal and subperitoneal spaces will manifest at CT as a soft-tissue process (tumorous or infiltrative) with or without associated fluid (ascites). Additional findings may include a cystic component or cystic necrosis, calcification, prominent contrast material enhancement, or fat attenuation. However, most CT findings are nonspecific, since there are a number of nonneoplastic and benign neoplastic peritoneal conditions that can closely mimic malignancy (1). Even in the absence of a known organ-based malignancy, an occult gastrointestinal, ovarian, or other organ-based cancer will likely be initially favored in the setting of findings that are suspicious for peritoneal carcinomatosis. However, primary neoplasms of peritoneal and subperitoneal origin, albeit relatively rare, must also be considered in this setting. Unfortunately, most of these primary tumors are malignant and often portend a poor prognosis.

In this article, we review anatomic considerations relating to the peritoneum and subperitoneum. We also discuss and illustrate CT technique and subsequent findings in a wide spectrum of primary peritoneal neoplasms, including peritoneal mesothelioma, papillary serous carcinoma, desmoplastic small round cell tumor, various benign and malignant mesenchymal tumors, and lymphoproliferative disorders (Table). We do not discuss desmoid tumor (fibromatosis) because it is not a true neoplasm (1).

Anatomic Considerations

The visceral and parietal peritonea 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 by means of the relatively unrestricted movement of fluid and cells. Often overlooked, however, is the potential for disease extension within the subperitoneal space, which lies deep to the surface lining of the visceral and parietal peritonea, the omentum, and the various peritoneal ligaments and mesenteries (2). Furthermore, the

Primary Peritoneal Neoplasms

Mesothelioma
Cystic mesothelioma
Papillary serous carcinoma
Desmoplastic small round cell tumor
Benign mesenchymal tumors
Hemangioma
Lymphangioma
Lipoma
Nerve sheath tumor
Gastrointestinal stromal tumor (GIST)
Leiomyomatosis peritonealis disseminata
Malignant mesenchymal tumors
Liposarcoma
Malignant fibrous histiocytoma
Leiomyosarcoma
GIST
Nerve sheath tumor
Fibrosarcoma
Angiosarcoma
Synovial sarcoma
Lymphoproliferative disorders
Peritoneal lymphomatosis
Leukemic infiltration
Granulocytic sarcoma
Extramedullary plasmacytoma

subperitoneal space has both intraperitoneal and extraperitoneal components that bridge the peritoneum and retroperitoneum, allowing bidirectional spread of disease processes. These components help explain the dual involvement that is sometimes encountered in these traditionally separate and distinct compartments. In this article, we focus on entities arising primarily within the peritoneum and the intraperitoneal portion of the subperitoneal space.

CT Technique

Prior to the advent of modern cross-sectional imaging, peritoneal disease was generally not detectable until the process had resulted in significant displacement of abdominal organs or bowel obstruction. Multidetector CT readily allows noninvasive evaluation of the peritoneum, but early detection of disease remains a diagnostic challenge, particularly in the absence of peritoneal fluid (ascites). In general, both orally and intravenously administered contrast material will increase the overall diagnostic accuracy of CT for detecting peritoneal abnormalities. Soft-copy CT image interpretation with real-time image scrolling has greatly enhanced the ability to reliably detect subtle peritoneal soft tissue and distinguish it from normal bowel. Two-dimensional multiplanar reformatted images may also be useful in selected cases.

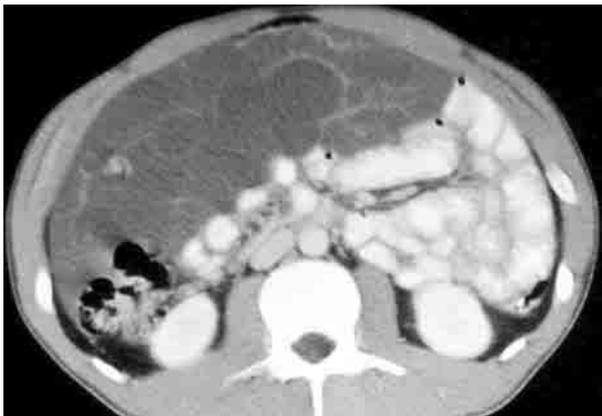


1.

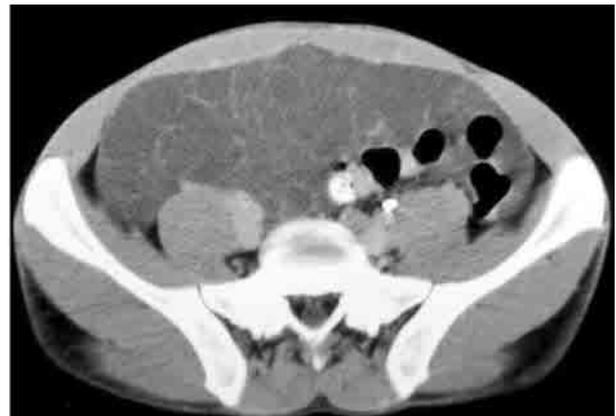


2.

Figures 1, 2. Malignant peritoneal mesothelioma. (1) Contrast material–enhanced CT scan shows confluent peritoneum-based masses of varying attenuation that scallop the liver. (2) Contrast-enhanced CT scan shows relatively uniform and diffuse thickening of both the visceral and parietal (arrowheads) peritonea with associated ascites. Omental infiltration is present, but no bulky masses or loculated fluid collections are seen.



a.



b.

Figure 3. Cystic mesothelioma in a young woman. CT scans (a obtained cephalad to b) show a large, peritoneum-based multilocular cystic mass. Note the thin cyst walls. No prominent soft-tissue component is present.

Types of Neoplasms

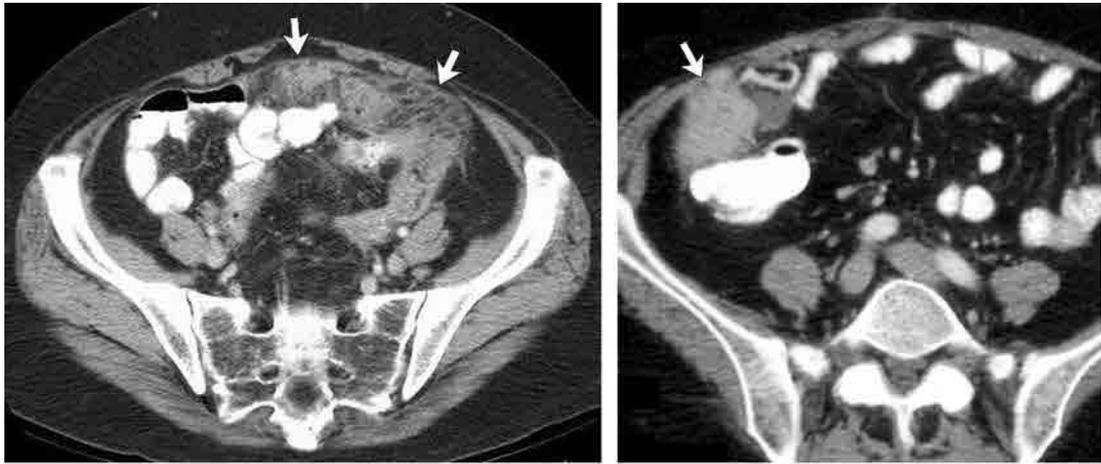
Mesothelioma

Malignant peritoneal mesothelioma is a rare but aggressive tumor that is derived from the peritoneal mesothelium. Although most mesotheliomas involve the pleural surface, approximately 30% arise solely from the peritoneum (3), and in only a minority of these cases will the patient have a history of significant asbestos exposure. CT features of peritoneal mesothelioma range from a “dry” appearance consisting of peritoneum-based masses, which may be large or confluent (Fig 1), to a “wet” appearance consisting of ascites and associated peritoneal thickening, which may be nodular or diffuse (Fig 2). Scalloping of or mass effect on adjacent abdominal organs may be seen (Fig 1). Calcification is uncommon, and other

causes of extensive calcification in a peritoneum-based tumor should be considered. Unlike in pleural mesothelioma, associated calcified peritoneal plaques are not commonly encountered.

Cystic Mesothelioma

Cystic mesothelioma is a benign neoplasm that is derived from the peritoneal mesothelium (4). This rare tumor has a distinct female predilection. Cystic mesothelioma has no malignant potential but is reported to recur in 25%–50% of cases. Typically, the lesion consists of multiple grape-like clusters of mesothelium-lined cysts separated by varying amounts of fibrous tissue. At CT, cystic mesothelioma appears as either a peritoneum-based multilocular cystic mass (Fig 3) or multiple



4.

5.

Figures 4, 5. Primary papillary serous carcinoma of the peritoneum. (4) Contrast-enhanced CT scan obtained in a middle-aged woman shows irregular soft-tissue infiltration of the omentum (arrows). No ovarian mass was present. The diagnosis was confirmed at ultrasonography-guided biopsy. (5) Contrast-enhanced CT scan obtained in a different middle-aged woman shows a solid mass involving the right aspect of the omentum (arrow). Note the small amount of adjacent peritoneal fluid and the prominent omental vessel. As in Figure 4, no ovarian mass was present.

distinct unilocular thin-walled cysts. Involvement of the pelvic region is characteristic. Absence of significant mass effect, increased attenuation, calcification, or a prominent soft-tissue component are CT features that can help distinguish cystic mesothelioma from malignant metastatic disease such as pseudomyxoma peritonei (5).

Papillary Serous Carcinoma

Primary papillary serous carcinoma of the peritoneum is a rare malignancy that predominantly affects postmenopausal women (6). Multicentric peritoneal involvement is typical, with omental involvement being particularly common (Figs 4, 5). Extensive calcification of omental caking is present in many cases and is a useful CT finding for excluding mesothelioma. The absence of an ovarian mass is critical for excluding metastatic papillary serous ovarian carcinoma, which otherwise has a similar appearance at CT (Fig 6) and is histologically identical to its primary peritoneal counterpart (6).

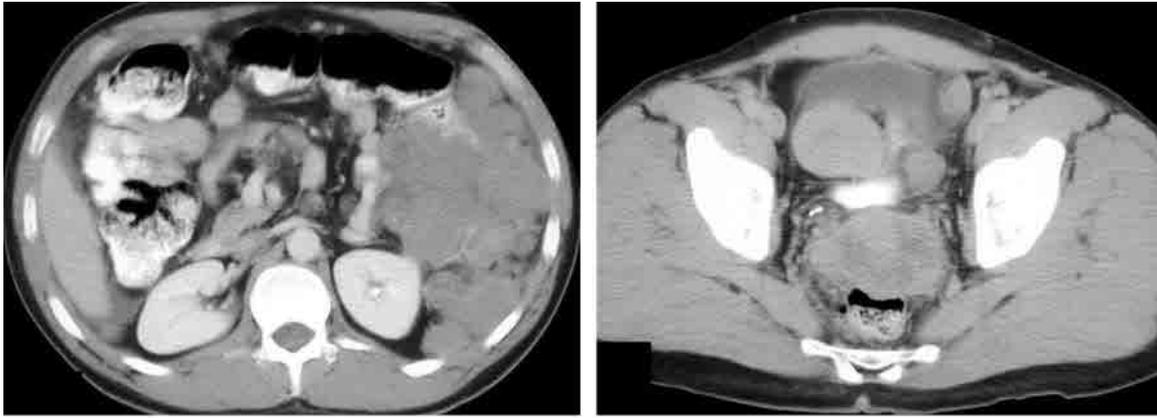
Desmoplastic Small Round Cell Tumor

Desmoplastic small round cell tumor is a highly aggressive malignancy that has only recently been described (7). The peritoneal cavity is the most



Figure 6. Metastatic papillary serous ovarian carcinoma. CT scan (bone window) shows densely calcified peritoneum-based metastases, which are considerably higher in attenuation than contrast material in the bowel. Primary peritoneal papillary carcinoma could have an identical appearance.

common site of involvement. Unlike most of the other primary peritoneal neoplasms discussed in this article, desmoplastic small round cell tumor most often affects adolescents and young adults. The most typical CT appearance is that of multiple rounded peritoneal masses (Fig 7) with or without ascites (6). Heterogeneous enhancement with central low-attenuation areas is common. The omentum and paravesical regions are often



a. **b.**
Figure 7. Desmoplastic small round cell tumor. Axial contrast-enhanced CT scans (**a** obtained cephalad to **b**) show multiple rounded peritoneum-based masses, which represent the most frequent manifestation of this disease.



Figure 8. Desmoplastic small round cell tumor. Axial contrast-enhanced CT scan shows diffuse peritoneal thickening with omental caking, findings that represent a less frequent manifestation of desmoplastic small round cell tumor.

involved. An infiltrative appearance with diffuse peritoneal thickening is a less common manifestation (Fig 8).

Benign Mesenchymal Tumors

Mesenchymal tumors that arise from the intraperitoneal portion of the subperitoneal space may be derived from lymphatic, vascular, neuromuscular, or fatty tissues. Malignant mesenchymal tumors outnumber benign neoplasms in most published series. However, the CT findings in benign mesenchymal neoplasms are more often suggestive of a specific diagnosis.



Figure 9. Lymphangioma. Contrast-enhanced CT scan shows a low-attenuation cystic lesion involving the gastrohepatic ligament (arrowheads). Note the vessel coursing between locules of the lesion.

Lymphangiomas may represent either congenital malformations of the lymphatic system or benign neoplasms (8) and typically appear as large, thin-walled, usually multiloculated cysts at CT. Mesenteric infiltration is common, and removal often necessitates bowel resection; an omental origin is less commonly seen. At CT, the cyst walls are often imperceptible, but vessels may be seen coursing between locules (Figs 9, 10). Cyst contents can have an attenuation less than that of water due to their chylous nature, which can be a key distinguishing feature.

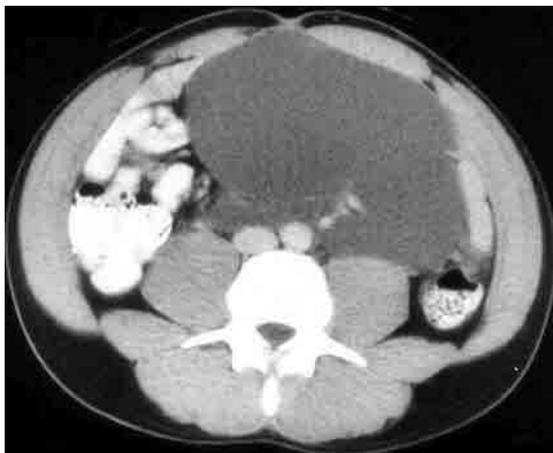


Figure 10. Mesenteric lymphangioma. Contrast-enhanced CT scan shows a large, low-attenuation cystic lesion involving the small bowel mesentery. Note the vessels coursing through the lesion (cf Fig 9).

Hemangiomas may be classified into cavernous, capillary, and venous types (9). Cavernous hemangiomas are the most common type to involve the mesentery and consist of relatively large blood-filled sinuses lined by endothelium. Unlike cavernous hemangiomas of the liver, mesenteric lesions generally lack a well-defined border. However, the presence of phleboliths is highly suggestive of the diagnosis (Fig 11).

Lipomas arising from subperitoneal adipose tissue are generally of no clinical concern but may be detected incidentally at CT (Fig 12). Uniform fat attenuation without a prominent soft-tissue component is a pertinent feature arguing against the possibility of liposarcoma (10).

Nerve sheath tumors arising from the subperitoneal space are uncommon. A mesenteric plexiform neurofibroma in the setting of neurofibromatosis type 1 (NF-1) likely represents the most common manifestation of subperitoneal involvement, although retroperitoneal involvement is much more common. At CT, nerve sheath tumors often have a multifocal, branching or coalescent appearance and may mimic low-attenuation lymphadenopathy or even a cystic lesion (Figs 13, 14) (11). These tumors often bridge the retroperitoneal and subperitoneal spaces (Figs 13, 14).



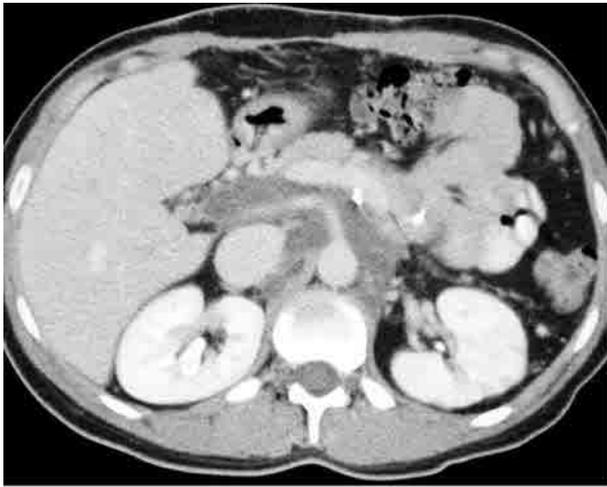
Figure 11. Mesenteric hemangioma. Contrast-enhanced CT scan shows a poorly circumscribed mesenteric soft-tissue mass (arrow) with punctate calcifications that represent phleboliths.



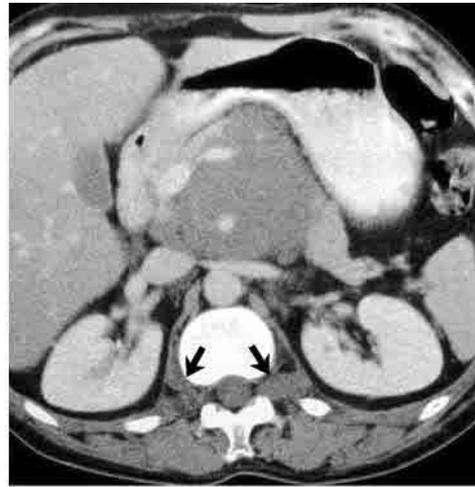
Figure 12. Mesenteric lipoma. Contrast-enhanced CT scan shows a well-circumscribed lesion with uniform fat attenuation (arrowheads). No soft-tissue component is present.

Associated nerve root lesions or other findings typical of NF-1 can strongly suggest the diagnosis (Fig 14). Although usually benign, these tumors may undergo malignant degeneration.

Leiomyomatosis peritonealis disseminata is a rare benign condition that primarily affects women of reproductive age and is associated with uterine fibroids, pregnancy, and contraceptive steroids (12). This disease entity is characterized by the presence of multiple subperitoneal nodules composed of smooth muscle cells (Fig 15).



13.



14.

Figures 13, 14. (13) Plexiform neurofibroma in NF-1. Contrast-enhanced CT scan shows confluent low-attenuation material surrounding the superior mesenteric and replaced right hepatic arteries, a finding that represents a plexiform neurofibroma. Contiguous involvement of the retroperitoneum underscores the continuity of the retroperitoneum with the subperitoneal space. (14) Multiple neurofibromas in NF-1. Contrast-enhanced CT scan shows a prominent low-attenuation mass near the root of the mesentery that surrounds the superior mesenteric vessels. Lymphadenopathy could also have this appearance; note, however, the presence of low-attenuation neurofibromas involving the paraspinal nerve roots (arrows).



a.



b.



c.

Figure 15. Leiomyomatosis peritonealis disseminata in a woman with acute abdominal symptoms from cecal volvulus. Contrast-enhanced CT scans incidentally show several enhancing peritoneum-based nodules (arrowheads in **a** and **b**). Note also the multiple pedunculated subserosal uterine fibroids. Leiomyomatosis peritonealis disseminata was proved at subsequent laparotomy for cecal volvulus.

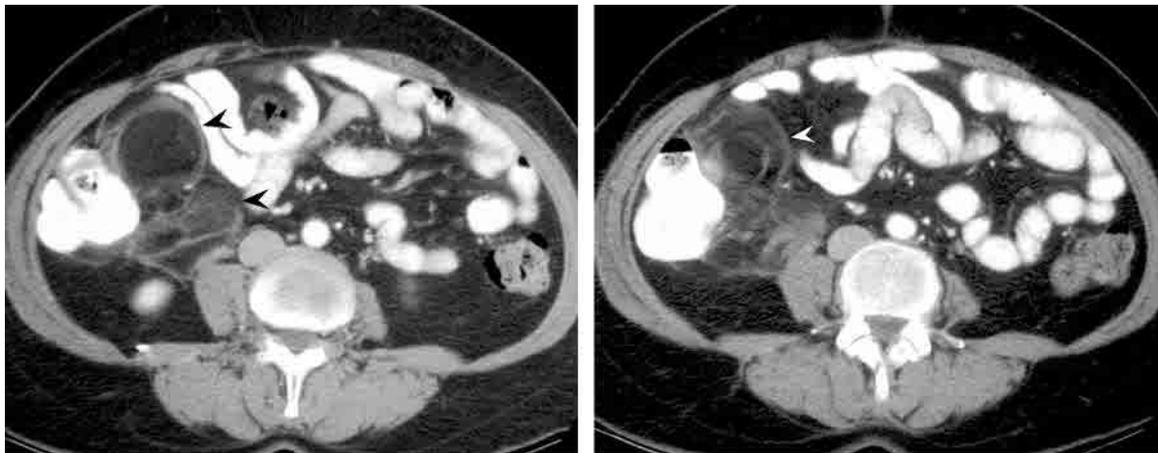
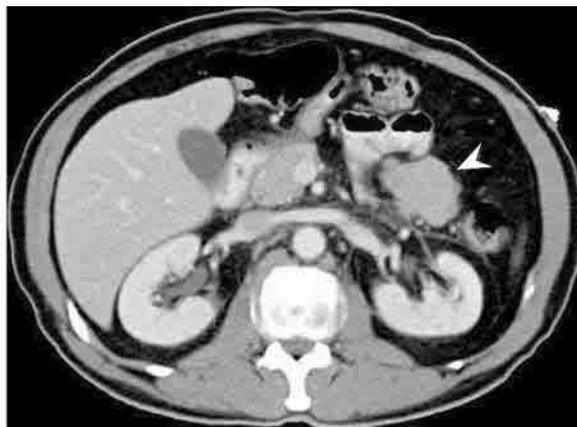


Figure 16. Mesenteric liposarcoma. Contrast-enhanced CT scans (**a** obtained cephalad to **b**) show a mesenteric lesion (arrowheads) that is predominantly fatty but contains heterogeneous soft-tissue elements.



17.



18.

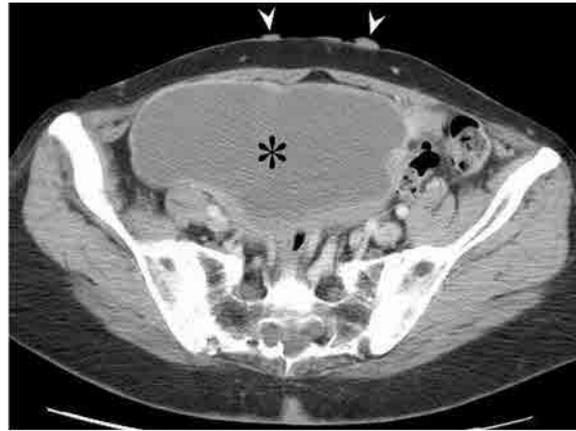
Figures 17, 18. Malignant fibrous histiocytoma. (17) Contrast-enhanced CT scan shows a large, heterogeneous soft-tissue mass, which is a typical appearance of peritoneal malignant fibrous histiocytoma. (18) Contrast-enhanced CT scan shows a relatively small mesenteric tumor (arrowhead), which is an uncommon appearance of peritoneal malignant fibrous histiocytoma.

Figure 19. Leiomyosarcoma. Axial contrast-enhanced CT scan shows a large, peritoneum-based soft-tissue mass with heterogeneous enhancement.





Figure 20. GIST. Contrast-enhanced CT scan shows a large, heterogeneous omentum-based mass, a finding that was believed to represent a primary peritoneal tumor.



a.

b.

Figure 21. GIST complicating NF-1 in a patient who was referred for preoperative staging of “ovarian cancer.” Contrast-enhanced CT scans show a large, heterogeneous midline abdominal mass (arrows in **a**) with a large cystic component extending inferiorly (* in **b**). Additional findings in NF-1 include mesenchymal dysplasia of the lumbar spine with an associated lateral meningocele (black arrowhead in **a**) and multiple cutaneous neurofibromas (white arrowheads). Given the CT findings, a diagnosis of malignant nerve sheath tumor or GIST rather than ovarian primary tumor was made, and the surgical approach was altered accordingly.

Leiomyomatosis peritonealis disseminata most often represents an unexpected finding at laparotomy performed for other reasons and is often initially assumed to represent carcinomatosis.

Malignant Mesenchymal Tumors

Primary sarcomas of the subperitoneal space such as liposarcoma, malignant fibrous histiocytoma, and leiomyosarcoma occur less frequently than their retroperitoneal counterparts (13). In some cases, extension of tumor results in involvement of both the intraperitoneal and extraperitoneal portions of the subperitoneal space, with the process most often originating in the latter. These tumors are generally seen in adults and are typically large at diagnosis.

Although liposarcoma is one of the most common primary retroperitoneal malignancies, peritoneal liposarcoma is relatively rare (14). However, unlike in the other primary peritoneal sarcomas, the CT findings can suggest this specific

diagnosis when the tumor contains areas of fat attenuation (Fig 16). Fat attenuation is less likely to be seen in higher-grade liposarcomas, such as the pleomorphic and round cell subtypes.

Apart from liposarcoma, the peritoneal sarcomas largely lack any distinguishing features and generally manifest at CT as large, solitary masses (13). Malignant fibrous histiocytoma is reported to be the single most common peritoneal sarcoma (Figs 17, 18) (15). Peritoneal involvement by leiomyosarcoma and GIST is most often due to metastatic spread from a primary gastrointestinal site, but primary peritoneal tumors do occur (Figs 19, 20) (16). In the setting of NF-1, a large peritoneal mass will most likely represent either a malignant nerve sheath tumor or a GIST, both of which occur more frequently in these patients (Fig 21). Fibrosarcoma of the mesentery and omentum in young patients can mimic inflammatory pseudotumor at

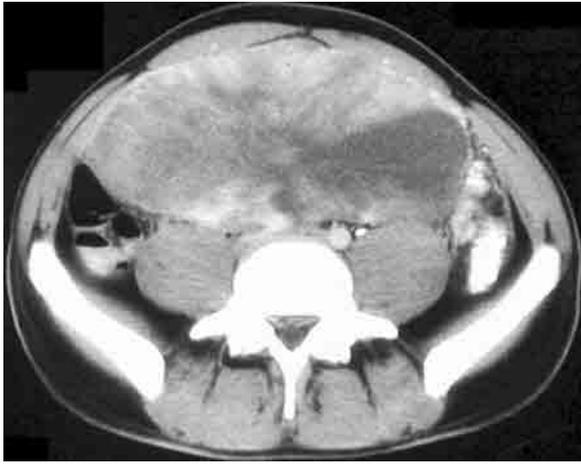


Figure 22. Inflammatory fibrosarcoma in a young patient. CT scan shows a large, peritoneum-based mass with heterogeneous enhancement. Differentiation from inflammatory pseudotumor can be difficult at both imaging and pathologic evaluation in such cases.

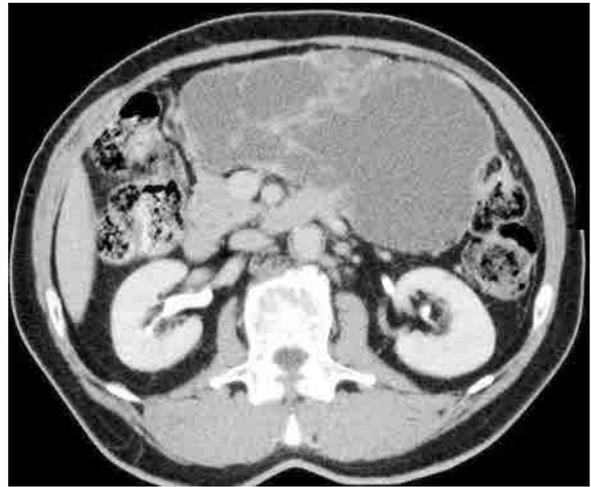
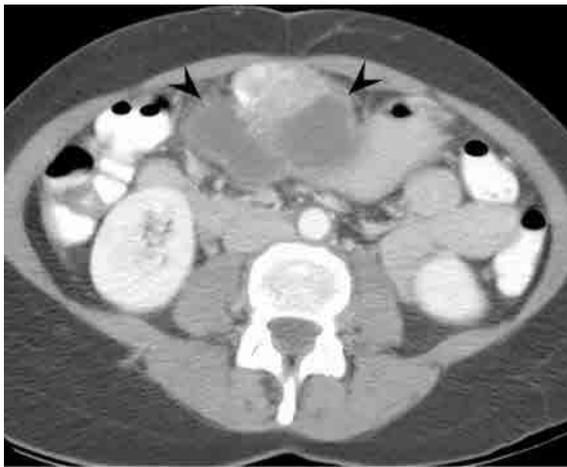


Figure 23. Angiosarcoma. Axial contrast-enhanced CT scan shows a large, complex cystic or necrotic mass involving the omentum. Angiosarcoma was subsequently proved at pathologic evaluation.



a.



b.

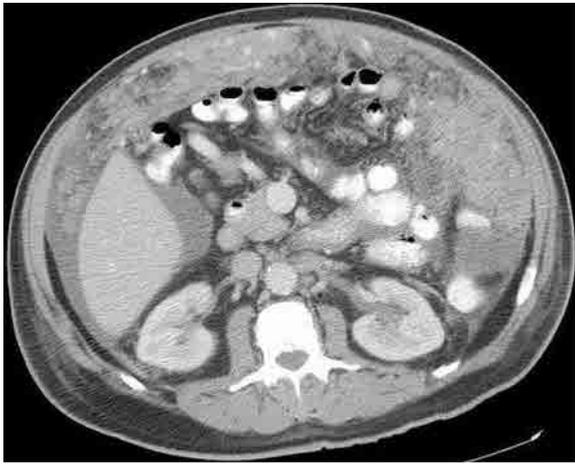
Figure 24. Synovial sarcoma. CT scans show a complex cystic-solid mass (arrowheads in **a**) centered in the gastrocolic ligament between the greater curve of the stomach and the transverse colon. Tumor calcification is also present, a finding that is unusual in peritoneal sarcomas.

both CT and pathologic evaluation (Fig 22) (17). Angiosarcoma can arise from vascular elements of the subperitoneal space (Fig 23). Even synovial sarcoma can arise within the peritoneum and may show dystrophic calcification (Fig 24) (18).

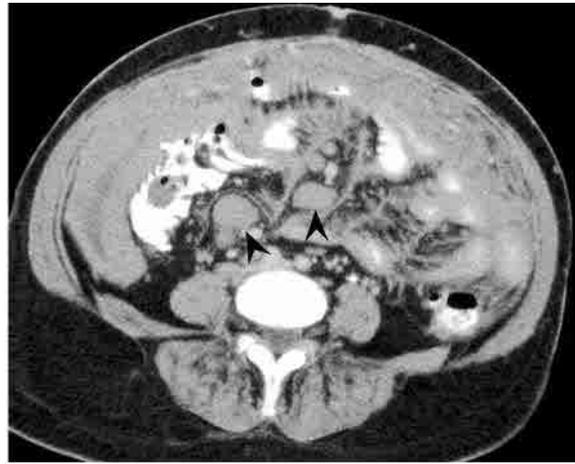
Lymphoproliferative Disorders

Lymphoproliferative neoplastic disease can rarely manifest as a primary subperitoneal process without additional sites of involvement at presentation.

Mesenteric lymphadenopathy is a relatively common CT manifestation of non-Hodgkin lymphoma. However, extensive lymphomatous infil-



25.



26.

Figures 25, 26. Peritoneal lymphomatosis. (25) Contrast-enhanced CT scan shows omental soft-tissue infiltration and ascites. No organ-based primary site was identified. Omental lymphoma was diagnosed at ultrasonography-guided biopsy. (26) Contrast-enhanced CT scan shows extensive peritoneal soft-tissue infiltration with omental caking and diffuse mesenteric thickening. Note also the presence of lymphadenopathy (arrowheads).



a.



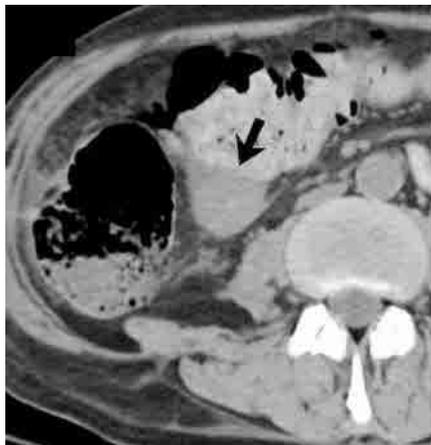
b.

Figure 27. Leukemic infiltration in a woman with acute lymphocytic leukemia. Contrast-enhanced CT scans show diffuse, irregular peritoneal thickening (arrowheads) and ascites. Uterine involvement is also present (* in b).

tration of the intraperitoneal portion of the subperitoneal space is uncommon and is referred to as peritoneal lymphomatosis (19). CT findings consist of omental caking, diffuse peritoneal thickening, and ascites (Figs 25, 26). Associated retroperitoneal and mesenteric adenopathy is sometimes present (Fig 26).

Leukemic infiltration of the peritoneum is also rare. As in peritoneal lymphomatosis, the CT

findings can closely mimic peritoneal carcinoma-tosis (Fig 27). Granulocytic sarcoma, sometimes called chloroma because of its greenish hue, represents a subset of extramedullary disease that is associated with acute myeloid leukemia (20). Peritoneal involvement can manifest at CT as a focal soft-tissue mass (Fig 28) or a diffuse infiltrative process (Fig 29).



28.

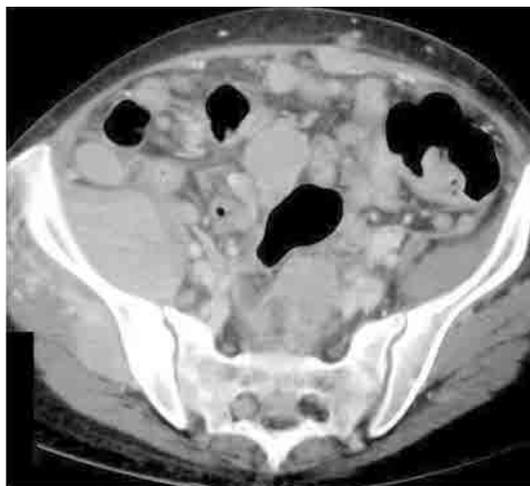


29.

Figures 28, 29. Granulocytic sarcoma (chloroma). (28) Contrast-enhanced CT scan obtained in a patient with acute myelogenous leukemia shows extramedullary peritoneal involvement consisting of a focal mesenteric mass in the ileocecal region (arrow). (29) Contrast-enhanced CT scan obtained in a different patient with acute myelogenous leukemia shows extensive and diffuse peritoneal involvement.



a.



b.

Figure 30. Extramedullary plasmacytomas in a patient with known multiple myeloma. Contrast-enhanced CT scans (a obtained cephalad to b) show multiple peritoneal and retroperitoneal soft-tissue masses. Renal, adrenal, and pancreatic involvement are also seen.

Extramedullary plasmacytoma can occur either with or without known underlying multiple myeloma of the marrow space. The retroperitoneum is a more common site of extramedullary involvement by plasmacytoma than the peritoneum, involvement of which is quite rare (Fig 30). In fact, peritoneal disease in these patients is perhaps just as likely to result from myeloma-associated amyloidosis (AL type).

Conclusions

Primary neoplasms arising from the peritoneum and subperitoneum occur much less frequently than metastatic peritoneal involvement from a known or occult primary tumor. Nevertheless, these rare primary lesions should be considered in the absence of a known or suspected organ-based malignancy. CT is useful not only for detection, characterization, and staging, but also for guiding biopsy for tissue diagnosis.

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