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Review

Masses in right side of the heart: spectrum of imaging findings

Silvia Pradella[1], Giulia Grazzini[1], Mayla Letteriello[1], Cristian De Amicis[1], Roberta Grassi[2], Nicola Maggialetti[3], Mattia Carbone[4], Pierpaolo Palumbo[5], Marina Carotti[6,7], Ernesto Di Cesare[8], Andrea Giovagnoni,[6,7] Diletta Cozzi[1], Vittorio Miele[1]

[1]Department of Radiology, Careggi University Hospital, Florence, Italy; [2]Department of Precision Medicine, University of Campania "Luigi Vanvitelli", Naples, Italy; [3]Department of Medicine and Health Sciences "V. Tiberio", University of Molise, Campobasso, Italy; [4]Department of Radiology, S. Giovanni and Ruggi D'Aragona Hospital, Salerno, Italy; [5]Department of Biotechnology and Applied Clinical Sciences, University of L'Aquila, L'Aquila, Italy; [6]Department of Clinical, Special and Dental Sciences, University Politecnica delle Marche, Ancona, AN, Italy; [7]University Hospital "Umberto I-Lancisi-Salesi", Department of Radiology, Ancona, Italy; [8]Department of Life, Health & Environmental Sciences, University of L'Aquila, L'Aquila, Italy

Summary. Primary heart tumors are rare, benign tumors represent the majority of these. If a cardiac mass is found, the probability that it is a metastasis or a so-called "pseudo-mass" is extremely higher than a primary tumor. The detection of a heart mass during a transthoracic echocardiography (TE) is often unexpected. The TE assessment can be difficult, particularly if the mass is located at the level of the right chambers. Cardiac Computed Tomography (CCT) can be useful in anatomical evaluation and Cardiac Magnetic Resonance (CMR) for masses characterization as well. We provide an overview of right cardiac masses and their imaging futures. (www.actabiomedica.it)

Keywords: Cardiac masses, right heart, tumors, MRI, CT

Right sided cardiac masses: introduction and overview of imaging modalities

Primary cardiac tumors are very rare and even less common are the malignant ones (about 25 percent of all) (1). Primary cardiac neoplasms count approximately 0.02 to 0.1% of autopsy (1, 2). In young people rhabdomyoma and fibromas are most common although in adults myxomas and sarcomas are more frequent(3, 4).Approximately 75% of myxomas arise from left atrium (LA) and only about 20% of cases from right atrium (RA), whereas cardiac sarcomas, most frequently diagnosed as angiosarcoma, occur predominantly in the right atrium (5). As reported in literature, intracardiac masses are most likely to be found in the LA and more unlikely in the right ventricle (RV) (1). Neoplastic masses include benign, malignant tumors and metastatic tumors (6). Metastases are the most common cardiac neoplasms (7). Counting all cardiac masses, thrombus is the most common entity (8). Clinical manifestations of heart masses depend on the size and location of the tumor and the infiltration of adjacent structures rather than the nature of the tumor itself (9). Cross-sectional imaging as Magnetic Resonance (MR) and Computed Tomography (CT) techniques, gained large application in the day practice, particularly in cardiovascular radiology. Right-sided cardiac masses do not have a uniform clinical presentation. Pulmonary embolization may occur as the initial clinical manifestation for right myxoma (10-29). Non-invasive imaging plays a central role in the diagnosis and management pathway of cardiac masses (8). Transthoracic echocardiography (TE) remains the first-line modality of investigation to evaluate a cardiac mass because of its availability, capability to get dynamic assessment of lesions, non-invasiveness, absence

of contrast material or radiation exposure (30). However, echocardiography provides limited assessment of soft-tissue characteristics and extracardiac structures, it may be limited by poor acoustic windows, particularly for right chambers evaluation mostly in obese patients and the uncooperative ones.

Different imaging modalities are available for anatomical evaluation and characterization of tissues and masses in various district, including the heart (31-50). Multimodality imaging such as cardiac MR prevents misdiagnosis which often leads to unnecessary cardiac surgery (51). Cardiac CT (CCT), CMR, and 18F-FDG PET/CT work synergistically with echocardiography. Non-invasive imaging can distinguish tumor from "tumor like" mass and highlight typical characteristics of benign tumors differentiating them from malignant ones (8). CMR is fundamental to differentiate malignant from benign tumors through the use of multiparametric sequences (52). CCT has a limited role in tissue characterization compared to the CMR imaging; but both can be used synergistically with echocardiography to detect the mass, define its exact location and its anatomical relationships with adjacent structures(6). Appropriate Use Criteria for CMR and CCT provide specific guidance on the approach to cardiac masses. Since CMR does not require the use of ionizing radiation, it is the modality of choice particularly for pediatric patients, along with echocardiography(4). CMR quality images is dependent on patient cooperation. Additionally, CMR is specifically contraindicated in patients with claustrophobia and implanted unsafe magnetic devices. Also, CMR may be inadequate for evaluating small mobile masses (e.g., papillary fibroelastoma or valvular vegetations) due to limitations in spatial resolution. CCT provides an optimal anatomical evaluation and, in the presence of ECG-gating, a detailed assessment of the coronary arteries, fundamental prior to surgery. ECG gating, however, it is used to reduce motion artifacts, which can preclude detecting small lesions and cause blurring of the margins of larger lesions. Prospective ECG-gating is preferred because it allows lesion mobility through the reconstruction of cine images. To obtain an appropriate opacification in the right heart chambers (not so dense as to create streak artifacts) is indicated an intravenous infusion of iodinated contrast medium (for adults about 60-70 mL at 5 mL/s) followed by a 50%:50% contrast: saline flush. A delayed phase (obtained 2–3 min later without additional contrast injection) can be performed to help with tissue characterization. Considering that the radiation dose is very low with the latest CT generation, the CT can be a valid alternative to CMR (53, 54). Transesophageal echocardiography (TEE), like three-dimensional studies, can be a useful tool in addition to TE to clarify cardiac mass lesions characteristics in selected patients (8).

Right sided cardiac masses: lesion characteristics and imaging approach

In clinical practice, whenever a cardiac mass is detected on echocardiography, a reasonable approach is to focus on the location first, and then on the imaging characteristics **(Table 1)**. The clinical context and symptom presentation must guide us into getting the correct diagnosis. Embolic phenomena into the pulmonary circulation are an example of right sided lesions symptom onset. A reduction of cardiac output can result from an intracavity obstruction or valvular dysfunction caused by a tumor hemodynamically expressive. Generally, features that suggest a malignant lesion include myocardium and pericardium invasion resulting in arrhythmias, heart failure and pericardial effusions. Features that suggest a benign lesion include a well-defined and single mass that involves a single cardiac chamber (more frequent left than right); small size (<5 cm) and smooth margin; the presence of a narrow transition zone or pedunculated appearance; absent or minimal enhancement; absence of local invasion, metastasis and pericardial effusion; rare calcification (except for small foci in fibroma, myxoma, or teratoma) **(Table 2)** (6, 55).

Primary cardiac tumors

Benign Cardiac tumors

-Myxoma. Approximately, myxomas have an incidence of 0.0017% in the general population and they count 25–50% of all primary cardiac tumors. They are most common from the third to sixth decade of life. Myxoma can arise in the LA (75%), but they may arise

Table 1. Right sided cardiac most common masses- main characteristics

Cardiac masses	Lesion category	Age and sex	Incidence	Common location	Imaging typical features
Myxoma	Benign tumor	Adult, female-to-male ratio 2.7:1	Most common primary cardiac tumor (50% of benign tumors)	Atria (95%): left (75%) right (20%	Spherical, mobile, hyperintense on T2 w CMR images, 10 % calcified
Papillary fibroelastoma	Benign tumor	Adult, female-to-male ratio 1:1	Third in the prevalence of benign cardiac tumors	Predominantly from the aortic or mitral valve (usually tricuspid valve)	Pedunculated usually mobile, solitary, small (10 mm), smooth, hyperintense on T2 w CMR images
Lipoma	Benign tumor	Middle-aged and older adults, female-to-male ratio 1:1	About 8% of primary cardiac tumors approximately 14% of benign cardiac masses	Any chamber, intra-myocardial or intracavitary	Encapsulated, well-circumscribed, may be mobile, signal dropout on STIR CMR sequences. Low-attenuation on CT
Rhabdomyoma	Benign tumor	Children	The most common benign pediatric cardiac tumor	Left ventricle and septum (70%), right ventricle and atrial wall (30%)	Single or multiple well circumscribed, hyperintense on T2w CMR images, no or minimal enhancement
Angiosarcoma	Malignant tumor	Adult, female-to-male ratio 1:2-3	Rare, 0.0001% in autopsy series	Right atrium	Broad base, irregular, heterogeneous, infiltrative, pericardial effusion, metastatic. Heterogeneous signal on CMR: isointense on T1w and hyperintense on T2w. Heterogenous CE with a "sun ray appearance"
Lymphoma	Malignant tumor	Adult, median age 60 years old (range of 13–90 years old)	Secondary cardiac involvement by lymphoma 25% of patients, primary cardiac lymphoma 2% of cardiac primary tumors	Right atrium	Ill-defined, infiltrative (encasing adjacent structures), often pericardial effusion. On CMR: isointense on T1 w and hyperintense on T2 w. Homogenous CE
Metastases	Malignant tumors	Adult, female-to-male ratio 1:1	9 % in patients with metastatic cancer. Intracavitary metastases are rare, making up 3% to 5% of cardiac metastases	Pericardial. Any one of the heart chambers (if intracavitary)	CMR: hypointense on T1 w and hyperintense on T2 w. Heterogenous CE
Right Intracardiac thrombus	Non-neoplastic	Any age and sex; depend on underlying cardiac disorder	Intracardiac thrombi are found in about 10% of cases of pulmonary thromboembolism (PTE).	Right atrium, right ventricle, or main pulmonary artery	Intracavitary and freely mobile, especially if recently formed thrombi. No CE in early contrast phases
Prominent crista terminalis	Non-neoplastic	Any age and sex	Occasional finding	Right atrium	A well-defined fibromuscular ridge on the posterolateral wall of the right atrium

Table 2. Suggestive lesion features

Mass characteristics	Benign	Malignant
Location	Left > right	Right > left
Dimension	Small (<5 cm), single mass	Large (>5 cm), several masses
Calcification (if present)	Small	Large
Shape	Well defined margins, pedunculate	Irregular margins, broad base, signs of infiltration
Contrast Enhancement	Usually absent	Weak to intense

in other sites, such as the right side of the heart (5-20%), especially the RA (56). The sizes of these tumors vary from 1 to 15 cm and, usually, they appear as well defined, smooth and lobular mass, pedunculated.

On CMR images, myxomas have a typical heterogeneous appearance with variable iso- or hypointensity on T1-weighted sequences and hyperintensity on T2-weighted sequences due to their water content **(Fig.1).** Their heterogeneous appearance is due to the presence of intralesional hemorrhagic foci, cysts, necrosis, fibrosis, and calcification. Myxomas show heterogeneous enhancement mostly hypointense on first-pass perfusion sequences, with areas iso- or hyperintense on late gadolinium enhancement (LGE) sequences (4, 30).

On CT images myxomas appear as a lobular mass with low attenuation (usually like water attenuation values) with intralesional calcification, especially in myxoma localized in the RA. After the administration of contrast medium, they show heterogeneous contrast enhancement (CE) (6, 57).

-Papillary fibroelastoma. Papillary fibroelastoma (PF) has an incidence of up to 0.33 % in autopsy series and it is the third most common primary benign cardiac tumor (58). PFs are more frequent among the 4th and 8th decades of life. PFs are most commonly on the surface of the valves (80%) and consist of connective tissue (58). All heart valves can be affected, though the aortic valve is the most frequently involved, followed by the mitral valve. Gowda et al. (58) reported in a review of 725 cases of PFs that tricuspid valve was involved in 11% of cases, and pulmonary valve in 7%; while in the 4% of the cases PFs arise from endocardial surface of right atrium, right ventricle, right atrial appendage and Eustachian valve. Most of the patients are asymptomatic and PFs are diagnosed accidentally; however, they can be associated with valve dysfunction, embolization from attached thrombi and fragmentation (59). They are usually small (average diameter of 10 mm) and they appear as pedunculated or mobile masses (59). PFs are not always easily visible on

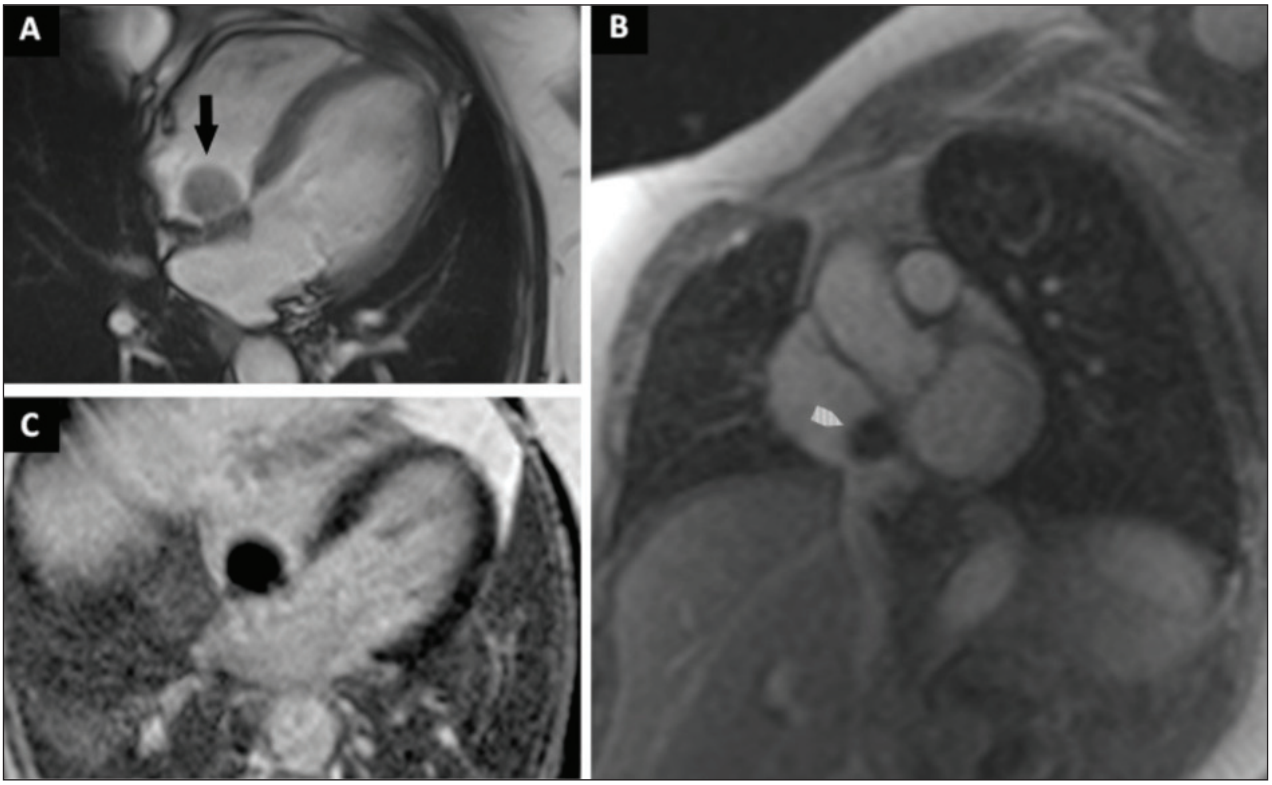

Figure 1. Right atrium myxoma- 70-year-old female. Rounded and well-defined mass (diameter about 22 mm) mobile, isointense to myocardial tissue on Trufi cine sequence (black arrow) (A), hypointense on first-pass perfusion sequences (white arrowhead) (B) and isointense to myocardial on LGE (C).

CMR due to the small size. On CMR cine-sequences they appear as small mobile and homogeneous valvular mass surrounding turbulent flow, with isointense T1 and hyperintense T2 signal intensity. Fat-saturation sequences are useful to differentiate fibroelastomas from lipomas, because the first one has no signal loss dropout. In cardiac CT, they appear small, hypodense attached through a thin stalk to valve surface (6).

-Lipoma. Lipomas represent about 8% of primary cardiac tumors and approximately 14% of benign cardiac masses, with an incidence of 0.2 and 0.4% reported in autopsy studies. Lipomas can occur in all age groups, but they're most likely to be found in the fifth and sixth decades of life(60). Lipomas are encapsulated and well-defined tumors containing neoplastic adipocytes (6). Typically, lipomas are localized in the RA and the left ventricle (LV) (60). In most cases, cardiac lipomas are discovered accidentally and the patients are asymptomatic. Rarely, very large lipomas can lead to symptomatic obstruction of the tricuspid valve or vena cava resulting in heart failure. They can be multiple in patients with tuberous sclerosis (61). On CMR they appear slightly hyperintense on T2-weighted, homogeneous hyperintense on T1-weighted, with loss of signal on fat suppression sequences, and with no CE (30). On CT imaging they appear with a homogeneous fat attenuation (density<-50 Hounsfield units), without CE (6).

-Rhabdomyoma. Rhabdomyoma is the most common benign pediatric cardiac tumor, frequently associated with tuberous sclerosis. These tumors involve children from fetal age to early infancy and tend to regress spontaneously (56). They are multiple in 90% of cases and most commonly localized in the LV and ventricular septum, although up to 30% of cases the atrial wall or RV are involved (62). These tumors may present clinically with arrhythmia and heart failure. Usually, echocardiography is adequate for making the diagnosis, but CMR can be useful in atypical presentations or for surgical planning (4). On imaging, they appear as single or multiple well-circumscribed, intramural or intracavitary masses. On CMR images, rhabdomyomas are homogeneous on all sequences, isointense to normal myocardium on T1-weighted images and hyperintense on T2-weighted images, and they show no or minimal enhancement on first-pass perfusion imaging and iso-intensity on LGE images (4). CCT is rarely performed because the affected population is mainly pediatric.

-Fibroma. Fibroma is the second most common type of pediatric cardiac tumor (63). It occurs rarely in adolescents or adults (about 15 %) (56). Cardiac fibromas typically arise in the LV (63). Although, cases of fibromas located in the RV (30%) and RA (10%) have been reported (64). The average diameter of this tumor is 5 cm and the clinical onset depends upon their location and size. On CMR imaging fibromas are slightly hypointense on T2-weighted images and isointense on T1-weighted images relative to muscle, due to their dense and fibrous nature. They have well-defined borders; little or no CE during the early phases, hyperenhancement on LGE and high extracellular volume (ECV) values (63, 64). On CT, fibroma appears as a homogeneous mass, with low attenuation and no or minimal enhancement. The characteristic presence of central calcification (patchy hypointense foci within the lesion) is useful to differentiate it from rhabdomyoma (6).

-Hemangioma. Hemangioma is a rare benign vascular tumor (2,8 % of primary cardiac tumors) that can arise from any cardiac chamber, with a predilection of RA and interventricular septum. In a review of 200 cases of hemangiomas, Weidong Li (65) reported that in the 44.1% of cases the right heart was involved (including RA, RV, tricuspid valve, and pulmonary valve). The average age of patients was 43 years old; however, pre-natal cases are also described with a range from the 20th gestational week up to 86 years (65). Hemangioma can be symptomatic in case of bleeding (56). On CMR images, hemangiomas appear heterogeneous with intermediate signal on T1 images and inhomogeneous hyperintense T2 images because of the presence of calcification and fibrous septa; after administration of gadolinium the mass shows intense enhancement due to its vascular structures. In the same way, on CT images hemangioma appears heterogeneous with intense enhancement (65).

Malignant cardiac tumors

-Sarcomas. Sarcomas are mesenchymal tumors and they are the most common cardiac primary malig-

nant neoplasms (3, 56). Usually, males are affected more often than females with a ratio of 2–3/1. The patients younger than 65 years old are the most affected (66).

Any type of sarcoma can affect the heart but angiosarcoma is the most frequent in the right chambers, especially in the RA (80% of cardiac angiosarcomas); while leiomyosarcoma, osteosarcoma and malignant fibrous histiocytoma are the most common in the left chambers. Sarcomas are generally large and they can frequently infiltrate adjacent structures (56). Hemorrhagic pericardial effusion is frequently present. Dyspnea is the most common presenting symptom; however, the patient may also present chest pain, embolic phenomena, peripheral edema, arrhythmias, syncope and sudden death (1). At the time of diagnosis patients can have metastases, most commonly to lungs, lymph nodes, bone and liver (56). CT shows the presence of large lesion, typically with a broad-based attachment and heterogenous CE because are present hemorrhagic and necrosis areas (6, 57).

On CMR T1-weighted images, the mass appears predominantly isointense to myocardium, eventually with areas of high signal due to the presence of intralesional hemorrhage. On T2-weighted images, especially angiosarcoma shows a heterogeneous predominantly hyperintense appearance **(Fig. 2)**. On LGE sequences the tumor has a "sunrise appearance" (peripheral enhancement and central areas of necrosis) (30, 67).

-Lymphoma. Cardiac primary lymphoma is generally a non-Hodgkin type. Secondary cardiac involvement by lymphoma is about 25% of patients with a disseminated lymphoma, while primary cardiac lymphoma is 2% of cardiac primary tumors. The age range of presentation is between 13–90 years old (56). Most commonly lymphomas arise from the right heart chambers, particularly the RA, followed by the RV

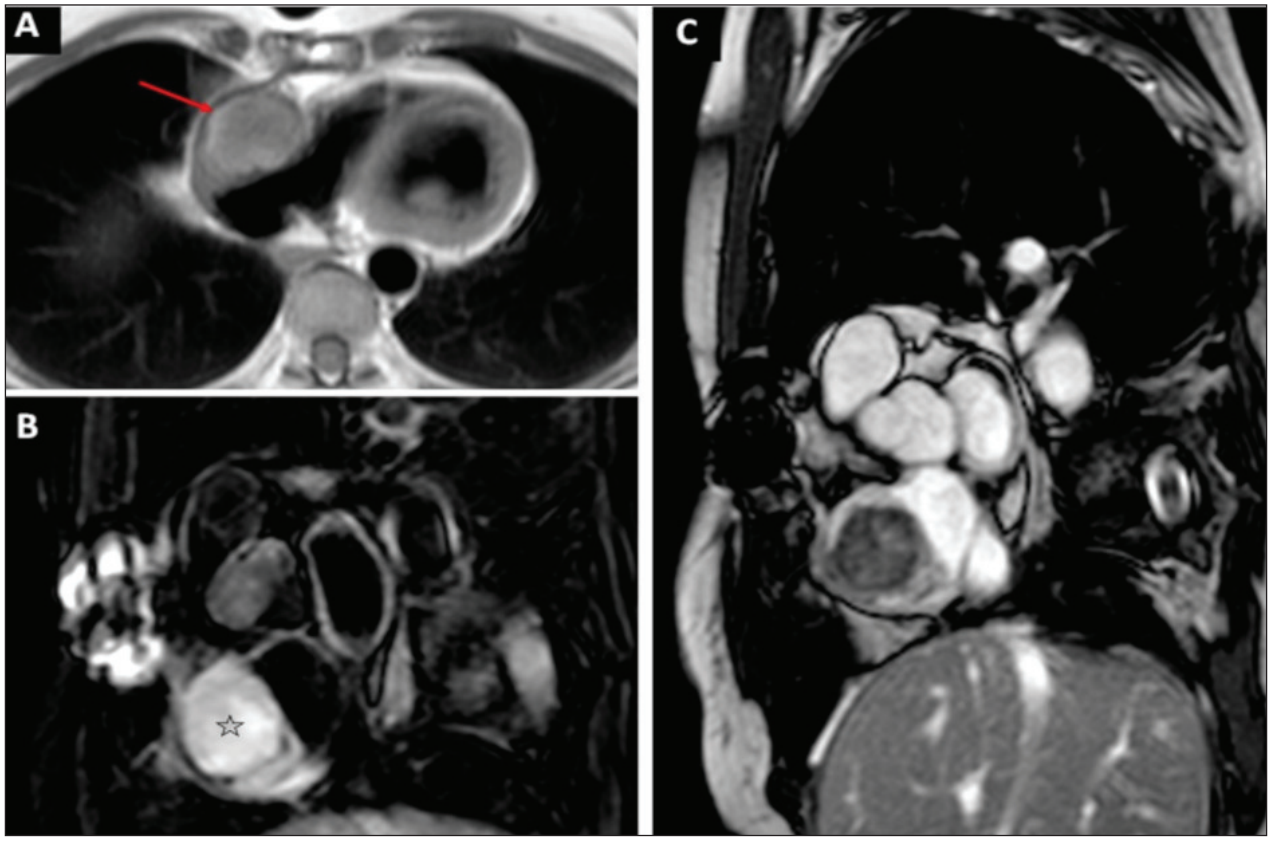

Figure 2. Right atrium angiosarcoma treated with radiotherapy- 57-year-old female. Large with a broad-based mass into the right atrium associated to thickening of the pericardium (red arrow), predominantly isointense to myocardium on T1-weighted image (A). On T2-weighted image, SPIR, the mass shows a typical and high signal hyperintensity (star) (B); heterogenous appearance on balance cine sequences (C).

(56). Often it is associated with pericardium effusion and invasion of the pericardium is frequent. On CT images lymphoma appears or as multiple solid masses attached to the myocardium, or, more commonly, as myocardial or epicardial infiltrative large mass, hypodense or isodense related to the myocardium; with a slight CE. A peculiar characteristic of lymphoma is its diffusion along the surface of the adjacent structures enclosing them (68). On CMR usually, lymphomas are homogeneous relatively hypointense on T1-weighted images and hyperintense on T2-weighted images. CE may be homogeneous or heterogeneous, but not so intense as other malignant tumors (30). The absence of necrosis or hemorrhage areas is useful to distinguish cardiac lymphoma from angiosarcoma.

-Metastasis. Cardiac metastases are more frequent than primary cardiac malignancy(30). Malignant tumors can metastasize to the heart through the direct extension of adjacent organs (lung, esophagus), or through blood vessels dissemination (melanoma) or venous extension into the right atrium within the inferior vena cava (renal cell carcinoma and hepatocellular carcinoma) and via mediastinal lymphatics. The most frequent location of metastasis is the pericardium (64-69%), followed by epicardium (25-34%) and myocardium (30%). Intracavitary metastases are rare, making up 3% to 5% of cardiac metastases **(Fig. 3)** (69). Usually, the imaging features of metastatic disease are not specific, but generally, they have heterogeneous CE, low signal intensity on CMR T1-weighted images and high signal intensity on T2-weighted images. The only exception is reserved for melanoma metastases which appear hyperintense on T1 because of the presence of melanin pigment and also hemorrhagic lesions can show hyperintense areas on T1- weighted images due to the blood degradation products. (30, 67).

Cardiac pseudo-masses

-Thrombosis. Thrombi are the most common intracavitary masses. Thrombus can occur in any of the

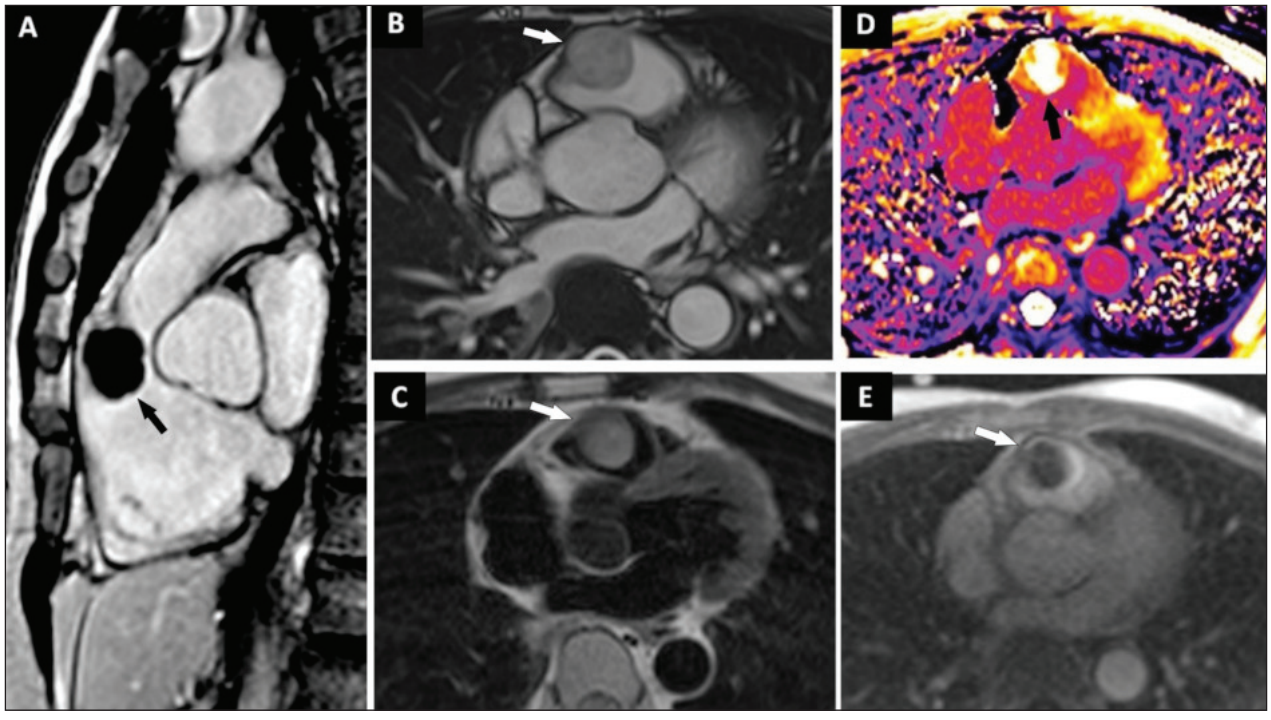

Figure 3. Right ventriculus chordoma metastasis -69-year-old male, with a history of sacral chordoma surgically removed about 10 years before. On TE accidentally discovered a mass in the right ventricular outflow tract (RVOT) (black arrow) (A). CMR shows a solid and rounded mass with regular edges, about 3 cm, intracavitary, slightly inhomogeneous on Trufi cine sequence (white arrow) (B); slightly hyperintense to myocardium on T2- STIR sequence (white arrow) (C). The mass shows different values on T1 map compared to normal adjacent myocardium (black arrow) (D) and progressive CE (white arrow) (E).

cardiac chambers but are typically found in the LA in patients with atrial fibrillation. However, they develop in the region where the flow is slowed or around foreign bodies (for example central venous catheter tip) **(Fig. 4)**. Thrombus is easily detected on CCT image, especially on delayed imaging, as a low-attenuation mass, without CE. Chronic thrombi may present peripheral enhancement because of the presence of a fibrous pseudo-capsule and may develop spotty calcifications. On CMR first pass enhancement images and LGE thrombus show no CE (30, 55).

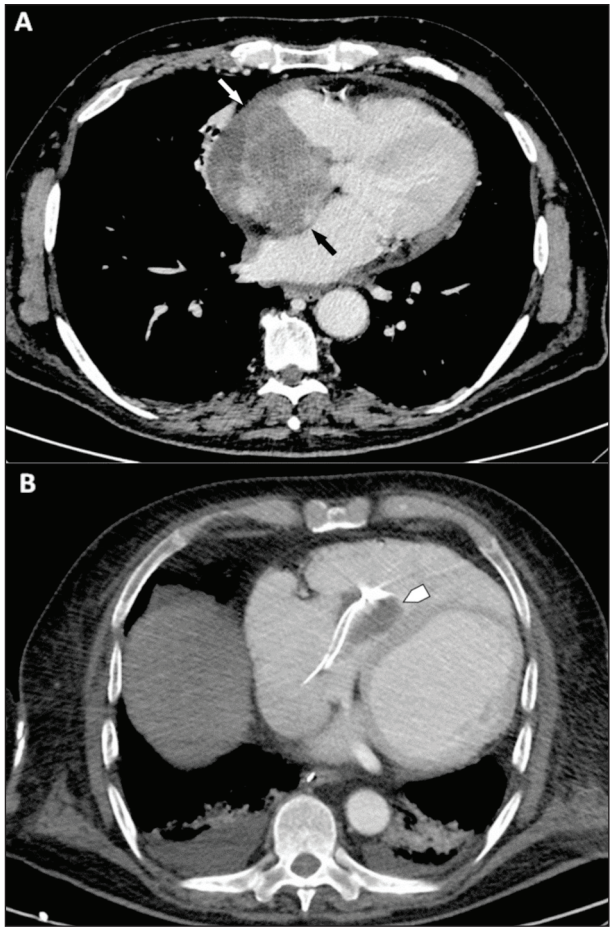

Figure 4. Voluminous sarcoma arising from the right atrium (A) vs thrombus in the right ventricle (B). In A the mass shows invasion of inter-atrial septum (black arrow), inferior vena cava, and pericardium (white arrow) (which appears thickened with a thin pericardial effusion); on CT venous phase, the mass shows heterogenous enhancement. In B in the right ventricle a catheter tip-related thrombosis (white arrowhead) shows hypodensity without CE on CT venous phase (B).

Pseudo-masses. Pseudo masses are normal structures misinterpreted as pathological findings. The orifice of the inferior vena cava (Eustachian valve) can simulate a mass. Into RA the "crista terminalis" or hypertrophic or accessories trabeculae (into RA and RV) may appear as a thrombus or as a mass. Also, lipomatous hypertrophy of the interatrial septum (LHIS) can simulate a tumor (70). LHIS is indistinguishable from lipoma except that the formers occur in the atrial septum with a typical distribution (generally sparing the fossa ovalis). They are asymptomatic and do not require resection (71, 72).

Conflict of interest: Authors declare that they have no commercial associations (e.g. consultancies, stock ownership, equity interest, patent/licensing arrangement etc.) that might pose a conflict of interest in connection with the submitted article.

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Correspondence:
Diletta Cozzi, MD
Department, of Radiology, Careggi University Hospital
L.go G.A. Brambilla, 3 - 50134 Florence – Italy
E-mail: dilettacozzi@gmail.com