

## **A PRIMARY EPITHELIOID CARDIAC SARCOMA – CASE REPORT AND REVIEW OF THE LITERATURE**

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**A PRIMARY EPITHELIOID CARDIAC SARCOMA. CASE REPORT AND REVIEW OF THE LITERATURE (Abstract):** The aim of the study was to characterize the clinical and histological picture of a rare primary malignant cardiac tumor and to assess the operative outcome. *Case report:* A case of primary cardiac sarcoma located in the right ventricle is described, using data collected from the medical record of the patient. The patient was admitted for symptoms and signs of right heart failure and the diagnosis was established by echocardiography. The patient was operated on through a median sternotomy but, due to extension of the tumor, only partial removal was possible. Although incomplete, surgical resection resulted in the improvement of the cardiac function and allowed the patient to be discharged one month post-operatively in a satisfactory clinical condition. The histological examination revealed a high grade pleomorphic, undifferentiated sarcoma. The patient survived three months. *Conclusions:* The advanced local stage of the tumour at the time of the diagnosis resulted in a short postoperative survival. However, irrespective of the histological subtype and completeness of surgery, malignant cardiac tumours are a poor prognosis.

**KEY WORDS:** CARDIAC SARCOMA, MESENCHYMOMA, MALIGNANT FIBROUS HISTIOCYTOMA

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### **INTRODUCTION**

Primary cardiac tumours (PCT) generate only 0.3% of all open heart operations. However, modern cardiac imaging explorations have turned primary cardiac tumours from a condition rarely diagnosed before autopsy to a potentially curable heart disease [1].

The most common primary malignant tumor of the heart and pericardium is sarcoma, but this tumors occur rarely, the most primary cardiac tumors are benign. In a series of surgical resections, Virmani reported that cardiac sarcomas represent 10% from all cardiac tumours [2,3]. Silver noted that the largest group of primary cardiac sarcomas demonstrate fibroblastic and myofibroblastic differentiation [4]. Classically, these are divided into malignant fibrous histiocytoma (comprising more than 50% of this group), fibrosarcoma and fibromyosarcoma. Undifferentiated pleomorphic sarcomas are often designated as malignant fibrous histiocytoma [3]. When occurring in the heart,

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these three types of sarcoma share similar histological and clinical features and may prove difficult to distinguish one from another both clinically and even pathologically.

The majority of cardiac myofibroblastic sarcomas are endocardially based, with a bulky tumor within the heart cavity. It was speculated that they originate from myofibroblastic cells that normally occur in the intima and that are analogous to intimal sarcomas of the great vessels [4].

The major differential diagnosis of undifferentiated sarcoma, especially right-sided lesions, is of metastatic tumor [5]. Malignant melanoma, metastatic carcinoma, and malignant mesothelioma should always be excluded if a pleomorphic tumor of the heart is encountered [6,7].

The objective of the present study was to describe a particular case of PCT in which the curative resection was not possible due to locally extensive tumor.

### CASE REPORT

A 36 year-old female patient with a recent history of Lyme disease, was admitted in our department in august 2005 for fatigue, progressive dyspnoea, medium intensity upper abdominal pain. The physical examination revealed signs of right cardiac failure, including pleural effusion, ascites and liver enlargement. Lab data pointed out hepatocytolysis.

Abdominal ecography confirmed the diffuse liver enlargement and the presence of medium quantity ascites. The routine chest X- ray confirmed the presence of bilateral pleural effusion and raised the suspicion of a right ventricular tumor. Echocardiography was requested both to confirm and identify tumor location as well as to monitor left ventricular function, filling status and valve function. A tumor infiltrating the ventricular septum and ventricular walls was diagnosed. Considering the rapid alteration of the heart function, the operation was decided on emergency basis. Following median sternotomy, the intraoperative exploration confirmed the advanced local stage of the lesion which infiltrated the ventricular walls and septum. Under these circumstances, the operative technique consisted in a partial right ventricular tumor resection, supported by extracorporeal circulation for releasing right ventricular outflow.

In the early postoperative period, the patient developed transient hepatic and renal failure, which were remitted under intensive treatment. She was discharged a month after surgery in satisfactory clinical condition. Lyme disease aggravated the patient clinical status, requiring long term antibiotherapy.

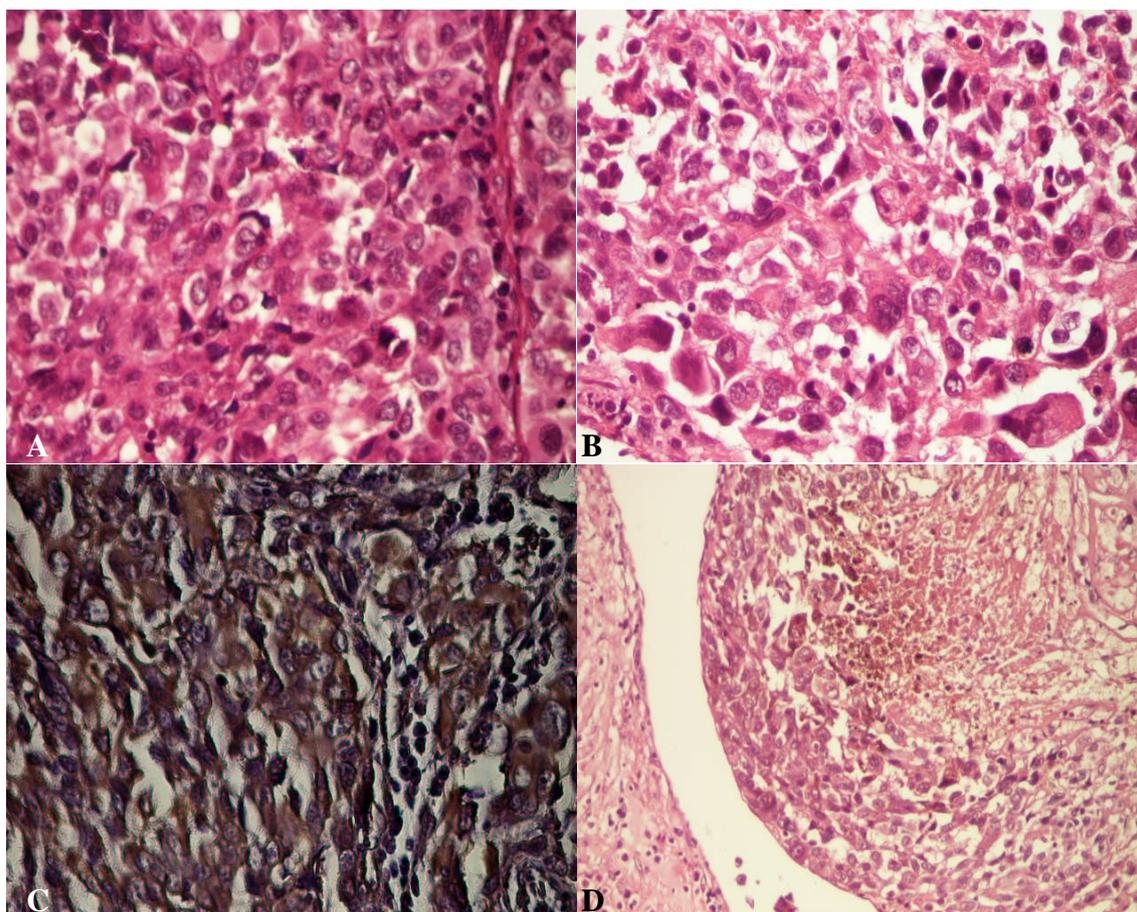
The pathological examination of the surgical specimen revealed, grossly, the tumor corresponded to a polypoid mass, with a sessile endocardial basis, infiltrating the ventricular septum, encircling the tricuspid orifice and projecting in the right ventricular cavity (RVC). The tumor mass distended RVC and impinged the pulmonary valve, obstructing the pulmonary outflow, as well. The tumor was variegated in appearance due to the presence of the hemorrhagic and necrotic areas.

The histological findings of the malignant tumor revealed a high-grade sarcoma, without clear evidence for a specific line of differentiation. The tumor was heterogeneous in appearance and hypercellular, the constituent cells being spindle or epitheloid (Fig. 1A), and focally, intermixed with multinucleated giant cells (Fig. 1B). The dominant tumoral feature was of marked pleomorphism and increased mitotic activity.

The immunohistochemical tests showed a positive immunoreactivity for vimentine (Fig. 1C) that strongly suggested a connective origin, a negative

immunoreactivity for desmin, a focal positive immunoreactivity for citokeratine and negative for mesothelial markers. The results are considered aspects of a pleomorphic undifferentiated sarcoma with "focal epitheloid habitus".

Another important feature was the tendency for tumoral embolism, evidenced by the presence of minute tumoral nodules (Fig. 1D) dislodged from the tumoral mass, without any evidence of metastases. There was also present a hypercoagulability state, as well, explaining the associated thromboembolism.



**Fig. 1 The histological findings of the cardiac sarcoma**

A Cardiac sarcoma with epitheloid appearance (HE x 20); B Cardiac sarcoma with cellular pleomorphism (HE x 20); C Cardiac sarcoma with positive immunoreactivity to vimentin (IMH x 20); D Thromboembolus (HE x10)

## DISCUSSION

Undifferentiated pleomorphic sarcoma is a malignant mesenchymal tumour with a high grade of malignancy showing fibroblastic or myofibroblastic differentiation and areas of marked cellular pleomorphism [3]. Primary sarcomas of the heart are rare, representing less than 25% of primary cardiac tumors and only 10% of surgically resected tumors. Virmani [8] observed that, from surgically resected tumors, angiosarcomas and undifferentiated sarcomas are commonest histological subtypes.

Considering the age, Burke appreciated there is no gender predilection and the mean age is around 45 years (range 20-80 years); only few cases have been reported in infants [2].

The tumor of the heart was detected as an abnormal finding on a chest radiogram. Taking into consideration the Davies' observations it results that, once detected a cardiac tumor, imaging exploration is needed to define: a) tumor location, extent and boundaries; b) relationships with adjacent key cardiac structures such as valves and coronary arteries; c) tumor type; and d) presence and degree of functional impairment [3,9]. Consistent with these guidelines, in this case the echocardiography was used to assess the ventricular tumor, without identifying the presence of metastases at the time of diagnosis. In this context, Pomerance considers that magnetic resonance imaging is helpful preoperatively to determine precise tumor size, location and adjacent tissue invasion, as well as post-operatively for assessment of recurrence [10].

Concerning the site of the tumor, we documented a cardiac sarcoma with a right ventricular location, while Fletcher found that majority of the cardiac sarcomas were located in the left atrium of the heart and interatrial septum [11]. In other reports, cardiac sarcomas were located, in order of frequency, in the left atrium, right atrium and ventricles [6,8].

In general, the clinical presentation of any primary tumors of the heart depends on its location. Although cardiac sarcomas can involve any cardiac chamber, in our case the tumoral intracavitary growth caused right ventricular outflow obstruction, obliterating progressively the right ventricle [12]. The right location of the sarcoma was associated with congestive heart failure as well. Because the tumor was interfering with valve function resulted in a progressive tricuspid regurgitation. In this case, cardiac symptoms were masked, for a long time, by associated Lyme disease. This severe infection was the cause of a progressive hepato-renal failure that was under therapeutical control.

Macroscopically, similarly with Robinson' considerations, we could appreciate that the appearance of the tumor was not suggestive for a malignant tumoral type [13]. Histologically, the tumor was corresponding with a high grade sarcoma, without a specific line of differentiation, resulting that undifferentiated pleomorphic sarcoma is a diagnosis of exclusion, and immunohistochemical studies are important in ruling out metastatic myogenic, melanocytic as well as sarcomatoid carcinomas. It was demonstrated that the cell of origin of cardiac sarcomas is a mesenchymal cell, a pluripotential cell, giving rise to all types of cardiac sarcomas [14]. In cardiac sarcomas, the histological types are identical with those found in extracardiac soft tissue tumors [8].

Discussing about the tumor grading and staging we can appreciate that there is no TNM classification for cardiac sarcomas. Due to low frequency of malignant cardiac tumors, there is no grading scheme, as well. Burke recommended the use of WHO's criteria for soft tissue tumors [3]. Although there is no grading system for cardiac sarcomas, the presence of the necrosis and high mitotic index is considered an indicative for a low survival rate. A high grade tumor is a tumor with more than one mitosis per HPF, in the most active mitotic area, but tumors with areas of necrosis are considered high grade tumors, whatever the mitotic rate [6].

Referring to the tendency of the right sided cardiac tumor for embolism to the lungs, Burke and Virmani appreciated that chronic embolization may also mimic a chronic thromboembolic disease associated with signs and symptoms of pulmonary

hypertension [3]. In our case this tromboembolic feature could be associated with a hypercoagulopathy state, either as a result of hepatic function disturbance or as a component of a paraneoplastic syndrome.

As in all soft tissue sarcomas, there is a poor survival rate in patients with cardiac sarcomas, suggested by the presence of a high mitotic rate and areas of necrosis [8]. Usually, the prognosis of the cardiac sarcomas is poor and it is measured in months. Silver, noted that pathologic findings associated with increased survival rate, include: left-sided tumors, a low mitotic rate and the absence of necrosis. Even with sarcomas of low mitotic rate that lack necrosis, the long-term outlook is poor and few patients survive 5 years [15].

In this case, the tumor was identified incidentally, because of the presence of embolism and right outflow obstruction. Surgery was undertaken on emergency basis to resolve the present life-threatening symptoms, consisting in a palliative resection in order to relieve obstruction. Burke considers that, the prognosis of PCT is very poor even if complete resection is attempted. Even if adjuvant chemotherapy and irradiation are used, these are not effective in most cases. An alternative treatment option for primary malignant cardiac tumors may be heart transplantation, favourable results being reported, despite immunosuppression [6].

## CONCLUSIONS

The cardiac sarcomas occur rarely and the advanced local stage of the tumour at the time of the diagnosis resulted in a short postoperative survival. However, irrespective of the histological subtype and completeness of surgery, malignant cardiac tumours are a poor prognosis.

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