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Sarcoma of the heart: survival after surgery†

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Abstract

OBJECTIVES: Malignant intracardiac tumours are rare, and consensus concerning the optimal therapeutic approach is lacking. We performed a retrospective medical analysis, identifying 9 patients having been operated for cardiac sarcomas. All of them had a complete postoperative long-term follow-up. To enhance understanding of the best therapeutic approach for future patients, it is crucial to reveal special medical problems and to analyse the potential impact they may have on disease course and survival rate in this specific patient group.

METHODS: Cardiac tumours operated on 2000 to the end of 2015 were reviewed. Late mortality during the follow-up period was determined. The impact of tumour extension, tumour localization, resection status (complete versus partial) and histopathological diagnosis on survival was analysed retrospectively.

RESULTS: Of all cardiac malignant tumours resected, sarcomas were, with an incidence of 0.14% (9 patients), the most frequent histological group admitted to cardiac surgery. All of the patients presented with cardiac symptoms. All of the patients survived the operation and all had relief or improvement of cardiac symptoms. The mean follow-up period was 17 ± 13 months. Five patients died after 6, 8, 12, 12 and 15 months, respectively. Four survivors (3 with a pulmonary artery tumour sarcoma and 1 with a left atrial sarcoma) had a mean follow-up of 26 ± 17 months. Macroscopically complete tumour resection, absence of metastatic spread and histological sarcoma type had an impact on follow-up survival.

CONCLUSIONS: Although cardiac sarcomas are rare, surgeons occasionally encounter them. A 1-year mortality rate of 44% reflects an unfavourable prognosis, but surgery seems to be a secure, reliable option in selected patients for treating cardiac symptoms and avoiding early cardiac-related deaths.

Keywords: Cardiac tumour • Sarcoma • Survival • Cardiac surgery

INTRODUCTION

Malignant intracardiac tumours are rare, and therapeutic guidelines are lacking. Few data are available on mid- and long-term outcomes after surgery for cardiac sarcoma. Surgery under these circumstances is often palliative, but it reduces tumour mass and may help to control symptoms. We present a series of 9 patients with sarcomas with complete follow-up after surgical resection of the tumour.

METHODS

We searched the database of our single-centre cardiac surgery unit for patients operated on for cardiac tumours between January 2000 and the end of December 2015. All patients with cardiac sarcomas were identified and included in this series. Major risk factors and their potential impact on mid- and long-term outcomes were analysed. The 1-year mortality rate and overall survival after surgery were determined. Corresponding factors that might have an impact on survival were identified. General health conditions at admission, tumour extension and metastatic spread at time of surgery, intracardiac or intravascular tumour localization, surgical technique, completeness of tumour resection and the histopathological type of sarcoma were assessed for their impact on overall survival and on local or distant tumour relapse.

Approval was granted by the institutional review board of the investigator's centre according to international recommendations.

RESULTS

During the observation period, about 6450 patients underwent cardiac surgery, 55 (0.85%) of whom had a cardiac tumour

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resected (Table 1). Sarcomas were, with an incidence of 0.14% (9 patients), after myxomas/fibroelastomas, the most frequently occurring malignant tumour and the 2nd most common cardiac tumour treated surgically. The mean age of the patients who had a sarcoma was 69 ± 12 years (range 45–85 years). All of them presented with cardiac symptoms at admission (mainly dyspnoea and rhythmic disorders) but were in acceptable general overall health, i.e. nobody presented with a major organic disorder (all had normal liver and kidney function) or cancer-related cachexia (body mass index 25 ± 3 kg/m^2; range 22–34 kg/m^2). All patients with sarcomas survived beyond 30 days postoperatively, and no cardiac complications were observed during the early follow-up period. All patients had significant improvement in or complete disappearance of their cardiac symptoms. Length of follow-up was 17 ± 13 (range 6–47) months. The 1-year mortality rate was 44%, with 4 patients dying 6, 8, 12 and 12 months after surgery, respectively. Another patient died 15 months after surgery. Four survivors had a mean follow-up period of 26 ± 17 (range 12–47) months. The maximum tumour diameter based on the resection specimen varied from 4 to 10 cm (mean 6 ± 2.7 cm).

Four (44%) patients had a macroscopically complete (R0/1) intracardiac tumour resection, and all of them survived during the follow-up period (Table 1). Three of them had intrapulmonary artery sarcomas and underwent a complete right ventricular outflow tract resection up to the pulmonary artery bifurcation and reconstruction using either a homo- or a xenograft (Contegra® pulmonary valved conduit, Medtronic, Minneapolis, MN, USA) (Fig. 1). The 4th patient had a left intra-atrial sarcoma attached to the mitral valve. A bioprosthetic mitral valve replacement allowed also a macroscopically complete tumour resection (Table 1).

Local relapse occurred in 3 (33%) patients, all of whom had a macroscopically incomplete tumour resection (R2) due to complex cardiac infiltrations of the free ventricular wall (2 patients) and of the septum and cardiac skeleton (1 patient) (Table 1). Coronary neovascularization of the sarcoma was documented by angiography in 1 of these patients (Fig. 2).

All 3 patients with known metastases on the preoperative computed tomography scan have died, 2 at 12 months and 1 at 15 months, respectively (Table 1).

Histopathological grading, i.e. undifferentiated versus well differentiated, may indicate prognosis. Among 4 patients with a histopathologically confirmed undifferentiated sarcoma, 3 died during the 1st year after surgery (Table 1).

DISCUSSION

Although they are rare, cardiac tumours pose an important, complex problem that cardiac surgeons have to manage. Malignant cardiac tumours are frequently one of the many different subtypes of sarcomas [1, 2]. We analysed the postoperative mid- and long-term follow-up survival rates of this series of 9 patients with sarcomas to determine if an aggressive therapeutic approach would be justified. In accordance with the available literature, at 1 year, nearly half of the patients were dead, reflecting the poor prognosis [3, 4]. However, serious, acute and life-threatening symptoms indicative of intrapericardial tumour compression or haemodynamic compromise disappeared or greatly improved, and the immediate postoperative outcomes were excellent in the selected patients who underwent cardiac surgery (all patients were otherwise healthy). Survival was associated mainly with the

Table 1: Sarcoma distribution, survival, surgical data and histopathology

No.	Leading cardiac complication	Follow-up (months)	Died ≤12 months postoperatively	Alive	Macroscopic tumour resection	Main location	Type of surgery	Late (follow-up) metastasis	Preoperative metastasis	Type of sarcoma
1	RV compression/pericardial tamponade	6	1	0	0	RV	Incomplete local resection	1	0	Rhabdomyosarcoma
2	Pulmonary vein occlusion	12	1	0	0	Left + right atrium, atrial septum	Partial left atrium + right pulmonary vein resection	1	1	Undifferentiated sarcoma
3	Myocardial ischaemia/ LAD compression	15	0[a]	0[a]	0	Antero-apical epicardium	Incomplete local resection + cover with xenopericardium	1	1	Synovial sarcoma
4	Recurrent pulmonary embolism	47	0	1	1	Pulmonary artery	RVOT resection + Contegra® graft	1	0	Angiosarcoma
5	LV compression	12	1	0	0	Anteroapical LV	Dor resection	1	1	Undifferentiated sarcoma
6	Mitral stenosis (mitral tumour prolapse)	12	0	1	1	Left atrium, mitral valve	Partial left atrium resection + mitral valve replacement	0	0	Angiosarcoma
7	Pulmonary stenosis	31	0	1	1	Pulmonary artery	RVOT resection + homograft	0	0	Undifferentiated sarcoma
8	Pulmonary stenosis	12	0	1	1	Pulmonary artery	RVOT resection + Contegra graft	0	0	Angiosarcoma
9	Pulmonary vein occlusion	8	1	0	0	Left atrium, pulmonary veins, mitral valve	Partial left atrium resection + mitral valve replacement	1	0	Undifferentiated sarcoma
Mean ± SD		17.2 ± 13.2	4	4	4			6	3	

[a]Patient died after 15 months follow-up.

Dark grey: patients died early, within the 1-year follow-up period; LAD: left anterior descending artery; light grey: potential long-term survivors; LV: left ventricle; RV: right ventricle; RVOT: right ventricular outflow tract; SD: standard deviation.

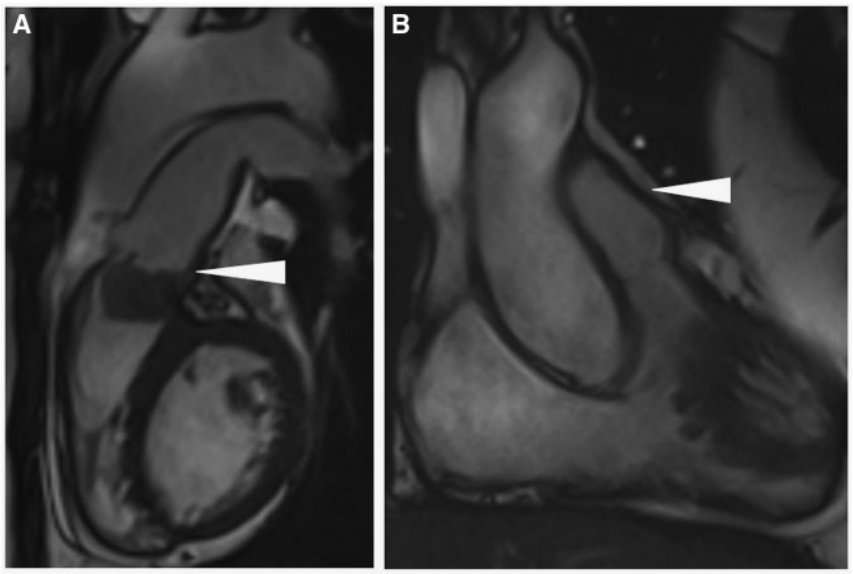

Figure 1: Cardiac magnetic resonance imaging scan showing an angiosarcoma (arrow) close to the pulmonary valve, before (**A**) and after (**B**, arrow) right ventricular outflow tract reconstruction.

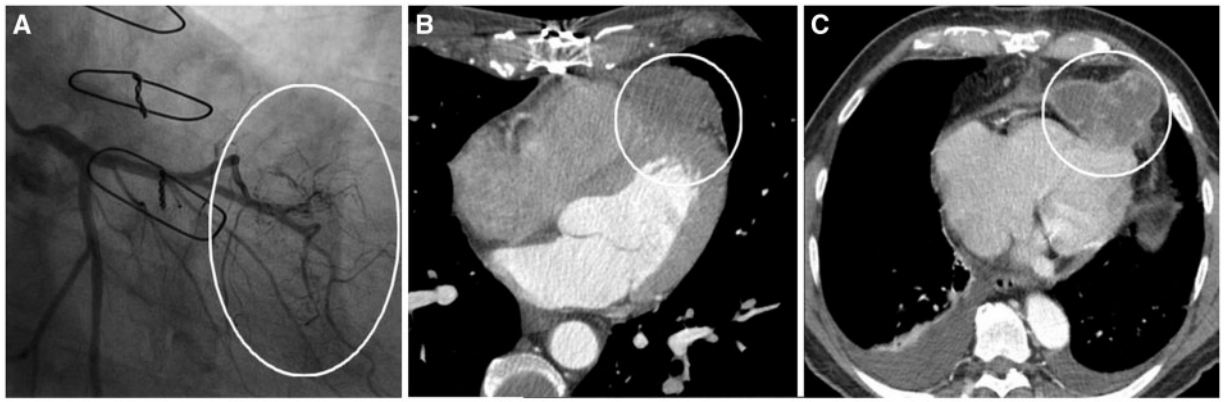

Figure 2: Angiography shows coronary tumour neovascularization, principally via the left anterior descending artery and its side branches (circle), of an infiltrating anterior apical synovial sarcoma (**A**). The same myocardial infiltrating tumour (circle) shown in the computed tomography scan (**B**), compared to a pericardial rhabdomyosarcoma (**C**, circle).

location of the intracardiac tumour and correlated with better results in cases of macroscopic complete resection. Pulmonary artery sarcomas may be accessible for a complete right ventricular outflow tract resection and reconstruction using conduits (Table 1, Fig. 1), which might explain in part, together with their tendency to metastasize late in the course of the disease, their good outcome [5]. In contrast, sarcomas infiltrating the ventricular myocardium have a worse prognosis because of the technical limitations of the surgery and faster metastasis (Fig. 2) [5]. Clearly, preoperative metastatic spread is associated with poor prognosis, and all patients in our series who had positive findings on their preoperative computed tomography scan died within 15 months after surgery [6].

Biological factors, i.e. histopathological tumour type and grade, may have a prognostic impact, with low-grade (well-differentiated) sarcomas having a better outcome than high-grade (undifferentiated) sarcomas; however, this is a matter of ongoing discussion [7, 8]. Our results also confirmed previously published data concerning the incidence and preferred location of frequently occurring subtypes of primary heart sarcomas. Angiosarcomas, which probably have a stronger affinity for right heart structures, and undifferentiated sarcomas, which frequently affect left cardiac structures, were the 2 predominantly encountered types, with the corresponding above-mentioned distribution frequency [2, 7, 9, 10].

Kim *et al.* [9], in one of the largest series with complete follow-up, reported a similar 1-year mortality rate of 41% (9 of 22 patients died during the 1st year after surgery). Information concerning early perioperative morbidity and mortality was not available.

Compared to published data, early surgical and clinical outcomes were excellent in our small series. Donsbeck *et al.* [8] reported a similar 40% complete intracardiac tumour resection rate, but also a high perioperative mortality rate of 20% ($n = 4/20$; 3 patients died within a day after surgery). Comparable results have been published by Zhang *et al.* [11], who reported an early mortality rate close to 24% ($n = 4/17$; 4 patients died within 5 days of the initial surgery) among their series of 17 patients operated on for cardiac sarcomas. The main difference concerning their observed outcome is the high incidence of cardiac-related deaths

during the early follow-up period caused by tumour recurrence, leading directly to heart compression and failure [11]. This outcome may indicate a well advanced tumour, similar to those in the series of Zhang *et al.*, which included 3 cases of cardiac transplantation as initial surgical treatment [8, 11].

Cardiac autotransplantations for complex left atrial reconstructions, as described by Shapira *et al.* [12], have not been performed. This complex and radical approach led to good results in the large series of Bakaeen *et al.* [13], with a reported early mortality rate of 7% among 27 patients: 8 patients had an autotransplantation for atrial reconstruction and all survived the operation, with a tumour-free survival period beyond 20 months in 4 of them.

Systemic therapies play a minor role in these patients, because their efficacy is limited, and symptoms necessitate immediate surgical action. However, in patients with metastatic disease, systemic treatment should follow the recommendations and guidelines based on the histological diagnosis [14].

The preceding series underline the importance of strict selection criteria when accepting patients with sarcomas for cardiac surgery. Good general health and nutritional conditions, despite a potentially advanced tumour and a primarily symptom-orientated surgical approach, may lead to a successful early outcome in these high-risk patients.

Limitations

The major limitations of the study result from its retrospective and selective design, explaining, as previously mentioned, at least partially the excellent early postoperative outcome in these severely diseased patients.

CONCLUSIONS

Based on current knowledge, surgical resection of an intracardiac sarcoma can be a reliable, effective option to successfully treat a symptomatic, life-threatening cardiac tumour, but only in selected patients. A curative surgical approach and technique may be possible and can assure survival beyond several years in selected patients, who otherwise usually have a bleak prognosis. Mid- and long-term outcomes depend on the location of the cardiac tumour and its accessibility for a complete resection. Low-grade tumours might be associated with a better prognosis, whereas metastatic disease always signifies a poor prognosis.

Conflict of interest: none declared.

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