

Case Report

Primary atrial fibrosarcoma of the heart

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Abstract

The primary fibrosarcoma of the heart is a rare tumor. There are no exact numbers about its incidence, but even among malignant cardiac tumors, only around 3% are fibrosarcomata. Symptoms are nonspecific with signs of right or left heart failure depending on localization. Diagnosis is thereby often delayed. Primary treatment of choice is surgery, followed by several possible postsurgical adjuvant strategies. Survival is poor with a mean of about 11 months. © 2008 Elsevier Inc. All rights reserved.

Keywords: Fibrosarcoma; Heart; Tumor

We present a 54-year-old female patient who presented with precordial pain and dyspnea.

After clinical examination, ECG, echocardiography, computed tomography (CT), and MRI were performed. A lobulated mass was seen in the left atrium, attached to the posterior leaflet of the mitral valve.

The patient was admitted for surgery with a tentative diagnosis of myxoma. After sternotomy, the left atrium was approached through the interatrial septum, showing the mass that macroscopically invaded the posterior leaflet of the mitral valve. The mitral valve and tumor were resected and replaced by a prosthetic valve.

The resected specimen weighed 18 g and consisted of two leaflets with chordae tendinae and the sessile tumor mass on one of the leaflets (Fig. 1). Histology showed a high-grade spindle cell proliferation with a fascicular growth pattern, with 13 mitoses per 10 HPF and focal tumor cell necrosis (Figs. 2 and 3). Immunohistochemically, the tumor was negative on expression of CD31, CD34, EMA, desmin, and myogenin. Since the tumor expressed multifocal cytoplasmatic positivity for α -SMA, diagnosis of fibrosarcoma was preferable.

As adjuvant therapy, adriamycin in monotherapy was chosen. Five months after surgery, local relapse was seen; hence, palliative chemotherapy was started. The patient

died 14 months postoperatively from generalized metastatic disease.

Cardiac fibrosarcoma is a rare malignant tumor originating from the mesenchymal structures of the heart. Based on an autopsy study, the incidence of primary cardiac (benign and malignant) tumors is estimated to be between 0.0017% and 0.35% [1–3]. The overall incidence of fibrosarcoma within the group of malignant cardiac tumors is estimated at about only 3.3% (Table 1). Symptoms are determined by anatomical localization, size, and intracavitary spread of the tumor, more than by histological type. Echocardiography, CT, and MRI applications give an accurate indication about localization, size, shape, point of insertion, mobility, and whether the lesion is sessile or pedunculated [1,2,4]. New techniques like PET scan are mostly used for staging purposes.

Characteristics of the tissue that point to malignant behavior are found on the ultrastructural level. Lesions with more than 10 mitoses per 10 HPF at $\times 400$ magnification and presence of necrotic cells are considered as high-grade malignant and correlate with a poor prognosis [5]. Immunohistochemical staining make further differentiation possible. A correlation between more advanced techniques to map cell proliferation, such as Ki-67 antigen expression or measurements of DNA content, and prognosis has to be further examined [5].

Surgery still stands as the primary treatment for cardiac fibrosarcoma; however, it is rarely curative. The

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Fig. 1. Resection specimen. Notice the yellow-brown tumor, sessile on a part of the posterior leaflet of the mitral valve. The valve apparatus has also been removed.

additional value of radiotherapy and chemotherapy is still doubtful [3,5]. Autotransplantation is a rather experimental technique where the heart is reimplanted after tumor excision [4].

As compared with other sarcomas elsewhere in the body, cardiac sarcomas have a poor prognosis due to the often incomplete resection of the tumor and the immediacy of other important structures. A fast relapse ratio has been described for cardiac lesions [5]. A third factor that contributes to the bad prognosis is the high incidence of local, lymphogenous, and hematogenous metastasis. The following are the most common sites where tumors spread (in order of importance): lungs, thoracic lymph nodes, mediastinum, and spine. Tumor spread (hematogenous in nature) to pericardium, liver, kidneys, suprarenal glands, pancreas, bone, spleen, bowel, brain, and thyroid gland is rather rare [1,5]. Overall median survival seems to vary between 6 and 9 months [2,5].

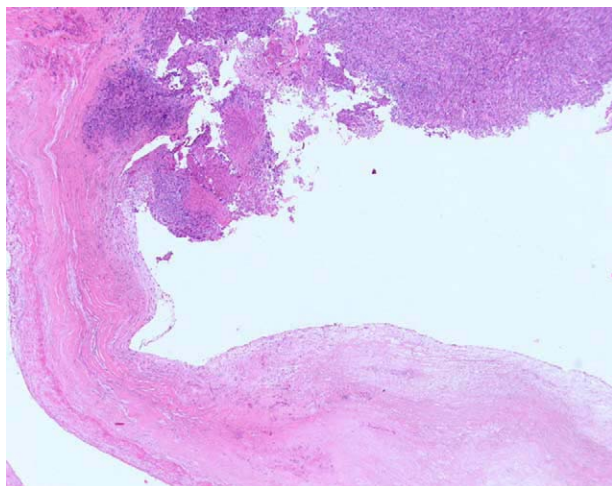


Fig. 2. Low-power view showing the base of the mitral valve (bottom) and the fibrosarcoma (H&E, ×25).

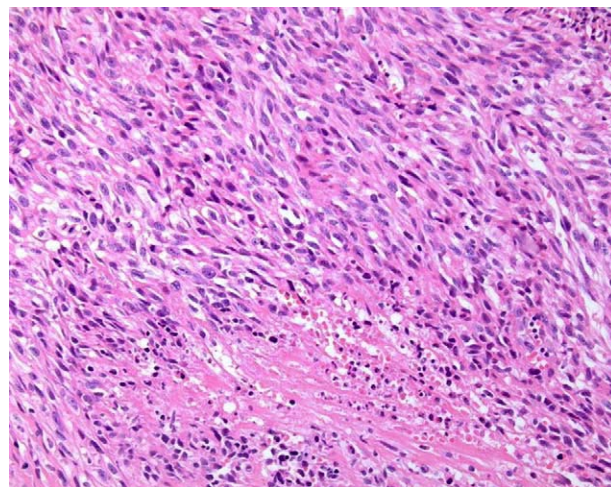


Fig. 3. High-power magnification of the fibrosarcoma, composed of spindle cells arranged in fascicles. Mitoses are present, as well as focal necrosis of the tumor (bottom) (H&E, ×200).

1. Summary

The primary fibrosarcoma of the heart is a rare tumor with an incidence of only around 3% within the group of malignant cardiac tumors. Diagnosis is often delayed because of nonspecific symptoms. In spite of surgery and several possible postsurgical adjuvant strategies, survival is poor.

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