Cardiac tumors in a tertiary care cancer hospital: clinical features, echocardiographic findings, treatment and outcomes

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Abstract

Cardiac tumors are a rare entity, comprised of tumors with diverse histology and natural history. We report the clinical characteristics, echocardiograhic findings, therapy and outcome of 59 patients with primary and metastatic cardiac tumors. Our institutional echocardiogram data base from 1993 through 2005 was reviewed to identify patients diagnosed with intra-cardiac tumor. A total of 59 patients with cardiac tumors were identified and included in the study. The patients' characteristics, presenting symptoms, diagnostic tests, location, histology of the tumor, treatment and the one year survival rate of this population was collected from the medical records. Of the 59 cardiac tumor cases, 16 (27%) were primary cardiac tumors and 43 (73%) were secondary cardiac tumors. The most common primary tumor was sarcoma affecting 13 (81%) of the 16 cases. Of these, 5 patients were angiosarcoma, 5 unclassified sarcoma, one myxoid sarcoma and 2 malignant fibrous histiocytoma. The mean age at presentation was 41.1 years, and the most common location was right atrium affecting 6 cases (37.5%). The most common symptom of dyspnea was present in 10 (62.5%) cases. Eleven (25.6%) of the 43 secondary cardiac tumors were metastasis from renal cell carcinoma. The mean age at presentation was 55.4 years. Right atrium was the most frequent location affecting 18 (42%) of the 43 patients. The most common presenting symptom was dyspnea in 15 (35%) cases. For both primary and secondary tumors, dyspnea was the most common symptom and right atrium was most frequently involved. Sarcoma was the most common primary cardiac tumor while metastasis from renal cell carcinoma was the most common secondary tumor.

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Introduction

Cardiac tumors are divided into primary and secondary tumors. Primary cardiac tumors are very rare, with an autopsy incidence of 0.001-0.03%.[1] Primary cardiac tumors include benign or malignant neoplasm that may arise from any tissue of the heart. Secondary or metastatic cardiac tumors are 30 times more common than the primary neoplasm with an autopsy incidence of 1.7-14%.[2] The clinical manifestation of cardiac tumor is variable and they are sometimes found on routine surveillance by echocardiogram. There are limited detailed reports on large series of cardiac tumors. We report 59 cases of cardiac tumors, 16 primary cardiac tumors and 43 secondary cardiac tumors.

Materials and Methods

We present a 12-year single-institution experience with cardiac tumors from the Department of Cardiology at the MD Anderson Cancer Center. The study was approved by the institutional review board. The echocardiogram data base from 1993 to 2005 was reviewed to identify patients diagnosed with intra-cardiac tumor. For the purpose of this study, for primary cardiac tumors both benign and malignant primary cardiac tumors of cardiac and pericardial origin were included in the study. Secondary cardiac tumors with pericardial metastases alone were excluded from the study, and those with intracardiac invasion with or without pericardial involvement were included. A total of 59 patients with cardiac tumors were identified and all were included in the study. Patients' characteristics, presenting symptoms, diagnostic tests, location, histology, treatment and 1-year survival rates were collected from the medical records. Response to chemotherapy was assessed based on revised RECIST criteria.[3]

Results

Of the 59 cardiac tumor cases, 16 (27%) were primary cardiac tumors and 43 (73%) were secondary cardiac tumors.

Primary cardiac tumors

Of the 16 primary cardiac tumors, 15 (94%) were malignant and one (6%) was benign (Table 1). Mean patient age was 41.1 years (range 20-63 years) with 7 (44%) females and 9 (56%) males.

The most common primary tumor was sarcoma affecting 13 (81.2%) of the 16 cases, followed by paraganglioma in 2 (12.6%) and myxoma in one (6.2%). Of the 13 cases of sarcomas, 5 (38.5%) were angiosarcomas, 5 (38.5%) were unclassified sarcomas, 2 (15.4%) malignant fibrous histocytoma and one (7.7%) was myxoid sarcoma.

The most common site was right atrium (RA) with 6 cases (37.5%), followed by left atrium (LA) 4 cases (25%), right ventricle (RV) 2 cases (12.5%), RA and RV one case (6.3%), LA and mitral valve (MV) one case (7.7%), LA/RA/MV in one case (6.3%) and intrapericardial one case (6.3%). The size of the tumor was known in 14 patients and ranged from 1-7.7 cm.

Of the 16 cases, 14 (87.5%) were sympto-

matic and in 2 (12.5%) cases this was an incidental finding. The most common symptom was dyspnea; this was presented in 10 (62.5%) cases, followed by chest pain in 4 (25%), dizziness in 2 (12.5%), cardiac tamponade in one (6.3%) and palpitations in one (6.3%). Pericardial rub was observed in 2 cases (12.5%) and systolic murmur was audible in 2 cases (12.5%).

Data regarding chemotherapy was available in only 11 patients. These patients were treated with various combinations of the following chemotherapy agents: doxorubicin, ifosfamide, gemcitabine, docetaxel and epirubicin. The most common front-line regimen was doxorubicin 75 mg/m^2 given over 72 h as continuous infusion in combination with ifosfomide 10 g/m^2 per day divided over 4-5 days. The second most common regimen was gemcitabine 675-900 mg/m^2 on Day 1 and Day 8 with docetaxel 60-100 mg/m^2 on Day 8 only. Angiosarcoma showed the best response to therapy compared to other histological subtypes, but due to small sample size the difference was not statistically significant. All 5 angiosarcoma treated with adriamycin-based therapies showed at least partial response. Other histological varieties showed a less robust response to chemotherapy.

A total of 7 (44%) patients died within one year of diagnosis. The mean survival for malignant primary cardiac tumor was 26.4±6.8 months (mean±SEM; 95% CI: 13.1-39.6). Half the patients survived for more than 24 months

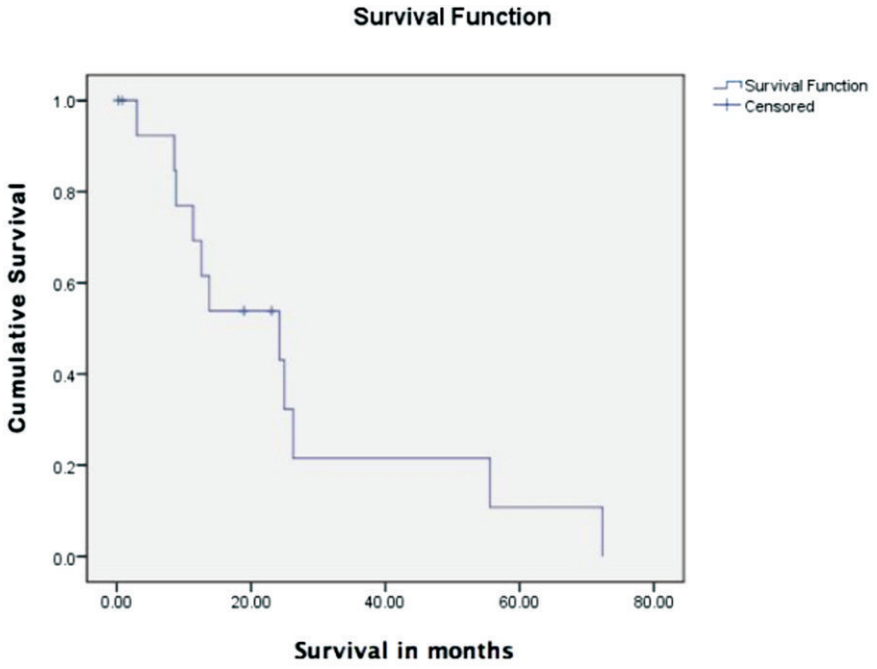

Figure 1. Survival of patients with primary cardiac tumor.

Table 1. Patients with primary cardiac tumors: characteristics, treatment and outcomes.

	A/S	Primary	Size of tumor (cm)	Location of tumor	History	Histology	Surgery Yes/No	Chemotherapy Yes/No	Died within 1 year of diagnosis
1	43/M	Angiosarcoma	5×5.5	RA	I	NA	No	Yes	Yes
2	20/F	Angiosarcoma	2.4×5	RV/RA	Dyspnea	Angiosarcoma	Yes	Yes	Yes
3	55/F	Angiosarcoma	4.1×2.6	RA	CP	NA	No	Yes	No
4	45/F	Angiosarcoma	2..1×4.0	RA	Dyspnea,Tamponade	Angiosarcoma	Yes	Yes	Yes
5	40/M	Angiosarcoma	1×2.5	RA	Dyspnea	Angiosarcoma	Yes	Yes	No
6	56/M	Sarcoma	2×4	LA	Dyspnea	Unclass. sarcoma	No	Yes	Yes
7	44/F	Sarcoma	7.0×2.0	LA/RA/MV	Dyspnea	Unclass. sarcoma	Yes	NA	Yes
8	32/M	Sarcoma	3.5×3.5	LA/MV	Dizziness	Unclass. sarcoma	Yes	Yes	No
9	34/M	Sarcoma	NA	RA	Dyspnea, CP, Dizziness	Unclass. sarcoma	Yes	Yes	No
10	35/F	Sarcoma	6×7.7	Pericardium	Dyspnea, CP	Unclass. sarcoma	No	NA	Yes
11	49/F	Sarcoma	NA	RV	Dyspnea	MFH	Yes	Yes	No
12	37/F	Myxoma	4.5×2.5	RA	Palpitations	Myxoma	Yes	-	No
13	26/M	Paraganglioma	4×4	LA	CP	Paraganglioma	Yes	-	No
14	63/M	Paraganglioma	6×4	LA	I	Paraganglioma	Yes	-	No
15	38/F	Myxoid sarcoma	7.5×5	RV	Dyspnea	Cardiac myxoid sar.	Yes	Yes	Yes
16	26/M	MFH	3.3×2.7	LA	Dyspnea, chest pain	High grade sarcoma, consistent with MFH	Yes	Yes	No

M, male; F, female; NA, data not available; RA, right atrium; LA, left atrium; RV, right ventricle; MV, mitral valve; I, incidental finding; TEE, ransesophageal echocardiogram; CT, computed tomography; MFH, malignant fibrous histocytoma; A, age; S, sex; unclass., unclassified; CP, chest pain. In Patient 6, specimen for biopsy was obtained *via* sternotomy. In Patient 10 the diagnosis was established by cytology from pericardial fluid.

(Figure 1). Of the 16 cases of primary cardiac tumor, 12 (75%) underwent surgical resection. Of the 12 patients who underwent surgery, 4 (33%) died within one year. Out of 4 patients who did not undergo surgery, 3 (75%) died within one year.

Secondary cardiac tumors

Of the 43 secondary cardiac tumors, 23 were males (53.5%) and 20 were females (46.5%), with a mean age of 54.4 years (range 24-82 years) (Tables 2 and 3). Eleven (25.6%) of the 43 secondary cardiac tumors were metastases from renal cell carcinoma, 6 (14%) from sarcomas (pleura, retroperitoneal, pelvis, endometrium, mediastinum and inferior vena cava), 6 (14%) from melanoma, 6 (14%) from lung cancer, 3 (7%) from breast cancer, 2 (4.7%) from carcinoid tumor.

The RA was the most frequent site affecting 18 (41.9%) of the patients, followed by LA in 13 (30.2%), left ventricle (LV) in 7 (16.3%), RV in 2 (4.7%), RA/RV in 2 (4.7%) and LA/RA in one (2.3%).

The size of the tumor was available in 35 patients and it ranged from less than 1 cm to 11 cm.

Of the 43 cases, 24 (56%) had symptoms and in 19 (44%) cases this was an incidental finding. The most common symptom was dyspnea, which was present in 15 (35%) cases, followed by palpitations/atrial fibrillation/atrial flutter in 5 (11.6%), chest pain in 3 (7%), ankle edema in 4 (9%), syncope/near syncope in 2 (4.7%), dizziness in one (2%) and hypotension in one (2%). Systolic murmur was observed in 11 cases (25.6%).

A total of 23 (53.4%) patients died within one year of diagnosis of secondary tumor. Of the 43 patients with secondary cardiac tumors, 23 (53.5%) underwent surgical resection, of which 13 (56.5%) were alive and 10 (43.5%) were dead at one year of diagnosis. Of the 20 patients who did not undergo surgery, 7 (35%) were alive and 13 (65%) were dead within one year of diagnosis.

Discussion

Primary cardiac tumors

The majority (>80%) of primary cardiac tumors are benign; myxoma is the most common.[4,5] The remaining 20% are malignant primary cardiac tumors, of which cardiac sarcomas are the most common.[4,5] However, in our series the most common primary cardiac tumor was malignant cardiac sarcoma. Our institution is a tertiary referral center for malignant disorders and this probably explains this population bias of predominance of malignant over benign cardiac tumors. Review of the literature reveals that primary cardiac sarcoma constitutes approximately 1% of all soft tissue sarcomas, with a median age of presentation of 39-44 years.[6]

Cardiac tumors cause disease in three separate mechanisms: embolization, obstruction and arrhythmias. Embolization occurs frequently, fragments of the tumor itself can migrate, this may mimic endocarditis or vasculitis, and larger particles can cause cerebrovascular events. Atrial tumors, when large enough, may result in obstruction which impedes valvular flow, resulting in obstructive symptoms like syncope or congestive heart failure (CHF). Similarly, ventricular tumors may block outflow tracts resulting in chest pain, shortness of breath or syncopal episodes, as well as CHF. Finally, tumors can cause intra-myocardial and intra-cavitary infiltration, and affect the conduction system resulting in heart block, arrhythmias and ventricular tachycardia, which may present as sudden death.[7] The most common presenting symptoms are dyspnea, followed by chest pain, cough, syncope, hemoptysis, sudden death, fever, embolic events, cardiac arrhythmias, hepatic vein and superior vena caval obstruction.[8,9] Physical findings such as systolic murmur, diastolic murmur, elevated jugular venous pressure and tumor plop may be present.[10] Cardiomegaly is a common radiological finding of cardiac sarcomas.[9] Electrocardiogram (ECG) changes are usually non-specific; however, heart block, ventricular hypertrophy, bundle branch blocks, atrial flutters and atrial tachycardia may be present in some cases.[9] In our series, 38 patients (64.4%) were symptomatic and, similar to other reports, the most common presenting symptom was dyspnea (66%) followed by chest pain (21%). In 21 of 59 (35.6%) cases this was an incidental finding (Table 4).

Nearly half of the primary tumors in the right atrium are malignant and are predominantly found in males. The majority of the left atrial primary tumors are benign.[5] Previously, primary cardiac tumors were identified at an advanced stage, but in the past two decades have been identified at an earlier stage due to better modalities of cardiac imaging. Echocardiogram is used in the initial diagnosis of primary cardiac tumors with transthoracic echocardiogram being the initial modality. However, there are several well known limitations, such as operator experience, restricted field of view due to bone and lung interference, and unfavorable body habitus such as chronic obstructive pulmonary disease or narrow rib spaces. Transesophageal echocardiogram and especially cross sectional imaging methods such as computerized tomography (CT scan) and magnetic resonance imaging (MRI) have roles in further assessment of cardiac neoplasm, especially in evaluation of myocardial invasion, involvement of mediastinal structures and tissue characterization and vascularity.[11-14] CT scan is useful in predicting extra cardiac extension of tumor, while MRI scan is better in identifying the amount of myocardial and great vessel involvement.[5] Complete resection of the tumor is possible in most of the benign primary tumors compared to malignant tumors, with a perioperative death of 1.4%.[5]

Cardiac sarcomas are the commonest of the malignant primary cardiac tumors. The age of presentation for cardiac sarcomas ranges from one to 76 years, with a mean age around 40 years.[9] Angiosarcomas and unclassified sarcomas account for 76% of all cardiac sarcomas, of which angiosarcomas are the most common followed by unclassified sarcomas.[15] In our study, 77% of the cardiac sarcomas were angiosarcomas and unclassified sarcomas. Rhabdomyosarcoma is the most common form of cardiac sarcoma in children. Leiomyosarcoma, synovial sarcoma, osteosarcoma, fibrosarcoma, myxoidsarcoma, liposarcoma, mesenchymal sarcoma, neurofibrosarcoma, malignant fibrous histocytoma are other cardiac sarcomas observed.[8,15] Angiosarcomas are predominantly found on the right side while osteosarcomas and unclassified sarcomas are predominantly found on the left side of the heart.[9,15] Pericardial angiosarcomas are rare. All 5 (100%) angiosarcomas in our series were found in the RA. Of the 5 unclassified sarcomas in our series, 3 (60%) were in the LA, one (20%) was in the RA and one (20%) was in the pericardium. About 29% of cardiac sarcomas have metastatic disease at the time of presentation.[8] The most common site of metastasis is lung.[8,9] In our series, 3 (23%) of the 13 cardiac sarcoma patients had metastasis at the time of presentation; two of them had metastases in the lung and one in the liver.

For treatment purposes, cardiac sarcomas are divided into three groups: right heart sarcomas, left heart sarcomas and pulmonary artery sarcomas.[16] The treatment for right heart and left heart sarcomas is surgery and chemotherapy. Radiotherapy is avoided in these patients as it may cause myocardial injury. Treatment of pulmonary artery sarcomas includes surgery, chemotherapy and radiotherapy. Radiotherapy can be used in these sarcomas as myocardium is shielded from the radiation field.[16] The most common chemotherapeutic regimen used for cardiac sarcomas is combined doxorubicin and ifosfamide. More than half of the cardiac sarcoma patients who received chemotherapy in our series received doxorubicin and ifosfamide. A combination of docetaxel and gemcitabine also showed good response in various sarco-

Table 2. Patients with secondary cardiac tumors: characteristics, treatment and outcome.

	A/S	Primary tumor	Size of tumor(cm)	Location of tumor	History	Histology	Surgery Yes/No	Died within 1 year of diagnosis
1	57/M	Renal cell carcinoma	NA	RA	I	NA	Yes	No
2	40/M	Renal cell carcinoma	NA	LA	I	RCC	Yes	Yes
3	69/M	Renal cell carcinoma	NA	RA	Ankle edema	RCC	Yes	No
4	72/M	Renal cell carcinoma	4×5	RA	I	RCC	Yes	No
5	43/F	Renal cell carcinoma	NA	RA	Dyspnea	Collecting duct ca.	Yes	Yes
6	33/M	Renal cell carcinoma	4×3	LA	I	NA	No	Yes
7	78/F	Renal cell carcinoma	1.5×1.4	RA	I	Sarcomatoid ca.	Yes	Yes
8	52/F	Renal cell carcinoma	3.5×2.2	LV	I	NA	No	Yes
9	65/M	Renal cell carcinoma	2.2×1.5	LA	Dyspnea	NA	No	No
10	56/F	Renal cell carcinoma	NA	RA/RV	Dyspnea, edema	RCC	Yes	Yes
11	60/M	Renal cell carcinoma	NA	RA	Dyspnea, edema	RCC	Yes	No
12	24/F	Pleural sarcoma	1.6×1.4	LV	Dyspnea	NA	No	Yes
13	50/M	Pleomorphic liposar.	6.3×2.7	LV	Dyspnea	Pleomorphic sar.	Yes	Yes
14	15/M	Undiff. sarcoma pelvis	0.7×0.8	RA/RV	I	Sarcoma	Yes	No
15	31/F	Endometrial sarcoma	NA	RA	Dyspnea	Sarcoma	Yes	No
16	54/F	Leiomyosarcoma (med.)	3.5×4.1	RA	I	Leiomyosarcoma	Yes	No
17	56/F	Leiomyosarcoma (IVC)	3.1×2.4	RA	I	Leiomyosarcoma	Yes	Yes
18	56/M	Malignant melanoma	1.9×1.9	RA	Dyspnea	NA	No	Yes
19	56/M	Malignant melanoma	2.5	LV	Hypotension	NA	No	Yes
20	39/F	Malignant melanoma	5.5×3	LA	Dyspnea, palpitation, syn.	Metastatic melanoma	Yes	No
21	32/M	Malignant melanoma	7×11	LA	Chest pain	Metastatic melanoma	Yes	Yes
22	67/M	Malignant melanoma	4.0×2.0	RA	I	NA	Yes	No
23	40/M	Malignant melanoma	3.3×3.8	RA	Chest pain	Metastatic melanoma	Yes	Yes
24	59/M	Sarcomatoid lung can.	1.3×0.6	RA	Dyspnea	NA	Yes	No
25	43/M	Unclass. large cell ca. lung	NA	RA	Dyspnea	NA	No	Yes
26	75/F	Non small cell ca. lung	2.1×2.5	LA	Palpitations	Poorly diff. adenoca.	Yes	Yes
27	51/F	SCC lung	3×2	LA	Dyspnea	SCC	Yes	Yes
28	71/M	SCC lung	3.3×1.6	LA	Atrial flutter	NA	No	Yes
29	46/M	Adenocar. lung	6.0-7.0	LA	I	Adenocarcinoma	Yes	No
30	42/F	Inf. ductal ca. breast	2.5×2.5	LA	Dyspnea	NA	No	Yes
31	44/F	Adenoca. breast	4.6×2.0	RV	I	NA	No	Yes
32	78/F	Inf. ductal ca. breast	1.5×1.5	RA	Dyspnea, edema, CHF	NA	No	No
33	56/F	Carcinoid tumor	3.7×3.9	RA/LA	I	NA	No	No
34	71/M	Carcinoid syndrome	3.0×3.0	RV	I	NA	No	No
35	49/F	MFH (colon)	3.3×2.4	LA	I	NA	No	No
36	43/F	Transitional and SCC (bl.)	2.6×1.0	LV	I	NA	No	Yes
37	82/M	Rectal adenocar.	2.5×4	RA	Dizziness, syncope	NA	No	Yes
38	80/M	SCC (tongue)	3.1×1.0	RA	Irregular heart rhythm	NA	No	No
39	62/F	Large B-cell lymphoma	1.0×1.0	LV	I	NA	No	Yes
40	57/M	Multiple myeloma	3.9×4.8	LA	I	NA	No	Yes
41	43/F	Leiomyoma (uterus)	4×3.8	RA	Dyspnea, palpitation	Smooth muscle tum.	Yes	No
42	42/F	Smooth cell tumor (thigh)	1.8×3.5	LV	Chest pain	NA	No	No
43	63/M	Carcinoid neuroendocrine tumor	3.5×4.3	LA	I	NA	Yes	No

A, age; S,sex; M, male; F, female; NA, not available; RA, right atrium; LA, left atrium; RV, right ventricle; LV, left ventricle; MV, mitral valve; I, incidental; TEE, transesophageal echocardiogram; CT, computed tomography; MRI, magnetic resonance imaging; IVC, inferior venacava; CHF, congestive heart failure; CA, carcinoma; SCC, squamous cell carcinoma; AML, acute myeloid leukemia; ca., carcinoma; sar., sarcoma; syn., syncope; diff., differentiated; Inf., infiltrative; unclass., unclassified; undiff., undifferentiated; RCC, renal cell carcinoma; adenocar., adenocarcinoma; tum., tumor; MFH, malignant fibrous histocytoma; med., mediastinum; bl., bladder.

mas and can be used as alternative chemotherapeutic agents.[17] Other treatment regimens include ifosfamide-epirubicin (doxorubicin) and cyclophosphamide, vincristine, doxorubicin and dacarbazine (CyVADIC).[6]

Unlike other sarcomas, cardiac sarcomas have a very poor prognosis with a median survival rate of 6-25 months after diagnosis.[9,15] Presence of tumor necrosis and metastases is associated with a poor prognosis.[8,9] A recent study showed that 14.8% of the resected tumors were low grade and all the patients were alive at follow up.[18] This underlies the importance of tumor grade in survival of post-operative patients. Sarcomas other than angiosarcomas, sarcomas on the left side of the heart and completely resected sarcomas have a better prognosis.[8,9] Angiosarcomas grow faster, infiltrate widely, and metastasize early; they therefore have a poor prognosis.

Secondary cardiac tumors

The autopsy incidence of secondary cardiac tumors ranges from 1.7 to 14% (average 7.1%) in cancer patients and 0.7 to 3.5% (average 2.3%) in the general population.[2] In comparison to older series, there is a significant increase in the incidence of cardiac metastases (CM) in cancer patients after 1970.[2] This is likely due to improvement in imaging modalities.

Sarcomas that affect the myocardium are mostly high grade and progress swiftly. Myocardial infiltration, outflow obstruction and distant metastasis result in death within a few weeks to two years of onset of symptoms, median survival ranging from 6-12 months. Different series documented the metastatic rate to be 26-43% at presentation and 75% at the time of death.[8,9,19-27] CM can occur either by direct extension, *via* blood stream, lymphatics or by intracavitary diffusion through inferior vena cava (IVC). Pericardial metastasis (69%) is the most common type of CM, followed by epicardial (34%), myocardial (32%) and endocardial metastases (5%).[28] Pericardium is most often involved due to direct invasion by the thoracic cancers. Myocardium or epicardium is most commonly involved through lymphatic spread and endocardial metastases through hematogenous spread. Abdominal and pelvic tumors may reach the RA through IVC. The most common tumor exhibiting this tendency is renal cell carcinoma. Three patients with renal cell carcinoma in our series had right atrial metastases via the IVC.

Table 3. Demographics of secondary cardiac tumors (n=43).

Primary tumor	N (%)
Renal cell carcinoma	11 (25.6)
Sarcoma	6 (14)
Malignant melanoma	6 (14)
Lung cancer	6 (14)
Breast cancer	3 (7)
Carcinoid tumor	2 (4.7)
Lymphoma	1 (2.3)
Leukemia	1 (2.3)
Multiple myeloma	1 (2.3)
Colon/rectal cancer	1 (2.3)
Leiomyoma	1 (2.3)
Squamous cell carcinoma of tongue	1 (2.3)
Neuroendocrine tumor	1 (2.3)
Smooth muscle tumor	1 (2.3)
Bladder cancer	1 (2.3)

N, numbers.

Table 4. Clinical symptoms of symptomatic cardiac tumors (n=38)*.

Clinical symptoms	P+S (38) N (%)	P (14) N (%)	S (24) N (%)
Dyspnea	25 (66)	10 (71)	15 (63)
Chest pain	8 (21)	5 (36)	3 (13)
Palpitation/atrial flutter/atrial fibrillation	6 (16)	1 (7)	5 (21)
Ankle edema	4 (10)	0	4 (17)
Dizziness	3 (8)	2 (14)	1 (4)
Syncope	2 (5)	0	2 (8)
Tamponade	1 (3)	1 (7)	0
Hypotension	1 (3)	0	1 (4)

*Of the 59 cases in the study, 38 were symptomatic; 21 were incidental findings. P, primary cardiac tumors; S, secondary cardiac tumors; N, numbers. Some patients had more than one of the above sympthoms.

Table 5. Reported primary origin of secondary cardiac tumors (including pericardial invasion).

	Author	Year	CM cases primary	Most common	N (%)	2nd most common primary	N (%)	Chamber/side involved
1	Bussani *et al.*[28]	2007	662	Lung cancer	263 (38%)	Leukemia/ malignant lymphoma	67 (10%)	NA
2.	Hanfling[29]	1960	122	Leukemia	34 (28%)	Lymphoma	27 (22%)	NA
3	Cates *et al.*[30]	1986	47	Lung cancer	25 (53%)	Leukemia/ lymphoma	5 (11%)	NA
4.	Butany *et al.*[31]	2005	264	Lung cancer	-	Leukemia/ multiple myeloma	-	NA
5	Rafajlovski *et al.*[32]	2005	79	Lung cancer	18 (23%)	Leukemia/ malignant lymphoma	16 (20%)	LV
6	Abraham *et al.*[33]	1990	95	Lung cancer	-	Lymphoma	-	NA
7	Karwinski *et al.*[34]	1989	130	Lung cancer	60 (46%)	Malignant melanoma	13 (10%)	NA
8	Goudie[35]	1954	126	Lung cancer	85 (67%)	Lymphadenoma/reticulosar.	10 (8%)	Lt side (37%) Rt.side (28%) Both (35%)
9.	Lockwood *et al.*[36]	1980	179	Lung cancer	108 (60%)	Kidney	14 (8%)	NA
10	Silvestri *et al.*[37]	1997	162	Melanoma	(48%)	Lung cancer	(29%)	NA
11.	Abioye *et al.*[38]	1975	64	Burkitt's lymphoma	30 (31%)	Hodgkin's disease	4 (6%)	RA(31.25%)
12	De Loach *et al.*[39]	1953	137	Lung cancer	22 (16%)	Reticulum cell sarcoma	18 (13%)	Rt side (86%), Right ventricle (46%)
13	Mac Gee *et al.*[40]	1991	57	Lung cancer	20 (40%)	Breast cancer	11 (22%)	LV (77%)
14	Manojlovic[41]	1990	39	Lung cancer	17 (44%)	Tongue and sublingual region	9 (23%)	Lt. side

CM, cardiac metastases; NA, not available; sar., sarcoma, Lt, Left; Rt, right;.LV, left ventricle, RA, right atrium.

Review of previous report shows that lung cancer is the most common cause of cardiac metastasis followed by hematologic malignancy. Only a couple of reports show a different trend (Table 5).[28-41] In our study, about 25.6% of the intra-cardiac metastases were from renal cancer followed by 14% from sarcomas, 14% each from malignant melanoma and lung cancer. Breast cancer and carcinoid tumor were the primary source for 7% and 4.7% of the secondary cardiac tumors, respectively. In the previous reports, the most common side/chamber of the heart involved with CM varied from study to study (Table 5).[28-41] In our study, both sides of the heart were equally affected and right atrium was the most commonly involved chamber of the heart.

The symptoms of CM are extremely variable, depending on the location of the tumor. Dyspnea, palpitations, syncope, chest pain and peripheral edema are common clinical presentations of CM.[28-29] Congestive heart failure, cardiac arrhythmias, heart blocks, acute myocardial infarction, myocardial rupture and systemic embolization are other manifestations of CM.[29] A new heart murmur of stenosis or any new ECG finding without any symptoms in a cancer patient should raise the suspicion of CM.[29-30] The ECG findings commonly found in CM are ST-T wave changes (mimicking myocardial ischemia or injury), new atrial fibrillation or flutter and low voltage ECG.[30] The ECG findings of myocardial injury have high specificity for CM.[30]

Treatment of metastatic cardiac tumors is usually palliative. Different series have shown that the median survival is 17-24 months for patients who can undergo complete resection and 6-10 months for patients unable to undergo complete resection.[8,12] Surgery with post-operative chemotherapy and/or radiotherapy to prevent local recurrence is indicated in patients with better prognosis and when they have only CM without disseminated disease.[2] Orthotopic heart transplantation is an option in selected patients, with improved survival. [42,43] In patients with disseminated disease, limited life expectancy and poor performance status, radiotherapy is the treatment of choice. Chemotherapy is recommended for tumors which are chemo-sensitive. In these patients, end of life care should be discussed and all efforts should be made to improve patient quality of life.

In conclusion, in our tertiary center, sarcoma was the most common primary cardiac tumor while metastasis from renal cell carcinoma was the most common secondary tumor. Dyspnea was the most common symptom and RA was most frequently involved. Overall prognosis for primary cardiac sarcoma is poor, with 54% patients dying within one year of diagnosis.

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