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Case Report

A cardiac intimal sarcoma mimicking infective endocarditis ☆

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ABSTRACT

Primary malignant cardiac tumors are rare and usually misdiagnosed because they can mimic more common intracardiac lesions, therefore, in clinical practice it is important to always consider even uncommon diseases in order to avoid delayed diagnosis and to plan the most appropriate therapeutic strategy in a timely fashion. We report a case of a 73-year-old man with clinical signs and imaging findings (echocardiography) suggesting infective bacterial endocarditis of the mitral valve. However, intraoperative evaluation raised suspicion that the mitral lesions had a different nature. Surgical removal of the mass was performed, and the final correct diagnosis was made through pathologic examination, revealing a mitral valve sarcoma thus allowing for the beginning of specific oncological treatment.

1. Introduction

Primary cardiac tumors are very rare entities with the incidence ranging between 0,001% and 0,003% [1]. About 10% of primary cardiac tumors are malignant and most of these are sarcomas. Cardiac sarcomas are often asymptomatic [2–4]; patients usually show signs and symptoms when the tumors have grown so large that they affect the patients' hemodynamic status. Metastatic lesions are usually rapidly proliferating and their presence negatively affects clinical prognosis (median survival is 5 and 15 months in patients with or without metastasis, respectively) [5]. In the reported case, the sarcoma was initially misdiagnosed, as the patient presented with signs of infection and the mass was described at echocardiography as an infective endocarditic vegetation. Only intraoperative evaluation and postoperative histopathologic examination allowed for correct identification of the nature of the lesion and to start the appropriate therapeutic strategy.

2. Case report

Clinical: A 73-year-old man was referred to the Emergency Room of our Hospital due to dyspnea, orthopnea, and persistent cough. A few months before, the patient was placed on oral antibiotics due to the presence of a bilateral basal pneumonia; following this he developed palpitations. At hospital admission, clinical evaluation showed no fever, no hypotension but a systolic murmur grade 4 (Levine scale) on the mitral position; he also had atrial fibrillation with rapid ventricular response that was treated with amiodarone and had the typical signs of congestive heart failure. Transthoracic echocardiography demonstrated an atrial mass (3 × 2 cm) attached to the posterior mitral leaflet causing severe mitral regurgitation. Trans-esophageal echocardiography confirmed the presence of a possible endocarditic vegetation of the posterior mitral leaflet with an abscess cavity (Fig. 1, Video 1-2). All blood cultures were negative and total body computed tomography (CT) scan did not identify systemic embolization nor other peripheral sites of bacterial infection. Preoperative coronary angiography showed normal coronary arteries and the report did not mention other significant abnormalities. The patient was scheduled for mitral valve replacement and mass removal. In the operating theatre after a median longitudinal sternotomy, cardiopulmonary bypass was routinely instituted and cold blood cardioplegic solution was administered via the aortic root after aortic cross clamping. The left atrium was opened through a longitudinal paraseptal incision. due to the bulky size of the mass and the difficulties in determining the anatomic relationships between the mass, the mitral apparatus, and the left atrial wall, and also for a better visualization of the mitral valve, the superior vena cava was transected and the atriotomy was extended to the left atrial roof. A fibrotic mass measuring about 3 × 3 cm involved the

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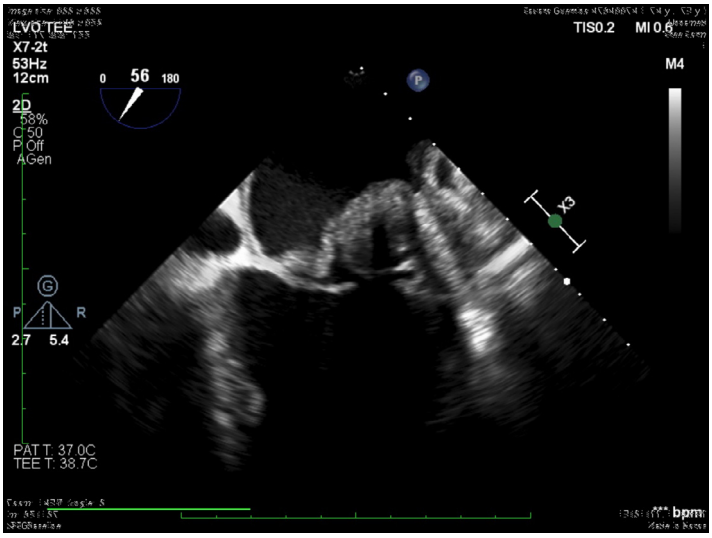

Figure 1. Preoperative transesophageal echocardiographic evaluation. This picture shows the mass of the posterior mitral leaflet with a suspected image of annular abscess.

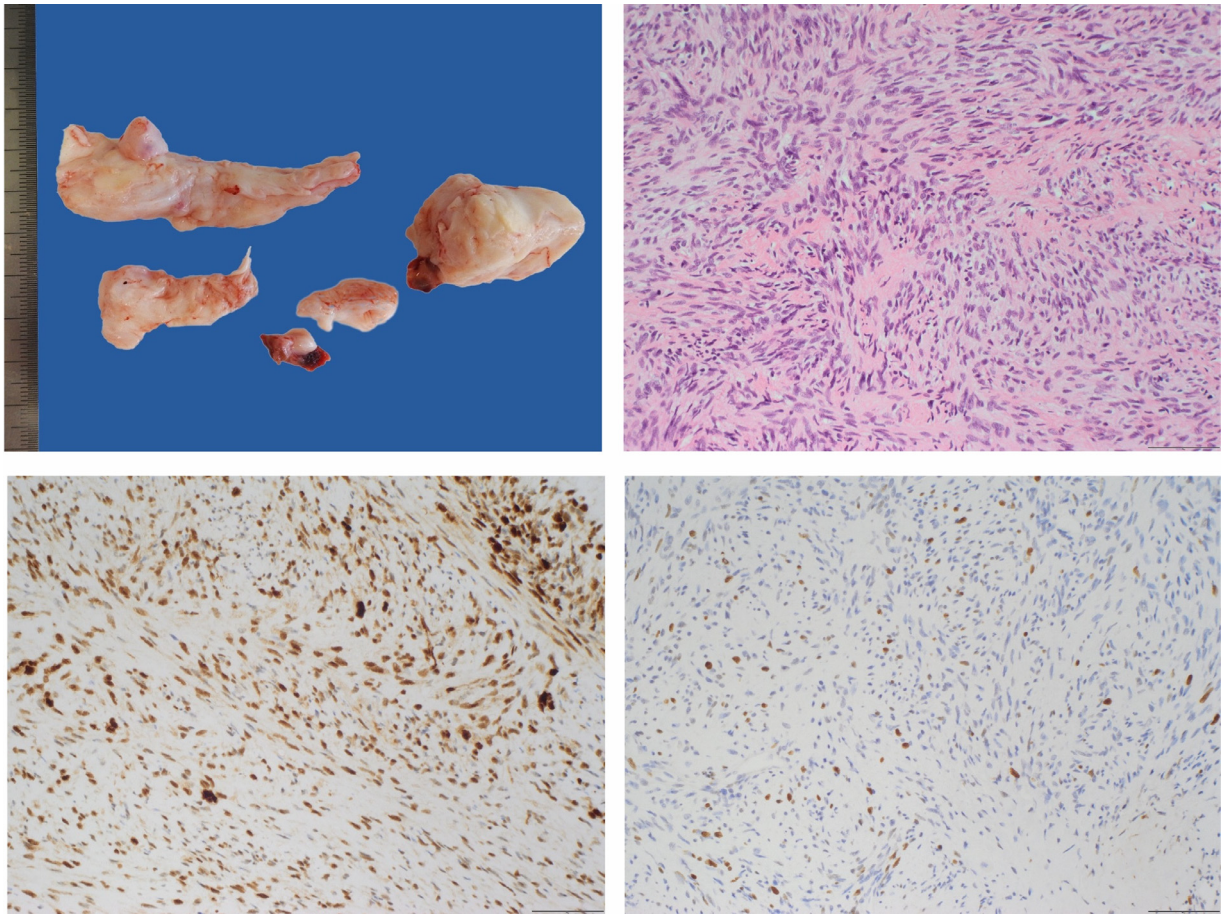

Figure 2. Pathologic evaluation (A) Macroscopic appearance of surgical specimen revealed lobulated, whitish and firm masses. (B) Histological examination showed a fascicular spindle cell proliferation with mild to moderate cellular pleomorphism (H&E, 100x). (C) On immunohistochemistry, the neoplastic cells were strongly positive for MDM2 (100x). (D) The Ki67 index was about 10% (100x)

entire posterior mitral leaflet and the subvalvular mitral apparatus, which also appeared also fibrotic and retracted. The anterior leaflet appeared thickened and fibrotic. Valve leaflets and the mass were completely removed, and a bioprosthesis (Epic 31 mm, Abbott, Abbott Park, IL, USA) was implanted with the standard technique (Video 3).

2.1. Clinical pathology

The microbiological analysis was negative. Macroscopically, the resected mass was gray-yellow in color, with a polylobed surface and a hard consistency. Microscopically, the mass was composed of moderately atypical spindle cells disclosing the presence of an in-

Table 1
Intracardiac intimal sarcomas: a literature review of case reports describing different intimal sarcomas localized in the great vessel or inside the heart.

Authors	Years	F/M	Age	Symptoms	Site of the lesion	Assumed diagnosis	Surgery	Diagnosis	Outcome
Xu et al [9]	2020	F	41	Chest tightness + cough + syncope	RVOT + PV + MPA + RPA + LPA	PTE	Pulmonary endoarterectomy + resection of the mass + tricuspidplasty	Pulmonary artery intimal sarcoma	Alive at FU (1 m)
Pu et al [8]	2017	F	36	Dyspnea + fatigue + syncope	MPA + RPA + LPA	PTE	Pulmonary thromboendoarterectomy	Pulmonary artery intimal sarcoma	Lost at FU
		F	67	Chest distress + palpitation	MPA + RPA + LPA	CTEPH	Pulmonary thromboendoarterectomy	Pulmonary artery intimal sarcoma	Lost at FU
		M	59	Chest pain + cough + hemoptysis	MPA + RPA	Pulmonary stenosis	Pulmonary thromboendoarterectomy	Pulmonary artery intimal sarcoma	Alive a FU (150 d)
Onagi et al [17]	2021	F	25	Dyspnea + palpitations	MPA + RPA + LPA	PA tumor (calcification)	Resection of the mass	Pulmonary artery intimal sarcoma (ISCOS)	Alive ar FU (16 m)
Morisaki et al [18]	2020	M	50	Cough + dyspnea	TV + PV + RPA + LPA + lungs	Spindle cell tumor (lung biopsy)	Palliative surgery (TVR + resection)	Pulmonary artery and tricuspid valve intimal sarcoma	Dead (9 m)
Alsoufi et al [7]	2006	M	76	Dyspnea, + weakness + malaise	RVOT + PV +MPA + RPA+ LPA	PTE	Pulmonary thromboendoarterectomy + PVR + tricuspid valve repair	Rhabdomyosarcoma	Alive at FU
Shu-I Lin et al [13]	2015	F	45	Epigastric pain + nausea	distal thoracic aorta	Abdominal aortic dissection	descending aorta excision	Aortic intimal sarcoma	Died (6 m)
Nanjo et al [19]	1996	F	54	fever + fatigue	aortic arch	tumor	total arch replacement	Aortic intimal sarcoma	Died (10 m)
Grant et al [6]	2020	F	31	left-sided weakness	left atrium	atrial myxoma	Resection of the mass	Intimal sarcoma	NR
Prol et al [12]	2021	F	83	Dyspnea	Left atrium + mitral valve	malignant cardiac tumor	Not suitable	Intimal sarcome (TEE-guided percutaneous transcatheter byopsy)	Died (3 m)
Todo et al [14]	2021	M	78	Heart failure	left atrium + mitral valve	non-bacterial thrombotic endocarditis	MVR + resection of the mass	Intimal sarcoma	NR
Zhang et al [20]	2016	F	38	Cough + chest pain	mitral valve	mitral stenosis	MVR + resection of the mass	Undifferentiated sarcoma	Alive at FU (3 m)
Abreu et al [21]	2018	F	70	Dyspnea + weight loss + +night sweat	left atrium + inferior pulmonary veins	NR	Partial resection	Intimal sarcoma	Died (1 m)
Chen et al [11]	2021	M	19	Dyspnea + abdominal distension + nausea	Right atrium + IVC	Tumor	Resection of the mass	Intimal sarcoma	Alive at FU (2 y)
Scheidl et al [22]	2010	M	40	Dyspnea + thoracic pain + hemptysis	PV + MPA + RVOT	CTEPH	Pulmonary thromboendoarterectomy + PVR + tricuspid repair	Pulmonary artery intimal sarcoma	Died (1 m)

RVOT = right ventricle outflow tract; PV = pulmonary valve; MPA = main pulmonary artery; RPA = right pulmonary artery; LPA = left pulmonary artery; PTE = pulmonary thromboembolism, FU =follow-up; CTEPH = chronic thromboembolic pulmonary hypertension; ISCOS = intimal sarcoma with chondroblastic osteosarcomatous differentiation; TV =tricuspid valve; TVR =tricuspid valve replacement; PVR = pulmonary valve replacement; TEE = trans-esophageal echocardiography; MVR = mitral valve replacement; IVC = inferior vena cava; d = days; m = months; y = years, NR = not reported.

timal sarcoma with areas of necrosis and chondroid differentiation (Fig. 2). Immunohistochemical analysis revealed positive expression of MDM2 and focal positivity for smooth muscle actin and sarcomeric actin in neoplastic cells, with a proliferation index (MIB1) of 10%.. Furthermore, targeting markers associated with neural (S100), endothelial (CD34), and epithelial (pan-cytokeratins) origins were found.

Final clinical descritpion: The following postoperative course was uneventful, and the patient was discharged home on the 11th postoperative day and immediately started anthracycline-based chemotherapy. During follow-up, multiple metastatic lesions quickly developed in the chest and in the liver showing severe and uncontrolled disease progression. The patient died 11 months after surgery.

3. Discussion

Primary cardiac sarcomas are extremely rare and due to their rapid proliferating and metastases, the prognosis is poor [5]. The median presenting age of primary cardiac sarcomas is 42 years. Intimal sarcomas were firstly described in 1923 by Mandelstamm during an autopsy and have no definite age or gender of occurrence but there is a slightly greater incidence in women over 40 years old. Intimal sarcomas are mesenchymal neoplasms that arise predominantly in the great vessels, but they can also be found in heart chambers or valves; the incidence of pulmonary artery with intimal sarcoma (0.001-0.03%) is twice that seen in the aorta [6–9]. These tumors are often misdiagnosed due to nonspecific symptoms suggesting other cardiac or pulmonary conditions; as a matter of fact, patients with intimal sarcoma located in the right ventricle or pulmonary artery present dyspnea, chest pain, cough, hemoptysis, and syncope and these symptoms can mimic those of acute or chronic pulmonary thromboembolism [10,9]. Otherwise, when the tumor is localized in right atrium or inferior vena cava, patients usually manifest dyspnea and abdominal symptoms [11]. If the sarcoma is localized in the left chambers of the heart, the patient usually shows heart failure symptoms while if it is located in different portions of the aorta, signs mimicking transient ischemic attack, claudication or aortic dissection can occur [12,10,6,13]. In one case report, a sarcoma mimicking a nonbacterial thrombotic endocarditis was described [14]. Table 1 summarizes the different locations and clinical presentations of intimal sarcoma reported in literature.

There are currently some treatment options aimed at prolonging survival: surgical excision, radiotherapy, chemotherapy, and heart transplantation. The effects of adjuvant chemotherapy remain uncertain [15]. In our case, the clinical history and the echocardiographic findings were highly suggestive of infective endocarditis since two of Dukes' three major criteria for the diagnosis of endocarditis were present (new valvular regurgitation and echocardiography showing endocardial vegetation and abscess). The intraoperative macroscopic aspect of the mass was not suggestive of endocarditis and, therefore, we decided to perform an extended resection of both mitral valve leaflets and of the entire mass. The microbiological and histopathological evaluations of the surgical specimen were crucial for the final correct diagnosis [16]. Retrospectively, we re-evaluated the preoperative coronary angiography of this patient and we found visible tumor vascularization from the left circumflex artery. This image was hard to evaluate at initial observation and was not reported on the cath-lab chart (Video 4). These situations should make physicians think beyond just vegetation and endocarditis whenever they encounter an intracardiac mass with unusual features. Early diagnosis is crucial to start the most appropriate therapeutic strategy in a timely fashion.

4. Conclusions

Primary cardiac sarcomas are often challenging to diagnose since they can mimic other cardiac pathologies. They should always be taken into consideration when an intracardiac mass with unusual characteristics is found. Timely assessment and extended resection are crucial to give patients chances of prolonged survival even if overall prognosis remains poor.

Ethical approval

IRB/EC approval has been waived since this is a single patient case report. This patient signed an informed consent for use of anonymous clinical data for scientific purposes and clinical research.

Declaration of competing interest

Nothing to declare in terms of financial, personal, political, intellectual, or religious interests.

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All authors listed have made a substantial, direct, and intellectual contribution to the work, and approved it for publication.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.carpath.2023.107598.

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