Primitive Heart Undifferenciated Sarcoma: A Case Report and Literature Review

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Article ID: WMC003579
Article Type: Case Report
Article URL: http://www.webmedcentral.com/article_view/3579
Subject Categories: CANCER
Keywords: Cardiopathy; Sarcoma; Primary Tumor.

How to cite the article: El Yacoubi H, Ouafki I, Inrhaouen H, Mrabti H, Errihani H. Primitive Heart Undifferenciated Sarcoma: A Case Report and Literature Review. WebmedCentral CANCER 2012;3(8):WMC003579

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Source(s) of Funding:
No funding resource

Competing Interests:
No competing interests
Primitive Heart Undifferentiated Sarcoma: A case Report and Literature Review

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Abstract

The primary malignant cardiac tumors are extremely rare, they represent 25% of all primitive cardiac tumors. We report a case of a man aged 45 years with a tumor of the left atrium, discovered by a dyspnea, hemoptysis and faintness, chest X-ray showed a cardiomegaly and pulmonary parenchymal overload. The echocardiography found a mass in the left atrium obstructing the mitral orifice at each diastole. A complete surgical resection was performed and pathological examination found an undifferentiated sarcoma, the patient was lost from view for 13 months. The reappearance of dyspnea related to local recurrence without metastasis has required a reoperation, immediate evolution was favorable. By the realisation of this observation, a literature review of the primary heart sarcoma was performed.

Introduction

The primitive cardiac tumors are extremely rare; their frequency varies from 0.001% to 0.02% based on autopsy series [1]. They are malignant in 25% of cases, represented almost one third by sarcomas [2]. Despite early medico-surgical care, the prognosis remains very serious. We report a case of a patient with primary cardiac sarcoma.

Case Report(s)

A man, aged 45 years presented 3 months before diagnosis dyspnea accompanied by palpitations and dizziness without loss of general condition, cardiac auscultation suggested rotation diastolic mitral valve, the chest radiograph showed cardiomegaly and pulmonary overload. On transthoracic echocardiography there was a mass of the left atrium measuring 60mm obstructing the mitral orifice at each diastole, strongly suggesting a myxoma. Intraoperatively, we discovered a tumor-like myxoma, measuring 75mm/55mm and very adherent to the roof of the left atrium [Illustration.1], resection was difficult but complete, and associated with mitral annuloplasty and rehabilitation of inter atrial septum. Histological examination found an undifferentiated sarcoma. The evolution was favorable and the patient was lost from view for 13 months. A recurrence of symptoms with congestive heart failure and several episodes of syncope, an ultrasound was done, it objectified a local recurrence of the tumor and absence of metastasis on thoraco-abdominal CT scanner. A reoperation was performed for the extirpation of nearly all of the tumor despite an invasion of inter atrial septum and inferior vena cava [Illustration.2]. The follow-up at 3 months, 16 months after the histological diagnosis is satisfactory. The patient was again lost sight and refused any adjuvant therapy.

Discussion

The soft tissue sarcomas are a group of relatively rare malignancies. They represent 1% of malignant tumors in adults and 15% in children. The primary cardiac sarcoma accounts for 30% of primary malignant tumors of the heart [3]. It can occur at any age, but especially between 30 and 50 years without sex preference. Most of the time, clinical examination is poor, the clinical signs are not specific, they depend on the location and extent of locoregional tumor. Their clinical expression may therefore take the mask of a cardiomyopathy, pericardial effusion or valvular heart disease as is the case in our observation. The chest radiograph is non specific, a cardiomegaly can be found as in our case, calcifications, or irregular contour of the cardiac silhouette. Transthoracic echocardiography and particularly transesophageal can usually reveal the existence of a cardiac tumor and suspected malignancy before the invasion of the vena cava, the hemopericardium, akinsia, infiltration of cardiac tissue and the invasion of multiple cavities [4]. MRI is the gold standard, it allows a very fine anatomical diagnosis and eliminate false positive ultrasound, the arguments in favor of a malignant character are heterogeneous after gadolinium injection [5]. Histologically, angiosarcoma is 30 to 40% of primary malignant tumors of the heart with right atrial location (60%) and pericardial (20%). Rhabdomyosarcoma is the primary malignant tumor of the heart the most common in children, he serves both right and left, it is often multifocal. Fibrosarcoma, leiomyosarcoma and liposarcoma are rare [6]. As is
the case with our patient some cardiac sarcomas remain unclassified, these are undifferentiated sarcomas. The differential diagnosis is with benign tumors of the heart and essentially the myxoma. Until 1950 cardiac tumors were not subjected to surgical treatment, it was proposed to chemotherapy and radiation therapy without many standardized protocol. With the introduction of cardiopulmonary bypass the interest of the surgical treatment of cardiac tumors has been developed. As soon as the diagnosis of tumor is established, surgical intervention is indicated to prevent sudden death by engagement of the tumor in the atrioventricular orifice, irreversible heart failure, systemic embolism or rhythm disorders. The surgery not only increase survival, it also improves the quality of life, and allows a beneficial effect on hemodynamics in patients with congestive heart failure or cardiogenic shock. The complete resectability is rare. It depends on the location of the tumor, the extension and the invasion in the myocardium. Heart transplantation may be proposed in cases of local recurrence or unresectable tumors without metastasis [7]. The role of chemotherapy is far from being codified in the management of primary malignant cardiac tumors. This information is retained for most series, for unresectable tumors, tumors metastatic stage in adjuvant or neo adjuvant. It is also often associated with radiotherapy [8]. The primary malignant cardiac tumors are not radiosensitive. Our patient has benefited, as soon as the diagnosis is established, a complete surgical resection without adjuvant therapy (which was refused by the patient himself), reoperation was necessary 13 months after the first intervention which has improved the quality of life for our patient. The cardiac sarcoma primary care, despite early diagnosis and advances in cardiac surgery, a terrible prognosis with a life expectancy of between 6 months and 12 months.

Conclusion

The cardiac sarcomas are rare, their diagnosis should be suspected when unexplained cardiac symptoms. The prognosis of these tumors is very poor, with survival not exceeding 2 years.

References

Illustrations

Illustration 1

Heart sarcoma
Illustration 2

heart sarcoma recurrence
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