#### **REVIEW ARTICLE**



# Diagnosis and treatment of cardiac tumors

Toru Imai<sup>1</sup> · Tatsunori Shimoi<sup>1</sup> · Akira Kawai<sup>2</sup> · Kan Yonemori<sup>1</sup>

Received: 14 November 2024 / Accepted: 4 March 2025 / Published online: 17 March 2025 © The Author(s) 2025

#### **Abstract**

Cardiac tumors, though rare, present significant diagnostic and therapeutic challenges due to their diverse nature and potential severity. These tumors, which can be primary or metastatic, are often detected incidentally through imaging modalities such as echocardiography or CT scans. Differentiating between benign and malignant forms is crucial for guiding appropriate management strategies. This review synthesizes current diagnostic approaches and treatment modalities for cardiac tumors, with a focus on the role of imaging techniques like UCG, CT, MRI, and PET in tumor characterization. Multidisciplinary treatment plans are necessary, including surgical resection for benign tumors, chemotherapy, and radiotherapy for malignant tumors, and novel targeted therapies such as MDM2 inhibitors for selected cases. While primary malignant tumors like sarcomas and mesotheliomas exhibit rapid progression and poor prognosis, recent advances in multimodal therapy offer potential improvements in survival. The incidence of primary cardiac tumors is low, with an autopsy-reported occurrence rate of 0.02%. Benign cardiac tumors, such as myxomas and fibromas, generally have favorable outcomes with surgical resection. In contrast, primary malignant tumors like sarcomas and mesotheliomas exhibit rapid progression and poor prognosis, necessitating aggressive treatment including surgery, chemotherapy, and radiotherapy. Metastatic cardiac tumors occur in approximately 10% of cancer patients at autopsy and are managed according to the treatment plan for the primary malignancy. The management of cardiac tumors requires a multidisciplinary approach tailored to tumor type, location, and systemic effects. While benign tumors often respond well to surgical management, malignant and metastatic tumors demand more complex strategies to optimize patient outcomes.

Keywords Primary cardiac tumors · Malignant soft tissue sarcoma · Metastatic cardiac tumors · Benign tumors

### **Abbreviations**

UCG Ultrasound cardiography
CT Computed tomography
MRI Magnetic resonance imaging
PET Positron emission tomography
PCTs Primary cardiac tumors
PFE Papillary fibroelastoma

# **Background**

Cardiac tumors, though rare as primary tumors, can manifest in both benign and malignant forms. Cardiac masses are frequently discovered incidentally during echocardiographic or CT scans. These masses are categorized into tumor and non-tumor lesions, with tumors further subdivided into primary and secondary (metastatic) tumors. When echocardiography detects an abnormal cardiac structure, it is essential to distinguish it from benign embryonic remnants and congenital anomalies. Subsequent differentiation involves ruling out degeneration, thrombosis, or papilloma, although primary cardiac tumors are exceptionally rare. According to data from 22 large autopsy series, the incidence of primary cardiac tumors is approximately 0.02%, equating to around 200 tumors per 1 million autopsies [1, 2]. Metastatic cardiac tumors, in contrast, are not uncommon, with autopsy studies in cancer patients revealing a prevalence of 10.7% [3]. Malignancies like pleural mesothelioma and malignant melanoma have a high propensity for cardiac metastasis, while



<sup>☐</sup> Tatsunori Shimoi tshimoi@ncc.go.jp

Department of Medical Oncology, National Cancer Center Hospital, Tsukiji 5-1-1, Chuo-ku, Tokyo 104-0045, Japan

Department of Musculoskeletal Oncology and Rehabilitation, National Cancer Center Hospital, Tsukiji 5-1-1, Chuo-ku, Tokyo 104-0045, Japan

metastases from lung and breast cancer are also frequently observed [4]. While benign cardiac tumors generally have a favorable prognosis, primary malignant cardiac tumors are associated with poor outcomes, with a 1-year survival rate of 44.3% and a 5-year survival rate of only 16.6% [5].

These tumors often present significant diagnostic challenges and are associated with severe clinical symptoms, necessitating a comprehensive understanding of their diagnosis and treatment. Effective management requires a collaborative, multidisciplinary approach. This paper provides a detailed overview of the epidemiology, diagnosis, and treatment of cardiac tumors, with a particular focus on the primary benign and malignant types.

# **Diagnosis**

Accurate diagnosis of cardiac tumors relies heavily on clinical information. Although biopsy is the gold standard for definitive diagnosis, its feasibility is often limited by case complexity or institutional constraints. Thus, imaging and clinical data are critical for differential diagnosis. Surgery often serves both therapeutic and diagnostic roles, and in some cases, a diagnosis can be inferred from non-invasive methods before being confirmed surgically. The following four key factors are essential for differentiation prior to biopsy or surgery:

(1) Epidemiology: The frequency of cardiac tumors varies by age. In individuals over 16, myxomas (45–50%), lipomas (20%), and papillary fibroelastomas (12–15%) are most common [6]. In children under 16, the most frequent primary tumors are rhabdomyomas (45%), fibromas (15%), myxomas (15%), and teratomas (15%) [7]. The incidence of cardiac tumors by age is presented in Table 1 and Table 2 [6, 8–10].

Table 1 Frequency of cardiac tumors in pediatrics

Tumor type	Frequency (%)
Benign	
Rhabdomyoma	45%
Teratoma	15%
Fibroma	15%
Myxoma	15%
Angioma	5%
Hemangioma	5%
Malignant	
Rhabdomyosarcoma	~2%
Fibrosarcoma	~2%
Malignant teratoma	Rare
Lymphoma	Rare



Table 2 Frequency of cardiac tumors in adults

Tumor type	Frequency (%)
Benign	90%
Myxoma	45-50%
Lipoma	20%
PFE	12-15%
Angioma	10–12%
Fibroma	3%
Rhabdomyoma	~2%
Malignant	10%
Sarcoma	_
Angiosarcoma	40%
Undifferentiated sarcoma	25%
Leiomyosarcoma	8–9%
Lymphoma	~1%
Mesothelioma	~1%

- (2) Clinical Symptoms and Medical History: Present and past medical history are vital for differential diagnosis. Information such as immune deficiency, presence of artificial valves (which heighten the risk of infective endocarditis), or a history of atrial fibrillation (which increases the risk of atrial thrombosis) is important for distinguishing non-tumor cardiac masses.
- (3) Tumor Location: Primary cardiac tumors tend to localize in specific areas and exhibit distinct patterns, such as being solitary or multiple. The most frequent tumors based on location within the heart should be considered.
- (4) Imaging Diagnostics Using Multiple Modalities: Some tumors exhibit characteristic imaging features, and combining various imaging techniques—such as echocardiography, CT, and MRI—can improve diagnostic accuracy.

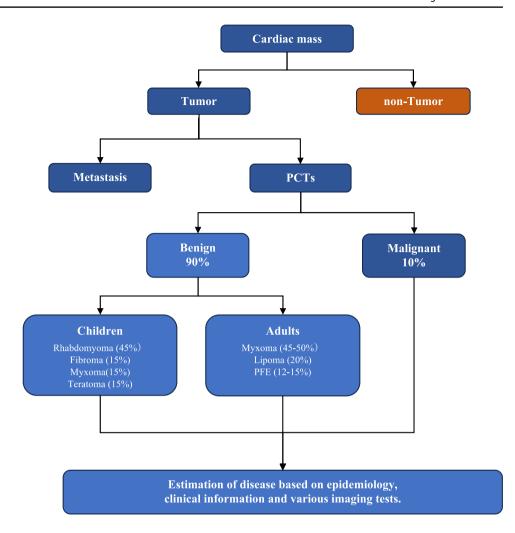
By considering these factors, the differential diagnosis becomes more precise, paving the way for surgical intervention or biopsy if needed. Figure 1 illustrates the diagnostic pathway leading to biopsy or surgery. The next section will discuss the critical role of imaging and pathological examinations in these diagnostic processes.

#### **Imaging test**

## Ultrasound cardiography: UCG

Transthoracic UCG is typically the first diagnostic modality employed when a cardiac tumor is suspected. It is non-invasive, cost-effective, and offers excellent spatial and temporal resolution, enabling the detection of even small mobile

Fig. 1 Diagnostic flow for cardiac tumors. Tumors are classified into malignant and benign categories and further classified as metastatic or primary. Approximately 90% of these tumors are benign, while 10% are malignant. The diagnosis is made by integrating clinical information such as the location of the tumor and the patient's age, along with imaging findings



tumors. However, its utility may be limited in patients with obesity or chronic lung disease, and it struggles to assess tumor extent or origin. Transesophageal UCG excels in imaging the left side of the heart and is particularly useful for evaluating the left atrium and mitral valve. It can also be employed intraoperatively to assess valvular function after tumor resection.

## Computed tomography: CT

CT provides high-resolution imaging of the heart and surrounding structures, making it ideal for evaluating potential cardiac metastases from extracardiac malignancies. It is particularly effective for visualizing calcifications and vascular structures. Coronary angiography CT plays an important role in surgical planning. However, the drawbacks include radiation exposure and the potential for contrast-induced nephropathy. CT is particularly useful for evaluating calcifications and vascular structures. For example, angiosarcomas show heterogeneous enhancement, while fibromas often appear as homogeneously calcified tumors.

#### Magnetic resonance imaging: MRI

Cardiac MRI offers detailed assessment of morphology, size, location, extent, and tissue characteristics (such as fat infiltration, necrosis, hemorrhage, calcification, and vascularity). It is less invasive than other techniques and is typically used following UCG. Limitations include long scan times, unsuitability for patients with unstable hemodynamics, and inapplicability in individuals with MRI-incompatible devices or claustrophobia.

## Positron emission tomography: PET

PET, in conjunction with CT or MRI, can assess cardiac tumors by detecting FDG uptake. Malignant and metastatic tumors often exhibit higher FDG uptake, helping differentiate benign from malignant masses [11]. However, PET's lower sensitivity compared to MRI and its susceptibility to false positives (e.g., benign lipomatous hypertrophy) limit its utility [12]. PET, in conjunction with CT or MRI, can assess cardiac tumors by detecting FDG uptake. Notably, cardiac



lymphomas often exhibit intense FDG uptake, whereas benign lipomatous hypertrophy may cause false positives.

Each of these imaging modalities has distinct advantages and limitations, but together they form an essential part of the diagnostic toolkit for cardiac tumors, aiding in the differentiation, planning, and management of treatment.

# pathological examination

## Cardiac cytoplasm cytology

Pericardial metastasis is common in primary cardiac malignancies, with a prevalence of 10–20% in autopsy cases. Cytological examination of pericardial fluid can aid in diagnosis if safe aspiration is possible. Aspiration should also be considered for cases presenting with symptoms of pericardial effusion.

## **Tissue biopsy**

Pathological diagnosis is critical for guiding treatment, as the prognosis for cardiac tumors varies significantly by pathological subtype. Biopsy is particularly important when malignancy is suspected and surgical excision is not feasible. However, biopsy procedures are often challenging due to the rarity of primary cardiac tumors (PCTs) and the limited experience many oncologists and cardiologists have with these procedures [13]. The risk of complications must also be considered. Biopsy methods include percutaneous, transvascular, and thoracotomy approaches, depending on tumor location. In cases of right-sided endocardial tumors where surgery is difficult, histological biopsy may be performed despite the high risk of complications [14].

## **Primary cardiac tumor: PCT**

# **Benign tumors**

The majority of PCTs are benign (90%). Since the incidence of benign PCTs varies between children and adults, it is essential to consider age when making a differential diagnosis. Surgery is generally recommended due to the risk of embolization, especially for left-side tumors. For right-sided tumors, assessing the presence of a patent foramen ovale or septal defect is crucial. On the other hand, subtypes such as rhabdomyosarcoma may regress spontaneously. Even when surgery is not indicated, regular echocardiographic monitoring is advisable. Thus, treatment plans should be confirmed based on the histological type and location of the tumors. Table 3 outlines the frequency, common sites, imaging findings, and treatment strategies for benign PCTs, which are relatively common in both pediatric and adult populations.

Table 3 Characteristics of benign primary cardiac tumors

	Frequency/preferred locations	Imaging findings	Treatment strategy
Myxoma	Most frequent in adults Middle-aged women, Left atrium(75%), right atrium (15%)	On UCG, it often appears as pedunculated and mobile. Shows high signal intensity on T2-weighted MRI images	Surgical resection. In cases of familial myxomas, there is a higher risk of recurrence
Lipoma	More frequent in middle-aged and older people, Right atrium most common, but can appear anywhere in the heart	On UCG, it often appears immobile with clear boundaries. It shows high signal intensity on both T1-weighted and T2-weighted MRI images, with no uptake of contrast agent	If there are no severe clinical symptoms, monitoring the progression is recommended
PFE*	Commonly occurs in individuals aged 70–80 years with no gender difference Often develops on the heart valves	On UCG, it exhibits mobility It appears isointense on T1-weighted MRI images and hypointense on T2-weighted MRI images	When tumors on the left side exceed 10 mm in diameter, there is an increased risk of stroke, therefore surgical removal is recommended
Rhabdomyoma	Rhabdomyoma Most frequent in children,  Most likely to occur in the ventricles, no difference between right and left ventricles	On UCG, it appears as high brightness On MRI, it is isointense on T1-weighted images and hyperintense on T2-weighted images	Up to 50% of cases may regress naturally. Surgical removal is considered based on clinical symptoms such as arrhythmias or heart failure
Fibroma	Most common in children, More likely to occur in the ventricles and ventricular septum	Calcifications are observed on CT and UCG It appears isointense on T1-weighted MRI images and hypointense on T2-weighted images, with no uptake of contrast agent	Surgical resection is recommended due to the risk of sudden cardiac death



#### Myxoma

Myxomas are the most common PCTs in adults, accounting for 50% of all PCTs. They predominantly occur in middle age, with 90% of cases between the ages of 30 and 60 years, and are more prevalent in women than men. In contrast, myxomas are rare in children, comprising about 10% of benign PCTs in this group. Approximately 75% of myxomas occur in the left atrium, particularly near the oval foramen of the atrial septum, while 10–15% are found in the right atrium. Clinically, myxomas can cause symptoms of left heart failure, embolism, and systemic symptoms resembling collagen disease, such as fever, joint pain, weight loss, and fatigue. The most frequent clinical manifestations were constitutional symptoms (74%), dyspnea (45%), and embolism (41%) [15].

For imaging, transthoracic and transesophageal echocardiography, cardiac CT, and MRI are commonly used. Diagnosis is typically made upon identifying a pedunculated, mobile mass via echocardiography. On MRI, most myxomas appear isointense on T1-weighted images and hyperintense on T2-weighted images relative to the endocardium. Surgical resection is the primary treatment option [16]. The recurrence rate for solitary myxomas is low, at less than 3%, but higher in familial myxomas (12%) and cases associated with the autosomal dominant Carney complex (22%). Regular echocardiographic follow-up is recommended post-resection.

#### **Fibroma**

Fibromas are the second most common PCTs in children, though they also occur in adults. They are mostly found in the ventricles and ventricular septum [17]. Clinical symptoms can include chest pain, pericardial effusion, heart failure, and arrhythmias. However, most patients with cardiac fibroma are asymptomatic [18]. Mutations in the PTCH1 gene, associated with the autosomal dominant Gorlin syndrome, are known to cause these tumors [19].

Echocardiographically, fibromas appear as homogeneously bright masses, often with calcification. CT scans reveal homogeneously calcified tumors, particularly in the central areas. On MRI, fibromas are isointense to normal myocardium on T1-weighted images and hypointense on T2-weighted images. Due to their avascular nature, they generally do not show enhancement after contrast administration. Surgical resection is recommended due to the risk of sudden cardiac death from arrhythmias caused by conduction pathway disruption, regardless of symptom presence [20].

#### Papillary fibroelastoma: PFE

PFEs are rare PCTs that most commonly affect heart valves, comprising approximately 11.5% of all PCTs. They most frequently affect the aortic valve (75% of cases), especially on its downstream side [21]. The typical onset is between 70 and 80 years, with no gender difference. While valve dysfunction can occur, embolic symptoms, such as stroke, are more common due to the left-sided valve involvement. However, a significant proportion of patients (approximately 60%) may not show symptoms [22].

Echocardiographically, PFEs are small, attached to the endocardium, and exhibit independent motion. Due to their small size, transesophageal echocardiography often has higher sensitivity than transthoracic echocardiography. On MRI, PFEs appear isointense on T1-weighted images and hypointense on T2-weighted images [23]. For right-sided PFEs, where hemodynamic obstruction risk is low, conservative management is often practiced. For left-sided PFEs or larger than 10 mm, surgical removal is recommended due to the increased risk of stroke [24]. In cases where surgery is not performed, antiplatelet medications may be considered, although data on this approach are limited [25].

#### Lipoma

Lipomas are rare benign PCTs that predominantly occur in middle-aged and elderly individuals. Often asymptomatic, they are usually discovered incidentally through imaging. Symptoms depend on the location: subendocardial (50%), subepicardial (25%), or myocardial (25%) [20]. Subepicardial lipomas may cause chest pain due to ischemia from coronary artery compression, while myocardial lipomas frequently cause arrhythmias.

Echocardiographically, these tumors appear non-mobile and well demarcated. On T1-weighted MRI images, they show high signal intensity, similar to T2-weighted images, while appearing markedly hypointense on fat-suppressed images. Due to their avascularity, contrast agents provide poor delineation. Surgical removal is generally not indicated unless the patient experiences significant clinical symptoms [26]. If a cardiac lipoma becomes symptomatic, surgical intervention is recommended to alleviate symptoms and prevent disease progression. However, there is currently no consensus on the optimal management approach for asymptomatic patients [27, 28].

## Rhabdomyoma

Rhabdomyomas are the most common tumors in pediatric PCTs, comprising 50–60% of cases. These tumors typically manifest within the first year of life [29]. They often appear as multiple tumors located in both atria and ventricles,



evenly distributed across the left and right sides of the heart [30]. Rhabdomyomas may be sporadic or associated with tuberous sclerosis, which occurs in 30–50% of cases. The tumors can cause arrhythmias, leading to symptoms such as palpitations and fainting. However, it is estimated that approximately 90% of cases remain asymptomatic [31].

Rhabdomyoma appears as bright masses on echocardiography, while on MRI, they are isointense on T1-weighted images and hyperintense on T2-weighted images. Since up to 50% of rhabdomyoma may regress spontaneously, regular monitoring with echocardiography is essential. Surgery is reserved for patients with intractable arrhythmias or heart failure [32]. In cases where surgical resection is not feasible despite the presence of symptoms, treatment with mTOR inhibitors may be considered, particularly in patients with tuberous sclerosis complex [31, 33].

## Malignant tumors

Malignant tumors constitute only 5-6% of PCT, making them exceedingly rare [34]. The most common malignant PCTs are outlined in Table 4, with sarcomas comprising 64.8%, followed by lymphomas (27%) and mesotheliomas (8%). Rapid clinical progression, the presence of nutritive vessels, intratumoral necrosis, and pericardial effusion are suggestive of malignancy. Prognosis for patients diagnosed with malignant PCT is extremely poor, with a reported five-year survival rate of 11.5% [35]. A favorable prognosis is associated with negative surgical margins [36]. While complete surgical resection is desirable, the operative mortality rate is 7.4%, and the recurrence rate post-surgery is 51.8% [37]. Given the high risks of recurrence and mortality, a multidisciplinary approach involving cardiac surgeons, cardiologists, and oncologists is essential. If complete resection is not feasible, palliative chemotherapy is an alternative. For malignancies such as sarcomas and lymphomas, anthracycline-based chemotherapy is often the first choice, though it carries the risk of cardiotoxicity. The new molecular targeted therapies are developing. The efficacy of targeted therapies, such as MDM2 inhibitors for intimal sarcomas, is currently being investigated in clinical trials, showing promising results. The decision to avoid anthracyclines is not mandatory given the cardiac origin of the tumor. Prognostic factors include age, comorbidities, and metastasis [35].

## **Angiosarcoma**

In 75% of cases, cardiac sarcomas occur in the right atrium, where they frequently invade adjacent structures [38]. At diagnosis, metastases are found in 47–89% of patients, with the lungs being a common metastatic site [39]. Angiosarcomas are more common in males, with peak onset in the 40 s. Angiosarcomas that appear on the scalp are often linked to underlying genetic abnormalities and are typically found in younger individuals [40]. Common symptoms include right heart failure and palpitations due to supraventricular arrhythmias. The most reported symptom in patients with cardiac angiosarcoma is dyspnea, occurring in 59% to 88% of cases [41].

On contrast-enhanced CT, these tumors usually show heterogeneous enhancement. MRI reveals a heterogeneous signal intensity pattern on both T1- and T2-weighted images, reflecting intratumoral necrosis and blood flow abnormalities. Surgical resection, if feasible, offers the best outcomes. The postoperative survival for patients undergoing surgery ranges from 2 to 55 months, with a median survival of 14 months [42]. If resection is not possible, the mean survival is approximately  $3.8 \pm 2.5$  months [43]. In cases with distant metastasis, chemotherapy regimens similar to those for vascular sarcomas (e.g., paclitaxel) are used. However, literature on the treatment of cardiac sarcomas remains

Table 4 Characteristics of malignant primary cardiac tumors

	Frequency/preferred locations	Imaging findings
Angiosarcoma	Originates primarily in the right atrium (75%)  More common in males, with the peak incidence in the 40 s	On contrast-enhanced CT, it shows heterogeneous enhancement. Both T1-weighted and T2-weighted MRI images also display heterogeneous signal intensities
Rhabdomyosarcoma	The most frequent malignant primary cardiac tumor (PCT) in children, with no predilection for either the left or right side	On MRI, it appears isointense on T1-weighted images, hyperintense on T2-weighted images, and shows increased signal intensity with contrast enhancement
Leiomyosarcoma	Comprises 8% of cardiac sarcomas, with the left atrium being the most common site of occurrence	On CT, it shows low attenuation with a lobulated appearance
Intimal sarcoma	Some reports suggest it is the most frequently occurring type It predominantly occurs in the left cardiac system	There are no consistent reports on imaging findings
Undifferentiated sarcoma	Accounts for about 24% of cardiac sarcomas, with an average onset age of 44 years and predominantly found in the left atrium	Imaging findings include the presence of necrosis and hemorrhage



limited to case reports and single-institution studies, and no standardized treatment protocol has been established [44].

## Leiomyosarcoma

Leiomyosarcomas account for 8–9% of cardiac sarcomas [45]. They most commonly occur in the posterior left atrium but can develop in other chambers of the heart. These tumors may remain asymptomatic until they reach an advanced stage, depending on their size and location [46]. A symptom of obstruction was the most frequent complaint at diagnosis [47].

On imaging, leiomyosarcomas typically appear on CT as irregular, lobulated areas of low attenuation. Prognosis may improve with a combination of complete surgical resection followed by chemotherapy or radiotherapy. The average survival without treatment is six months, while palliative chemotherapy or post-surgical adjuvant therapy can extend survival to 10–18 months [45].

## Rhabdomyosarcoma

Rhabdomyosarcomas are the most common malignant tumors in pediatric cardiac cases, accounting for 4–7% of all cardiac sarcomas [48]. Originating from the myocardium, these tumors often infiltrate the valve apparatus. There is no significant difference in frequency between the left and right sides of the heart [49]. Rhabdomyosarcomas are aggressive, with rapid proliferation. Dyspnea is the most common symptom, reported in 59% of cases, followed by chest pain (19%) and conduction disorders (19%) [50].

On CT, these tumors typically appear as areas of low attenuation in the ventricles. On MRI, they are isointense on T1-weighted images and hyperintense on T2-weighted images, with contrast enhancement noted. The standard treatment for cardiac rhabdomyosarcoma is multidisciplinary, combining surgery, chemotherapy, and radiation therapy [51]. Radiation therapy may provide symptom relief and local tumor control but carries the risk of both acute and chronic cardiac toxicity [52]. The standard chemotherapy regimen includes vincristine, actinomycin-D, and cyclophosphamide [53]. Despite comprehensive treatments, the average survival for patients with cardiac rhabdomyosarcoma is less than one year.

## Intimal sarcoma

Among primary cardiac soft tissue sarcomas, angiosarcomas are frequently seen in adults, while rhabdomyosarcomas are more common in children. Intimal sarcomas also occur frequently, particularly in left-sided cardiac structures [54, 55]. Cardiac intimal sarcoma is often detected due to nonspecific symptoms such as dyspnea, palpitations, and a sensation of chest pressure [56]. Diagnostic imaging for intimal sarcomas is often inconsistent, leading to frequent misdiagnosis

as myxomas or thrombi. Most reports on this tumor type remain limited to case studies [57].

Though complete surgical resection is preferred, the high invasiveness of intimal sarcomas makes it challenging [58]. The amplification of MDM2 has led to reports suggesting that MDM2 inhibitors may be effective [59].

#### Undifferentiated sarcoma

Undifferentiated sarcomas represent less than 24% of cardiac sarcomas, with an average onset age of 44 years and no gender preference [60]. These tumors most commonly affect the left atrium, causing diffuse wall infiltration. Clinical signs and symptoms are associated with the left atrium and may include dyspnea, palpitations, and manifestations of heart failure, as well as nonspecific constitutional symptoms such as fever and weight loss [61].

MRI findings often reveal lesions with necrosis and hemorrhage. The prognosis for undifferentiated sarcomas is poor, with treatment primarily consisting of surgery and palliative chemotherapy [62].

#### Mesothelioma

Most cases of cardiac mesothelioma originate from the pericardium, leading to pericardial effusion. Primary pericardial mesothelioma is an extremely rare disease, accounting for 0.8% of all mesotheliomas and 2% to 3% of all pericardial tumors [63, 64]. It is more common in males, particularly between 50 and 70 years [65]. Most cases are diagnosed at autopsy[66], with reported cases presenting with constrictive pericarditis, cardiac tamponade, or heart failure[67–69].

On CT, these tumors may show high attenuation. Pericardiocentesis for cytological examination can be performed, but obtaining a definitive diagnosis is often difficult [70]. A pericardial biopsy is the most reliable method for confirming the diagnosis.

There is no established standard treatment, but if the tumor is localized, surgical resection may be performed. Chemotherapy may be used in conjunction with surgery, though its effectiveness is not clearly established. In Japan, chemotherapy combinations such as cisplatin with pemetrexed or nivolumab with ipilimumab are used, and nivolumab is covered by insurance based on the results of the Viola trial [71]. The prognosis is extremely poor, with minimal efficacy observed for chemotherapy or radiotherapy. The average survival time is approximately 4 to 6 months [72].



**110** Page 8 of 10 Medical Oncology (2025) 42:110

#### Metastatic cardiac tumor

It is estimated that 10% of cancer patients exhibit metastasis to the heart or pericardium at autopsy, making metastatic cardiac tumors 20–40 times more common than PCTs [73]. Malignant pleural mesothelioma and malignant melanoma have the highest propensity to metastasize to the heart [74]. Additionally, cancers originating in the thoracic cavity, such as breast cancer, lung cancer, and esophageal cancer, are also prone to metastasizing to the heart.

## **Diagnosis**

When patients with a history of malignancy present with cardiovascular symptoms or signs, it is crucial to consider the possibility of cardiac metastasis and to conduct appropriate diagnostic tests with this in mind. Generally, an echocardiogram, which is a straightforward and accessible procedure, should be performed first to assess for metastases. In cases of pericardial effusion, pericardiocentesis and biopsy can provide a definitive diagnosis if feasible. However, due to the invasive nature of these procedures, they are often avoided unless the results would significantly impact the treatment plan.

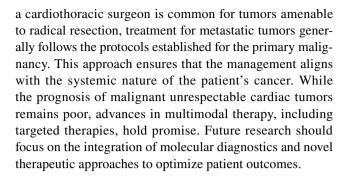
#### **Treatment**

Cardiac metastases are treated according to the management plan for the primary tumor, as they indicate a Stage IV condition. Surgical removal of metastatic lesions in the heart is generally not performed. However, under certain circumstances, surgical intervention may be considered for symptom relief [75], including:

- (1) Metastatic lesions that significantly impair hemodynamics, potentially leading to heart failure.
- (2) Isolated cardiac metastasis in patients where the primary tumor is under control and a favorable prognosis is expected.

## Conclusion

Cardiac tumors are often discovered incidentally during echocardiography or CT scans. They are classified into primary and metastatic types, with metastatic tumors being found in approximately 10% of cancer patients at autopsy. Diagnosis relies heavily on imaging studies and clinical information, with epidemiology, clinical symptoms, tumor location, and imaging results being key factors for differential diagnosis. Treatment strategies vary depending on the tumor type and condition. While surgical resection by



#### Acknowledgements None.

**Author contributions** T.I. and T.S. designed the study and wrote the paper. T.S, A.K., and K.Y. supervised, wrote, reviewed, and edited the manuscript's final version. All authors reviewed the paper and approved its content.

**Funding** This review received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

**Data availability** No datasets were generated or analyzed during the current study.

#### **Declarations**

**Conflicts of interest** No potential conflict of interest relevant to this review was reported.

Ethical approval Not applicable.

Consent for publication Not applicable.

Open Access This article is licensed under a Creative Commons Attribution-NonCommercial-NoDerivatives 4.0 International License, which permits any non-commercial use, sharing, distribution and reproduction in any medium or format, as long as you give appropriate credit to the original author(s) and the source, provide a link to the Creative Commons licence, and indicate if you modified the licensed material. You do not have permission under this licence to share adapted material derived from this article or parts of it. The images or other third party material in this article are included in the article's Creative Commons licence, unless indicated otherwise in a credit line to the material. If material is not included in the article's Creative Commons licence and your intended use is not permitted by statutory regulation or exceeds the permitted use, you will need to obtain permission directly from the copyright holder. To view a copy of this licence, visit http://creativecommons.org/licenses/by-nc-nd/4.0/.

## References

- Habertheuer A, Ehrlich M, Wiedemann D, Mora B, Rath C, Kocher A. A rare case of primary cardiac B cell lymphoma. J Cardiothorac Surg. 2014;9:14.
- McAllister HA Jr, Hall RJ, Cooley DA. Tumors of the heart and pericardium. Curr Probl Cardiol. 1999;24(2):57–116.
- 3. Klatt EC, Heitz DR. Cardiac metastases. Cancer. 1990:65(6):1456-9.
- Bussani R, De-Giorgio F, Abbate A, Silvestri F. Cardiac metastases. J Clin Pathol. 2007;60(1):27–34.



- Bui Q, Ngo TNM, Mazur J, Pham V, Palmer C, Truong BQ, Chung ES, Vuong HG, Truong VT. Long-term outcomes of primary cardiac malignant tumors: Difference between African American and Caucasian population. Cancer Med. 2021;10(24):8838–45.
- 6. Kumar P, Gandhi I, Sharma P, Ambasta R. Why our heart is not prone to cancer? American J Res Com. 2013;1:143–51.
- Tyebally S, Chen D, Bhattacharyya S, Mughrabi A, Hussain Z, Manisty C, Westwood M, Ghosh AK, Guha A. Cardiac tumors: JACC cardiooncology state-of-the-art review. JACC CardioOncol. 2020;2(2):293–311.
- 8. Casavecchia G, Lestuzzi C, Gravina M, Corrado G, Tusa M, Brunetti ND, Manuppelli V, Monte IP. Cardiac tumors. J Cardiovasc Echogr. 2020;30(Suppl 1):S45–53.
- Uzun O, Wilson DG, Vujanic GM, Parsons JM, De Giovanni JV. Cardiac tumours in children. Orphanet J Rare Dis. 2007;2:11.
- Viswanath D. Cardiac tumors in children- a review. Int J Med Med Sci. 2014;6:53–8.
- Rahbar K, Seifarth H, Schäfers M, Stegger L, Hoffmeier A, Spieker T, Tiemann K, Maintz D, Scheld HH, Schober O, Weckesser M. Differentiation of malignant and benign cardiac tumors using 18F-FDG PET/CT. J Nucl Med. 2012;53(6):856–63.
- Klein MA, Scalcione LR, Youn T, Shah RA, Katz DS, Sung WW, Yung EY. Intensely hypermetabolic lipomatous hypertrophy of the interatrial septum on 18-FDG PET with MRI and CT correlation. Clin Nucl Med. 2010;35(12):972–3.
- Dong J, Wang L, Salvatore J, Lau MW. Difficulty in diagnosing rare cardiac tumors: a case series. Cureus. 2023;15(1): e33367.
- Poterucha TJ, Kochav J, O'Connor DS, Rosner GF. Cardiac tumors: clinical presentation, diagnosis, and management. Curr Treat Options Oncol. 2019;20(8):66.
- Gabe ED, Rodríguez Correa C, Vigliano C, San Martino J, Wisner JN, González P, Boughen RP, Torino A, Suárez LD. Cardiac myxoma. Clinical-pathological correlation. Rev Esp Cardiol. 2002;55(5):505–13.
- Griborio-Guzman AG, Aseyev OI, Shah H, Sadreddini M. Cardiac myxomas: clinical presentation, diagnosis and management. Heart. 2022;108(11):827–33.
- Zheng XJ, Song B. Left ventricle primary cardiac fibroma in an adult: a case report. Oncol Lett. 2018;16(4):5463–5.
- Tao TY, Yahyavi-Firouz-Abadi N, Singh GK, Bhalla S. Pediatric cardiac tumors: clinical and imaging features. Radiographics. 2014;34(4):1031–46.
- Burke AP, Rosado-de-Christenson M, Templeton PA, Virmani R. Cardiac fibroma: clinicopathologic correlates and surgical treatment. J Thorac Cardiovasc Surg. 1994;108(5):862–70.
- Rajiah P, Kanne JP, Kalahasti V, Schoenhagen P. Computed tomography of cardiac and pericardiac masses. J Cardiovasc Comput Tomogr. 2011;5(1):16–29.
- Mariscalco G, Bruno VD, Borsani P, Dominici C, Sala A. Papillary fibroelastoma: insight to a primary cardiac valve tumor. J Card Surg. 2010;25(2):198–205.
- Klarich KW, Enriquez-Sarano M, Gura GM, Edwards WD, Tajik AJ, Seward JB. Papillary fibroelastoma: echocardiographic characteristics for diagnosis and pathologic correlation. J Am Coll Cardiol. 1997;30(3):784–90.
- 23. Bruce CJ. Cardiac tumours: diagnosis and management. Heart. 2011;97(2):151–60.
- 24. Boodhwani M, Veinot JP, Hendry PJ. Surgical approach to cardiac papillary fibroelastomas. Can J Cardiol. 2007;23(4):301–2.
- Tamin SS, Maleszewski JJ, Scott CG, Khan SK, Edwards WD, Bruce CJ, Oh JK, Pellikka PA, Klarich KW. Prognostic and bioepidemiologic implications of papillary fibroelastomas. J Am Coll Cardiol. 2015;65(22):2420–9.
- Mankad R, Herrmann J. Cardiac tumors: echo assessment. Echo Res Pract. 2016;3(4):R65–77.

- Rocha RV, Butany J, Cusimano RJ. Adipose tumors of the heart. J Card Surg. 2018;33(8):432–7.
- Wang H, Hu J, Sun X, Wang P, Du Z. An asymptomatic right atrial intramyocardial lipoma: a management dilemma. World J Surg Oncol. 2015:13:20.
- Tzani A, Doulamis IP, Mylonas KS, Avgerinos DV, Nasioudis D. Cardiac tumors in pediatric patients: a systematic review. World J Pediatr Congenit Heart Surg. 2017;8(5):624–32.
- 30. Kocabaş A, Ekici F, Cetin I, Emir S, Demir HA, Arı ME, Değerliyurt A, Güven A. Cardiac rhabdomyomas associated with tuberous sclerosis complex in 11 children: presentation to outcome. Pediatr Hematol Oncol. 2013;30(2):71–9.
- Yıldırım S, Aypar E, Aydın B, Akyüz C, Aykan HH, Ertuğrul İ, Karagöz T, Alehan D. Cardiac rhabdomyomas: clinical progression, efficacy and safety of everolimus treatment. Turk J Pediatr. 2023;65(3):479–88.
- 32. Günther T, Schreiber C, Noebauer C, Eicken A, Lange R. Treatment strategies for pediatric patients with primary cardiac and pericardial tumors: a 30-year review. Pediatr Cardiol. 2008;29(6):1071–6.
- Sugalska M, Tomik A, Jóźwiak S, Werner B. Treatment of cardiac rhabdomyomas with mTOR inhibitors in children with tuberous sclerosis complex-a systematic review. Int J Environ Res Public Health. 2021;18(9):4907.
- 34. Cresti A, Chiavarelli M, Glauber M, Tanganelli P, Scalese M, Cesareo F, Guerrini F, Capati E, Focardi M, Severi S. Incidence rate of primary cardiac tumors: a 14-year population study. J Cardiovasc Med (Hagerstown). 2016;17(1):37–43.
- Sultan I, Bianco V, Habertheuer A, Kilic A, Gleason TG, Aranda-Michel E, Harinstein ME, Martinez-Meehan D, Arnaoutakis G, Okusanya O. Long-term outcomes of primary cardiac malignancies: multi-institutional results from the national cancer database. J Am Coll Cardiol. 2020;75(18):2338–47.
- 36. Isambert N, Ray-Coquard I, Italiano A, Rios M, Kerbrat P, Gauthier M, Blouet A, Chaigneau L, Duffaud F, Piperno-Neumann S, Kurtz JE, Girard N, Collard O, Bompas E, Penel N, Bay JO, Guillemet C, Collin F, Blay JY, Le Cesne A, Thariat J. Primary cardiac sarcomas: a retrospective study of the French Sarcoma Group. Eur J Cancer. 2014;50(1):128–36.
- Bakaeen FG, Jaroszewski DE, Rice DC, Walsh GL, Vaporciyan AA, Swisher SS, Benjamin R, Blackmon S, Reardon MJ. Outcomes after surgical resection of cardiac sarcoma in the multimodality treatment era. J Thorac Cardiovasc Surg. 2009;137(6):1454–60.
- 38. Yanagawa B, Mazine A, Chan EY, Barker CM, Gritti M, Reul RM, Ravi V, Ibarra S, Shapira OM, Cusimano RJ, Reardon MJ. Surgery for tumors of the heart. Semin Thorac Cardiovasc Surg. 2018;30(4):385–97.
- 39. Butany J, Yu W. Cardiac angiosarcoma: two cases and a review of the literature. Can J Cardiol. 2000;16(2):197–205.
- Urbini M, Astolfi A, Indio V, Nannini M, Pizzi C, Paolisso P, Tarantino G, Pantaleo MA, Saponara M. Genetic aberrations and molecular biology of cardiac sarcoma. Ther Adv Med Oncol. 2020;12:1758835920918492.
- Riles E, Gupta S, Wang DD, Tobin K. Primary cardiac angiosarcoma: a diagnostic challenge in a young man with recurrent pericardial effusions. Exp Clin Cardiol. 2012;17(1):39–42.
- Antonuzzo L, Rotella V, Mazzoni F, Doni L, Bianchini D, Garbini F, Maio V, Di Costanzo F. Primary cardiac angiosarcoma: a fatal disease. Case Rep Med. 2009;2009: 591512.
- Patel SD, Peterson A, Bartczak A, Lee S, Chojnowski S, Gajewski P, Loukas M. Primary cardiac angiosarcoma - a review. Med Sci Monit. 2014;20:103–9.
- Look Hong NJ, Pandalai PK, Hornick JL, Shekar PS, Harmon DC, Chen YL, Butrynski JE, Baldini EH, Raut CP. Cardiac angiosarcoma management and outcomes:



**110** Page 10 of 10 Medical Oncology (2025) 42:110

20-year single-institution experience. Ann Surg Oncol. 2012;19(8):2707-15.

- Jellis C, Doyle J, Sutherland T, Gutman J, Macisaac A. Cardiac epithelioid leiomyosarcoma and the role of cardiac imaging in the differentiation of intracardiac masses. Clin Cardiol. 2010;33(6):E6–9.
- Gierlak W, Syska-Sumińska J, Zieliński P, Dłużniewski M, Sadowski J. Cardiac tumors: leiomyosarcoma - a case report. Kardiochir Torakochirurgia Pol. 2015;12(3):251–4.
- Wang JG, Cui L, Jiang T, Li YJ, Wei ZM. Primary cardiac leiomyosarcoma: an analysis of clinical characteristics and outcome patterns. Asian Cardiovasc Thorac Ann. 2015;23(5):623–30.
- 48. Grebenc ML, Rosado de Christenson ML, Burke AP, Green CE, Galvin JR. Primary cardiac and pericardial neoplasms: radiologic-pathologic correlation. Radiographics. 2000;20(4):1073–103.
- Gilkeson RC, Chiles C. MR evaluation of cardiac and pericardial malignancy. Magn Reson Imaging Clin N Am. 2003;11(1):173-86.
- Ahmed S. Abstract 17945: characterization and treatment of primary cardiac rhabdomyosarcoma in the last three decades. Circulation. 2023;148(Suppl\_1):A17945-A.
- Panda SP, Chinnaswamy G, Vora T, Prasad M, Bansal D, Kapoor G, Radhakrishnan V, Agarwala S, Laskar S, Arora B, Kaur T, Rath GK, Bakhshi S. Diagnosis and management of rhabdomyosarcoma in children and adolescents: ICMR consensus document. Indian J Pediatr. 2017;84(5):393–402.
- 52. Chen CF, Lin MH, Chu KA, Liu WS, Hsiao SH, Lai RS. Effective cardiac radiotherapy relieved life-threatening heart failure caused by advanced small cell lung cancer with cardiac metastasis: a case report. J Thorac Dis. 2018;10(4):E250–4.
- Crist WM, Anderson JR, Meza JL, Fryer C, Raney RB, Ruymann FB, Breneman J, Qualman SJ, Wiener E, Wharam M, Lobe T, Webber B, Maurer HM, Donaldson SS. Intergroup rhabdomyosarcoma study-IV: results for patients with nonmetastatic disease. J Clin Oncol. 2001;19(12):3091–102.
- Chan EY, Ali A, Zubair MM, Nguyen DT, Ibarra-Cortez SH, Graviss EA, Shapira OM, Ravi V, MacGillivray TE, Reardon MJ. Primary cardiac sarcomas: Treatment strategies. J Thorac Cardiovasc Surg. 2023;166(3):828-38.e2.
- 55. Neuville A, Collin F, Bruneval P, Parrens M, Thivolet F, Gomez-Brouchet A, Terrier P, de Montpreville VT, Le Gall F, Hostein I, Lagarde P, Chibon F, Coindre JM. Intimal sarcoma is the most frequent primary cardiac sarcoma: clinicopathologic and molecular retrospective analysis of 100 primary cardiac sarcomas. Am J Surg Pathol. 2014;38(4):461–9.
- Nistor C, Carsote M, Cucu AP, Stanciu M, Popa FL, Ciuche A, Ciobica ML. Primary cardiac intimal sarcoma: multi-layered strategy and core role of MDM2 amplification/co-amplification and MDM2 immunostaining. Diagnostics (Basel). 2024;14(9):919.
- Ye N, Lan L, Hu H, Liu J, Xu H. Case report: the diagnostic challenge of primary cardiac intimal sarcoma. Front Cardiovasc Med. 2023;10:1089636.
- Tamaki T, Suzuki M, Umezu R, Utanohara Y, Mahara K, Takanashi S, Murai T, Tomoike H. Clinicopathological features of biventricular cardiac intimal sarcoma-Report of an autopsy case. J Cardiol Cases. 2017;16(4):113–5.
- 59. Koyama T, Shimizu T, Kojima Y, Sudo K, Okuma HS, Shimoi T, Ichikawa H, Kohsaka S, Sadachi R, Hirakawa A, Yoshida A, Ando RM, Ueno T, Yanagaki M, Matsui N, Nakamura K, Yamamoto N, Yonemori K. Clinical activity and exploratory resistance mechanism of milademetan, an mdm2 inhibitor, in intimal sarcoma with

- MDM2 amplification: an open-label phase Ib/II study. Cancer Discov. 2023;13(8):1814–25.
- Lestuzzi C, De Paoli A, Baresic T, Miolo G, Buonadonna A. Malignant cardiac tumors: diagnosis and treatment. Future Cardiol. 2015;11(4):485–500.
- 61. Board WHOCWHOCTE. Thoracic Tumours: International Agency for Research on Cancer; 2021.
- Watson R, Frye J, Trieu M, Yang MX. Primary undifferentiated pleomorphic cardiac sarcoma with MDM2 amplification presenting as acute left-sided heart failure. BMJ Case Rep. 2018. https:// doi.org/10.1136/bcr-2018-226073.
- Gemba K, Fujimoto N, Kato K, Aoe K, Takeshima Y, Inai K, Kishimoto T. National survey of malignant mesothelioma and asbestos exposure in Japan. Cancer Sci. 2012;103(3):483–90.
- Kaul TK, Fields BL, Kahn DR. Primary malignant pericardial mesothelioma: a case report and review. J Cardiovasc Surg (Torino). 1994;35(3):261–7.
- Eren NT, Akar AR. Primary pericardial mesothelioma. Curr Treat Options Oncol. 2002;3(5):369–73.
- Nambiar CA, Tareif HE, Kishore KU, Ravindran J, Banerjee AK. Primary pericardial mesothelioma: one-year event-free survival. Am Heart J. 1992;124(3):802–3.
- Choi WS, Im MS, Kang JH, Kim YG, Hwang IC, Lee JM, Lee S, Shin HS, Lee SP, Cho GY. Primary malignant pericardial mesothelioma presenting as acute pericarditis. J Cardiovasc Ultrasound. 2012;20(1):57–9.
- Kayatta MO, Dineen SP, Sica G, Puskas JD, Pickens A. Primary pericardial mesothelioma in a 19-year-old presenting as pericarditis. Ann Thorac Surg. 2013;96(2):680–1.
- Massicotte G, Bernier M, Piché ME, Cantin L, Couture C. Case report of aggressive primary pericardial mesothelioma presenting as a constrictive pericarditis. Circ Cardiovasc Imaging. 2019;12(3): e008621.
- Mezei G, Chang ET, Mowat FS, Moolgavkar SH. Epidemiology of mesothelioma of the pericardium and tunica vaginalis testis. Ann Epidemiol. 2017;27(5):348-59.e11.
- Kuribayashi K, Igeta M, Daimon T, Ibu M, Suna S, Okamoto R, Kijima T. Clinical efficacy and safety of nivolumab in malignant non-pleural mesothelioma: a multicenter, open-label, single-arm, Japanese phase II trial (Viola) protocol. Oncology. 2023;101(4):257–61.
- Oliveira GH, Al-Kindi SG, Hoimes C, Park SJ. Characteristics and survival of malignant cardiac tumors: a 40-year analysis of >500 patients. Circulation. 2015;132(25):2395–402.
- Butany J, Nair V, Naseemuddin A, Nair GM, Catton C, Yau T. Cardiac tumours: diagnosis and management. Lancet Oncol. 2005;6(4):219–28.
- Patel JK, Didolkar MS, Pickren JW, Moore RH. Metastatic pattern of malignant melanoma. A study of 216 autopsy cases. Am J Surg. 1978;135(6):807–10.
- Palaskas N, Thompson K, Gladish G, Agha AM, Hassan S, Iliescu C, Kim P, Durand JB, Lopez-Mattei JC. Evaluation and management of cardiac tumors. Curr Treat Options Cardiovasc Med. 2018;20(4):29.

**Publisher's Note** Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

