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ORIGINAL ARTICLE

Primary malignant cardiac tumors: Surgical results☆

Joana Saraiva[a], Pedro Engrácia Antunes[a], Lina Carvalho[b], Manuel Jesus Antunes[a,*]

[a] *Centro de Cirurgia Cardiotorácica, Centro Hospitalar Universitário e Faculdade de Medicina, Coimbra, Portugal*
[b] *Departamento de Anatomia Patologia, Centro Hospitalar Universitário e Faculdade de Medicina, Coimbra, Portugal*

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KEYWORDS
Primary malignant cardiac tumors;
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Abstract
Objective: To characterize primary malignant cardiac tumors operated on in our center and to analyze patient survival.
Methods: Between January 1994 and August 2014, 123 patients with cardiac tumors underwent surgery, of which 12 (9.8%) were primary malignant tumors – eight sarcomas (67%), three B-cell lymphomas (25%) and one epithelioid hemangioendothelioma (8.3%). The tumor affected the left atrium in five cases (42%), the right atrium in four (33%), the right ventricle in two (17%) and the pulmonary valve in one (8%). Patients' mean age was 55.4±16.9 years, 67% were female and 75% presented in NYHA class III–IV.
Results: Resection was complete (negative margins) in five cases and partial in seven (five sarcomas and two lymphomas), and 11 patients needed adjuvant therapy, surgery alone being curative in only one (epithelioid hemangioendothelioma). Mean follow-up was 41.7±61.3 months: 24.8±30.0 months (3.8–95.7) for sarcomas, 70.1±118.0 months (1–206.3) for lymphomas and 91.9 months for the epithelioid hemangioendothelioma. During follow-up, 10 patients died (83%) and two were alive (17%). Overall survival at 30 days, six months, one year and two years was 91.7%, 66.7%, 58.3% and 41.7%, respectively. In the sarcoma group, 1-year and 2-year survival were 62.5% and 37.5%, respectively.
Conclusions: Resection of primary malignant cardiac tumors, even partial, is safe, provides relief of obstructive symptoms and improves quality of life, but is rarely curative and has a low survival rate. Due to the rarity of such tumors, a multicenter database could improve knowledge and help clarify the indications for cardiac surgery as a treatment option.

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* Corresponding author.
E-mail address: antunes.cct.chuc@sapo.pt (M.J. Antunes).

PALAVRAS-CHAVE
Tumor cardíaco maligno primário;
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Tumores cardíacos primários malignos: resultados cirúrgicos

Resumo
Objetivo: Caracterizar os tumores malignos cardíacos operados no nosso centro e analisar a sobrevida dos doentes.
Métodos: De janeiro/1994 a agosto/2014, 123 doentes com tumores cardíacos foram submetidos a cirurgia, dos quais 12 revelaram ser malignos (9,8%) – oito sarcomas (67%), três linfomas de células B (25%) e um hemangioendotelioma epitelioide (8,3%). A AE estava afetada em cinco casos (42%), a AD em quatro (33%), o VD em dois (17%) e a válvula pulmonar em um (8%). A idade média era 55,4 ± 16,9 anos, 67% do sexo feminino e 75% apresentavam-se em classe III-IV da NYHA.
Resultados: A ressecção foi completa (margens negativas) em cinco casos e parcial em sete (cinco sarcomas, dois linfomas). Onze doentes necessitaram de terapia adjuvante, sendo a cirurgia curativa em apenas um (hemangioendotelioma epitelioide). O tempo de seguimento médio foi de 41,7 ± 61,3 meses; 24,8 ± 30,0 (3,8-95,7 meses) para sarcomas, 70,1 ± 118,0 (1-206,3 meses) para linfomas e 91,9 meses para o hemangioendotelioma epitelioide. Durante o seguimento, dez doentes faleceram (83%). A sobrevida global aos 30 dias, seis meses, um e dois anos foi de 91,7, 66,7, 58,3 e 41,7%, respetivamente. No grupo dos sarcomas, a sobrevida a um e dois anos foi de 62,5 e 37,5%.
Conclusões: A ressecção de tumores malignos primários, mesmo que parcial, é segura, providencia alívio sintomático, podendo melhorar a qualidade de vida, mas é raramente curativa e tem baixa sobrevida. Dada a raridade, uma base de dados multicêntrica poderia melhorar o conhecimento e ajudar a clarificar as indicações cirúrgicas.

Introduction

Primary cardiac tumors are rare, the incidence on autopsy varying between 0.0017% and 0.03%. Around 25% are malignant and most (75%) are sarcomas.[1]

Cardiac tumors are generally asymptomatic until they are large enough to cause valve or chamber obstruction. Although they are easily detected by echocardiography, malignancy and specific histological type cannot always be determined or even suspected before surgical intervention.[2]

The main treatment for primary malignant cardiac tumors, particularly sarcomas, is still complete surgical resection, combined with chemotherapy.[3,4] Nevertheless, cardiac sarcomas have a poor prognosis, with mean survival of 11–17 months.[1,5,6]

The aim of this study was to characterize primary malignant cardiac tumors operated on in our center and to analyze patient survival.

Methods

This was a retrospective study of patients operated between January 1994 and August 2014. Information was collected from patients' medical records, together with data on follow-up from hospital records and telephone contact with patients or relatives. During the study period, 23 010 patients underwent cardiac surgery. Of these, 123 (0.53%) had cardiac tumors, but only 12 had primary malignant tumors, corresponding to 9.8% of all cardiac tumors and an incidence of 0.05% among patients undergoing cardiac surgery.

Patients' mean age was 55.4±16.9 years (21–79), 67% were female and 75% presented in NYHA class III–IV. The tumor was located in the left atrium in five cases (42%), the right atrium in four (33%), the right ventricle in two (17%) and the pulmonary valve in one (8%) (Table 1).

Eight of the tumors (67%) were sarcomas, three (25%) B-cell lymphomas and one (8.3%) epithelioid hemangioendothelioma. In the sarcoma group, there were three cases of angiosarcoma (38%) and one each of rhabdomyosarcoma, leiomyosarcoma, myxofibrosarcoma, undifferentiated sarcoma and myxochondrosarcoma.

Only two patients had a histological diagnosis prior to surgery: one with an epithelioid hemangioendothelioma, previously biopsied by right thoracotomy, who had no known metastases and had not undergone neoadjuvant therapy; and the other with an angiosarcoma, diagnosed by mediastinoscopy, with lymph node metastasization and pericardial effusion, who had undergone neoadjuvant chemotherapy, resulting in reduction of the mass and resolution of the pericardial effusion. All the other tumors were diagnosed through anatomopathological analysis of the surgical specimens.

The myxochondrosarcoma, myxofibrosarcoma and one of the lymphomas presented with pericardial effusion. One patient with angiosarcoma had liver metastases preoperatively and the patient with leiomyosarcoma presented diffuse lung metastases. One of the lymphomas presented as vena cava syndrome. The other cases had no known