Primary peritoneal tumors are uncommon lesions that arise from the mesothelial or submesothelial layers of the peritoneum. Primary malignant mesothelioma, multicystic mesothelioma, primary peritoneal serous carcinoma, leiomyomatosis peritonealis disseminata, and desmoplastic small round cell tumor are the most prominent of these rare lesions. Primary malignant mesothelioma is a highly aggressive malignancy that occurs most commonly in older men and that has a strong association with high levels of asbestos exposure. It manifests most often as diffuse sheetlike or nodular thickening of the peritoneal surfaces, but it may occasionally be a localized mass. Multicystic mesothelioma occurs most frequently in women and has benign or indolent biologic behavior in the majority of patients. It is a multilocular cystic mass that arises from the pelvic peritoneal surfaces. Primary peritoneal serous carcinoma occurs almost exclusively in women. It is histologically identical to ovarian serous carcinoma and may be indistinguishable from metastatic ovarian carcinoma at imaging studies. Leiomyomatosis peritonealis disseminata is a rare, benign proliferative process that also occurs exclusively in women and is characterized by multiple smooth muscle nodules throughout the peritoneum. Desmoplastic small round cell tumor is a highly aggressive malignancy of unknown origin that occurs most often in the peritoneal cavity of young men. This unusual group of tumors is linked together by a common site of origin and imaging manifestations that mimic those of peritoneal carcinomatosis. Knowledge of the spectrum of imaging findings in this group of primary peritoneal tumors, along with their clinical and pathologic characteristics, is important in the evaluation of patients with diffuse peritoneal disease.

Abbreviation: H-E = hematoxylin-eosin

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**Introduction**

Primary tumors of the peritoneum are a rare, fascinating, and often confusing group of benign and malignant tumors that affect diverse patient populations. These tumors share a common anatomic site of origin in the peritoneal cavity and, as such, have similar clinical manifestations. Regardless of the underlying tumor histologic characteristics, patients frequently report abdominal discomfort, abdominal distention, gastrointestinal complaints, and, less commonly, pelvic pain or a palpable mass. When peritoneal masses are discovered, the principal diagnostic concern is metastatic disease, which is the most frequently encountered neoplastic process that involves the peritoneal cavity. However, primary peritoneal tumors should be included in the differential diagnosis, particularly when there is no evidence of a visceral primary malignancy.

The nomenclature of many of the primary peritoneal tumors is confusing because a variety of names have been applied to these tumors as our understanding of their pathogenesis and biologic behavior has evolved. Consequently, there are many synonyms for the same tumor. In addition, disparate clinicopathologic entities share similar names. For example, peritoneal malignant mesothelioma is a highly aggressive, lethal malignancy that occurs predominantly in older men and has a strong association with asbestos exposure. Well-differentiated papillary mesothelioma is a low-grade malignancy with a good prognosis that occurs predominantly in women and has no association with asbestos exposure. Multicystic mesothelioma is a benign, indolent, multilocular cystic lesion that arises most commonly from the pelvic peritoneum in women. Primary peritoneal serous carcinoma also occurs most frequently in older men and is derived from extraovarian mesothelium. It is histologically identical to serous adenocarcinoma of the ovary. Primary peritoneal borderline tumor is a rare lesion of low malignant potential that resembles serous borderline tumor of the ovary.

Knowledge of primary peritoneal tumors is important so that they can be appropriately included in the differential diagnosis in patients presenting with diffuse or focal peritoneal disease processes. This article summarizes the current literature and our recent experience with primary peritoneal tumors from the Radiologic Pathology Archives at the Armed Forces Institute of Pathology. The clinical, pathologic, and imaging features of peritoneal malignant mesothelioma, multicystic mesothelioma, primary peritoneal serous carcinoma, leiomyomatosis peritonealis disseminata, and desmoplastic small round cell tumor are emphasized.

**Normal Peritoneal Anatomy**

The peritoneum is a thin, translucent serosal membrane of mesodermal origin that covers the surface of the peritoneal cavity and its mesenteries. The peritoneum partially or completely covers the visceral organs contained within the peritoneal cavity. The peritoneum is continuous in males, resulting in a closed peritoneal cavity. In females, the peritoneum is discontinuous at the ostia of the oviducts, allowing communication between the peritoneal cavity and extraperitoneal pelvis.

The purpose of the peritoneum is to provide a frictionless surface over which the viscera can move. It also serves as a site of fluid transport. Consequently, a thin layer of hyaluronate coats the peritoneal surface. The peritoneal cavity normally contains a small amount of sterile fluid that serves as a lubricant as well as a local bacterial defense (1).

The peritoneum is divided into visceral and parietal components. The visceral peritoneum covers the intraperitoneal organs (stomach, jejunum, ileum, transverse colon, sigmoid colon, liver, and spleen), omenta, and mesenteries. The parietal peritoneum lines the anterior, lateral, and posterior abdominal walls; undersurface of the diaphragm; anterior surface of the retroperitoneal viscera (duodenum, ascending and descending colon, pancreas, and portions of the adrenal glands and kidneys); and the pelvis.

The intraperitoneal organs are suspended and supported within the peritoneal cavity by peritoneal ligaments and mesenteries. Two layers of peritoneum that invest blood vessels, lymphatics, nerves, adipose tissue, and connective tissue form the peritoneal ligaments and mesenteries. The peritoneal ligaments (coronary, gastrohepatic, hepatoduodenal, falciform, gastrocolic, duodeno-colic, gastroplenic, splenorenal, and phrenicocolic) and mesenteries (transverse mesocolon, small bowel mesentery, and sigmoid mesentery) subdivide the peritoneum into interconnected compartments that dictate the location and routes of spread of primary and secondary malignancies and infection within the peritoneal cavity (2).

The peritoneal reflections and recesses in the upper abdomen and in the pelvis are significant locations for the spread and localization of inflammatory and neoplastic peritoneal diseases (peritoneal reflections are the points or lines of...
folding where the visceral peritoneum becomes the parietal peritoneum; peritoneal recesses are the spaces formed by the reflections). The subphrenic submesothelial lymphatics communicate with subpleural lymphatics and provide the majority of the lymphatic clearance from the peritoneal cavity (3). Consequently, the subphrenic peritoneal surfaces and the visceral peritoneal surface of the liver and spleen become major sites of dissemination of primary and secondary peritoneal malignancies. The subphrenic space is separated into right and left by the falciform ligament. The right subphrenic space is limited posteriorly and inferiorly by the right coronary ligament, which forms the boundary of the bare area of the liver (Fig 1). In general, the left subphrenic space is not limited by the triangular and coronary ligaments of the left hepatic lobe because they are small and anatomically insignificant (2). However, the phrenicocolic ligament, which is a peritoneal fold extending from the splenic flexure of the colon to the diaphragm, limits the spread of peritoneal processes in the left upper quadrant because it partially separates the perisplenic space from the left paracolic gutter (2). In the clinical setting of primary and secondary peritoneal malignancies, careful evaluation of the peritoneal recesses is necessary to locate occult disease in those patients who are undergoing CT staging and planning for debulking procedures.

In the pelvis, the dependent peritoneal recesses are the rectovaginal pouch in the female (also called the cul-de-sac or pouch ofDouglas), the retrovesical pouch in the male, and the lateral paravesical recesses in both sexes (Fig 2). Primary and secondary peritoneal malignancies commonly

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**Figure 1.** Normal peritoneal anatomy and recesses in the upper abdomen. (a) Sagittal drawing of the right upper abdomen shows the right subphrenic space, bare area of the liver (arrow), and right subhepatic space. (b) Axial drawing of the upper abdomen at a level inferior to the bare area of the liver shows the right and left subphrenic spaces separated by the falciform ligament (arrow). The right subhepatic space (arrowhead) and the superior recess of the lesser sac (⋆) are shown.

**Figure 2.** Normal peritoneal reflections in the pelvis. (a) Sagittal drawing of the female pelvis shows the peritoneum lining the superior surfaces of the bladder, uterus, and anterior upper one-third of the rectum. The rectovaginal pouch is the space between the vagina and rectum (arrow). (b) Sagittal drawing of the male pelvis shows the peritoneum lining the superior surface of the bladder and anterior rectum. The retrovesical space (arrow) is between the bladder and rectum.
seed the rectovaginal and retrovesical spaces because these spaces are the most caudal, posterior, and dependent portions of the peritoneal cavity.

**Normal Peritoneal Histologic Characteristics**

The peritoneum is composed of a single layer of mesothelial cells that rest on a basal lamina; the lamina separates the mesothelial cells from a submesothelial layer of connective tissue, which consists of variable amounts of collagen, elastic fibers, fibroblast-like cells, arteries, veins, and lymphatics (Fig 3) (1). Mesothelial cells are typically flat, have abundant cytoplasm, and have a centrally located round nucleus with a single, small nucleolus. In terms of ultrastructural anatomy, mesothelial cells have long, slender, surface microvilli, which are more abundant on those mesothelial cells that cover tissues or organs that move actively (4). The microvilli entrap hyaluronate, which reduces friction at these sites.

**Tumor Classification**

Primary peritoneal tumors are defined as tumors with primary manifestation in the peritoneum in the absence of a visceral site of origin (5). They arise from mesothelial cells, submesothelial mesenchymal cells, and uncommitted stem cells (1). Because the origins of some primary peritoneal tumors are obscure, these lesions are difficult to classify precisely. Based on the proposed histogenesis, we have loosely stratified the primary peritoneal tumors into mesothelial, epithelial, smooth muscle, and uncertain origin groups (Table 1). Soft-tissue sarcomas may also arise from submesothelial tissues, but they are not traditionally classified with tumors that arise from serosal surfaces. The differential diagnosis for primary peritoneal tumors includes secondary neoplastic and nonneoplastic diseases listed in Table 2.

**Peritoneal Malignant Mesothelioma**

Malignant mesothelioma is an uncommon malignant neoplasm that arises from mesothelial cells or multipotential subserosal mesenchymal cells of the pleura, peritoneum, pericardium, or tunica vaginalis of the testis. The majority of malignant mesotheliomas originate in the pleura. Peritoneal primary mesotheliomas account for 6%–10% of malignant mesotheliomas (6). Classifying malignant mesotheliomas into diffuse and localized subtypes has prognostic significance. Diffuse malignant mesotheliomas are highly aggressive and, with a few exceptions such as well-differentiated papillary mesotheliomas that occur in women, are incurable. In contrast, patients with localized malignant mesotheliomas usually have a good prognosis following complete surgical excision of the lesions (1).
Clinical Features

The association between malignant mesothelioma and asbestos exposure is well established. In men, asbestos exposure has been shown to be the etiology in 90% of pleural malignant mesotheliomas and 60% of peritoneal malignant mesotheliomas (7). Peritoneal malignant mesothelioma usually develops in individuals exposed to higher levels of asbestos, whereas lower levels of asbestos exposure are associated with pleural malignant mesothelioma. In women, asbestos exposure plays a smaller role in the development of malignant mesothelioma. Only 23% of women who develop peritoneal malignant mesothelioma have been exposed to asbestos (1). Other etiologic factors implicated in the development of malignant mesothelioma include exposure to erionite (a mineral fiber found in Turkey), therapeutic irradiation, exposure to simian virus 40, and, in rare cases, chronic pleural or peritoneal irritation (6,8). Occasionally, malignant mesothelioma is seen in young patients with no exposure history.

The majority of all malignant mesotheliomas occur in men, with a median age at presentation of 60 years (1). In women, peritoneal malignant mesothelioma occurs in a slightly younger age group (mean age, 50 years) and, in general, has a better prognosis. Patients with diffuse peritoneal malignant mesotheliomas commonly complain of abdominal pain or discomfort, abdominal distention or increasing abdominal girth, nausea, anorexia, and weight loss. Gastrointestinal complications such as bowel obstruction may occur with advanced disease. Patients with localized peritoneal malignant mesotheliomas may complain of localized abdominal pain or a palpable abdominal or pelvic mass.

Pathologic Features

At inspection of gross specimens, diffuse peritoneal malignant mesothelioma is often indistinguishable from peritoneal carcinomatosis. It is characterized by multiple, firm, gray or white nodules scattered along the peritoneal surfaces of the mesenteries, omenta, and serosal surfaces of the viscera (Fig 4) (6). The tumor may spread along the parietal and visceral peritoneal surfaces and form a continuous tumor rind that encases the peritoneal cavity and intraperitoneal organs. The small bowel may become fixed, rigid, and
immobile, which are signs of advanced disease (1).

Diffuse peritoneal malignant mesothelioma may invade the retroperitoneum, extend into the abdominal wall, and grow through the diaphragm into the pleural cavity. Similarly, pleural mesotheliomas may grow into peritoneal cavity. When malignant mesotheliomas occur on both sides of the diaphragm, the origin of the tumor is usually clear because the bulk of the tumor will be in the peritoneal cavity if the tumor originates there or in the pleural space if the tumor is pleural in origin (1).

Localized peritoneal malignant mesothelioma forms a focal, circumscribed mass (Fig 4). It may invade locally and extend into adjacent organs, but typically it does not spread diffusely throughout the peritoneal cavity. Both localized and diffuse malignant mesotheliomas classically form solid masses, but cystic and mucoid regions within the tumor may occur and create a heterogeneous consistency on the cut surface of the tumor.

The histologic, immunohistochemical, and ultrastructural features of diffuse and localized malignant mesotheliomas are the same. Morphologically, malignant mesothelioma can be divided into three forms: epithelial, sarcomatous, and mixed (1,5). The mixed form is often referred to as biphasic or bimorphic. Mixed tumors consist of both epithelial and sarcomatous components. Distinction between the morphologic subtypes is prognostically significant because patients with pure epithelial mesotheliomas have a better prognosis than do those with sarcomatous or mixed tumors (9,10).

Epithelial malignant mesotheliomas are composed of cells that resemble normal mesothelial cells. Architecturally, the cells form a tubulopapillary or trabecular pattern. Flattened or cuboidal cells that contain monotonous nuclei line the papilla or tubules (Fig 5). Mitotic figures are uncommon. The tumor infiltrates submesothelial connective tissue, fat, and muscle. The finding of submesothelial tumor infiltration is important because it may be difficult to distinguish malignant mesothelioma from benign mesothelial hyperplasia solely on the basis of cellular features. Mesothelial hyperplasia will not infiltrate submesothelial tissues (5). Epithelial malignant mesotheliomas may also exhibit other characteristics, such as prominent secretory change, microglandular patterns, signet-ring cell structure, or desmoplastic responses, that make these tumors diffi-
cult to differentiate from adenocarcinomas based on routine histologic analysis alone. Less commonly, epithelial malignant mesotheliomas are composed of either monotonous small hyperchromatic cells that resemble small cell carcinoma or flattened, attenuated cells lining glands that resemble adenoid cystic carcinomas of the salivary glands (6,11).

The sarcomatous pattern of malignant mesothelioma is typically composed of closely packed spindled cells (Fig 6) (1). The cells may be long and slender with sparse cytoplasm, or they may have a slightly rounded, epithelioid configuration. Mitotic figures may be present. Malignant osteoid, chondroid, or muscular elements may be present within the tumor. Calcification related to the malignant osteoid and chondroid elements may be detectable at computed tomography (CT).

Although there are immunohistochemical markers to assist in identifying mesothelial cells, no immunohistochemical markers are specific for malignant mesothelioma. A panel of immunohistochemical markers (calretinin, thrombomodulin, and cytokeratin 5/6) is generally used to help differentiate malignant mesotheliomas from more common tumors such as metastatic adenocarcinomas and soft-tissue sarcomas that may have similar histologic appearances. In some cases, electron microscopy is useful to show ultrastructural features that help differentiate epithelial malignant mesothelioma from adenocarcinoma. Electron microscopy is less helpful for the differentiation of sarcomatous mesothelioma from soft-tissue sarcomas.

**Imaging Features**

Diffuse peritoneal malignant mesothelioma produces two distinct patterns on cross-sectional images that reflect its gross pathologic appearance: (a) diffuse involvement of the peritoneal cavity and (b) focal intraperitoneal masses. The diffuse pattern is characterized by tumor infiltrating and thickening the peritoneum in a sheetlike fashion (Fig 7). Consequently, there is irregular and nodular thickening of the peritoneum. The focal pattern is characterized by dominant, moderate to large-sized intraperitoneal masses with associated peritoneal studding (12–15). In addition to the primary tumor, omental caking and ascites are usually present. Omental caking may manifest as fine, nodular, soft-tissue studding (Fig 8) or coalescent, masslike soft tissue within the omentum (Fig 7). The amount of ascites associated with diffuse malignant mesothelioma is quite variable, ranging from massive, diffuse ascites to focal, small, loculated collections of fluid (12).

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**Figure 6.** Sarcomatous peritoneal malignant mesothelioma. (a) Photomicrograph (original magnification, ×40; H-E stain) shows closely packed spindle cells that infiltrate the peritoneum. (b) Photomicrograph (original magnification, ×200; H-E stain) shows polymorphic spindle cells.
Ros et al (14) correlated the patterns of malignant mesothelioma to cellular morphology and found that epithelial malignant mesothelioma produced diffuse peritoneal thickening or multiple peritoneal nodules, sarcomatous malignant mesothelioma produced a focal mass, and those with mixed morphology produced both patterns. In contrast, Kebapci et al (16) found no correlation between cellular morphology and imaging pattern. Although the number of cases was small in both of these series, our experience matches the findings of Kebapci et al (16): We have seen both patterns in all subtypes of peritoneal malignant mesothelioma.

Malignant mesothelioma may infiltrate the small bowel mesentery, thickening the leaves of the mesentery and producing a pleated or stellate appearance on cross-sectional images (Fig 9) (15). Tumor infiltration of the small bowel mesentery fixates the small bowel and its mesentery, straightening the course of the mesenteric vessels. The straight mesenteric vessels, as well as linearly oriented tumor in the mesentery, produce the pleated appearance (Fig 10). The term *stellate*

**Figure 7.** Epithelial peritoneal malignant mesothelioma in a 70-year-old man who complained of weight loss, early satiety, and abdominal distention. (a, b) Axial CT scans obtained after intravenously injected (a) and orally administered (b) contrast material show ascites, sheetlike thickening of the peritoneum (arrows in a); peritoneal nodules (arrows in b); and a large, heterogeneously enhancing mass in the greater omentum (*). (c) Photograph of the resected omental mass shows confluent white-yellow tumor nodules. Scale is in centimeters.

**Figure 8.** Epithelial diffuse peritoneal malignant mesothelioma in a 63-year-old man who complained of abdominal pain. Intravenous contrast material–enhanced CT scan shows fine, nodular soft tissue within the greater omentum (arrows) and no ascites. There is a small pneumoperitoneum and gas in the anterior abdominal wall from a previous open biopsy.
Figures 9, 10.  (9) Epithelial diffuse peritoneal malignant mesothelioma in a 55-year-old man who formerly worked in a shipyard and complained of abdominal discomfort. (a, b) Intravenous contrast-enhanced CT scans show complex ascites, serosal implants on the liver margin (black arrows in a), extension of the tumor across the diaphragm (white arrow in a), and extensive tumor encasement of the small bowel (arrows in b). The tumor also extends throughout the small bowel mesentery. (c) Photograph of the small bowel and mesentery at autopsy shows thick, nodular tumor on the serosal surface of the bowel and throughout the mesentery. The opened segment of the small bowel (arrow) shows tumor thickening the bowel wall. (10) Epithelial diffuse peritoneal malignant mesothelioma in a 73-year-old man who had a long history of asbestos exposure and complained of weight loss. (a) Intravenous contrast-enhanced CT scan shows sheetlike thickening of the anterior peritoneum (arrows), encasement of the small bowel, and pleating of the small bowel mesentery (arrowheads). (b) Image from a barium examination shows separation of small bowel segments and irregular fold thickening of small bowel segments in the left lower abdomen.
mesentery refers to thick perivascular bundles, which produce prominent vessels that, when viewed in cross section adjacent to mesenteric fat, appear similar to stars in the sky. In addition, the sheetlike pattern of growth exhibited by diffuse malignant mesothelioma may extend to the visceral peritoneal surfaces of the small bowel, encasing it. When such encasement occurs, the intestinal wall appears thick on cross-sectional images (Figs 9, 10). Because the small bowel becomes rigid and fixed in position, it may be oriented in a radial distribution that may be adherent to the parietal peritoneal surfaces. During a barium examination, the small bowel segments are separated from one another and do not show normal peristaltic movements fluoroscopically (Fig 10). Irregular small bowel fold thickening may also be present.

Calcification within diffuse peritoneal malignant mesothelioma is considered rare (12,17). However, calcified pleural plaques and other asbestos-related changes such as pleural thickening and masses may be present within the chest (Fig 11). Nodal metastases are uncommon in malignant mesothelioma. Therefore, the presence of lymph node enlargement in a patient with diffuse peritoneal disease suggests another etiology, such as diffuse peritoneal carcinomatosis, lymphomatosis, or tuberculous peritonitis.

Localized peritoneal malignant mesothelioma appears as a heterogeneous, solid intraperitoneal mass on cross-sectional images. The margins are often irregular. In our experience, invasion of adjacent visceral structures such as the liver, spleen, or pelvic organs may occur (Fig 12). Localized, loculated ascitic fluid may be present, but manifestations of diffuse peritoneal involvement, such as generalized ascites, omental caking, and peritoneal nodularity, do not typically occur. Calcifications have been reported in localized malignant mesothelioma of the pleura (18).

The imaging patterns and features of malignant mesothelioma may be shown equally well with CT or magnetic resonance (MR) imaging. The peritoneal nodules and masses produced by malignant mesothelioma typically show enhancement at CT performed after intravenous injection of iodinated contrast material. They may be homogeneous or heterogeneous in attenuation. The heterogeneous pattern of enhancement occurs when there is intratumoral degeneration or necrosis. Cystic areas may occur within the tumor.

Figure 11. Epithelial diffuse peritoneal malignant mesothelioma in a 68-year-old man with a history of asbestos exposure and who complained of increasing abdominal girth and early satiety. Intravenous contrast-enhanced CT scans show calcified asbestos-related pleural plaques (arrows in a) and massive thickening of the peritoneum (* in b).

Figure 12. Localized peritoneal malignant mesothelioma with mixed histologic features in a 47-year-old man who complained of left-sided pain and who had no history of asbestos exposure. Intravenous contrast-enhanced CT scan shows a heterogeneously enhancing mass (*) arising in the region of the gastrospenic ligament that is adherent to the splenic flexure of the colon. The mass invades the abdominal wall (arrowhead).
when it contains a mucinous component or cystic degeneration. MR imaging is not used as commonly as CT to evaluate patients who complain of abdominal pain and distention. Axial MR images of the pelvis show ascites and nodular tumor masses that are isointense relative to muscle with T1 weighting (arrows in a) and slightly hyperintense relative to muscle with T2 weighting (arrows in b).

As with MR imaging, sonography is not typically used to evaluate the entire abdomen and peritoneal cavity. However, it may be used in the initial evaluation of a patient with abdominal pain or a pelvic mass. Sonography is very sensitive for the detection of ascitic fluid. The finding of unexplained ascites should prompt a more thorough search of the abdomen, and the presence of ascites facilitates sonographic identification of peritoneal and omental masses (Fig 14). Masses and nodules arising from the pelvic peritoneal surfaces of women are generally easily identified sonographically as hypoechoic or intermediate echotexture masses in the cul-de-sac or along the peritoneal surfaces of the uterus.
Differential Diagnosis
The presence of peritoneal masses with or without ascites in the abdominal cavity is most suggestive of peritoneal carcinomatosis in the majority of patients. Malignant mesothelioma should be considered in the differential diagnosis of solid peritoneal masses when the predominant imaging finding is sheetlike thickening of the peritoneum and when there is imaging evidence of asbestos exposure. Peritoneal malignant mesothelioma is indistinguishable from carcinomatosis when the predominant imaging findings are multifocal peritoneal nodules and omental caking. Ancillary findings to support the diagnosis of peritoneal malignant mesothelioma include the lack of evidence of a primary malignancy or metastasis elsewhere and the lack of lymphadenopathy within the abdomen. Lymphomatosis and peritoneal infections such as tuberculosis and histoplasmosis may have similar appearances, but typically they are associated with enlarged lymph nodes within the small bowel mesentery or retroperitoneum.

Well-differentiated Papillary Mesothelioma
Well-differentiated papillary mesothelioma is a rare clinicopathologic entity distinct from malignant mesothelioma (19–21). It occurs predominantly in women and most often arises from the peritoneal surfaces of the pelvis, but it has been reported to occur in the pleura, pericardium, and tunica vaginalis (1). The tumor is often discovered incidentally during pelvic examination or surgery. It has no reported association with asbestos exposure. In the largest study to date, patients with well-differentiated papillary mesothelioma had numerous nodules that ranged from 0.5 to 2.0 cm in diameter and that studded the peritoneum (19). At histologic analysis, the tumors have well-developed papillary architecture with a uniform, flat or cuboidal epithelium lining the papilla (19,21). Psammomatous calcifications may be present (19).

Our knowledge of the imaging appearances of well-differentiated papillary mesothelioma is limited to a few case reports in the medical literature. Lovell and Cranston (17) reported the CT findings of small, multifocal, calcified peritoneal nodules in an 11-year-old girl with well-differentiated papillary mesothelioma. Gong et al (22) reported observing a solitary, 4.2-cm mass arising from the visceral peritoneal surface of the liver. Patients with well-differentiated papillary mesotheliomas have a good prognosis. The tumors are typically cured with complete surgical resection or follow an indolent course with long survival.

Multicystic Mesothelioma
Multicystic mesothelioma is an unusual, multilocular cystic tumor that most commonly arises from the pelvic surfaces of the peritoneum. It has benign or indolent biologic behavior in the majority of patients. Multicystic mesothelioma has many alternative names, including peritoneal inclusion cyst, multilocular inclusion cyst, and benign multicystic mesothelioma. The variable nomenclature arises from an ongoing debate regarding the origin of this lesion. Some authors consider it to be a mesothelial neoplasm because it may recur locally and in rare cases may show malignant transformation (23,24). Other authors believe that it is a nonneoplastic, reactive mesothelial proliferation (25).

Clinical Features
Multicystic mesothelioma occurs predominantly in women (mean age, 37 years). Men (mean age, 47 years) represent 16% of cases (24). The majority of patients come to clinical attention with complaints of chronic or intermittent lower abdominal or pelvic pain. Other signs and symptoms include abdominal distention, tenderness, palpable mass, dyspareunia, constipation, and urinary hesitancy and frequency (24,25). Occasionally, multicystic mesothelioma is discovered incidentally at surgery, laparoscopy, or cross-sectional imaging. Women with multicystic mesothelioma often have a history of prior pelvic surgery, endometriosis, or pelvic inflammatory disease, which some authors consider support for a nonneoplastic origin of the lesion (25). Multicystic mesothelioma has an indolent course in the majority of patients, but 50% have recurrences 1–27 years after the initial diagnosis (5) and malignant transformation has been reported (23).

Pathologic Features
Multicystic mesothelioma is composed of multiple, translucent, fluid-filled cysts that grow along the pelvic peritoneum in grapelike clusters (Fig 15). It is usually large at the time of diagnosis (mean diameter, 13 cm) (25). In women, the tumor is typically located in the cul-de-sac and along the peritoneal surfaces of the uterus and rectum. In men, it most commonly arises along the peritoneal surface of the bladder and rectum.
When large, multicystic mesothelioma extends into the upper portions of the peritoneal cavity. Multifocality, freely floating cysts, and unilocular cysts have been reported (24).

At histologic analysis, multicystic mesothelioma is composed of multiple, thin-walled, irregularly spaced cysts lined by flattened or cuboidal mesothelial cells (Fig 16). The cysts may be filled with eosinophilic, serous fluid. Inflammatory cells and fibrous elements may be found within the stroma between the cysts (24). Focal reactive mesothelial changes such as hobnail-shaped cells and foci of mesothelial hyperplasia may also be present (5). With immunohistochemical analysis, multicystic mesotheliomas stain positive for calretinin and cytokeratins, results that confirm their mesothelial origin; however, they do not contain lymphocytes and lymphoid aggregates, which help distinguish them from lymphangiomas.

**Imaging Features**

Multicystic mesotheliomas occurring in women are often initially detected at pelvic sonography. Typically, they are multiseptated, cystic structures that have an intimate anatomic association.
with the uterus and ovaries (Fig 17). The fluid within the cysts is generally anechoic, but the cysts may contain echoes from debris or hemorrhage (Fig 17a). The number and complexity of septations, as well as the size of the cyst locules, are quite variable (26). Some may have few and incomplete septations, whereas others have innumerable septations of variable thickness. Multicystic mesothelioma may completely surround the ovaries such that the ovaries appear entrapped within the cystic lesion (27). Calcification has not been described in multicystic mesothelioma. Nodular excrescences or thick, polypoid, incomplete septations may be present, findings that simulate features more commonly associated with cystic ovarian neoplasms.

CT and MR imaging are useful modalities for defining the anatomic extent of the lesion and for surveying the upper abdomen for multifocal lesions (Fig 18). In addition, CT and MR imaging aid in the differential diagnostic evaluation by allowing the exclusion of visceral primary neoplasms that may produce pseudomyxoma peritonei and thus resemble multicystic mesothelioma. On CT scans, the cysts of multicystic mesothelioma have fluid attenuation values. The septa enhance with intravenous administration of contrast material (Fig 18). Innumerable small cysts and thick-walled cysts may appear as soft-tissue attenuation lesions, necessitating peritoneal carcinomatosis and pseudomyxoma peritonei to be included in the differential diagnosis (Fig 19a).

MR imaging is an excellent modality for visualizing pelvic anatomic relationships in patients with complex, cystic masses. The cysts in multicystic mesothelioma generally have homogeneous

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**Figure 17.** Multicystic mesothelioma in a 65-year-old woman with chronic pelvic pain. (a) Midline sagittal endovaginal sonogram of the pelvis shows a complex, multiseptated cystic mass (⊕) posterior to the uterus in the cul-de-sac. (b) Sagittal endovaginal sonogram of the right adnexal region demonstrates the complex cystic mass (arrow) adjacent to the right ovary.

**Figure 18.** Multicystic mesothelioma in a 35-year-old man who complained of increasing abdominal girth. Intravenous contrast-enhanced CT scans of the abdomen (a) and pelvis (b) show a cystic mass with enhancing septa (arrow in a) that extend from the superior margin of the bladder (⊕ in b).
fluid signal intensity that is hypointense on T1-weighted images and hyperintense on T2-weighted images (26). The septations enhance with intravenous administration of gadolinium-based contrast material (Fig 20).

Multicystic mesothelioma is not often considered in the differential diagnosis of a complex pelvic or abdominal cystic mass in a man because it...
occurs less commonly in men compared with women and because a pelvic origin may not be present or as clearly evident in male patients. In men, multicystic mesothelioma typically arises from the peritoneal surface along the superior margin of the bladder or in the retrovesical space (Fig 19b).

**Differential Diagnosis**
Pseudomyxoma peritonei (mucinous peritoneal metastasis) should be considered in the differential diagnosis of multicystic mesothelioma. Pseudomyxoma peritonei can be distinguished from multicystic mesothelioma when there is coexisting omental caking, soft-tissue peritoneal nodules, and scalloping of the serosal margins of the liver or spleen. These findings are not present in multicystic mesothelioma. However, freely floating cysts may be seen away from the dominant cyst and as such may mimic metastatic disease. Lymphangioma, complex loculated ascites, and other mesenteric cysts such as duplication cysts may have a similar appearance. When multicystic mesothelioma is located solely in the pelvis in women, tubo-ovarian abscess, hydrosalpinx, and cystic ovarian neoplasms should be considered in the differential diagnosis.

**Adenomatoid Tumor**
Adenomatoid tumor of the peritoneum is a very rare, benign mesothelial tumor that is usually incidentally discovered. It is typically small, less than 2 cm in diameter, and is characterized histologically by epithelioid cells that form vacuoles and tubular spaces within a fibrous stroma (5). It has been reported to occur in conjunction with multicystic mesothelioma as a composite tumor (28). To our knowledge, there are no reports in the medical literature describing the cross-sectional imaging appearance of peritoneal adenomatoid tumors.

**Primary Peritoneal Serous Carcinoma**
Primary peritoneal serous carcinoma is an epithelial tumor that arises from the peritoneum. At histopathologic analysis, it resembles a malignant ovarian surface epithelial stromal tumor. Primary peritoneal serous carcinoma is thought to arise from extraovarian mesothelium that has müllerian
potential, making it a unique clinicopathologic entity distinct from its ovarian counterpart (29). The nomenclature surrounding primary peritoneal serous carcinoma is confusing, and a number of different names have been used: serous surface papillary carcinoma, primary peritoneal carcinoma, extraovarian pelvic serous carcinoma, primary serous papillary carcinoma, and psammomacarcinoma.

Clinical Features
Primary peritoneal serous carcinoma almost always occurs in women (mean age, 56–62 years) (30,31). There are a few case reports of primary peritoneal serous carcinoma developing in men (32). Patients typically present with complaints of abdominal distention, pain, and fullness; increasing abdominal girth; and gastrointestinal symptoms such as nausea and vomiting (31,33). Clinically, the majority of patients have ascites and elevation of serum levels of cancer antigen CA-125. According to recent studies, primary peritoneal serous carcinoma is more common than previously thought, with 15% of epithelial ovarian carcinomas actually being primary peritoneal serous carcinomas (5). Patients with the breast cancer gene BRCA1 may develop a BRCA1-related primary peritoneal serous carcinoma, which may occur many years after prophylactic oophorectomy (34). Studies suggest that BRCA1-related primary peritoneal serous carcinoma has a unique molecular pathogenesis compared with BRCA1-related ovarian carcinoma (30).

Pathologic Features
Primary peritoneal serous carcinoma is indistinguishable from metastatic serous ovarian carcinoma at gross, histopathologic, and immunohistochromical examination. The primary gross characteristic is multiple nodules on the peritoneal surface and omentum. Omental caking may occur with confluent and large masses. Large masses may also have a papillary appearance grossly (1). The tumors are histologically composed of irregular, interconnecting clusters of malignant cells that show solid, cribriform, or cystic architecture (31). Psammoma bodies are commonly present and may be numerous, leading some authors to refer to this tumor as psammomacarcinoma (Fig 22) (1).

The following criteria have been established to make the diagnosis of primary peritoneal serous carcinoma: (a) both ovaries are normal; (b) the involvement of extraovarian sites must be greater than the involvement on the surface of either ovary; or (c) the ovarian involvement is limited to the ovarian surface epithelium, either without stromal invasion or involving the cortical stroma with tumor size less than 5 × 5 mm (5).

Imaging Features
Ascites, peritoneal nodules and thickening, and omental nodules and masses are the most common cross-sectional imaging features of primary peritoneal serous carcinoma (Fig 23) (35–38). A minority of patients have either no ascites or only
a small amount at initial presentation (Fig 24). The peritoneal recesses of the upper abdomen, particularly the subphrenic spaces, should be carefully evaluated for the presence of disease because these areas are a major site of lymphatic clearance of the peritoneum (3). Peritoneal and omental nodules and masses enhance with intravenous contrast material at CT and MR imaging. Calcifications within peritoneal and omental nodules represent psammoma bodies histopathologically. Psammomatous calcification has been reported to occur in 30% of cases (Fig 24) (35,38).

No detectable adnexal mass is identified at CT in the majority of patients with primary peritoneal serous carcinoma (35,38). In the series reported by Morita et al (37), five of 11 patients had unilateral or bilateral ovarian enlargement on CT or MR images that correlated histologically to microscopic tumor invasion of the superficial ovarian parenchyma.

**Differential Diagnosis**
The cross-sectional imaging features of primary peritoneal serous carcinoma overlap with those of other diffuse neoplastic processes in the peritoneum, such as peritoneal carcinomatosis, metastatic ovarian carcinoma, and malignant mesothelioma. However, primary peritoneal serous carcinoma should be suggested in the differential diagnosis when the findings of ascites and peritoneal and omental nodules and masses are identified in a female patient with no evidence of a visceral primary or ovarian mass.

**Primary Peritoneal Serous Borderline Tumor**
Primary peritoneal serous borderline tumor (also known as peritoneal serous micropapillomatosis) is a rare lesion of low malignant potential that is histologically identical to borderline surface epithelial stromal tumors of the ovary. It is histologically differentiated from primary peritoneal serous carcinoma because the tumor cells do not invade into the submesothelial layers of the peritoneum or omental fat.
Primary peritoneal serous borderline tumors occur in female patients from 16 to 67 years of age (mean, 33 years) and are most often discovered incidentally during laparotomy or laparoscopy as focal or diffuse miliary nodules on the peritoneal and omental surfaces (39). Patients are generally treated by surgical resection (omentectomy, hysterectomy, and oophorectomy) and have a good long-term prognosis (5). To our knowledge, there are no reports in the medical literature describing the cross-sectional imaging appearance of primary peritoneal serous borderline tumor.

**Leiomyomatosis Peritonealis Disseminata**

Leiomyomatosis peritonealis disseminata (also called diffuse peritoneal leiomyomatosis) is a rare, benign entity characterized by innumerable smooth muscle nodules throughout the peritoneal cavity. The smooth muscle nodules are derived from metaplasia of submesothelial cells. Leiomyomatosis peritonealis disseminata is usually discovered incidentally during surgery or imaging examinations of women of childbearing age who have uterine leiomyomas. It may be associated with high estrogen states, caused by pregnancy and oral contraceptive use (40), but rare cases in postmenopausal women and men have also been reported (41,42). The majority of patients have a benign clinical course, with spontaneous regression of the leiomyomas or regression following withdrawal of ovarian hormones or oophorectomy. In rare cases, sarcomatous degeneration may occur (43).

**Pathologic and Imaging Features**

At gross examination, well-demarcated, firm nodules of varying size are observed throughout the peritoneum. The nodules of leiomyomatosis peritonealis disseminata are histologically composed of closely packed eosinophilic spindle cells in the background of omental or mesenteric adipose tissue (Fig 25). The spindle cells have bland, uniform nuclei, and mitotic figures are absent (1).

Cross-sectional imaging studies show numerous well-circumscribed solid masses in the peritoneal cavity that vary in size from several millimeters to many centimeters (Fig 26). The masses are often heterogeneous in CT attenuation and enhance similar to uterine leiomyomas (Fig 26) (43–45). At MR imaging, the masses of leiomyomatosis peritonealis disseminata are isointense relative to muscle with T1-weighted sequences, heterogeneously enhance following intravenous administration of gadolinium, and are low signal intensity with T2-weighted sequences (Fig 26) (43).

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**Figure 25.** Leiomyomatosis peritonealis disseminata. (a) Photomicrograph (original magnification, ×40; H-E stain) shows multiple circumscribed nodules of smooth muscle (arrows). (b) Photomicrograph (original magnification, ×100; H-E stain) shows interlacing fascicles of benign smooth muscle cells.
Figure 26. Leiomyomatosis peritonealis disseminata in a 31-year-old woman who had a history of uterine fibroids and who complained of lower abdominal pain. (a, b) Intravenous and oral contrast-enhanced CT scans of the abdomen (a) and pelvis (b) show multiple enhancing masses throughout the peritoneum (arrows). There are innumerable masses in the pelvis. (c) Axial T1-weighted MR image of the pelvis shows that the uterus (arrow) and peritoneal masses (arrowheads) are isointense relative to skeletal muscle. (d) Axial gadolinium-enhanced T1-weighted MR image of the pelvis illustrates heterogeneous enhancement of the uterus (arrow) and peritoneal masses (arrowheads). (e) Axial T2-weighted MR image shows low-signal-intensity peritoneal masses (arrowheads) and multiple discrete fibroids within the uterus (arrow). A small amount of ascites is also present.
Differential Diagnosis
Although, the finding of multiple peritoneal nodules is nonspecific and overlaps with the appearances of peritoneal carcinomatosis, malignant mesothelioma, and primary peritoneal serous carcinoma, leiomyomatosis peritonealis disseminata can be suggested as the diagnosis when the patient has coexisting uterine leiomyomas and no evidence of omental caking and ascites. The typical signal intensity patterns on MR images can be used to confirm the diagnosis.

Desmoplastic Small Round Cell Tumor
Desmoplastic small round cell tumor is a rare malignancy of unknown histogenesis that occurs predominantly in adolescent and young adult males. It is a distinctive clinicopathologic entity in the family of primitive pediatric tumors that are composed of small, round, blue cells, a group that includes Wilms tumor, Ewing sarcoma, peripheral primitive neuroectodermal tumor, and Askin tumor. Patients with desmoplastic small round cell tumor have a universally poor prognosis. The 3-year survival rate, even with treatment, is less than 30% (46,47).

Clinical Features
Desmoplastic small round cell tumor most commonly arises in the peritoneal cavity of young men with a mean age of 19 years (48). Primary para-testicular, pleural, and nonserosal desmoplastic small round cell tumors have been reported (1). Occasionally, the tumor may develop in older and female patients. The most frequent presenting symptom is crampy abdominal pain. Less common symptoms include abdominal distention, constipation, weight loss, diarrhea, dysphagia, hematemesis, jaundice, and hematuria (47).

Pathologic Features
At gross inspection, desmoplastic small round cell tumor may be a solitary or multifocal, gray to white, firm nodule or mass that arises from the peritoneal surface. Discrete and confluent peritoneal and omental nodules are often present. The histopathologic characteristics of desmoplastic small round cell tumor are quite distinctive. Cords and nests of undifferentiated, uniform, small, round, malignant cells are surrounded by dense, collagenous stroma (Fig 27) (49). The tumor cells have scant cytoplasm and large, hyperchromatic nuclei with dispersed nuclear chromatin. The presence of numerous mitotic figures and single cell necrosis is characteristic. Identification of the Ewing tumor family chromosomal translocation t(11;22)(p13;q12) is useful to confirm the histopathologic diagnosis (48,49).

Figure 27. Desmoplastic small round cell tumor. (a) Photomicrograph (original magnification, ×40; H-E stain) depicts cords of undifferentiated, uniform, small, round cells surrounded by a dense collagenous stroma (arrows). (b) Higher-power photomicrograph (original magnification, ×400; H-E stain) shows the tumor cells with scant cytoplasm and large, hyperchromatic nuclei (arrows).
Imaging Features
Desmoplastic small round cell tumor spreads diffusely throughout the peritoneal surfaces. Consequently, the primary imaging finding is the presence of peritoneal thickening, nodules, and masses. However, a solitary peritoneal mass may be the only finding seen at initial presentation (50). Intraperitoneal primary masses may be large and bulky (51,52). The majority of patients in the series reported by Bellah et al (51) had a dominant tumor mass larger than 10 cm (Fig 28). The masses are characteristically heterogeneous in CT attenuation or have centrally located low-attenuation regions, which reflect intratumoral necrosis or hemorrhage (Fig 28) (50,52). The tumor masses may contain small, punctate calcifications, visible on CT scans (Fig 29). Malignant ascites is frequently present. Complications such as bowel obstruction or ureteral obstruction may occur. The latter is particularly common in patients with dominant pelvic masses (51). At MR imaging, the tumors are typically heterogeneous in signal intensity but predominantly hypointense with T1-weighted sequences and hyperintense with T2-weighted sequences (53,54). Imaging after intravenous administration of gadolinium shows heterogeneous enhancement (Fig 30). Careful attention should be paid to all bones, solid organs, and lymphatic structures because hematogenous and lymphatic metastases are common at initial presentation as well as during disease progression. Fifty percent of the cases reported by Quaglia and Brennan (47) had distant metastasis at the time of presentation. Hematogenous metastasis to the liver, lung, and bone may occur.

Differential Diagnosis
Desmoplastic small round cell tumor should be strongly considered in the differential diagnosis for a young male patient who has multifocal or solitary peritoneal masses. Peritoneal carcinomatosis, lymphomatosis, and benign conditions such as splenosis should also be considered. However, the presence of a single or multiple dominant masses within the diffuse process is more characteristic of desmoplastic small round cell tumor compared with the other lesions. In addition, imaging evidence of tumor heterogeneity, calcification, or intratumoral degeneration is also suggestive of desmoplastic small round cell tumor.

Solitary Fibrous Tumor
Solitary fibrous tumors are submesothelial tumors of uncertain origin that most commonly arise in the pleura. Reports of extrapleural solitary fibrous tumors are unusual; in very rare cases, these tumors are reported to occur in the abdomen (55).
Figure 29. Desmoplastic small round cell tumor in a previously asymptomatic 16-year-old boy who was imaged following a motor vehicle accident and found to have a splenic laceration, hemoperitoneum, and peritoneal masses. Intravenous and oral contrast-enhanced CT scans of the abdomen (a) and pelvis (b) show the hemoperitoneum and multiple solid intraperitoneal masses (arrows in a). Some of the masses contain calcifications (arrow in b).

Figure 30. Desmoplastic small round cell tumor with bone metastasis in a 31-year-old man who complained of pelvic pressure and difficulty in urinating. (a) Intravenous contrast-enhanced CT scan of the pelvis shows a large heterogeneous mass (arrow) posterior to the bladder and anterior to the rectum. (b) Coronal T1-weighted MR image demonstrates that the pelvic mass is hypointense and reveals an infiltrative hypointense mass within the marrow space of the left proximal femur. (c) Coronal T2-weighted MR image shows that the pelvic mass has heterogeneous high signal intensity and that the left proximal femur metastasis has associated high signal marrow edema. (d) Coronal gadolinium-enhanced T1-weighted MR image shows heterogeneous enhancement of the primary pelvic tumor as well as of the femoral metastasis.
Conclusions

Primary peritoneal tumors are an uncommon group of diverse pathologic disorders that share a common anatomic site of origin and have overlapping imaging features, yet are distinctly different clinically. Their imaging appearances overlap with those of diffuse peritoneal metastatic disease and, less commonly, lymphomatous or infectious involvement of the peritoneum. Differentiating primary peritoneal tumors from metastatic disease is important clinically so that patient management is appropriate.

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References


Diffuse peritoneal malignant mesothelioma produces two distinct patterns on cross-sectional images that reflect its gross pathologic appearance: (a) diffuse involvement of the peritoneal cavity and (b) focal intraperitoneal masses.

Multicystic mesothelioma may completely surround the ovaries such that the ovaries appear entrapped within the cystic lesion (27).

However, primary peritoneal serous carcinoma should be suggested in the differential diagnosis when the findings of ascites and peritoneal and omental nodules and masses are identified in a female patient with no evidence of a visceral primary or ovarian mass.

Leiomyomatosis peritonealis disseminata is usually discovered incidentally during surgery or imaging examinations of women of childbearing age who have uterine leiomyomas.

Desmoplastic small round cell tumor is a rare malignancy of unknown histogenesis that occurs predominantly in adolescent and young adult males. It is a distinctive clinicopathologic entity in the family of primitive pediatric tumors that are composed of small, round, blue cells, a group that includes Wilms tumor, Ewing sarcoma, peripheral primitive neuroectodermal tumor, and Askin tumor.