

Cardiac Sarcomas

An Update

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Abstract: Primary cardiac sarcomas are rare and represent 20% of all primary cardiac tumors. Symptoms depend on the chambers and the cardiac structures involved. Transthoracic echocardiography is commonly used to identify a cardiac mass. The diagnosis of cardiac sarcoma requires adequate sampling and the careful use of ancillary diagnostic techniques. In the most recent histologic classification, angiosarcoma is the most common malignant tumor of the heart with recognizable differentiation. Undifferentiated sarcomas account for one-third of all cardiac sarcomas and have been incorporated in the malignant fibrous histiocytoma/pleomorphic sarcoma subgroup. Elective cardiac sarcoma therapy includes complete surgical excision when possible, followed by radio- and chemotherapeutic regimens, the latter preferably containing anthracyclines, ifosfamide, or taxanes. Prognosis of cardiac sarcomas is very poor, with mean survival ranging from 9.6 to 16.5 months. A less-aggressive course seems related to the left atrium location, a low histologic grading with scarce cellular pleomorphism and low-mitotic activity, absence of necrosis, myxoid tumor appearance, and absence of metastasis at diagnosis.

Key Words: Tumors of the heart, Classification, Diagnosis, Therapy, Pleomorphic sarcoma, Angiosarcoma.

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Primary cardiac sarcomas are malignant neoplasms deriving from mesenchymal cells and confined to the heart at the time of diagnosis. Most cardiac sarcomas have the same histologic appearance similar to their soft tissue counterpart. Consequently, an extremely rare metastasis from a preexisting noncardiac sarcoma should be excluded. Nevertheless, primary heart sarcomas are exceptionally rare. In a large study by the American Medical Association, prevalence of primary cardiac sarcomas at autopsy was 0.0017%,¹ with an incidence from 0.001 to 0.030%² and a general incidence of 0.07%.³ Cardiac metastases at autopsy occur 20 to 40 times

more frequently than primary neoplasms⁴ and originate mainly from cutaneous melanoma, lung, and breast carcinomas. Sarcomas represent approximately 10 to 20% of total primary cardiac tumors^{5,6} and occur without any gender predilection. Cardiac sarcomas may arise in any area of the heart, although angiosarcoma is typically found in the right atrium,⁷ whereas fibrosarcoma and undifferentiated sarcoma in the left atrium.^{5,8} Transthoracic echocardiography is the most common method of instrumental investigation.⁹ Elective therapy includes complete surgical excision when possible, followed by radio- and chemotherapy. Despite these therapeutic strategies, prognosis of cardiac sarcomas is very poor, with median survival of less than 1 year.¹⁰

CLASSIFICATION

For many years, no universally accepted classification of primary cardiac sarcomas existed. The difficulty derived from the rarity and lack of systematic studies, with only a few series of cardiac sarcomas reported in the literature. Virtually, all soft tissue sarcoma types have also been found to arise in the heart. However, it remains difficult to apply the same histologic classification, because most cardiac sarcomas have limited areas with morphologically recognizable differentiation. Moreover, despite a careful immunohistochemical study, they frequently lack tissue-specific antigens. The recent World Health Organization classification system of cardiac tumors proposed in 2004¹¹ is largely based on the soft tissue classification counterpart and only the most frequent malignant entities are listed (Table 1). Major changes compared with previously reported classification include the incorporation of undifferentiated sarcoma, previously considered a separate entity,⁵ in the malignant fibrous histiocytoma (MFH)/undifferentiated pleomorphic sarcoma subtype. Moreover, malignant mesenchymoma, a sarcoma showing two or more lines of differentiation, is no longer mentioned. In accordance with the soft tissue tumors classification, malignant mesenchymomas would nowadays be recognized as examples of other better defined malignant tumors, such as dedifferentiated liposarcoma, malignant peripheral nerve sheath tumor, or undifferentiated pleomorphic sarcoma displaying heterologous elements.¹² Undifferentiated sarcomas and angiosarcoma are the most frequent primary malignant tumors of the heart, accounting for up to 66% of all sarcomas in the two large series published in 1996⁵ and 2004.¹¹ MFH/undifferentiated pleomorphic sarcoma accounts for more than one-third

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TABLE 1. Classification of Cardiac Sarcomas

Recognizable Line of Differentiation	Uncertain Line of Differentiation
Angiosarcoma	Pleomorphic malignant fibrous histiocytoma/pleomorphic undifferentiated sarcoma
Fibrosarcoma (and myxosarcoma)	
Rhabdomyosarcoma	
Leiomyosarcoma	
Liposarcoma	Synovial sarcoma

Modified from cardiac sarcoma classification in "tumors of lung, heart and pericardium," edited by WHO in 2004.

of all malignant tumors of the heart¹¹ but only 5% of all cases are soft tissue tumors.¹² In the 2004 World Health Organization classification of cardiac sarcomas, exceptionally rare entities such as chondrosarcoma and osteosarcoma are no longer mentioned. A histologic revision of the few described cases of cardiac chondrosarcoma revealed the paucity of this finding and the presence of other better morphologically defined sarcomas.¹¹ Osteosarcomas of the heart are preferably reported as cases of undifferentiated pleomorphic sarcomas with prominent osteosarcomatous differentiation.¹¹ Finally, myxofibrosarcoma has been classified as a myxoid variant of fibrosarcoma.¹¹

Histogenesis and Genetic Features of Cardiac Sarcomas

The classic histogenetic hypothesis presumed that most cardiac sarcomas derive from a pluripotential mesenchymal cell.⁵ At present, molecular histogenesis of cardiac sarcomas is poorly known, and there are no specific genetic mutations reported. This is likely due to the rarity and consequent absence of a systematically characterized large series of cardiac sarcomas. *K-ras* mutation and the absence of p53 mutations have been reported in three cases of angiosarcomas and two cases of rhabdomyosarcomas of the heart.¹³ Naka et al.¹⁴ reported p53 mutations in two primary cardiac angiosarcomas. Finally, synovial sarcomas typically harbor t(X; 18; p21.2;q11.2) resulting mainly in SS18-SSX1 fusion transcripts, included in the examined case of cardiac localization.¹⁵

Clinical Presentation

Cardiac sarcomas may arise at any age without gender predominance. Reported mean age at clinical presentation is 41 years.⁸ At diagnosis, most patients are symptomatic and two-third in class III/IV of the New York Health Association functional classification system for staging heart failure. One-third of patients with primary cardiac sarcomas show metastasis at diagnosis.¹⁶ Symptoms depend on the cardiac structures or chambers involved in tumor growth. Dyspnoea and orthopnoea from pulmonary venous hypertension and frank pulmonary edema are the most common symptoms; systemic embolization is also quite frequent.¹⁷ Other symptoms include fever, arrhythmias, chest pain, pericardial tamponade, and congestive heart failure. Although found in all chambers of the heart, 50% of cardiac sarcomas are located in the left

atrium, in particular, 75% of leiomyosarcomas¹⁸ and 81% of undifferentiated pleomorphic sarcomas.¹⁹ Rare previously reported cardiac osteosarcomas²⁰ and malignant mesenchymoma²¹ were also invariably observed to origin within the left atrium. Exceptionally, rare biatrial involvement from undifferentiated sarcomas has been reported.²² For all these tumors, dyspnoea is the most common symptom. Angiosarcoma is typically observed in the right atrium of male patients^{7,23} and symptoms include dyspnoea, pulmonary edema, liver and spleen enlargement, and ascitis. Because of the propensity of cardiac angiosarcoma to invasion, pericardial constriction and cardiac tamponade are frequent, and lung, bone, liver, and spleen metastasis is reported⁷; malignant pericardial effusion can be the first clinical evidence of a cardiac sarcoma.²⁴ Rhabdomyosarcoma may arise throughout the entire myocardium, mostly in male children.²⁵ Finally, liposarcoma may involve both atria.²⁶

Imaging

Most cardiac sarcomas are diagnosed by transthoracic echocardiography, eventually followed by a presurgical transesophageal echocardiography.^{10,27} The presence of invasion and metastasis is documented by total body computed tomography (CT) and magnetic resonance imaging (MRI). These techniques provide useful information on tumor size and localization and involvement of cardiac anatomic structures, with particular reference to valves. MRI after contrast enhancement allows three-dimensional high-resolution tomography of the thoracic cavity including mediastinum, lung and large vessels, and tumor invasion of pericardic, intramural or intracavitary space can be distinguished. Finally, MRI better defines the lipomatous, myxoid, and thrombotic composition of an intracardiac mass.^{28,29} Coronary angiography is performed in patients with symptoms of coronary artery disease and/or possible coronary involvement by a cardiac mass. The combination of echocardiography, CT, or MRI generally provides an excellent anatomic definition for preoperative planning.

Histopathologic Diagnosis

An exhaustive sampling of a surgical specimen clinically suspected to be a cardiac sarcoma is needed to reach the correct diagnosis and grading. All areas that appear macroscopically heterogeneous (i.e., solid, myxoid, hemorrhagic, and necrotic areas) should be sampled and microscopically examined. If necessary, ultrastructural examination can be performed. Microscopic evaluation helps to recognize important diagnostic features, such as architectural (storiform, herringbone, whirling, or diffuse), cellular (spindle, small, large, and pleomorphic cells), and microvasculature pattern (curvilinear, hyaline, or chicken wire), the presence of stromal reaction (myxoid or sclerotic), hemorrhage, and necrosis if surgical resection was not preceded by therapeutic regimens (see later). At higher magnification, cytologic atypia and specific differentiation (i.e., the presence of rhabdomyoblasts, pseudolipoblasts, or blunt-ended cigar-shaped nuclei) should be recognized. It is worth noting that primary cardiac tumors can be polymorphic with heterogeneous areas of differentiation. Finally, a careful immunohistochemical study is useful

TABLE 2. Immunohistochemical Panel of Main Antibodies for the Diagnosis of Cardiac Sarcomas

	CD34	FVIII	CD31	S100	α -SMA	Desmin	CK	Vim	Myo
Angiosarcoma	+	+	+	-	-	-	R ^a	+	-
Undifferentiated pleomorphic sarcoma	-	-	-	-	R	-	R	+	-
Rhabdomyosarcoma	-	-	-	-	+	+	-	+	+
Fibrosarcoma	-	-	-	-	R	-	-	+	-
Leiomyosarcoma	-	-	-	-	+	+	R	-	+
Synovial sarcoma	-	-	-	-	R	-	+	+	-
Liposarcoma	-	-	-	+	-	-	-	+	-

^a Diffusely positive in epithelioid variant.

FVIII, factor VIII; α -SMA, α -smooth muscle actin; CK, cytokeratins; Myo, myogenin; Vim, vimentin; R, rare and focal; +, positive; -, negative.

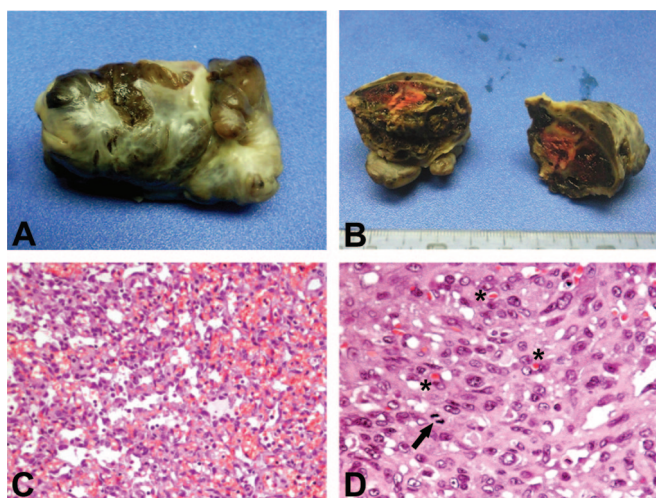


FIGURE 1. A, Example of cardiac angiosarcoma; (B) macroscopically, cut surface appears reddish-brown from alternating solid and hemorrhagic areas. Microscopically, the tumor shows (C) well-differentiated areas with vascular channel lined by atypical endothelial cells, and (D) solid areas from accumulation of large epithelioid cells with marked pleomorphism, mitosis (arrow) and atypical cells with vacuolized cytoplasm containing erythrocytes (*) (Hematoxylin and Eosin).

to reach a correct diagnosis and grading, in particular, of differentiated cardiac sarcomas. A panel of main immunohistochemical reactivity is schematically reported in Table 2.

Angiosarcoma

Angiosarcoma is typically found in the right atrium.^{7,30} Macroscopically, angiosarcoma appears as a gray-brown mass with hemorrhagic areas (Figures 1A, B). In two-third of cases, angiosarcoma is microscopically characterized by vascular differentiation with channels covered by endothelial cells exhibiting marked atypia (Figure 1C); endothelial papillary structures can be also observed. Solid areas of spindle or epithelioid cells with vacuoles containing erythrocytes can be detected (Figure 1D). Atypia and mitosis are fundamental in the differential diagnosis with other tumors sharing a vascular differentiation, such as hemangioma and heman-gioendothelioma. In some cases, angiosarcomatous areas with spindle cell pattern must be differentiated from those

present in fibrosarcoma, MFH, and Kaposi sarcoma. The latter can involve the heart, as reported in 17 to 19% of classic, endemic, and AIDS-related cases.³¹ Immunodetection of factor VIII or CD34 positive neoplastic cells represents useful markers of endothelial differentiation; moreover, cytokeratins can be diffusely positive in epithelioid areas of angiosarcoma.³²

Unclassified Sarcoma/Pleomorphic MFH

A cardiac sarcoma with no specific histologic pattern is classified as undifferentiated.⁵ Nowadays, undifferentiated sarcoma is considered synonymous with pleomorphic MFH.^{11,12} Undifferentiated sarcomas have previously been distinct in pleomorphic, epithelioid, or small-cell subtypes.⁵ Macroscopically, MFH appears yellowish-white with areas of necrosis and widely infiltrating myocardium and cardiac structures (Figures 2A, B). Microscopically, unclassified sarcoma/pleomorphic MFH displays great heterogeneity in appearance and variable cellularity, with alternated areas of spindled and epithelioid cells, sometimes with abundant eosinophilic cytoplasm and intermixed giant cells (Figures 2C, D). The presence of storiform arrangement, marked pleomorphism, high mitotic activity, and foci of necrosis is commonly reported. In some cases, undifferentiated sarcomas are characterized by the presence of small cells (Figure 2E) with phenotypical and ultrastructural features of embryonal or primordial mesenchymal cells and no further recognizable differentiation.²² Chondromatous differentiation in MFH/pleomorphic sarcoma can also be detected (Figure 2F), but the term chondrosarcoma is no longer reported in the classification of cardiac sarcomas and a diagnosis of MFH/pleomorphic sarcoma with heterologous elements is preferable. Nevertheless, the recognition of mature cartilaginous tissue in a cardiac tumor is diagnostic of malignancy.¹¹ The finding of true primitive osteosarcomas of the heart is debated. When osteosarcomatous areas are present, the diagnosis of MFH/pleomorphic sarcoma with osseous differentiation is preferred.¹¹ Undifferentiated sarcoma must be distinguished from embryonal rhabdomyosarcoma and metastatic small cell cancer. Immunostaining is crucial, being epithelial, neural, or endothelial markers are usually negative and vimentin typically positive; high-grade undifferentiated sarcomas can exhibit focal α -smooth muscle positive areas, but the latter are generally limited in their extension.³²

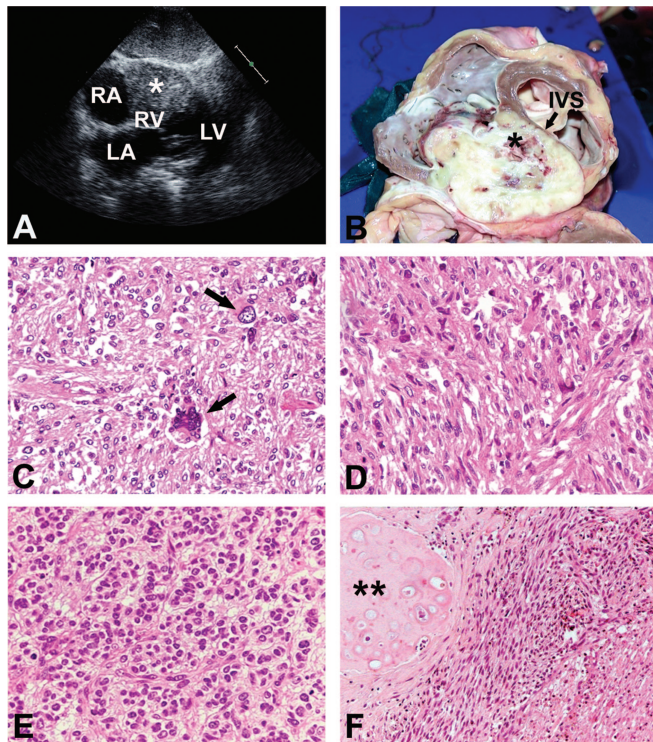


FIGURE 2. *A*, Two-dimensional echocardiography reveals a large undifferentiated pleomorphic sarcoma/MFH (*) occupying the right ventricle (RV). *B*, Autoptic examination of the heart of the patient shows a white-yellowish mass with areas of necrosis (*) widely infiltrating the myocardium and the interventricular septum (IVS). *C* and *D*, Histologically, the undifferentiated pleomorphic sarcoma/MFH is constituted by highly pleomorphic spindle or epithelioid cells with abundant eosinophilic cytoplasm and interspersed giant or multinucleated cells (arrows) arranged in vaguely storiform fashion. *E*, Undifferentiated pleomorphic sarcoma/MFH, small-cell subtype. *F*, Undifferentiated pleomorphic sarcoma/MFH with areas of chondroid differentiation (**). (Hematoxylin and Eosin). LA, left atrium; LV, left ventricle; RA, right atrium; MFH, malignant fibrous histiocytoma.

Leiomyosarcoma

Leiomyosarcoma is a malignant tumor showing phenotypic and ultrastructural smooth muscle differentiation. It has been described in all cardiac chambers. When located in the left atrium, it can mimic myxosarcoma.³³ Leiomyosarcoma is microscopically characterized by the presence of a bundle of atypical spindle cells with characteristic blunt-ended or “cigar-shaped” nuclei (Figures 3*A, B*) and often Periodic Acid-Schiff-positive intracytoplasmic vacuoles. Pleomorphic areas, necrosis, and multinucleated cells may be encountered. Immunostaining for desmin and smooth muscle actin are usually diffusely positive (Figures 3*C, D*), whereas epithelial, vascular, and neural markers are negative.³⁴

Fibrosarcoma and Myxosarcoma

Fibrosarcoma by definition is microscopically composed of atypical elongated fibroblast-like cells with interspersed collagen and the classic herringbone pattern.¹¹ Fibro-

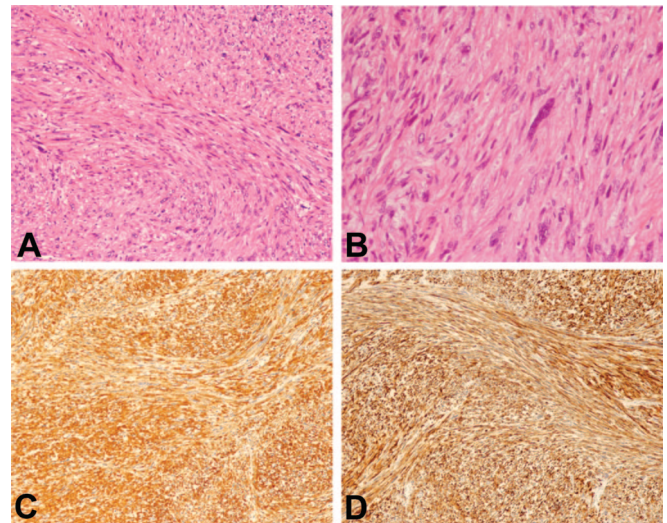


FIGURE 3. *A* and *B*, An example of cardiac leiomyosarcoma, microscopically composed of variably oriented bundles of pleomorphic spindle cells with blunt-ended nuclei (Hematoxylin and Eosin). Immunohistochemistry reveals the presence of diffuse (C) α -smooth muscle actin and (D) desmin-positive cells.

sarcoma is frequently located in the left atrium, mainly in young-adult women.⁸ Myxofibrosarcoma, previously considered a myxoid variant of MFH, has now been moved to the fibroblastic categories^{11,12} and considered a variant of fibrosarcoma. Myxofibrosarcoma is characterized by a broad spectrum of microscopic appearance, with hypocellular areas with spindle hyperchromatic cells, abundant myxoid matrix (Figure 4*A*), prominent elongated, curvilinear thin-walled blood vessels (Figure 4*B*) alternated with hypercellular areas, showing a herringbone growth pattern (Figure 4*C*), similar to classic fibrosarcoma. When hypocellular myxoid areas are diffuse, differential diagnosis with cardiac myxoma can be difficult. An exhaustive examination is needed to recognize the presence of areas with nuclear atypia and mitosis. The absence of typical myxomatous “ring structures” is also requested for a correct diagnosis of myxosarcoma.

Recently, a presumed primary cardiac low-grade fibromyxoid sarcoma has been reported.³⁵ This is a rare tumor arising in the soft tissue of proximal extremities, trunk, and thoracic cavity. Because low-grade fibromyxoid sarcoma seems to have a more favorable course compared with classic fibrosarcoma, it must be included in its differential diagnosis.

Rhabdomyosarcoma

By definition, a malignant tumor showing striated muscle differentiation is classified as rhabdomyosarcoma.¹¹ It usually arises in the ventricular wall,⁸ rarely in the myocardium of the left atrium.³⁶ Unlike the extracardiac counterpart, primary cardiac rhabdomyosarcoma is almost always of embryonal type and occurs in children or young people. Rhabdomyosarcoma is microscopically composed of small cells with the presence of recognizable rhabdomyoblasts. Masson’s trichrome revealing the characteristic cell cross-stria-

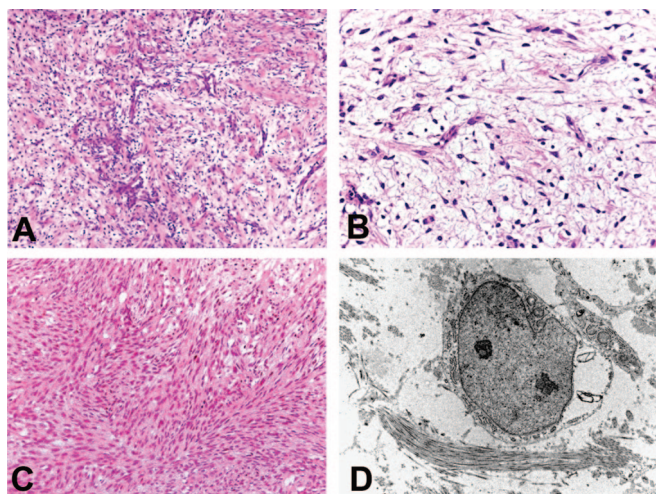


FIGURE 4. An example of cardiac myxosarcoma, microscopically characterized by (A) prominent elongated, curvilinear thin-walled blood vessels and (B) hypocellular areas with spindle hyperchromatic cells embedded in abundant myxoid matrix; (C) in other areas, myxosarcoma seems hypercellular with herringbone-like growth pattern (Hematoxylin and Eosin). D, Ultrastructural features of a myxosarcomatous cell, with scarce cytoplasm, prominent nucleoli, and collagen bundles intimately associated to the plasmalemma (transmission electron microscopy).

tions and a diffuse desmin-positive immunostaining help for a correct diagnosis.^{8,36} Rhabdomyosarcoma can express other muscle markers, including myogenin, myoD1, sarcomeric actin, muscle-specific actin, and myoglobin.³⁷ Among these, myogenin and myoD1 seem to be more specific.³⁸

Synovial Sarcoma

Synovial sarcoma is a biphasic malignant tumor usually located in the atria or pericardial surface. The latter localization should be considered for distinction from malignant mesothelioma. In the heart, the monophasic variant of synovial sarcoma is more common. A differential diagnosis between malignant mesothelioma and synovial sarcoma may be challenging on the basis of morphologic aspect and immunophenotype, both being positive for cytokeratins, calretinin, and vimentin. Nevertheless, distinction may be reached considering that malignant mesothelioma does not occur in the atria and generally infiltrates the heart, whereas synovial sarcoma usually seems localized as a rather circumscribed mass.¹¹ Synovial sarcoma must also be differentiated from solitary fibrous tumor, which usually expresses CD34. Synovial sarcoma typically harbors t(X; 18; p21.2;q11.2) resulting mainly in SS18-SSX1 fusion transcripts.¹⁵ This cytogenetic feature represents a useful diagnostic tool and can be detected by using reverse-transcription polymerase chain reaction or fluorescence in situ hybridization techniques.

Grading

As for their soft tissue counterpart, histologic grading of malignancy of cardiac sarcomas has the greatest correlation with survival,³⁹ with the important exception of angio-

sarcoma. Although two-third of cases of cardiac angiosarcoma display a variable evidence of vascular differentiation with well-formed vascular channels and papillary structures, this tumor is highly invasive, with a mean survival of few months.¹¹ The French Federation Nationale des Centres de Lutte Contre le Cancer (FNCLCC) classification is the grading system that seems to better correlate with survival of soft tissue sarcomas. The FNCLCC system score (grade 1–3) is obtained by evaluating three main histologic parameters of the tumor (i.e., differentiation, mitotic rate, and percentage of necrosis).⁴⁰ The FNCLCC grading is applicable for untreated sarcomas and requires an exhaustive postsurgical tissue sampling.

Therapy and Prognosis

The gold standard therapy for eligible patients with cardiac sarcoma without metastasis at diagnosis is a complete surgical removal of the tumor. Patients undergo operation by way of median sternotomy with minimal touch technique and cardiopulmonary bypass under cardioplegic arrest.⁴¹ Unfortunately, a complete surgical resection is often complicated because of the diffuse invasion of cardiac structures, or almost impossible for tumor localization in those anatomic sites that require extremely complex reconstruction. Pericardial patches are used to replace removed atrial wall or large vessel segments, and sometimes valve replacement is also required.^{41–43} If complete surgical resection of cardiac sarcoma is impossible, more than 90% of patients die within 1 year despite any postsurgical therapy.⁴⁴ When extracardiac metastasis is identified, surgical resection is reserved for palliative indication. In a series of 34 cases from Mayo Clinic, median overall survival was 17 months for patients undergoing complete surgical excision and 6 months for those in whom complete tumor removal could not be performed, and the difference was statistically significant.⁴⁵ Finally, cardiac transplantation can be also considered, because evidence of long-term benefit in selected patients has been reported.^{46,47}

There is no general agreement on efficacy of postsurgical therapy for cardiac sarcomas. Chemotherapy after surgical resection is recommended, even when clear surgical margins are obtained because of the high probability of missed microscopic disease and likely aggressive course.⁴² Other authors did not find any evidence supporting adjuvant chemotherapy as beneficial.⁴⁸ Because there is an absence of specific clinical trials due to the rarity of the localization for eligible patients, chemotherapy protocols are derived from extracardiac soft tissue counterpart data. The most common regimen for soft tissue sarcoma treatment is combined doxorubicin and ifosfamide but anthracycline chemotherapy is both cardiotoxic and a radiation sensitizer.⁴⁹ Nevertheless, an initial large meta-analysis of adjuvant chemotherapy trials has shown no survival benefit of adjuvant chemotherapy for adult soft tissue sarcomas.⁵⁰ Recently, gemcitabine activity in soft tissue sarcomas has been described.⁵¹ Gemcitabine and docetaxel in a schedule-dependent combination provided promising results in patients with leiomyosarcoma⁵² and with other soft tissue sarcomas.⁵³ Weekly paclitaxel has been shown to be highly effective in angiosarcomas, with reported

overall tumor control rate of 70%.⁵⁴ Nakamura-Horigome et al.⁵⁵ reported the case of a 49-year-old man with a primary cardiac angiosarcoma of the right atrium and cardiac tamponade who responded to a combination therapy with docetaxel and radiotherapy and a good treatment toleration, with only grade 1 esophagitis recorded. Tumor regression was minimal on CT scan; however, fluorodeoxyglucose positron emission tomography, initially positive for tracer uptake in the tumor mass, was negative after combination therapy. No evidence of progression was documented after 12 months of follow-up. Neoadjuvant chemotherapy with cytoreductive intent can be pursued to facilitate cardiac surgery.⁵⁶

Postoperative radiotherapy is an effective adjuvant to complete excision for extracardiac soft tissue sarcomas.⁵⁷ Its value in the treatment of cardiac sarcomas is limited by the sensitivity of the heart to radiation injury; in fact, exposure of ventricular wall to a radical radiation dose can cause serious cardiomyopathy and/or chronic pericarditis.^{42,58–62} Therefore, the role of radiotherapy in the adjuvant setting or as primary treatment is uncertain, because rarely used and only anecdotal responses are reported.^{63,64} In one case, Movsas et al.⁶³ documented the complete response of an unresectable high-grade sarcoma with hyperfractionated radiotherapy (total dose of 7050 centigrays) with concomitant administration of iododeoxyuridine as radiosensitizer. Nevertheless, the overall prognosis for primary cardiac sarcomas remains very poor, with reported mean survival of 16.5 and 9.6 months in two series of 24 and 17 cases, respectively.^{16,65}

CONCLUSION

For their intrinsic rarity, diagnosis, classification, and treatment of primary cardiac sarcomas are largely based on their soft tissue counterparts. Additional studies, including biomolecular and genetic characterization, are needed to better define the origin and biology of these highly aggressive tumors. Collection and analysis of larger series will ameliorate and better address therapeutic strategies to improve prognosis of cardiac sarcomas.

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