Neoplastic Pericardial Disease

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KEYWORDS

• Tumors • Masses • Cardiac • Epicardium • Thoracic

KEY POINTS

- Pericardial neoplasms are rare and may be primary or secondary, the latter much more common.
- Finding a pericardial mass should always prompt a careful history and may warrant further imaging and/or sampling.
- Imaging plays a critical role in characterizing pericardial masses and can help to narrow the differential diagnosis.

Pericardial tumors include a range of both neoplastic and non-neoplastic entities. Neoplasms of the pericardium can be broadly dichotomized into those that arise within the pericardium (primary pericardial neoplasms) and those that metastasize to involve the pericardium (secondary pericardial neoplasms). Although secondary pericardial neoplasms are, by definition, malignant processes, primary pericardial neoplasms may be either benign or malignant.

In general, involvement of the heart or pericardium by metastatic disease is substantially more common than involvement by a primary neoplastic process. It is estimated that metastatic lesions are greater than 100 times more common, and the pericardium is the most common site of said involvement.[1] In high-stage cancer patients, cardiac involvement has been demonstrated in as many as 14% of cases.[2] Primary pericardial processes, on the other hand, have been estimated to account for between 6% and 10% of primary cardiac tumors but are typically benign.[3]

Involvement of the pericardium by malignant processes can affect cardiac function several important ways, such as pericardial effusion (with or without tamponade physiology), direct myocardial invasion, pericardial constriction, or pericarditis.[4] In general, the most common of these is effusion. Even benign tumors can be large enough to incite hemodynamic compromise by virtue of their size, location, or number.[5–8]

Pericardial tumors may present with a wide range of clinical presentations, ranging from asymptomatic, incidental diagnosis to sudden death. Patients with significant tumor burden in the pericardium may present with dyspnea, orthopnea, pain, edema, or even hemoptysis.[9] Clinical examination can reveal cyanosis, venous distention, hepatomegaly, pleural or pericardial effusion (distant heart sounds or rub), murmur, or electrocardiographic changes.

This review provides an overview of the spectrum of neoplastic pericardial disease and discusses the defining pathologic characteristics as well as the expected findings at imaging. Only brief mention of non-neoplastic entities are made, primarily to round out discussion of the differential diagnosis of pericardial mass-lesions.

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IMAGING

Pericardial neoplasms may be found incidentally at imaging, surgery, or autopsy or they may be discovered during work-up for symptoms that are considered to have a cardiac etiology. Once discovered, imaging is often integral to the diagnostic work-up. Although plain film radiography and echocardiography are considered the cornerstone of cardiovascular imaging, CT and MRI should be considered frontline in the assessment of pericardial anatomy. Echocardiography may disclose the presence of increased pericardial thickness, nodularity, or discrete mass as well as secondary hemodynamic effects of the lesion, including presence of cardiac tamponade. Beyond these features, the role of echocardiography in the diagnostic evaluation of pericardial tumors remains limited.

CT and MRI have the clear advantage of in their ability to delineate the exact location of the neoplasm, its effects on neighboring cardiac and noncardiac structures; particularly in the setting of CT imaging, the ability to stage disease that can assist in therapeutic planning and surveillance. Despite the advances in cardiovascular imaging, it is important to recognize that in most cases, the evaluation of a primary pericardial tumor requires tissue sampling and remains a pathologic diagnosis.

PERICARDIAL METASTASES

Pericardial metastases are, as discussed previously, the most common cause of pericardial mass-lesions and any findings of a pericardial mass should, therefore, prompt a careful evaluation for underlying malignancy. They may involve the visceral and/or parietal layers. In a series of 60 cases of pericardial metastasis, approximately half of cases had significant cardiovascular compromise as a result.[9] Mechanistically, this is often the result of effusion, but myocardial invasion and tumor encasement were also involved (**Fig. 1**).[10]

The most common malignancies diagnosed, either at biopsy or cytologic sampling, are carcinomas arising from the lung and breast (the most common primary site of origin in women).[11] Involvement of the pericardium is also not uncommon in cases of leukemia/lymphoma. Metastasis from gastrointestinal, genitourinary, and gynecologic malignancies may also occur as well as from extracardiac sarcomas.

Metastatic involvement of the pericardium has nonspecific imaging findings and is often suggested by the diagnosis of the primary noncardiac neoplasm. Imaging findings include presence of effusion, irregular thickening of the pericardium or nodularity, and distinct pericardial masses (see **Fig. 1**).[12] On MRI, most secondary neoplasms to the pericardium have low signal intensity on T1-weighted imaging with the exception of metastatic melanoma, which may have high signal intensity secondary to paramagnetic metals bound by melanin.[13,14] Postcontrast delayed imaging typically discloses pericardial enhancement in the regions of metastases.[12]

Pericardial metastases are often diagnosed in cytologic specimens, rather than biopsy (tissue) specimens.[15] The primary differential, both cytologically and histologically, is between reactive mesothelial hyperplasia and carcinoma — which can have some morphologic overlap. Therefore, it is common to use immunohistochemical studies to help characterize the antigenicity and nature of the cells. If proved a carcinoma, immunohistochemistry

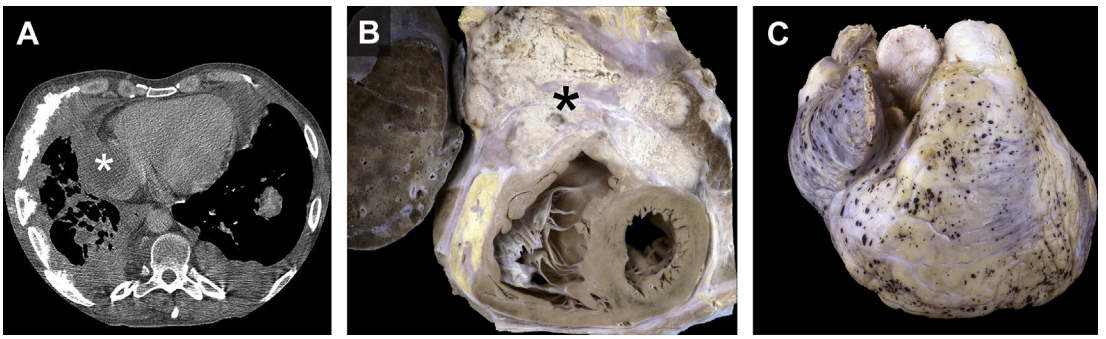

Fig. 1. Pericardial metastases. (*A*) CT scan performed with intravenous iodinated contrast (poor contrast opacification), demonstrating a large pericardial mass (*white asterisk*) with compression of the right atrium with pleural and pulmonary metastases and osteoblastic metastases involving multiple right-sided ribs. The primary tumor was rectal carcinoma. (*B*) Autopsy-derived specimen of a pulmonary adenocarcinoma (*black asterisk*) with direct extension to involve the anterior pericardium (visceral and parietal layers) and the subjacent myocardium. (*C*) Autopsy-derived specimen showing numerous visceral pericardial (epicardial) metastases by metastatic malignant melanoma.