Primary pericardial tumors are rare and may be classified as benign or malignant. The most common benign lesions are pericardial cysts and lipomas. Mesothelioma is the most common primary malignant pericardial neoplasm. Other malignant tumors include a wide variety of sarcomas, lymphoma, and primitive neuroectodermal tumor. When present, signs and symptoms are generally nonspecific. Patients often present with dyspnea, chest pain, palpitations, fever, or weight loss. Although the imaging approach usually begins with plain radiography of the chest or transthoracic echocardiography, the value of these imaging modalities is limited. Cross-sectional imaging, on the other hand, plays a key role in the evaluation of these lesions. Computed tomography and magnetic resonance imaging allow further characterization and may, in some cases, provide diagnostic findings. Furthermore, the importance of cross-sectional imaging lies in assessing the exact location of the tumor in relation to neighboring structures. Both benign and malignant tumors may result in compression of vital mediastinal structures. Malignant lesions may also directly invade structures, such as the myocardium and great vessels, and result in metastatic disease. Imaging plays an important role in the detection, characterization, and staging of pericardial tumors; in their treatment planning; and in the posttreatment follow-up of affected patients. The prognosis of patients with benign tumors is good, even in the few cases in which surgical intervention is required. On the other hand, the length of survival for patients with malignant pericardial tumors is, in the majority of cases, dismal.

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Abbreviations: ADC = apparent diffusion coefficient, MIBG = metaiodobenzylguanidine, PET = positron emission tomography
Primary cardiac neoplasms are rare, with a prevalence of approximately 0.02%–0.056% (1,2); however, those arising primarily from the pericardium are even less common. Meng et al (3) and Patel et al (4) determined that primary pericardial neoplasms accounted for only 6.7%–12.8% of all primary tumors arising in the cardiac region. On the basis of these data, the prevalence of primary pericardial neoplasms ranges from about 0.001 to 0.007%. Neoplastic involvement of the pericardium by direct invasion or metastatic disease is, on the other hand, 100–1000 times more prevalent (5). The most common benign pericardial mass is a pericardial cyst, followed by lipoma. Mesothelioma is the most common primary pericardial malignancy. Other primary malignant neoplasms include a wide variety of sarcomas as well as lymphoma (4).

The patient’s clinical history is highly valuable in narrowing the differential diagnosis. In patients with known neoplastic disease, a pericardial lesion will most likely be metastatic. Pericardial fibrosis may be seen in patients who have undergone irradiation of the chest. In patients with a history of sternotomy, a hematoma or infectious process may manifest as a mass. In addition, diffuse processes such as pericarditis may also mimic a neoplasm.

Patients with primary pericardial neoplasms present with diverse symptoms. Clinical signs and symptoms are usually the result of associated pericardial effusion, pericarditis, or invasion of adjacent structures and are therefore nonspecific. Symptoms include exertional dyspnea, chest pain, cough, palpitations, fatigue, night sweats, fever, and facial or lower-extremity edema. Occasionally, a primary pericardial tumor may be found incidentally in an asymptomatic patient, during work-up for an unrelated illness (5–8).

With such a broad spectrum of clinical symptoms and findings, imaging plays an important role in the evaluation of patients with primary pericardial neoplasms. Most often, the diagnostic approach begins with plain radiography of the chest or transthoracic echocardiography. Radiographic findings include an enlarged cardiac silhouette, an abnormal mediastinal contour, or a discrete mediastinal mass (5). Echocardiography, although commonly performed, has a limited role in the evaluation of primary pericardial neoplasms. Frequently, the only finding may be a thickened pericardium or pericardial effusion. Transesophageal echocardiography offers increased sensitivity compared with transthoracic echocardiography and may be a useful tool (8). Inherent limitations to this modality include its invasiveness and its limitations in the assessment of structures that are distant to the transducer, including the mediastinum, diaphragm, and lung parenchyma. Computed tomography (CT) demonstrates the location of the tumor, its relationship with neighboring structures, and its invasion of vital structures. CT also aids in narrowing the differential diagnoses, as some of these lesions may be characterized on the basis of their attenuation values or pattern of enhancement. In addition, CT can help confirm the presence of locoregional or distant metastatic disease. Magnetic resonance (MR) imaging provides higher contrast resolution compared with CT and, as a result, is a more powerful tool for characterizing the lesion. Another advantage of MR imaging is its ability to demonstrate myocardial invasion, as well as give insight into the functional impact of the neoplasm (5,8–11). Positron emission tomography (PET)/CT may be useful as a staging tool, as it may demonstrate distant or locoregional spread of disease (12).

Although in most cases the diagnosis of a primary pericardial tumor is ultimately reached through tissue sampling, knowledge of the patient characteristics is fundamental for the radiologist to offer a pertinent differential diagnosis on the basis of imaging appearance, because clinically, many of these lesions behave similarly. The radiologist also plays an important role in assessing the resectability of these lesions and evaluating the patient for associated complications. Some of the commonly seen complications include invasion of mediastinal structures, regional or distant metastases, pericardial effusion, cardiac tamponade, compression of vascular structures or cardiac chambers, encasement of vital structures, involvement of coronary arteries, myocardial infarction, diastolic dysfunction, and constrictive physiology.

In this article, the spectrum of primary pericardial tumors is discussed, with emphasis on their imaging features, the patients’ clinical presentation, and the wide array of associated cardiovascular complications.
Pericardial Cyst

Pericardial cysts, also called mesothelial cysts of the pericardium, are congenital lesions that arise as a result of aberrant formation of the somatic cavities (13). These cysts are usually unilocular, and the cyst walls are lined with a single layer of mesothelial cells, whereas the remainder of the wall is composed of a thicker layer of connective tissue (14). Most commonly, pericardial cysts are located in the right anterior cardiophrenic angle, with the next most frequent location being the left anterior cardiophrenic angle. Occasionally, a pericardial cyst may arise elsewhere in the mediastinum (13). In more than half of the cases, pericardial cysts result in no symptoms. When present, patients’ symptoms are often vague and include chest pain, cough, fever, and arrhythmias (15). Symptoms may also result from compression of adjacent structures. Serious complications are rare, but they may include erosion into vascular structures, pericarditis, obstruction of the right ventricular outflow tract, pulmonary stenosis, or sudden death (15).

Pericardial cysts are most often found incidentally at chest radiography or echocardiography. At chest radiography, a pericardial cyst appears as a well-defined, smooth, anterior mediastinal mass. Echocardiography demonstrates the cystic nature of this lesion (13,14).

CT and MR imaging are better at characterizing a pericardial cyst and its relationship with adjacent structures. Typically, pericardial cysts appear on CT images as well-defined, nonenhancing, homogeneous fluid-attenuation lesions that contain no internal septa (16). Occasionally, the cyst may contain proteinaceous fluid, in which case the lesion will demonstrate intermediate attenuation at CT. Pericardial cysts with hemorrhagic contents appear with hyperattenuation at CT.

MR imaging offers further characterization of the internal contents of the cysts. Usually, pericardial cysts demonstrate intermediate to low signal intensity on T1-weighted images and high signal intensity on T2-weighted images (Fig 1). Pericardial cysts with proteinaceous content show high signal intensity with T1-weighted sequences and intermediate to low signal intensity with T2-weighted sequences. Diffusion-weighted imaging may be useful in these cases, as pericardial cysts do not demonstrate restricted diffusion and show high signal on apparent diffusion coefficient (ADC) maps (Fig 2) (16,17). Hemorrhagic cysts are hyperintense with T1-weighted sequences and demonstrate susceptibility with gradient recalled echo sequences. Surgical resection or percutaneous drainage is reserved for symptomatic individuals, when complications are present, or when the diagnosis is uncertain and pathologic analysis is needed (Fig 3) (15).

**Figure 1.** Pericardial cyst. (a) Contrast material–enhanced CT image of the chest demonstrates a well-defined lesion of fluid attenuation in the right anterior cardiophrenic angle. (b) Coronal T2-weighted MR image shows the well-defined hyperintense lesion in the right anterior cardiophrenic angle.
Figure 2. Proteinaceous pericardial cyst. (a) T1-weighted MR image demonstrates a high-signal-intensity well-defined lesion in the anterosuperior aspect of the pericardium. (b) T2-weighted MR image obtained at the same level shows intermediate to low signal intensity in the lesion. (c) ADC map reveals high signal intensity in the mass, a finding that confirms the cystic nature of the lesion. Findings on T1- and T2-weighted images are in keeping with proteinaceous content.

Figure 3. Acute inflammation in a chronically inflamed pericardial cyst. Contrast-enhanced CT images (a obtained at a higher level than b) demonstrate a large, peripherally calcified, fluid-attenuation lesion along the left aspect of the pericardium that causes displacement of the left pulmonary artery. There is mass effect on the left atrium (arrows in b) and, to a lesser extent, on the left ventricle. Aspiration of the lesion yielded an exudate, and after surgical resection, pathologic analysis revealed acute on chronic inflammation of a pericardial cyst.

Pericardial Lipoma
Pericardial lipomas are benign lesions that most commonly grow in an insidious fashion, therefore resulting in few symptoms (18). As with subcutaneous lipomas, pericardial lipomas are encapsulated; however, unlike their subcutaneous counterparts, lipomas in the pericardium tend to be less circumscribed: Their contours are determined by the space they occupy (Figs 4, 5). Symptoms, when present, are usually related to compression of adjacent structures (Fig 5).
Figures 4, 5. (4) Pericardial lipoma. Noncontrast CT image of the chest demonstrates a large, low-attenuation intrapericardial lesion that insinuates into the pericardial recesses and conforms to the pericardial space. (5) Large pericardial lipoma. Contrast-enhanced CT scan of the chest shows a large low-attenuation lesion confined to the pericardium that conforms to the pericardial space and its recesses. The mass demonstrates no enhancing components. There is associated mass effect on the right and left atria (arrows).

At echocardiography, pericardial lipomas appear echogenic. CT findings are usually diagnostic, as these lesions demonstrate homogeneously low attenuation (<0 HU) without enhancing components (Figs 4, 5). A capsule may or may not be identified (Fig 6). MR imaging may be useful because of its superior contrast resolution. Lipomas appear as nonenhancing lesions that are hyperintense on T1-weighted MR images and intermediate to high signal intensity on T2-weighted MR images. Use of fat-saturated sequences to confirm the fatty nature of the lesion is helpful, as these sequences will result in a uniform drop in signal (Fig 6) (19).

Pericardial Lipoblastoma

Lipoblastomas are rare benign tumors that arise from embryonic fat and are almost exclusively seen in infants and children less than 3 years of age (20). The term lipoblastoma is usually reserved...
Pericardial Lipoblastoma

In young boys, pericardial lipoblastomas present as a large, well-defined, fatty pericardial mass with scattered enhancing septa. The patient’s age at presentation favors a diagnosis of lipoblastoma over liposarcoma. Deep lipoblastomas tend to be unencapsulated and diffuse, and they may demonstrate a more infiltrative behavior. When these features are present, the proper term is lipoblastomatosis (21,22). Occasionally, lipoblastomas may arise in the pericardium.

The imaging features of pericardial lipoblastoma are similar to those of pericardial lipomas. These lesions appear as an echogenic mass at echocardiography. On CT images, they are predominantly low in attenuation and may demonstrate septations of soft-tissue attenuation. At MR imaging, lipoblastomas follow subcutaneous adipose tissue signal and therefore appear hyperintense with T1-weighted sequences and moderately intense to hyperintense with T2-weighted sequences. The most characteristic feature is the presence of septations and intratumoral streaks that represent the fibrovascular network (Fig 7). Occasionally, cystic changes may be present (21,22).

Pericardial Paraganglioma

Intrapericardial paragangliomas are slow-growing tumors derived from neural crest cells and most often are nonfunctioning (23,24). Their functioning counterpart is exceedingly rare (25). Although most paragangliomas are benign, a minority of these neoplasms may exhibit locally invasive behavior or distant metastatic disease (24). As with paragangliomas in other anatomic locations, when intrapericardial paragangliomas are functioning, they result in symptoms related to elevated plasma levels of catecholamines, such as tremors, sweating, palpitations, headaches, and hypertension. Nonfunctioning intrapericardial paragangliomas are either incidentally discovered at imaging or are found as the culprit lesion when patients present with symptoms derived from compression of mediastinal structures. Intrapericardial paragangliomas most commonly arise adjacent to the left atrium or anterior to the aortic root near the origin of the coronary arteries.

On chest radiographs, an intrapericardial paraganglioma commonly resembles left atrial enlargement and is seen as a mass that splays the carina (23). CT and MR imaging are useful in further localizing and characterizing the lesion and play important roles in surgical planning. On contrast-enhanced CT images, the paraganglioma appears as a hyperenhancing vascular mass (Fig 8). In approximately one-half of the patients, a central area of low attenuation that most likely represents necrosis or degeneration is seen (23). The characteristic MR imaging feature is hyperintense signal with T2-weighted sequences. Contrast-enhanced T1-weighted images demonstrate findings similar to those seen on CT images. Cross-sectional imaging is useful for assessing the relationship of the tumor to adjacent structures, in particular invasion of the adjacent atrium (23). Patients may undergo imaging with indium 111 pentreotide, 131I MIBG, or 123I MIBG scintig-
raphy as part of their work-up. Scintigraphy demonstrates an intrathoracic focus of increased tracer activity and is especially useful when the lesion remains occult with other imaging modalities (Fig 8) (5,23,26). 111In pentreotide is the nuclear medicine agent of choice, with a sensitivity of 94%. The sensitivity of MIBG scintigraphy is lower, as it accumulates in only functioning paragangliomas (26). An intrathoracic focus of increased tracer activity is present in more than 90% of patients (Fig 8) (23). Conventional coronary angiography may also play an important role if surgery is planned. In some paragangliomas, the blood is supplied by vessels originating from the coronary arteries, and resection could result in myocardial ischemia unless appropriate presurgical planning is conducted (24).

Germ Cell Tumors
Germ cell tumors may arise from the pericardium. These tumors are composed of tissue from at least two of the three germ cell layers. The majority of germ cell tumors are located anteriorly and to the right. Most of them manifest in the newborn period or infancy. Diagnosis is increasingly made in utero at prenatal ultrasonography. Mature teratomas are generally cystic and are typically attached by a fibrous pedicle to the heart or great vessels. When these lesions are detected in utero, the main finding is that of fetal hydrops (fetoplacental anasarca, pleural or pericardial effusion) (8). The presence of an associated pericardial effusion in neonates may result in a surgical emergency if tamponade physiology is found. When present in young adults, germ cell tumors are usually asymptomatic.

On CT images, germ cell tumors are usually well-defined, heterogeneous, multilocular masses. Typically, the cystic component predominates. Calcifications are common, and fat is seen in about 75% of patients. On MR images, teratomas are heterogeneous masses with signal intensity characteristics that correspond to those of their components (Fig 9) (5,8,27).
Pericardial Hemangioma

Pericardial hemangiomas can occur in any age group and are commonly found incidentally at autopsy. The histologic classification includes capillary, cavernous, or venous type. When arising from the pericardium, hemangiomas are most often of the cavernous type and they usually arise from the visceral pericardium (28,29). Their size ranges from 1 to 8 cm, and although most pericardial hemangiomas are solitary, cases of multiple lesions have been described (8). Occasionally, patients will present with hemopericardium and findings of cardiac tamponade. It has been suggested that these conditions may develop from friction that ruptures neoplastic vessels (30,31).

As with hemangiomas that occur elsewhere in the body, pericardial hemangiomas appear as heterogeneous lesions on noncontrast-enhanced CT images and may contain foci of calcification. When contrast material is administered, they avidly enhance. At MR imaging, the hemangioma appears as a mass with intermediate T1-weighted signal intensity and high T2-weighted signal intensity. Upon contrast material administration, hemangiomas demonstrate nodular areas of enhancement with progressive filling on delayed images. Hemangiomas show benign features, such as well-circumscribed borders and no evidence of invasion of neighboring structures (8,28,31,32). These features help differentiate these lesions from angiosarcoma.

Pericardial Fibroma

Pericardial fibromas are benign tumors. They may grow to a large size and result in very minor symptoms. At CT, a fibroma appears as a soft-tissue attenuation mass. Fibromas are characteristically hypointense on T2-weighted MR images and isointense on T1-weighted MR images (33). On MR images obtained after the administration of gadolinium, fibromas demonstrate little enhancement, and when enhancement occurs, it is often heterogeneous (16).

Inflammatory Pseudotumor

Inflammatory pseudotumor is a benign process most commonly seen in the lung and orbit, but it can arise anywhere in the body, including the pericardium (34,35). The cause of inflammatory pseudotumor is unknown. Some cases develop secondary to surgery or trauma; some have been associated with IgG4-related sclerosing disease. Some authors argue in favor of a low-grade neoplastic process. A subset of cases has been seen in association with an infectious agent such as Mycoplasma and Nocardia bacteria. Inflammatory pseudotumor is most often seen in children and young adults, but cases in older patients have been described as well (34,36).
Figure 10. Inflammatory pseudotumor in a 35-year-old man. (a, b) Short-axis balanced steady-state free precession (a) and sagittal black blood half-Fourier acquired single-shot turbo spin-echo (b) MR images show a homogeneous, intermediate- to low-signal-intensity pericardial mass sitting on the anterolateral wall of the left ventricle and encasing, but not obstructing, the left anterior descending artery (arrow). (c) Contrast-enhanced short-axis volumetric interpolated breath-hold examination (VIBE) MR image demonstrates brisk and homogeneous enhancement (*). (d) Axial PET/CT image demonstrates high FDG uptake within the mass. Results of pathologic analysis demonstrated an idiopathic fibroinflammatory process.

At imaging studies, inflammatory pseudotumor mimics a neoplasm. On CT images, these lesions have a variable appearance. Typically, a mass of heterogeneous attenuation and enhancement is seen (34). An associated pericardial effusion may be present (35). On T1- and T2-weighted MR images, inflammatory pseudotumor usually has low signal intensity due to the fibrotic nature of the lesion. After administration of gadolinium contrast material, these lesions may show homogeneous or heterogeneous enhancement. On PET/CT images, these lesions show increased FDG uptake (Fig 10) (34,36).

**Primary Malignant Pericardial Mesothelioma**

Primary malignant pericardial mesothelioma is extremely rare, with a reported prevalence of 0.0022% at autopsy series. It is, however, the most common primary malignancy of the pericardium (37,38). Pericardial mesothelioma occurs more
and similar to those of other pericardial malignancies. Manifestations include constrictive pericarditis, pericardial effusion (Fig 11), tamponade physiology, and myocardial infiltration, which may result in conduction abnormalities (42). Pericardiocentesis commonly reveals hemorrhagic fluid, with malignant cells seen in only 20% of the cases (37).
The sensitivity of echocardiography in the detection of pericardial mesothelioma is low (8). CT and MR imaging offer an advantage, as these modalities better depict the extent of disease. On both CT and MR images, pericardial mesothelioma appears as a heterogeneously enhancing mass that involves both the parietal and visceral layers of the pericardium, with possible invasion of the adjacent vascular and anatomic structures (Figs 11, 12). MR imaging is helpful in providing information regarding the degree of constriction (42) and diastolic impairment (Fig 12). At FDG/PET, these neoplasms demonstrate hypermetabolic activity (Fig 11). Metastatic disease is found in approximately 50% of the cases, most commonly in local mediastinal lymph nodes and the lungs (43).

Resection may be curative in cases of localized disease. Most often, these patients require a pericardiectomy for palliation purposes, as this procedure prevents cardiac tamponade and relieves symptoms from pericardial constriction. Radiation therapy and chemotherapy have a limited role in cases of nonresectable disease (39,42,44).

Primary Pericardial Sarcomas
Primary pericardial sarcomas are uncommon neoplasms with a wide range of histologic appearances. Histopathologic classification is imperative for guiding therapy. Subtypes include angiosarcoma, synovial sarcoma, fibrosarcoma, liposarcoma, rhabdomyosarcoma, and undifferentiated sarcoma among others (45). The prognosis for patients with this group of neoplasms is uniformly poor, with a dismal length of survival ranging between 3 and 12 months (45).

Figure 13. Pericardial angiosarcoma arising about the right atrium in a 54-year-old man. (a) Coronal black blood half-Fourier acquired single-shot turbo spin-echo image demonstrates a heterogeneous lesion with foci of increased intensity, findings consistent with areas of necrosis. (b) Contrast-enhanced horizontal long-axis balanced steady-state free precession image reveals compression and invasion of the right atrium.

Angiosarcoma
Pericardial angiosarcomas are vascular, highly aggressive neoplasms that frequently manifest with associated hemopericardium (46–48). Echocardiography commonly demonstrates a pericardial effusion, but it may fail to show a mass, because the depiction depends on a good acoustic window (47,48). Pericardiocentesis will often yield bloody fluid that may not contain malignant cells (48).

CT reveals the vascular and aggressive nature of angiosarcomas. These lesions are usually lobulated, vegetated masses that heterogeneously enhance at CT with areas of necrosis (7,47). On T1-weighted MR images, the signal intensity of angiosarcoma is heterogeneous and variable, depending on the degree of hemorrhage and necrosis in the tumor; however, angiosarcoma is predominantly isointense relative to myocardium. On T2-weighted and steady-state free precession images, these masses appear heterogeneous and hyperintense relative to myocardium (Fig 13). Angiosarcoma is usually associated with the right atrioventricular groove, parasitizing blood flow from the right coronary artery. Large vascular channels may be seen as intraluminal flow voids (46,49). On contrast-enhanced images, angiosarcomas may show avid enhancement with a “sun ray” appearance (50). Both CT and MR imaging play an important role in depicting the degree of tumor invasion of adjacent structures. MR imaging may demonstrate constrictive physiology with diffuse thickening of
Primary pericardial synovial sarcoma in a 30-year-old man who presented initially with progressive dyspnea and lower-extremity edema. Axial (a) and coronal (b) contrast-enhanced CT images show a heterogeneously enhancing pericardial mass along the right atrium that causes significant compression. Pathologic analysis revealed monophasic synovial sarcoma with chromosomal translocation. Initial transthoracic echocardiography (not shown) demonstrated a large pericardial effusion with tamponade physiology. At pericardiocentesis, 2 liters of hemorrhagic fluid was drained.

At CT, synovial sarcomas appear as heterogeneous soft-tissue attenuation masses. MR imaging demonstrates the high degree of vascularization of these neoplasms, and cross-sectional imaging in general is helpful for depicting their invasive nature. Synovial sarcomas may result in invasion of neighboring structures, infiltrating the epicardium and myocardium, as well as encasing vessels (Fig 14). Metastatic involvement of mediastinal lymph nodes has also been described (Fig 15). Management includes a combination of surgery, radiation therapy, and chemotherapy (51).

Fibrosarcoma

Only a few cases of pericardial fibrosarcoma have been described in the medical literature (56,57). As with other pericardial malignancies, fibrosarcomas often manifest with a large hemorrhagic pericardial effusion. At CT, their imaging appearance is that of an infiltrative mass that may demonstrate central areas of necrosis. On T1- and T2-weighted MR images, they demonstrate heterogeneous signal intensity, reflecting some degree of necrosis and hemorrhage. Heterogeneous enhancement can be seen after the administration of intravenous gadolinium (Fig 16) (57–59).
Figure 15. Primary pericardial synovial sarcoma. Fused PET/CT (a) and maximum intensity projection PET (b) images, obtained in the same patient as in Figure 14 a few months later, show an interval increase in size of the mass. The mass has a central area of necrosis, as demonstrated by the peripheral rim of increased FDG uptake seen at PET/CT. Additional multiple foci of increased uptake are seen in the pericardium and mediastinum, findings consistent with locoregional spread of disease.

Figure 16. Primary pericardial fibrosarcoma in a 14-year-old girl. (a) Transthoracic echocardiogram demonstrates a large, predominantly echogenic, heterogeneous pericardial mass along the free wall of the left ventricle (arrows). (b) Coronal black blood MR image reveals the large, centrally necrotic, intrapericardial mass. (c) Coronal contrast-enhanced CT image shows the large mass along the left aspect of the pericardium. The low attenuation of the mass is in keeping with the extent of necrosis. Pathologic analysis revealed a pericardial fibrosarcoma. (Case courtesy of J. Delgado, MD, IATM, Medellin, Colombia.)
Figure 17. Primary pericardial rhabdomyosarcoma. Axial contrast-enhanced CT images of the chest obtained at the level of the pulmonary arteries (a) and heart (b) demonstrate a heterogeneous pericardial mass that encases the heart and great vessels. The mass invades the epicardial fat as well as the myocardium. Bilateral pleural effusions are also present.

**Liposarcoma**

About a dozen cases of primary pericardial liposarcoma have been described in the literature (60–63). These neoplasms are slow-growing tumors that are often large at presentation. Their large size and invasive nature limit complete surgical excision (61,63). Metastatic disease has been described (62,64). At imaging, pericardial liposarcomas appear similar to liposarcomas that occur elsewhere in the body. On CT images, pericardial liposarcomas appear as predominantly fatty to heterogeneous masses with areas of low-attenuation values. MR imaging also depicts their fatty nature and is useful for assessing the degree of invasion (61).

**Rhabdomyosarcoma**

On rare occasions, a rhabdomyosarcoma may arise from the pericardium (65), and most of these cases have been observed in pediatric patients (66). Rhabdomyosarcoma is a highly heterogeneous tumor (65). On CT images, rhabdomyosarcoma appears as a large, solid, infiltrative enhancing mass that can be homogeneous or heterogeneous, depending on the degree of necrosis and cavitation (Fig 17) (10). Foci of calcification may be seen (66). At MR imaging, they appear predominantly isointense with T1-weighted sequences and hyperintense with T2-weighted sequences, with homogeneous to heterogeneous enhancement after the administration of gadolinium (10).

**Undifferentiated Sarcoma**

Undifferentiated sarcomas are malignant neoplasms without specific histologic features (67). Patients usually present with symptoms related to a large pericardial effusion (67–69). On CT images, an undifferentiated sarcoma may appear as a discrete, infiltrative enhancing mass, which can infiltrate the myocardium and invade adjacent vascular structures. On T1- and T2-weighted MR images, they usually appear isointense relative to myocardium and demonstrate a heterogeneous enhancement pattern (58). Treatment includes a combination of surgery, chemotherapy, and radiation therapy (67–69).

**Pericardial Lymphoma**

Most primary pericardial lymphomas are diffuse, large B-cell lymphomas (70). Pericardial lymphoma most frequently manifests as an ill-defined, infiltrative soft-tissue mass with an associated pericardial effusion.

On CT images, the soft-tissue component of pericardial lymphoma may appear iso- to hypoattenuating with heterogeneous enhancement (Fig 18). Pericardial lymphoma is hypermetabolic at PET/CT (Fig 19). At MR imaging, it appears hypointense with T1-weighted sequences and iso- to hyperintense with T2-weighted sequences with variable heterogeneous enhancement. On occasion, a discrete mass is not seen, and the sole imaging finding may be diffuse pericardial thickening or effusion, which is commonly hemorrhagic (7,58,71,72). Hemorrhagic pericardial effusion appears as hyperattenuation on CT images. On T1-weighted MR
images, hemorrhagic fluid demonstrates high signal intensity; it has intermediate signal intensity on T2-weighted images. Strands of fibrin and coagulated blood, with or without locula-

Figure 18. Primary pericardial lymphoma. Contrast-enhanced CT image of the chest demonstrates a lobulated, heterogeneously enhancing soft-tissue mass, predominantly along the anterior aspect of the pericardium. Note the obliteration of the epicardial fat (arrow) in keeping with epicardial invasion.

Figure 19. Primary pericardial lymphoma. (a) Contrast-enhanced CT image demonstrates a homogeneously enhancing mass along the anterior aspect of the pericardium that extends into the right atrioventricular groove and encases the right coronary artery (arrow). (b) PET/CT image at the same level shows homogeneous FDG uptake throughout the mass. (c) PET/CT image obtained 1 year later, after chemotherapy, demonstrates complete resolution of the hypermetabolic mass. 

ations, can be seen on steady-state free precession images (73). In patients with human immunodeficiency viral infection or acquired immunodeficiency syndrome (AIDS), a large pericardial effusion should raise suspicion for primary effusion lymphoma. These are human herpesvirus-8–associated large B-cell lymphomas that account for approximately 4% of AIDS-related lymphomas (74).
Primary Pericardial Primitive Neuroectodermal Tumor

Primary pericardial primitive neuroectodermal neoplasms arise from cells of the primitive neuroectoderm. The extraosseous variety characteristically occurs in young adults (75). The majority of cases demonstrate the chromosomal translocation t(11;22)(q24;q12) (76). To our knowledge, until now, only three cases of primary cardiac primitive neuroectodermal tumors have been reported in the English literature, of which only one arose from the pericardium (75).

On CT images, primary pericardial primitive neuroectodermal tumors are heterogeneous in appearance with foci of necrosis. They generally demonstrate aggressive features, with invasion of the myocardium and extension into adjacent mediastinal and vascular structures (Fig 20). These tumors are generally iso- to hypointense on T1-weighted MR images, are intermediate to hyperintense on T2-weighted MR images, and demonstrate homogeneous to heterogeneous enhancement after the administration of intravenous gadolinium.

Summary

Primary pericardial neoplasms are rare but relevant tumors, because even when they are benign, these neoplasms may result in significant cardiovascular complications because of their anatomic location. The most common benign lesion is a pericardial cyst. Patients with malignant pericardial neoplasms have a dismal prognosis, which is partially because they present at a late stage in their disease, often after invasion of vital mediastinal or cardiac structures has occurred. The most common malignant primary pericardial neoplasm is mesothelioma.

The radiologist can play a key role in the care of patients with pericardial neoplasms. CT and MR imaging are fundamental in the evaluation of these neoplasms and may, in some occasions, allow full characterization of their features. Most important, the role of the radiologist centers in aiding the clinical team to guide therapy. Attention should be directed at the location of these lesions and the extent of disease, including invasion of adjacent structures, involvement of the coronary arteries, compression of cardiac chambers, pericardial effusion, constrictive physiology, and locoregional or distant metastatic disease.


References


Patients with primary pericardial neoplasms present with diverse symptoms. Clinical signs and symptoms are usually the result of associated pericardial effusion, pericarditis, or invasion of adjacent structures and are therefore nonspecific. Symptoms include exertional dyspnea, chest pain, cough, palpitations, fatigue, night sweats, fever, and facial or lower-extremity edema. Occasionally, a primary pericardial tumor may be found incidentally in an asymptomatic patient, during work-up for an unrelated illness.

Some of the commonly seen complications include invasion of mediastinal structures, regional or distant metastases, pericardial effusion, cardiac tamponade, compression of vascular structures or cardiac chambers, encasement of vital structures, involvement of coronary arteries, myocardial infarction, diastolic dysfunction, and constrictive physiology.

Typically, pericardial cysts appear on CT images as well-defined, nonenhancing, homogeneous fluid-attenuation lesions that contain no internal septa. Occasionally, the cyst may contain proteinaceous fluid, in which case the lesion will demonstrate intermediate attenuation at CT. Pericardial cysts with hemorrhagic contents appear with hyperattenuation at CT. MR imaging offers further characterization of the internal contents of the cysts. Usually, pericardial cysts demonstrate intermediate to low signal intensity on T1-weighted images and high signal intensity on T2-weighted images.

Lipomas appear as nonenhancing lesions that are hyperintense on T1-weighted MR images and intermediate to high signal intensity on T2-weighted MR images. Use of fat-saturated sequences to confirm the fatty nature of the lesion is helpful, as these sequences will result in a uniform drop in signal.

Primary malignant pericardial mesothelioma is extremely rare, with a reported prevalence of 0.0022% at autopsy series. It is, however, the most common primary malignancy of the pericardium.