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, echocardiographic 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.

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 1.7-1.4%.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 histiocytoma 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/RV/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/m² given over 72 h as continuous infusion in combination with ifosfamide 10 g/m² per day divided over 4-5 days. The second most common regimen was gemcitabine 675-900 mg/m² on Day 1 and Day 8 with docetaxel 60-100 mg/m² 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.

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   | Angiosarcoma     | 5x5.5              | RA                | I                   | NA              | No             | Yes                 | Yes                             |
| 2   | Angiosarcoma     | 2.4x3              | RV/RA             | Dyspnea            | Angiosarcoma    | Yes            | Yes                 | Yes                             |
| 3   | Angiosarcoma     | 4.1x2.6            | RA                | CP                 | NA              | No             | Yes                 | No                              |
| 4   | Angiosarcoma     | 2.1x4.0            | RA                | Dyspnea,Tamponade  | Angiosarcoma    | Yes            | Yes                 | Yes                             |
| 5   | Angiosarcoma     | 1x2.5              | RA                | Dyspnea            | Angiosarcoma    | Yes            | Yes                 | No                              |
| 6   | Sarcoma          | 2x4                | LA                | Dyspnea            | Unclass. sarcoma| No             | Yes                 | Yes                             |
| 7   | Sarcoma          | 7.8x2.0            | LA/RA/MV          | Dyspnea            | Unclass. sarcoma| Yes            | NA                  | Yes                             |
| 8   | Sarcoma          | 3.5x3.5            | LA/MV             | Dizziness          | Unclass. sarcoma| Yes            | Yes                 | No                              |
| 9   | Sarcoma          | NA                 | RA                | Dyspnea,CP, Dizziness| Unclass. sarcoma| Yes            | Yes                 | No                              |
| 10  | Sarcoma          | 6x7.7              | Pericardium       | Dyspnea,CP         | Unclass. sarcoma| No             | NA                  | Yes                             |
| 11  | Sarcoma          | NA                 | RV                | Dyspnea            | MFH             | Yes            | Yes                 | No                              |
| 12  | Myxoma           | 4.5x2.5            | RA                | Palpitations       | Myxoma          | Yes            | -                   | No                              |
| 13  | Paraganglioma    | 4x4                | LA                | CP                 | Paraganglioma   | Yes            | -                   | No                              |
| 14  | Paraganglioma    | 6x4                | LA                | I                  | Paraganglioma   | Yes            | -                   | No                              |
| 15  | Myxoid sarcoma   | 7.5x5              | RV                | Dyspnea            | Cardiac myxoid sar. | Yes      | Yes                 | Yes                             |
| 16  | MFH              | 3.3x2.7            | 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, transesophageal echocardiogram; CT, computed tomography; MFH, malignant fibrous histiocytoma; 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.
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were alive and 13 (65%) were dead within one year of surgery. Of the 12 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, retroperitoneum, 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 syncope episodes, as well as CHF. Finally, tumors can cause intra-myocardial and intra-cavity 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 cava obstruction.8 Physical findings such as systolic murmur, diastolic murmur, elevated jugular venous pressure and tumor plop may be present.9 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 39 (53.8%) 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, myxosarcoma, liposarcoma, mesenchymal sarcoma, neurofibrosarcoma, malignant fibrous histiocytoma are other cardiac sarcomas observed.4,5 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.16 The most common site of metastasis is lung.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 | Died within 1 year of diagnosis |
|------|--------------------------------|-------------------|-------------------|---------|-----------|---------|--------------------------------|
| 1    | 57/M Renal cell carcinoma      | NA                | RA                | I       | NA        | Yes/No  | No                             |
| 2    | 40/M Renal cell carcinoma      | NA                | LA                | I       | RCC       | Yes/No  | No                             |
| 3    | 69/M Renal cell carcinoma      | NA                | RA                | ankle edema | RCC    | Yes     | No                             |
| 4    | 72/M Renal cell carcinoma      | 4x5               | RA                | I       | RCC       | Yes     | No                             |
| 5    | 43/F Renal cell carcinoma      | NA                | RA                | Dyspnea Collecting duct ca. | Yes   | Yes    | Yes                            |
| 6    | 33/M Renal cell carcinoma      | 4x3               | LA                | I       | NA        | No      | No                             |
| 7    | 78/F Renal cell carcinoma      | 1.5x1.4           | RA                | I       | Sarcomatoid ca. | Yes     | Yes                            |
| 8    | 52/F Renal cell carcinoma      | 3.5x2.2           | LV                | I       | NA        | No      | Yes                            |
| 9    | 65/M Renal cell carcinoma      | 2.2x1.5           | 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           | 1.6x1.4           | LV                | Dyspnea | NA        | No      | Yes                            |
| 13   | 50/M Pleomorphic liposar.       | 6.3x2.7           | LV                | Dyspnea Pleomorphic sar. | Yes   | Yes    | No                             |
| 14   | 15/M Undiff. sarcoma pelvis    | 0.7x0.8           | RA/RV             | I       | Sarcoma   | Yes     | No                             |
| 15   | 31/F Endometrial sarcoma       | NA                | RA                | Dyspnea Sarcoma | Yes   | No      | No                             |
| 16   | 54/F Leiomyosarcoma (med.)     | 3.5x4.1           | RA                | I       | Leiomyosarcoma | Yes     | No                             |
| 17   | 56/F Leiomyosarcoma (IVC)      | 3.1x2.4           | RA                | I       | Leiomyosarcoma | Yes     | Yes                            |
| 18   | 56/M Malignant melanoma        | 1.9x1.9           | RA                | Dyspnea | NA        | No      | Yes                            |
| 19   | 56/M Malignant melanoma        | 2.5               | LV                | Hypotension | NA    | No      | Yes                            |
| 20   | 39/F Malignant melanoma        | 5.5x3             | LA                | Dyspnea, palpitation, syn. | Metastatic melanoma | Yes     | No                             |
| 21   | 32/M Malignant melanoma        | 7x11              | LA                | Chest pain Metastatic melanoma | Yes   | Yes    | Yes                            |
| 22   | 67/M Malignant melanoma        | 4.0x2.0           | RA                | I       | NA        | No      | No                             |
| 23   | 40/M Malignant melanoma        | 3.3x3.8           | RA                | Chest pain Metastatic melanoma | Yes     | Yes    | Yes                            |
| 24   | 59/M Sarcomatoid lung can.     | 1.3x0.6           | 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   | 2.1x2.5           | LA                | Palpitations Poorly diff. adenocarc. | Yes     | Yes    | Yes                            |
| 27   | 51/F SCC lung                  | 3x2               | LA                | Dyspnea SCC | Yes     | Yes    | Yes                            |
| 28   | 71/M SCC lung                  | 3.3x1.6           | 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    | 2.5x2.5           | LA                | Dyspnea | NA        | No      | Yes                            |
| 31   | 44/F Adenocar. breast          | 4.6x2.0           | RV                | I       | NA        | No      | Yes                            |
| 32   | 78/F Inf. ductal ca. breast    | 1.5x1.5           | RA                | Dyspnea, edema, CHF | NA    | No      | No                             |
| 33   | 56/F Carcinoid tumor           | 3.7x3.9           | RA/LA             | I       | NA        | No      | No                             |
| 34   | 71/M Carcinoid syndrome        | 3.0x3.0           | RV                | I       | NA        | No      | Yes                            |
| 35   | 49/F MPF (colon)               | 3.3x2.4           | LA                | I       | NA        | No      | No                             |
| 36   | 43/F Transitional and SCC (bl.)| 2.6x1.0           | LV                | I       | NA        | No      | Yes                            |
| 37   | 82/M Rectal adenocar.          | 2.5x4             | RA                | Dizziness, syncope | NA    | No      | Yes                            |
| 38   | 80/M SCC (tongue)              | 3.1x1.0           | RA                | Irregular heart rhythm | NA    | No      | No                             |
| 39   | 62/F Large B-cell lymphoma      | 1.0x1.0           | LV                | I       | NA        | No      | Yes                            |
| 40   | 57/M Multiple myeloma          | 3.9x4.8           | LA                | I       | NA        | No      | Yes                            |
| 41   | 43/F Leiomyoma (uterus)        | 4x3.8             | RA                | Dyspnea, palpitation Smooth muscle tum. | Yes     | Yes    | No                             |
| 42   | 42/F Smooth cell tumor (thigh) | 1.8x3.5           | LV                | Chest pain | NA    | No      | No                             |
| 43   | 63/M Carcinoid neuroendocrine tumor | 3.5x4.3 | 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; M, mitral valve; CHF, congestive heart failure; DA, 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; MPF, malignant fibrous histiocytoma; med, mediastinum; bl, bladder.
mas and can be used as alternative chemotherapeutic agents. Other treatment regimens include ifosfamide-epirubicin (doxorubicin) and cyclophosphamide, vincristine, doxorubicin and dacarbazine (CyVADIC).

Unlike other sarcomas, cardiac sarcomas have a very poor prognosis with a median survival rate of 6-12 months after diagnosis.9,15 Presence of tumor necrosis and metastases is associated with a poor prognosis. A recent study showed that 14.8% of the resected tumors were low grade and all the patients were alive at follow up. 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. 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. In comparison to older series, there is a significant increase in the incidence of cardiac metastases (CM) in cancer patients after 1970. 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. 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 epidermal (34%), myocardial (32%) and endocardial metastases (5%). CM 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) |

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

| Clinical symptoms            | P+S (38) | P (14) | S (24) |
|------------------------------|----------|--------|--------|
| 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) |
| Atrial edema                 | 4 (10)   | 0      | 4 (17)  |
| Dizziness                    | 3 (8)    | 2 (14) | 1 (4)   |
| Syncope                      | 2 (5)    | 0      | 2 (8)   |
| Hypotension                  | 1 (3)    | 1 (7)  | 0       |

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

| Author et al. Year, CM cases | Most common N (%) | 2nd most common primary N (%) | Chamber/side involved |
|------------------------------|--------------------|-------------------------------|-----------------------|
| 1 Bussani et al. 28          | Lung cancer 263 (38%) | Leukemia/malignant lymphoma 67 (10%) | NA                    |
| 2 Hanfling 95                | Leukemia 34 (28%) | Lymphoma 27 (22%) | NA                    |
| 3 Cates et al. 30            | Lung cancer 25 (53%) | Leukemia/lymphoma 5 (11%) | NA                    |
| 4 Butany et al. 11           | Lung cancer - | Leukemia/multiple myeloma - | NA                    |
| 5 Rafajlović et al. 52       | Lung cancer 18 (23%) | Leukemia/malignant lymphoma 16 (20%) | LV                    |
| 6 Abraham et al. 33          | Lung cancer - | Lymphoma - | NA                    |
| 7 Karwinski et al. 14        | Lung cancer 60 (46%) | Malignant melanoma 13 (10%) | NA                    |
| 8 Goudie 15                  | Lung cancer 85 (67%) | Lymphadenoma/reticular sarcoma 10 (8%) | Lt side (37%), Rt side (28%), Both (35%) |
| 9 Lockwood et al. 34         | Lung cancer 108 (60%) | Kidney 14 (8%) | NA                    |
| 10 Silvestri et al. 37       | Melanoma 48 (12%) | Lung cancer 29 (29%) | NA                    |
| 11 Abioye et al. 38          | Burkitt’s lymphoma 30 (31%) | Hodgkin’s disease 4 (8%) | RA (31,25%) |
| 12 De Losach et al. 20       | Lung cancer 22 (16%) | Reticulum cell sarcoma 18 (13%) | Rt side (86%), Right ventricle (40%) |
| 13 Mac Gee et al. 40         | Lung cancer 20 (40%) | Breast cancer 11 (22%) | LV (77%) |
| 14 Manoijovik 41             | 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.
Radiotherapy is the treatment of choice.

J and R was most frequently involved.

Nom was the most common secondary quality of life.

Tumor while metastasis from renal cell carcinoma was the most common primary cardiac tumor. 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).

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. Congestive heart failure, cardiac arrhythmias, heart blocks, acute myocardial infarction, myocardial rupture and systemic embolization are other manifestations of CM.

A new heart murmur of stenosis or any new ECG finding without any symptoms in a cancer patient should raise the suspicion of CM. 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. The ECG findings of myocardial injury have high specificity for CM.

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. 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. Orthotropic heart transplantation is an option in selected patients, with improved survival.

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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