

Unique Case of Primary Malignant Fibrous Histiocytoma of the Right Ventricle with Moderator Band Involvement

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ABSTRACT

Malignant primary tumors of the heart are very rare and in most cases are located in the left side of the heart, but involvement of the moderator band of the right heart has not yet been described in the literature. We report the case of a 22-year-old woman who presented after an episode of transient sudden loss of consciousness. The transthoracic echocardiography (TTE) and transesophageal echocardiography evaluations showed an echogenic mass that originated in the right ventricle and protruded into the tricuspid valve during systole. While operating on the patient, we found a sessile tumor that originated exclusively from the moderator band of the right ventricle. The tumor was completely resected through the tricuspid valve. An immunohistochemistry examination of the tumor confirmed the diagnosis of a malignant fibrous histiocytoma (MFH) (undifferentiated pleomorphic sarcoma). To our knowledge, this case is the first of moderator band involvement among the 50 cases of primary MFH of the heart that have been reported in the literature. After a year and a half, TTE, computed tomography (CT), and positron emission tomography CT (PET-CT) showed the absence of cardiac tumor, a normal tricuspid function, and no metastasis. The completeness of the surgical resection and the absence of local recurrence >1 year after surgery are encouraging signs for the patient's outcome.

INTRODUCTION

Tumors of the heart are rare. Reports of autopsy series have indicated a combined incidence of 0.0017% to 0.28% for benign and malignant primary cardiac tumors [Lam 1993; Reynen 1996]. Malignant tumors, the majority of which are sarcomas, constitute up to 25% of cardiac neoplasms [Silverman 1980]. Sarcomas of the heart may consist of a variety

of histologic types. Undifferentiated primary cardiac sarcomas usually develop on the left side of the heart, especially in the left atrium. Right heart involvement is rare, and moderator band involvement has not yet been described in the literature.

We report a case of primary undifferentiated pleomorphic sarcoma that was situated in the right ventricle and originated from the moderator band. To our knowledge, this case is the fourth of right heart involvement and the first of moderator band involvement among the 50 cases of primary malignant fibrous histiocytoma (MFH) of the heart that have been described in the literature.

CASE REPORT

A 22-year-old woman presented with a history of abdominal pain and nausea of 4 days' duration. She also related an episode of transient sudden loss of consciousness 3 days previously. On her admission, the results of a clinical examination, blood tests, and chest radiography were unremarkable.

Transthoracic echocardiography (TTE) results showed a moderate pericardial effusion and an echogenic mass that originated in the right ventricle and protruded into the tricuspid valve during systole. A transesophageal echocardiography evaluation confirmed the presence of a 40 × 50 mm mass located in the right ventricular outflow tract that protruded into the tricuspid valve. The pulmonary valve and pulmonary artery appeared normal. There was a moderate pericardial effusion without signs of pericardial tamponade.

A venous Doppler examination excluded a deep vein thrombosis, and an abdominal echocardiogram revealed no abdominal tumors or adenopathy. A computed tomography (CT) scan of the chest and abdomen also excluded the presence of other thoracic and/or abdominal masses. The differential diagnosis was either a ventricular myxoma or a malignant tumor.

We performed the operation through a median sternotomy and with standard, moderately hypothermic cardiopulmonary bypass. When we opened the right atrium, a reddish yellow mass that protruded through the tricuspid valve into the right atrium came into view (Figure 1). The tumor, which was sessile and originated exclusively from the moderator band of the right ventricle, was completely resected

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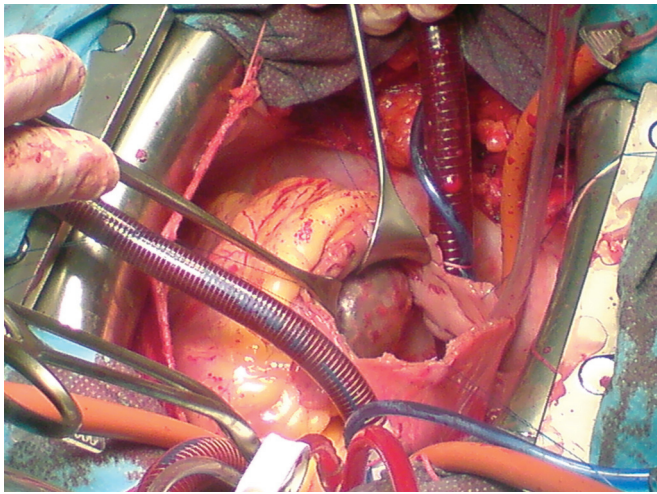


Figure 1. Transatrial view of the tumor.

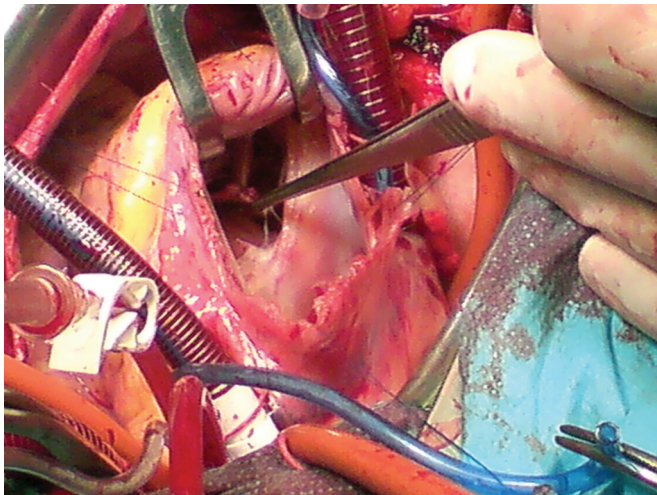


Figure 2. Site of tumor implantation.

through the tricuspid valve (Figure 2). The excised mass measured 50 × 45 × 35 mm and weighed 32 g. The patient's postoperative course was uneventful, and she was discharged on the seventh postoperative day.

An extemporaneous histopathologic examination performed during surgery revealed the presence of a fibrosarcoma, which was completely resected with a good clear margin. A subsequent histopathologic examination with hematoxylin and eosin staining (Figure 3) and an immunohistochemistry examination (Figure 4) confirmed the diagnosis of an MFH (undifferentiated pleomorphic sarcoma). Therefore, chemotherapy treatment was initiated immediately.

One year after the surgery, we performed TTE and positron emission tomography CT (PET-CT) examinations, which demonstrated the absence of cardiac tumor, a normal tricuspid function, and no metastasis. After another 6 months, we repeated the TTE evaluation and the CT scan and obtained the same good result.

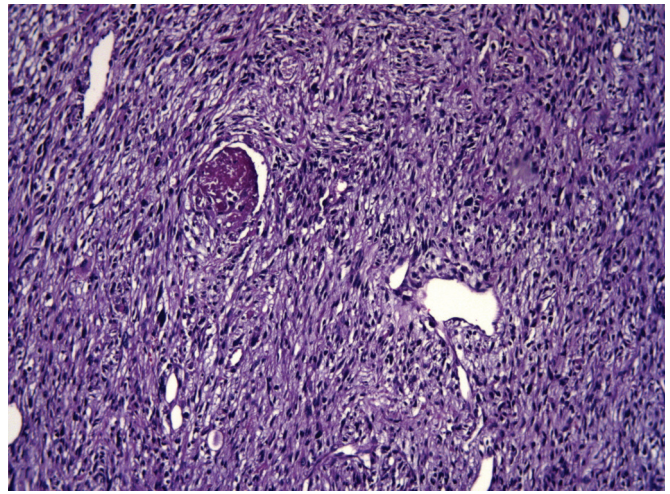


Figure 3. Hematoxylin and eosin stain. Tumoral proliferation consisting of spindle cells arranged in a focally storiform pattern, some with marked pleomorphism.

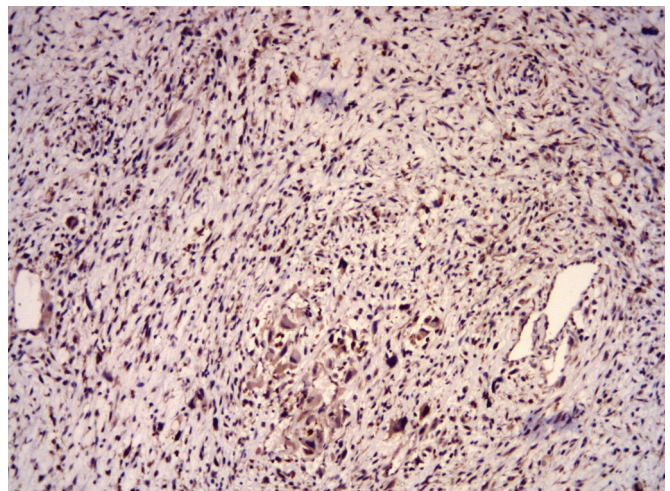


Figure 4. Immunohistochemical stain for S100 protein. Positive reaction in scattered histiocytoid cells with nuclear atypia.

DISCUSSION

Malignant primary tumors of the heart represent <40% of primary heart tumors [Burke 1995]. Most malignant tumors are sarcomas [Burke 1995], which include angiosarcomas, rhabdomyosarcoma, fibrous histiocytoma, myxosarcoma, spindle cell sarcoma, and unclassified sarcoma [Meng 2002]. MFH is regarded as synonymous with undifferentiated sarcoma [Burke 2004], whereas undifferentiated pleomorphic sarcoma is a high-grade malignancy neoplasm that shows fibroblastic or myoblastic differentiation and areas of marked cellular pleomorphism.

To our knowledge, this case is the 50th of primary MFH of the heart to be described in the literature and is the first reported case of involvement of the moderator band of the right heart.

Of the 49 previously reported cases, 39 tumors (81%) were located in the left atrium (including 1 case of a multicentric tumor also found in the left ventricle [Donsbeck 1999]). In other cases, the tumor was located in the pericardium in 4 cases [Tanprasert 1981; Fujio 1986; Wilk 1998; Matsukuma 2008], in the right atrium in 1 case [Donsbeck 1999], and in proximity to an atrial septal defect in another case [Ozawa 1997].

Right heart involvement has been described for only 3 other cases [Kern 1985; Bear 1987; Nawa 1997]. In one of these cases, an MFH of the heart located on the pulmonary valve caused thrombotic occlusion of branches of the left pulmonary artery. In the other 2 cases, the tumor was located in the right outflow tract and the main pulmonary artery [Bear 1987; Nawa 1997], but the moderator band of the right ventricle was involved in neither of these cases, making the present case unique.

The mean age of presentation is approximately 40 years [Burke 1992] with no sex predilection. The symptoms depend on the location and extent of the tumor. Usually, patients are admitted to the hospital after a variable period of symptoms that range from a few weeks to several months. Dyspnea is the most common symptom, followed by chest pain, congestive cardiac failure, palpitations, weakness, fever, and anemia [Putnam 1991].

To our knowledge, this case is the first of sudden onset of the symptoms with a syncopal episode. This type of onset was probably due to the location of the tumor, which almost blocked the blood flow across the tricuspid valve.

Echocardiography is the primary tool for diagnosing intracardiac mass lesions [Edwards 1994; Alam 1995]. Although a number of features help in differentiating sarcomas from benign myxomas, the picture is not easy to interpret [Kim 2003].

CT and magnetic resonance imaging scans of the chest and abdomen complement echocardiography in demonstrating the extracardiac extent of the tumor and the presence of metastasis. MFH tumors usually metastasize, as confirmed by the findings of >20 cases in the literature in which metastatic lesions secondary to probably hematogenous dissemination were observed [Okamoto 2001]. In our case, no metastatic lesions were observed in a CT examination performed at admission.

Complete surgical excision is the only mode of therapy that has been shown to prolong survival [Putnam 1991], which depends on the location of the tumor and the extent of myocardium involvement. In our case, as in 55% of the cases in which involvement was limited to a small part of the ventricle, we managed to perform a complete resection of the tumor.

Survival times for patients with cardiac MFH have ranged from <1 month to 6 years after diagnosis, with a mean survival time of 1 year [Okamoto 2001]. Only 1 patient survived for 6 years, and this individual underwent 5 additional surgeries. In our case, the patient is in very good condition a year and a half after the operation, and TTE, CT, and PET-CT examinations have revealed no recurrences.

Although the patient underwent chemotherapy treatment after the operation with very good results, the role of adjuvant therapy remains undefined in the literature [Llombart-Cussac 1998; Milicic 2007].

CONCLUSIONS

We have described a unique case of MFH with involvement of the moderator band of the right heart. In this case, it is too early to predict the outcome, but the completeness of the surgical resection and the absence of local and general recurrence at a year and a half after the surgery are encouraging signs for the patient's outcome.

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