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CT and MRI of pericardial and cardiac neoplastic disease

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Abstract

This article reviews the pathological classification of cardiac and pericardial neoplasms, the incidence of the various tumor types, and the role of CT and MRI, including their major differences and clinical impact on patient management.

Keywords: *Cardiac; neoplasm; computed tomography; magnetic resonance imaging; diagnosis.*

Introduction

Cardiac and pericardial neoplasms are relatively rare, affecting 1–3 per 10,000 patients in an autopsy series[1,2]. A review of 533 cases of primary heart tumors revealed that 60% were benign, 17% were pericardial and bronchogenic cysts and 23% were malignant primary tumors[1]. An autopsy study in a total of 343 patients showed the frequency distribution of the most common benign and malignant primary cardiac tumors in both adults and children (Table 1)[2]. Although the majority of these tumors are histologically benign, the clinical course is often complicated due to cardiac arrhythmias and compromise of ventricular function due to mass effect or tamponade[3]. Hence, histologically benign tumors can result in fatal outcomes if not adequately diagnosed and treated[1,2]. A relatively large review of primary cardiac neoplasms demonstrated approximately 10% mortality in myxomas, 30% mortality in non-myxoma benign tumors, and 100% mortality within 3 years in all patients with malignant tumors[4]. Thus, pre-surgical diagnosis should focus on determining whether a tumor is benign or malignant, and more specifically whether a tumor is a myxoma or not.

Echocardiography is usually the first diagnostic test in patients with tumor-related cardiac symptoms, such as tamponade, heart failure or systemic emboli. However, the introduction of multi-detector row (ECG-gated) computed tomography (CT) and innovations in magnetic resonance imaging (MRI) have expanded the diagnostic capabilities of imaging beyond that of echocardiography. These modalities have improved our ability to diagnose and delineate the tumors themselves, assess their impact on cardiac function and plan for surgical intervention, which has become increasingly sophisticated and feasible over the past three decades.

More recently, modalities such as combined positron emission tomography (PET) and CT are being applied to assess tumors of the heart and pericardium[5]. In the present review, the focus is on demonstrating the use of CT and MRI with relevant patient examples.

Magnetic resonance imaging

A study of 216 consecutive patients referred for cardiac MRI revealed tumors in 55, with 33 lesions histologically confirmed benign and 22 lesions malignant[6]. Benign tumors could be predicted based on location (tumors in the left chambers were benign in 19/33 and malignant in 3/10) and based on tissue composition (50% of benign tumors were homogeneous vs 15% of malignant tumors), whereas signal intensity and contrast enhancement were not different between the two groups. Other important predictors of malignancy included signs of myocardial infiltration, mass effect, pericardial and pleural effusions[6].

Two reviews on the use of MRI (and CT) for the diagnosis of benign and malignant primary cardiac tumors demonstrate the capability of this modality for determining tumor extent, differentiation, and common

Type of tumor	Total (%)	Pediatric (%)
Benign[a]		
Myxoma	57	10
Rhabdomyoma	10	35
Fibroma	10	25
Hemangioma	8	4
AV nodal tumor	5	4
Malignant[b]		
Sarcoma	95	100
Lymphoma	5	0

[a]Based on a total of 199 heart tumors, of which 45 were in children 15 years or younger.
[b]Based on a total of 144 heart tumors, 11 of which were in children 15 years or younger.

pitfalls[7,8]. The finding that most strongly suggests a diagnosis of myxoma on MRI is the presence of a narrow base of attachment to the interatrial septum[8]. Most malignant tumors demonstrate inhomogeneous enhancement with gadolinium, infiltration through tissue layers and frequent pericardial effusions[7].

MRI has proven extremely useful for assessment of pericardial disease, as it gives excellent tissue contrast and allows for clear visualization of the entire pericardial cavity[9,10].

Computed tomography

The clinical utility of computed tomography for cardiac imaging increased dramatically with the introduction of ultrafast electron beam CT, which enabled high-resolution imaging of the heart and its chambers as well as cinematic imaging for assessment of cardiac function and even tumor motion[11]. However, the recent developments with ECG-gated multidetector row CT and dual source CT systems have led to another quantum leap in performance, enabling widely available clinical CT scanners to evaluate the heart with incredible detail[12,13].

Few large studies have systematically explored the imaging findings of cardiac and pericardial neoplasms using multidetector row or electron beam CT. This is probably related to both the small number of cases seen in any one institution and the short time between introduction of novel CT technology and writing this review.

In order to give the reader some insight into the imaging findings of the most common cardiac and pericardiac neoplasms, we will proceed with an overview of the most salient findings at CT and MR.

Benign cardiac neoplasms

Myxoma

This is the most common benign cardiac neoplasms (Fig. 1), accounting for 50–60% of benign tumors as described in larger series[1–4]. Symptoms range from asymptomatic coincidental detection in approximately 12% of subjects, to systemic embolic complications, dyspnea, palpitations and non-specific symptoms such as fatigue, weight loss or fever[3,14]. Among patients with myxoma, 10% are inherited in autosomal dominant fashion[15]. These patients tend to present at a younger age and have less female predominance compared to sporadic cases. Finally, familial myxoma more commonly occurs in the ventricular cavity and with multiple cardiac tumors in multiple locations.

In the AFIP series of 83 patients, 81 myxomas were fully assessable: 47 were in the left atrium, 23 in the right atrium, 7 in right ventricle, 2 biatrial and 2 in multiple sites[14]. A review of 19 CT studies was performed and correlated with pathological findings. Of 21 myxomas visualized using CT, 20 were spherical or ovoid in shape, 16 (76%) were lobulated and 5 (24%) were smooth. Contrast-enhanced CT demonstrated lower attenuation than myocardium in 17 (81%) and equal attenuation in
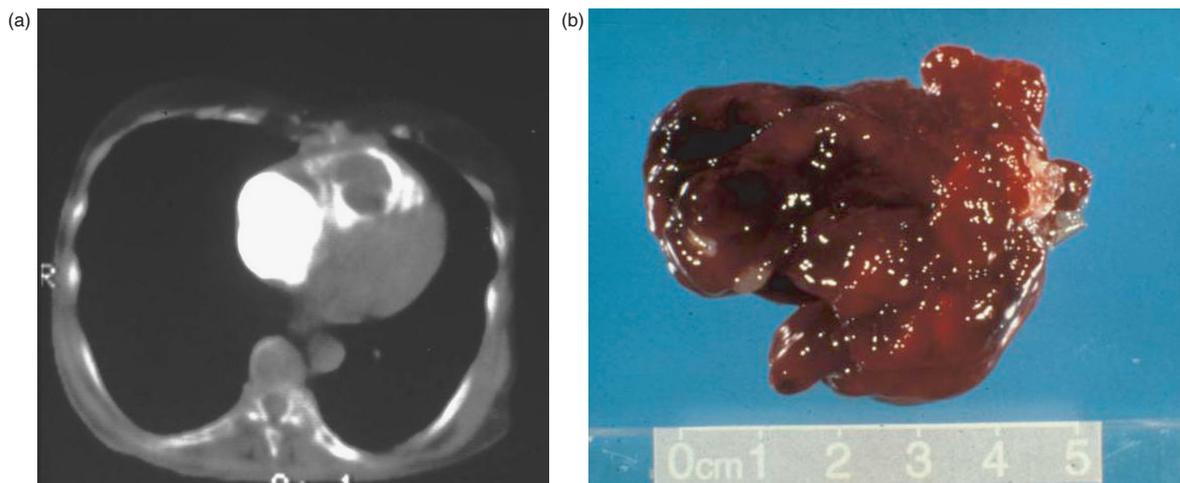
Figure 1 **Patient with right ventricular lobulated hypo-attenuating mass at electron beam CT (a) and the corresponding macroscopic specimen (b) demonstrating myxoma.**

the remaining four; none of the lesions showed hyper-enhancement. Heterogenous enhancement was seen twice as often (67%) and calcification was infrequent (3/21; 14%).

The same study also evaluated 30 myxomas with MR[14]. All but one were spherical or ovoid in shape; 26 (87%) had lobular contours. The vast majority (90%) showed heterogeneous signal intensity and 23 (79%) were hypo-intense compared to myocardium on T1-weighted sequences. Only six of the MR studies included gadolinium-enhanced sequences of which four myxomas showed heterogenous tumor enhancement[14]. Cinematic studies were performed in 10 myxomas, revealing a tumor prolapsing across the atrioventricular valve in 40%.

Rhabdomyoma

This is by far the most common benign pediatric cardiac neoplasm, accounting for 90% of benign primary tumors[2,16,17]. These lesions usually present within the first year of life, and up to 50% of patients have tuberous sclerosis[18,19]. Most cardiac rhabdomyomas regress spontaneously, and surgical excision is only required in patients with life-threatening symptoms secondary to left ventricular outflow tract obstruction, arrhythmias or transient hypoxic spells[15,17,20,21].

MR imaging allows for better definition of tumor margins and can evaluate the multiplicity of these lesions, which are iso-intense to myocardium on T1-weighted images and hyper-intense on T2-weighted images[8,19]. CT currently has little or no role to play in the management of patients with these lesions.

Fibroma

Fibroma typically affects children and infants, but can present at a later age in 15% of cases[22]. There is an association with basal cell nevus (Gorlin) syndrome, with a 10–15% prevalence of cardiac fibroma[2].

Clinically, patients present with heart failure, arrhythmias and sudden death (in up to 30% of patients), due to a combination of ventricular failure and invasion and/or compression of the conduction system[8,15,19].

Most fibromas are located within the ventricular wall, producing compression or obstruction of the ventricular lumen (Fig. 2). Tumor calcification may be present.

Computed tomography is able to delineate the heterogenous mural mass and calcifications, and 3D evaluation is feasible using multidetector row CT (Fig. 2a). Intravenous contrast enhancement is unpredictable; small pericardial effusions may be present (see above)[22].

MR imaging will demonstrate the mural mass, which appears iso- or hyperintense on T1-weighted and hypo-intense of T2-weighted sequences in comparison to myocardium and demonstrates variable enhancement with gadolinium (Fig. 2b and c)[8,19].

Hemangioma

These tumors may occur at any age and produce a characteristic tumor blush on arterial phase imaging, either using coronary angiography, CT or MRI[8,15,19]. They may occur as part of the Kasabach–Merritt syndrome[23], but are more often incidental findings in asymptomatic patients[1,2].

Lipoma

These lesions are very rare, but can become symptomatic due to their size, their location on valves (both leading to obstructive symptoms) or due to conduction defects and resulting arrhythmias[8,12,15,19,24].

Lipomatous tissue is easily distinguished by CT as hypodense tissue with characteristic attenuation of −20 to 50 HU, while MRI will demonstrate high signal intensity on T1 and T2-weighted imaging, with complete signal drop-out using fat-saturation imaging. Lipomas tend to appear as well-defined masses and should not be confused with interatrial septal lipomatous hypertrophy as demonstrated in Fig. 3[25].

Malignant cardiac neoplasms

Sarcoma

Although these tumors are exceedingly rare in children, they represent the only primary cardiac malignancy in this age group. Typically, sarcomas are seen in adults, presenting around the 4th decade. The majority of these tumors are located in the right heart chambers, leading to right heart symptoms including cardiac tamponade with direct tumor extension into the pericardial sac.

Angiosarcoma constitutes more than 1/3 of all cardiac sarcomas and has a predilection for the right atrium[26,27]. Clinically, males are affected twice as often as females. These tumors tend to cause flow obstruction and heart failure. Hemorrhagic pericardial tamponade indicates tumor infiltration through the myocardium[15,26,27].

The clinical course of angiosarcoma is rapid, as demonstrated in two chest radiographs 6 weeks apart (Fig. 4a and b). Rapid tumor growth, the mechanical effects of tumor mass on heart function, tumor invasion through tissue planes into the pericardium, and pulmonary metastases usually lead to a rapidly fatal course.

CT imaging demonstrates a large right atrial mass, often with invasion into the pericardial sac and (hemorrhagic) pericardial effusion (Fig. 4c and d). MR imaging will demonstrate the extent of the mass as well as significant (often heterogeneous) enhancement following gadolinium due to the high vascularity of this tumor type (Fig. 4e and f).

Rhabdomyosarcoma is the second most frequent primary sarcoma of the heart, also with a male predominance. Unlike angiosarcoma, this tumor type
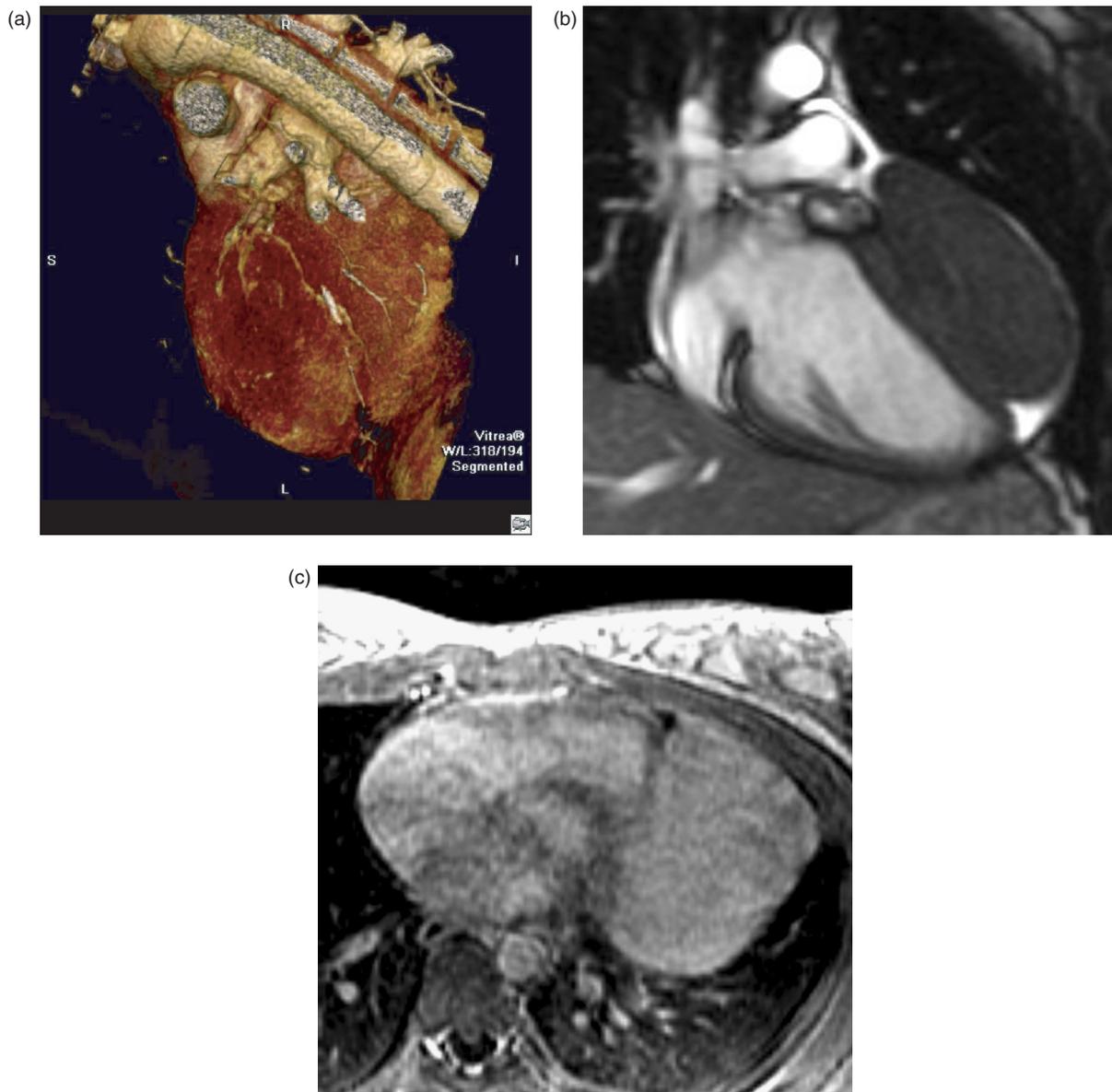
Figure 2 **Left ventricular fibroma on 3D reconstructed CT (a) and MRI. Frame from a coronal T2-weighted cine TrueFISP MR sequence (b) shows a large epicardial mass which is isointense to myocardium and expands into the pericardial space. Note the small pericardial effusion. Mass shows modest contrast enhancement after IV gadolinium on axial fat suppressed T1-weighted image (c).**

does not have an obvious predilection for any one cardiac chamber, and is seen slightly more frequently in the left atrium[2,15,27].

Several other types of sarcomas have been identified, including undifferentiated sarcoma, malignant fibrous histiocytoma, leiomyosarcoma and osteosarcoma. Some distinguishing features include a slight female predominance in malignant fibrous histiocytoma and the earlier presentation of leiomyosarcoma (in the 3rd decade)[7,19,28,29], but ultimately tissue sampling is required to demonstrate the true histology of these tumors.

MR is the method of choice for imaging of sarcomas, as it provides tissue characterization as well as local extent of the tumor. Furthermore, MRI produces information on tumor-induced compromise of cardiac function.

CT is a good secondary modality, particularly to assess for metastatic spread of tumors (mainly pulmonary metastases), but also for the demonstration of great vessel, myocardial, pericardial and mediastinal invasion.

Lymphoma

Primary cardiac lymphoma is very rare, and only occurs in adults. Histologically, these lymphomas are of the non-Hodgkin type. At diagnosis, they only involve the heart and pericardium. This lymphoma variety occurs more
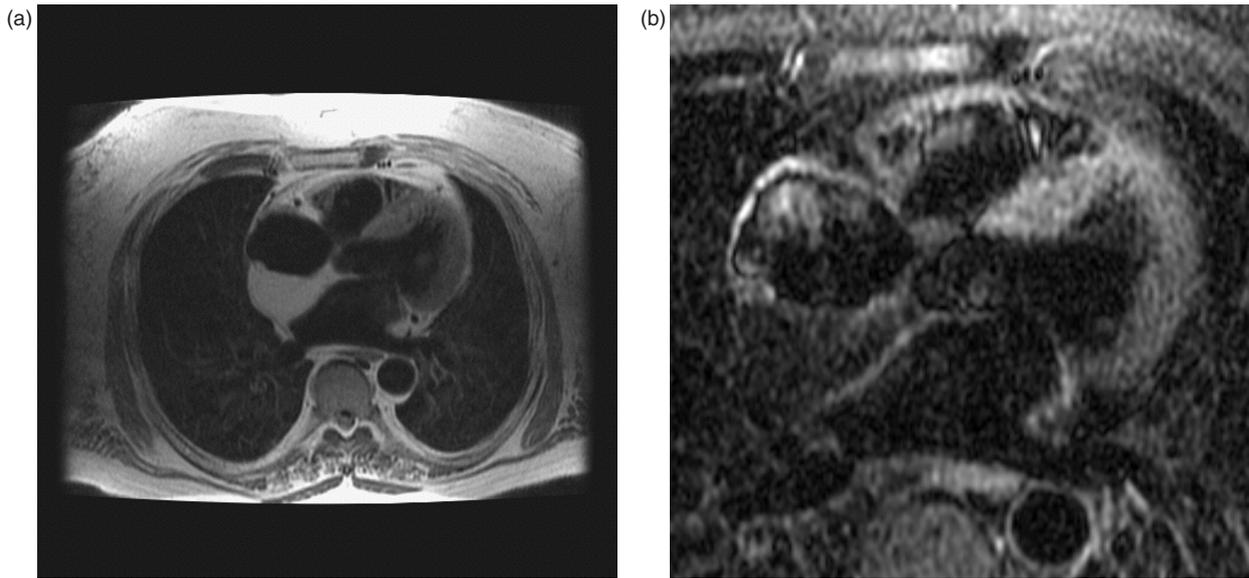
Figure 3 **Lipomatous hypertrophy of interatrial septum. Axial T1-weighted double inversion recovery MR image (a) shows marked thickening and high signal intensity of the interatrial septum. The thickened septum shows a signal void after inversion recovery fat suppression is applied (*B*).**

often in immunocompromised patients, and in the 6th decade of life[2,8,19,30]. Patients present with heart failure, signs of vena cava obstruction and chest pain, and the prognosis is invariably poor[30].

In the vast majority of patients, multiple cardiac chambers are involved. Invasion into the pericardium is common with some lymphomas and is associated with coronary artery invasion and myocardial infarction (Fig. 5).

CT demonstrates low-attenuation lesions with heterogeneous contrast enhancement, usually in association with a pericardial effusion[31].

On MR imaging, these lesions appear poorly delineated and heterogenous with low to iso-intense signal on T1-weighted images and iso-intense signal on T2-weighted images in comparison to myocardium. Gadolinium enhancement is heterogeneous. Inversion recovery imaging will demonstrate a pathological high signal in the pericardial fluid to indicate malignant effusion (Fig. 5).

Malignant secondary cardiac tumors

Metastatic disease to the heart is much more common than primary cardiac malignancy[2,15]. Cancers most likely to metastasize to the heart are described in Table 2. Melanoma is still a relatively rare tumor, although the incidence has been rising rapidly over the past decade. In 30–50% of melanoma patients, cardiac metastases will be found at autopsy. In absolute numbers, the most common cardiac metastases are caused by breast cancer and lung cancer (Fig. 6). The latter gives rise to both direct pericardial invasion and metastases to the left cardiac chambers. Other cancers that frequently involve

the heart include leukemia, lymphoma and esophageal cancer.

Pericardial neoplasms

Although pericardial neoplastic disease remains relatively rare, they are more frequently identified than cardiac neoplasms. The initial clinical evaluation should focus on making the distinction between neoplastic and non-neoplastic causes for pericardial filling defects (such as cysts, hematomas or loculated effusions). Echocardiography is often the first imaging method, but both CT and MRI are capable of yielding important information for clinical management[9,10].

Benign pericardial neoplasms

The most common benign tumors include lipoma, teratoma, fibroma and hemangioma[10]. Most of these lesions exhibit fairly typical features, including well-defined margins. Lipomas demonstrate the characteristic low attenuation on CT (−20 to 50 HU), while MRI will demonstrate high signal intensity on T1- and T2-weighted images with signal void of short tau inversion recovery or frequency selective fat-saturation sequences. On CT, teratomas of the pericardium exhibit the characteristic features of bone, soft tissue, fat and calcification. Fibromas appear as low intensity T2-weighted MR images with little or no enhancement. Hemangiomas, however, demonstrate an intense blush upon CT or MR contrast enhanced imaging.
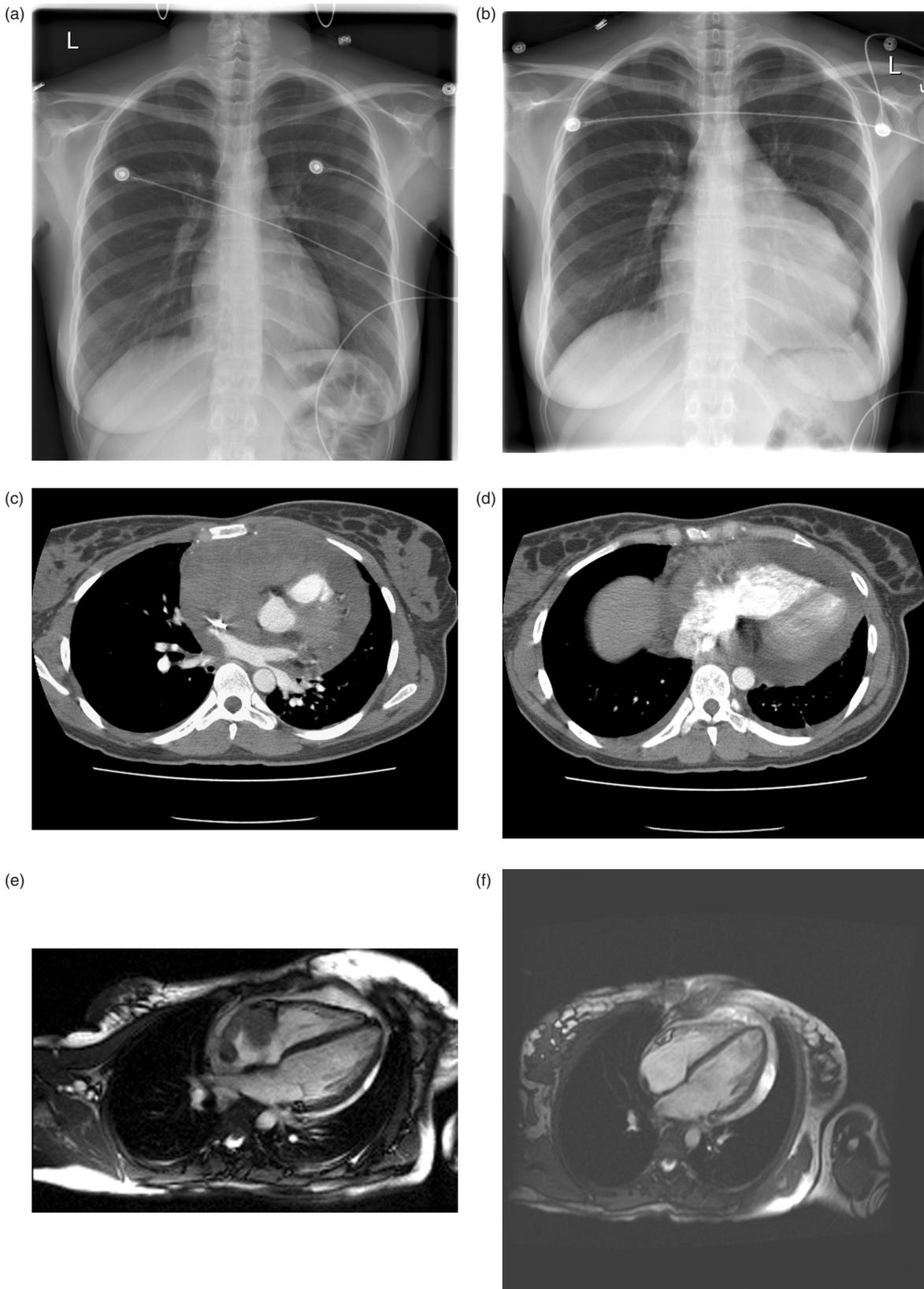
Figure 4   **Angiosarcoma showing rapid growth on two subsequent chest radiographs 2 months apart (a and b), tumor extent and invasion on CT (c and d) and MRI (e and f). Frame from a short axis T2-weighted cine TrueFISP MR sequence (e) shows a mass of low–intermediate signal intensity in the anterior aspect of the right atrium. Post-gadolinium axial fat suppressed T1-weighted image (f) shows nodular pericardial enhancement indicative of a malignant pericardial effusion.**

(a)

(b)
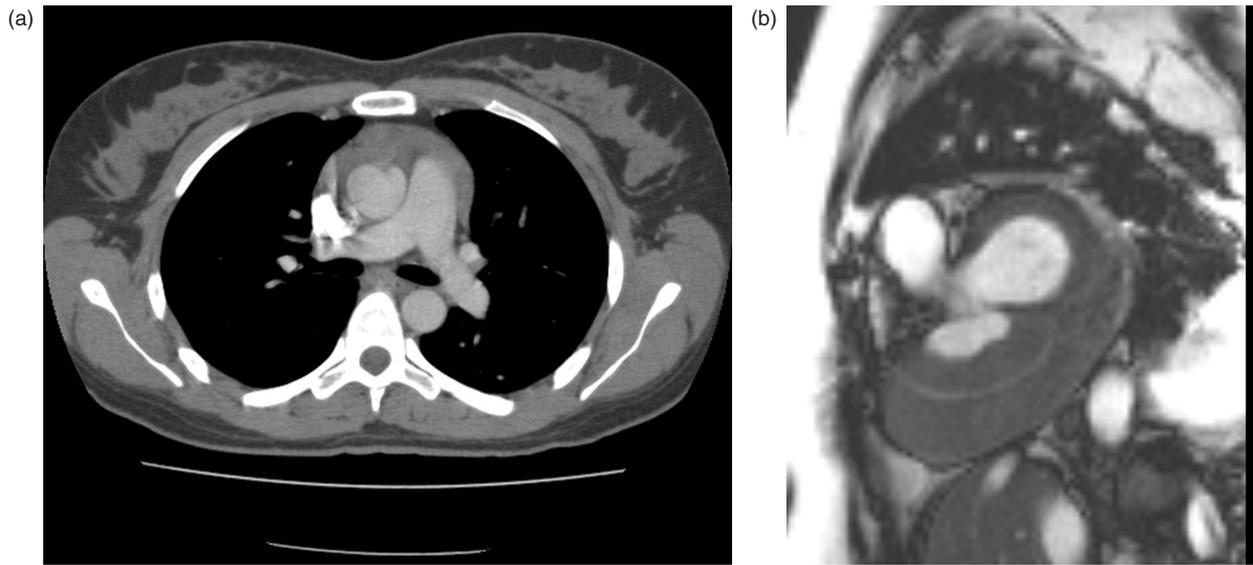
Figure 5 **Invasive pericardial lymphoma. CT demonstrates a soft tissue mass in the anterior pericardial space filling in the space of right coronary artery origin (a). Frame from a short axis T2-weighted cine TrueFISP MR sequence (b) shows a soft tissue mass of low–intermediate signal intensity filling the atrioventricular groove and encasing the right coronary artery.**

Table 2   Eight most common tumor types which metastasize to the heart (modified from Roberts[15]

Type	% with heart metastases	% of total heart metastases
Melanoma	46	1
Germ cell tumor	38	0.3
Leukemia	33	3.2
Lymphoma	17	6.2
Lung cancer	17	17
Sarcoma	15	2.5
Esophagus	13	4.7
Kidney	11	1.8
Breast	10	11

Figure 6 **Patient with proven lung cancer, with CT demonstrating direct invasion through the pericardium into the left atrium.**

Malignant pericardial neoplasms

Pericardial metastases are frequently encountered at autopsy[32]. Most commonly, they are related to breast cancer and lung cancer, either through direct extension (Fig. 6) or via lymphangitic or hematogenous spread[33].

Malignant mesothelioma will present with pericardial effusion and nodular thickening of the pericardium. More often, it is related to pleural mesothelioma, which has a propensity to invade into the pericardial sac.

Concluding remarks

Pericardial neoplastic disease is much more frequently encountered than cardiac neoplasms. Most often, pericardial neoplasms arise from hematogenous or lymphatic metastatic spread or direct invasion of a non-pericardial primary malignancy, with lung cancer, breast cancer and melanoma most frequently encountered.

The diagnosis is often made initially through echocardiography, but both CT and MRI have additional value in demonstrating the extent of the tumor and likely etiology. With future developments, it is quite likely that combined modality imaging, such as PET-CT or PET-MRI will gain greater applicability in the work-up of (peri-) cardiac neoplasms.

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