Early Recurrence of Malignant Fibrous Histiocytoma of the Heart

YELDA BAŞARAN, M.D., MUZAFFER DEĞERTEKIN, M.D., MEHMET BALKANAY, M.D.,* ESAT AKINCI, M.D.,* ÇIĞDEM ÇELIKEL, M.D.,** HANDAN DIZDAR, M.D.,** FIKRET TURAN, M.D., and CEVAT YAKUT, M.D.*
Departments of Cardiology and *Cardiovascular Surgery, Koşuyolu Heart and Research Hospital; and **Department of Pathology, Marmara University Medical Faculty, İstanbul, Turkey

Malignant fibrous histiocytoma is a pleomorphic soft tissue sarcoma, which constitutes only 2% of all cardiac malignancies and is typically located in the left atrium. We report a young male patient with malignant fibrous histiocytoma located on the right side of the heart. Early recurrence was observed after extensive surgical resection to relieve symptoms of outflow tract obstruction. Noninvasive evaluation and management with regard to the literature are discussed. (ECHOCARDIOGRAPHY, Volume 13, March 1996)

malignant fibrous histiocytoma

Primary cardiac tumors are uncommon. In large autopsy series, their incidence has ranged between 0.01% and 0.28%. Malignant tumors constitute only 25% of them; 75% are sarcomas that are more commonly found on the right side of the heart. Metastatic tumors occur 20 times more frequently than primary ones.[1-5] According to previous reports, angiosarcoma is the most frequent type of tumor, followed by malignant fibrous histiocytoma (MFH), mesothelioma, rhabdomyosarcoma, leiomyosarcoma, and primary lymphoma.[4-8]

Although MFH is the most frequently observed soft tissue sarcoma in adults, its occurrence in the heart is extremely rare. When it does occur, MFH arises almost exclusively in the left atrium, more frequently in the younger population with female predominance. The tumor is considered to originate from primitive mesenchymal cells composed of spindle and round forms arranged in a storiform pattern.[9-14] Sequential malignant transformation of cardiac myxoma to MFH and cardiac metastasis from soft tissue form have been reported.[15,16]

Symptomatology differs according to the size and location of the tumor in the heart. If the tumor obstructs cardiac outflow, it causes symptoms such as dyspnea and chest pain, eventually leading to congestive heart failure.

Malignant cardiac tumors are associated with poor long-term survival despite surgery, radiation, and/or chemotherapy. However, surgical resection is often required to relieve symptoms and may contribute to better survival in certain patient subgroups.[4-6,8] The overall actuarial survival for patients with primary cardiac sarcomas has been reported as 14% at 24 months with a median survival of 11 months.[4]

Case Report

In April 1994, a 24-year-old male was admitted to our hospital with symptoms of progressive exertional dyspnea, chest pain, and increasing fatigue for 3 months. On physical examination, his body temperature was 37.5°C, pulse rate 96 beats/min regular, and blood

Address for correspondence and reprints: Yelda Başaran, M.D., Koşuyolu Heart and Research Hospital, 81020 Kadiköy, İstanbul, Turkey. Fax: 90-216-339-04-41.

pressure 90/70 mmHg. His jugular veins were distended.

On chest examination, there was basal dullness on the left side with depression of respiratory sounds but no rhonchi. Cardiac examination was unremarkable. The liver was palpated 3 cm below the costal margin, and minimal ascites was found on abdominal examination.

The ECG revealed sinus rhythm with incomplete right bundle branch block, low voltage, and flat T waves in the precordial leads. Teleroentgenogram showed mild cardiac enlargement and moderate left-sided pleural effusion.

Computed tomography of the thorax revealed right minimal, left moderate pleural, and 1.5 cm of pericardial effusion all around the heart. The right ventricle was filled with an irregular mass that extended to the conus pulmonalis. The lower lobe of the right lung was collapsed. Mediastinum was not enlarged, and heart size was considered within normal limits.

On two-dimensional echocardiographic examination, the right ventricle was obliterated from view by a nonhomogeneous mass that was partially obstructing the right ventricular outflow tract (Fig. 1). Pericardial effusion was measured 1.45 cm posterior and 1.96 cm on the anterior wall of the heart. Color Doppler showed turbulent flow as a mosaic of colors at the right ventricular inflow and outflow tracts (Fig. 2). The tricuspid valve was considered to be functioning normally.

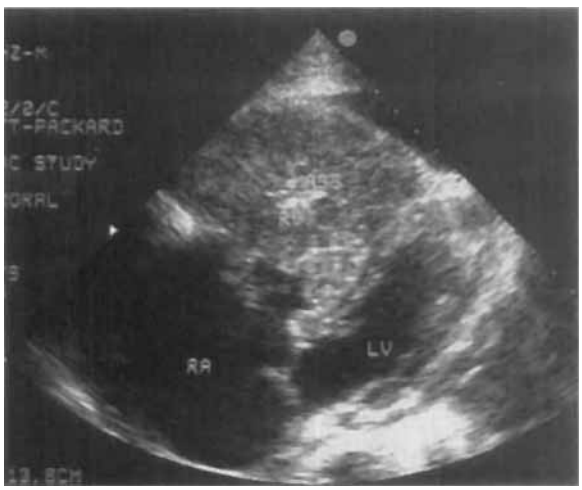

Figure 1. *Apical four-chamber view showing obliteration of the right ventricle (RV) by a nonhomogeneous mass. RA = right atrium; LV = left ventricle.*

Abdominal ultrasonography revealed hepatomegaly and ascites but no sign of metastasis. Scintigraphic bone survey was normal.

Blood chemistry studies showed high transaminase and alkaline phosphatase levels. Erythrocyte sedimentation rate was within normal limits.

At operation, multiple organized thrombotic foci located on the atrial wall were excised through right atriotomy. All three leaflets of the tricuspid valve were infiltrated by the tumorous mass protruding from the right ventricle. The right ventricle itself was filled with a brownish gelatinous fragile mass. Complete resection of this mass was attempted, and the tricuspid valve replaced with a #23 Biocor prosthesis.

Microscopic examination of the tumor revealed a neoplasm composed of tapering spindle cells arranged in interlacing bundles and forming whorls in cartwheel and storiform patterns (Fig. 3). The tumor cells showed moderate nuclear pleomorphism and obvious mitotic activity (Fig. 4).

Collagen and reticulin fibers were uniformly distributed but scarce (Fig. 5). Myxoid change was not detected with Alcian Blue pH 2.5 stain.

Immunohistochemical stains using the streptavidin-biotin alkaline phosphatase method were performed for cytokeratin, s-100 protein, desmin, and alpha-1-antichymotrypsin. All immunostains except for alpha-1-antichymotrypsin were negative (Fig. 6).

Postoperative echocardiographic evaluation was satisfactory (Fig. 7), and the patient was then referred to the oncology center.

The patient presented 2 months later with symptoms of congestive heart failure. No other therapeutic procedures had been performed during that interval.

Echocardiographic examination showed a normally functioning bioprosthesis in the tricuspid position. The right ventricular mass appeared almost the same as it had preoperatively (Figs. 8 through 11).

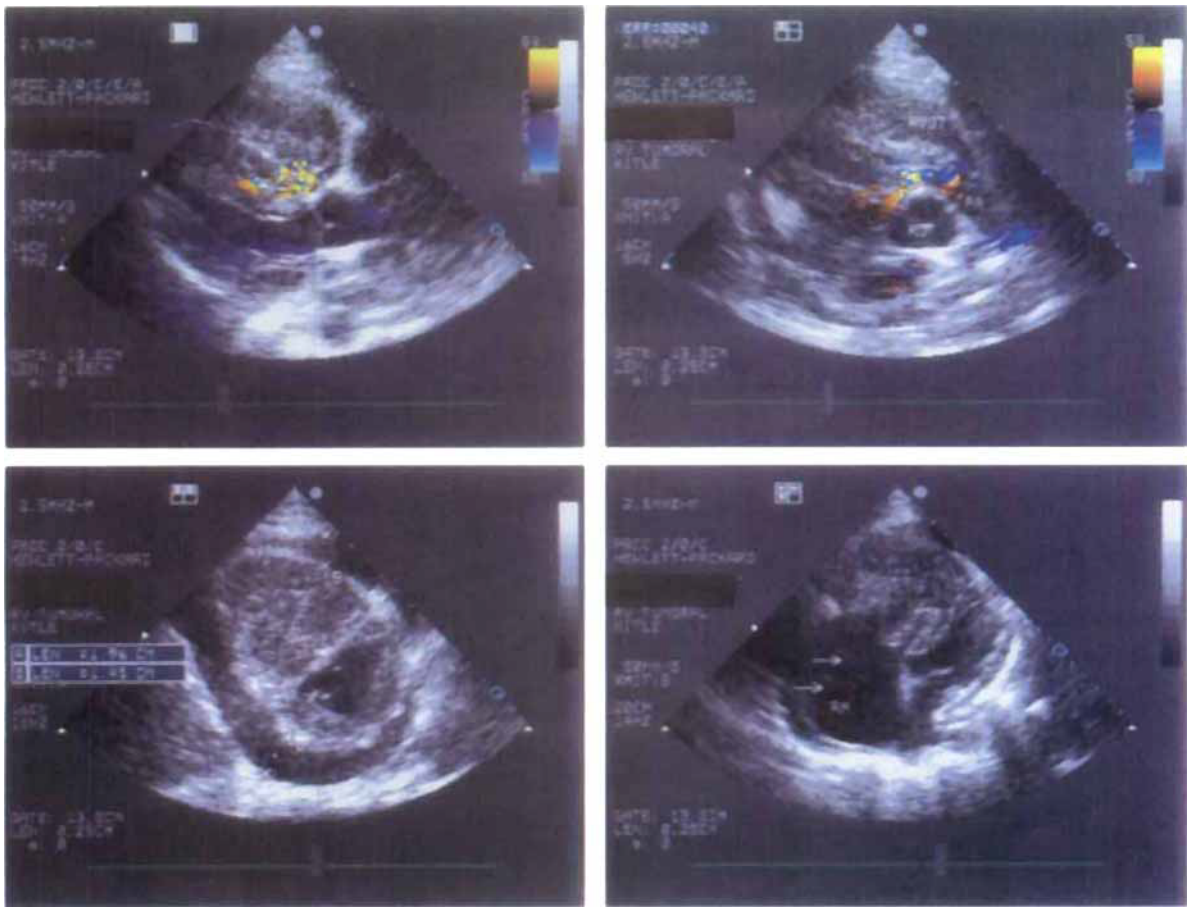

Figure 2. *Parasternal long- and short-axis views showing different appearances of the right ventricular mass with pericardial effusion. Ao = aorta; PA = pulmonary artery; RVOT = right ventricular outflow tract.*

Magnetic resonance imaging revealed almost complete occlusion of the right ventricle by the tumorous mass with left ventricular compression (Fig. 12). Despite supportive therapy, the patient died from progressive disease on the fourth day of his second admission.

Discussion

Cardiac tumors may produce hemodynamic disturbances depending on their anatomical location. Symptoms and physical findings are related to pericardial, myocardial, and endocardial involvement. The degree of intracavitary obstruction determines the severity of symptoms.[1-5] Two-dimensional echocardiography has become the most important method for noninvasive detection of cardiac tumors with very high sensitivity.[1-5] Although most cardiac tumors are diagnosed, they are usually inoperable due to their location or infiltrative growth into one or more cardiac chambers.[1-6]

Computed tomography and magnetic resonance imaging can provide full presentation of the heart and thorax, including the extent of invasion into surrounding structures.[4,17,18] Echocardiographically, the sarcomas are visualized as intramural and intracavitary masses, and it may be difficult to distinguish them from their counterparts. Direct biopsy is usually necessary for accurate histopathological diagnosis.[4,5]

Because of limited experience in the treatment of primary cardiac sarcomas, general strategies for management are not clear.[4,5,8] Since they produce symptoms related to obstruction, aggressive resection in these pa-

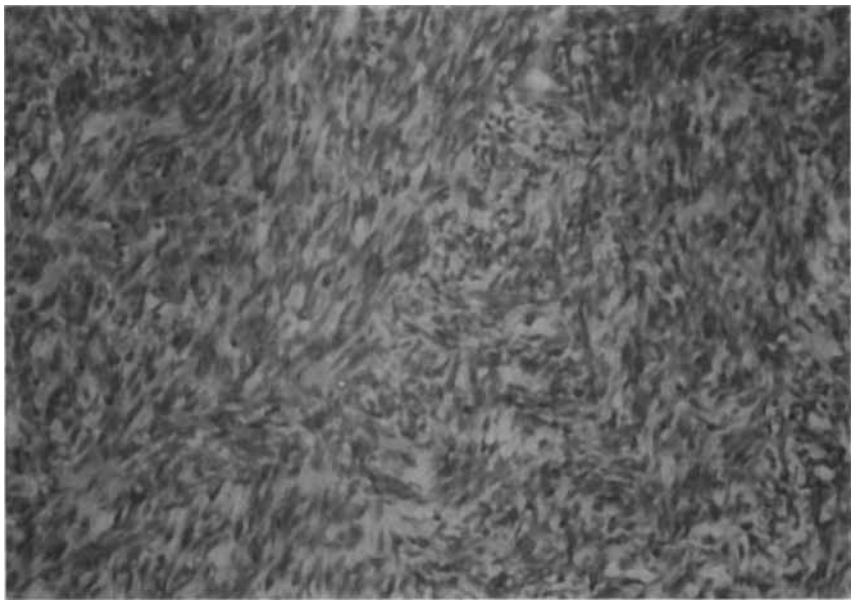

Figure 3. *Storiform pattern of fusiform cells with tapered nuclei is seen (hematoxylin-eosin stain, × 200).*

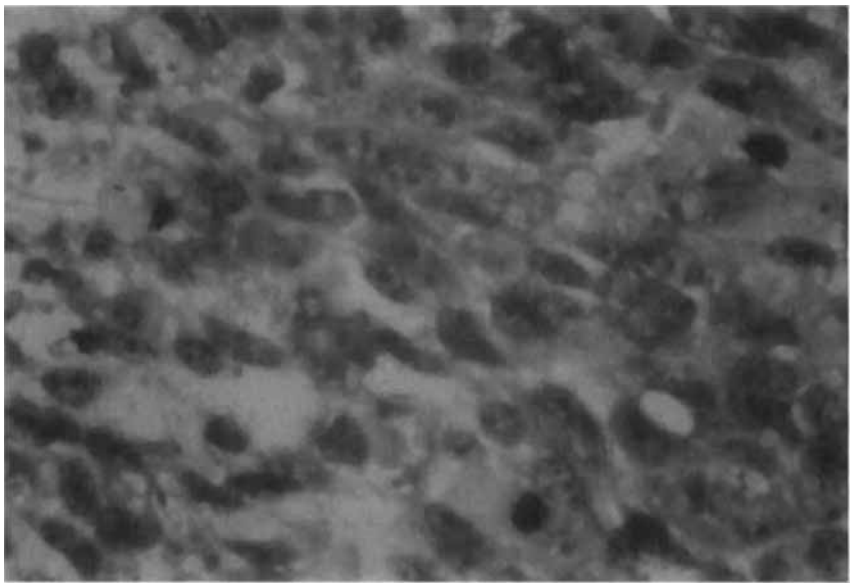

Figure 4. *The neoplastic cells show moderate pleomorphism but a rather high mitotic rate, which are indicative of malignancy (hematoxylin-eosin stain, × 200).*

tients is inevitable.[3-5] According to previous reports, radiation and chemotherapy are not sufficient to relieve symptoms.[3-6,8] In the absence of randomized clinical trials, it is not known whether adjuvant chemotherapy is beneficial in patients in whom curative surgery has been

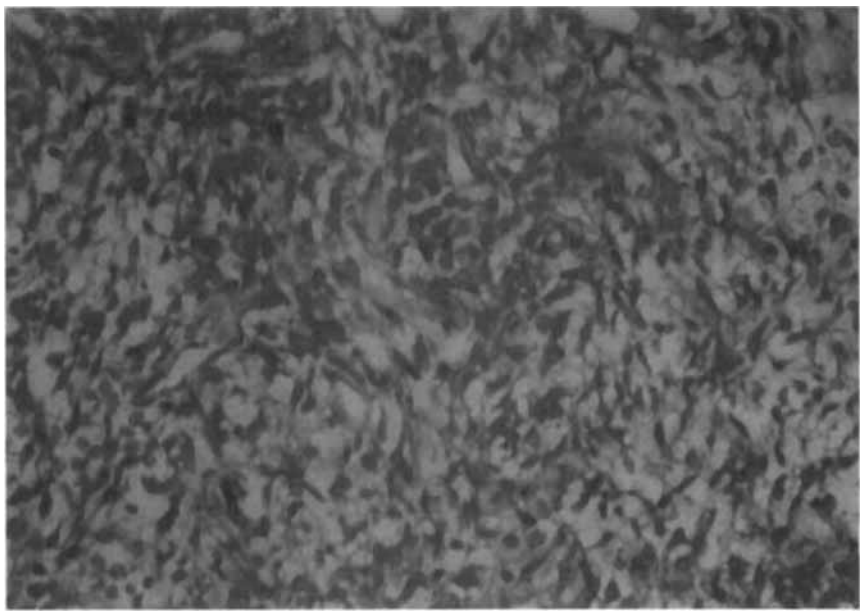

Figure 5. *Storiform pattern, moderate cellular pleomorphism, and scarce collagenous stroma are seen (Mason trichrome, × 200).*

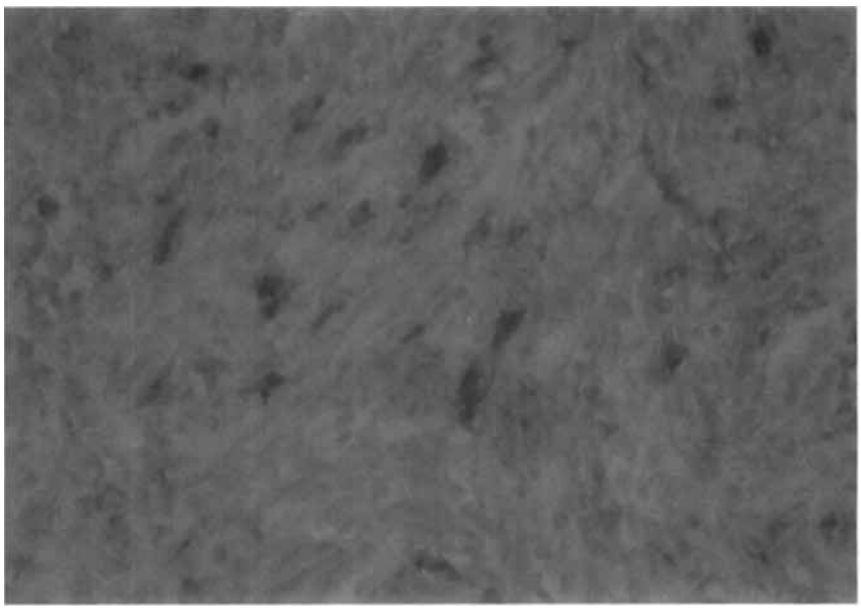

Figure 6. *Positive immunoreaction with alpha-1-antichymotrypsin is seen (× 200).*

performed. The prognosis of intracardiac MFH is poor despite various regimens of surgery, radiation, and/or chemotherapy.[8-12] According to Burke et al.,[5] age, gender, presence of differentiation, and histopathological type do not affect prognosis. Low mitotic activity, absence

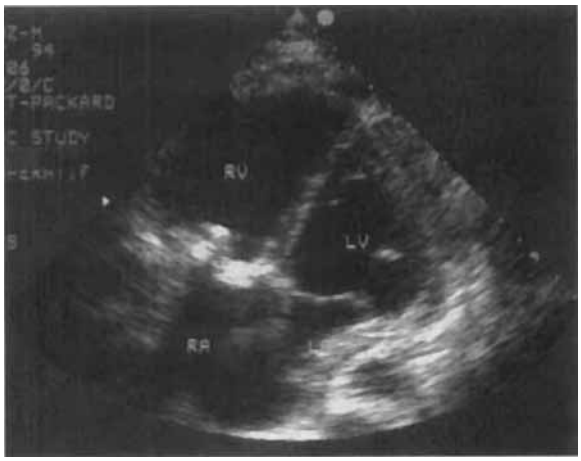

Figure 7. *Apical four-chamber view taken on the third postoperative day showing complete resection and bioprosthesis in the tricuspid position. LA = left atrium. Other abbreviations as in previous figures.*

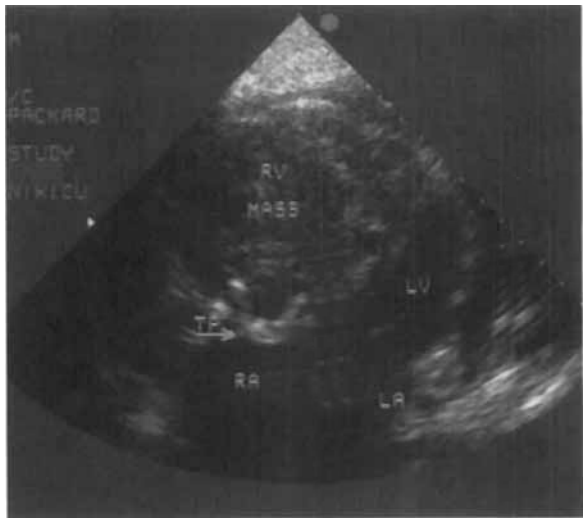

Figure 8. *Apical four-chamber view showing recurrence of the right ventricular mass with the prosthesis in the tricuspid position.*

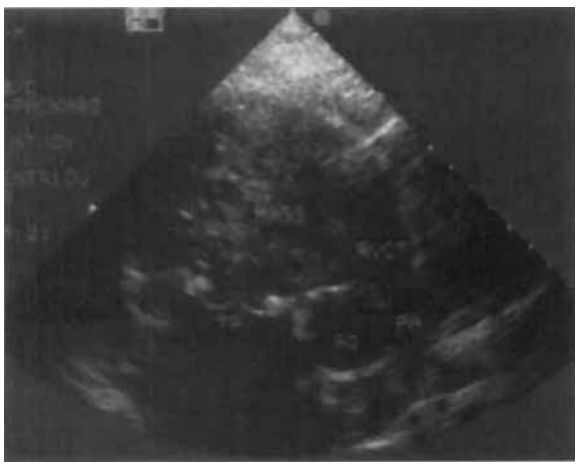

Figure 9. *Postoperative image of the mass extending into the right ventricular outflow tract (RVOT).*

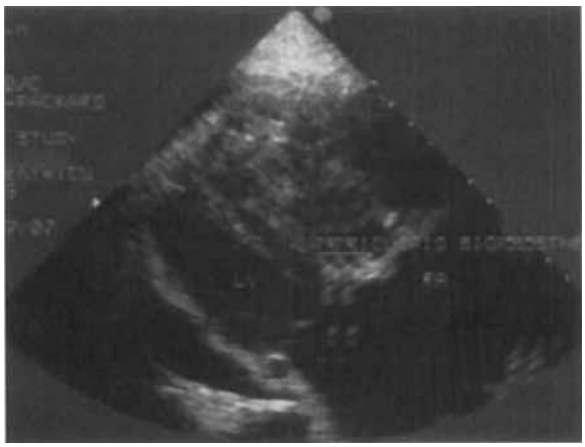

Figure 10. *Postoperative appearance of the right ventricular inlet showing the mass and bioprosthesis.*

of necrosis, and any type of therapeutic intervention can significantly improve survival rate.[5,8,9]

The rapid course and early recurrence in our case demonstrate that extensive resection did not render a satisfactory result. Our clinical experience and previous reports highlight that complete resection of malignant cardiac tumors can provide substantial relief of symptoms but cannot provide good long-term prognosis. Thus, we suggest that more radical approaches such as transplantation be performed on unresectable malignant cardiac tumors.

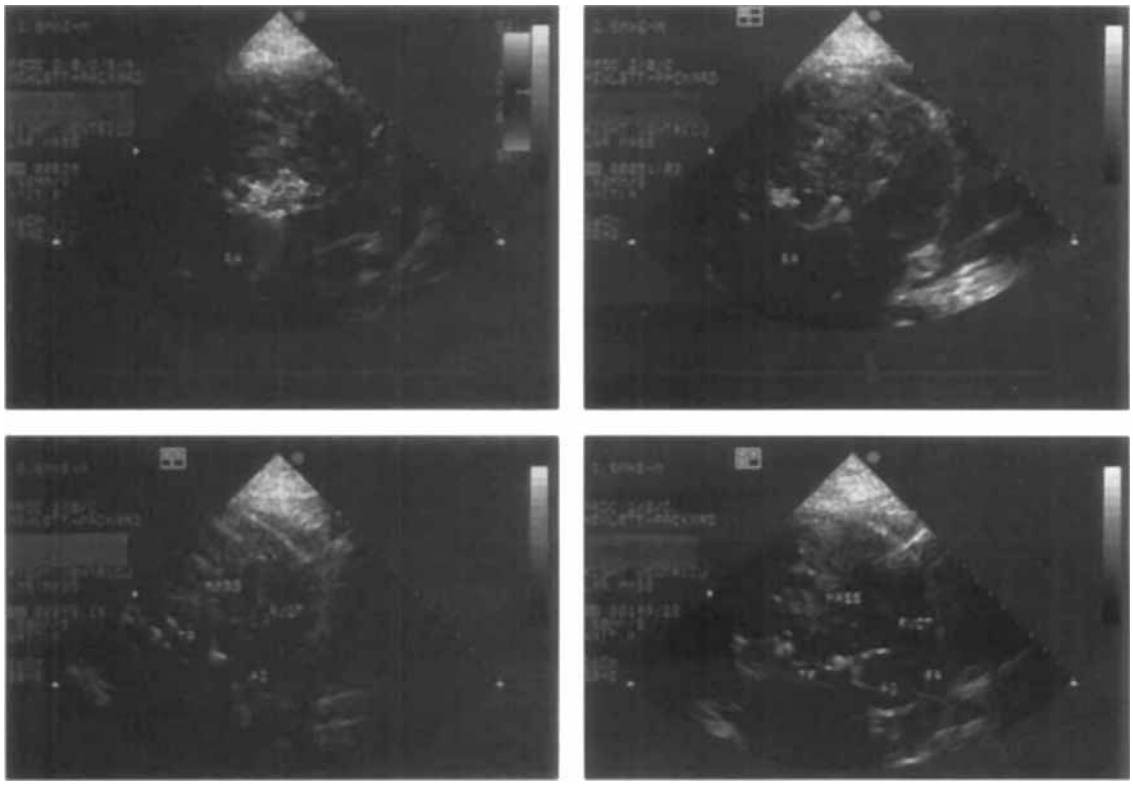

Figure 11. *Different postoperative images of the mass and bioprosthesis.*

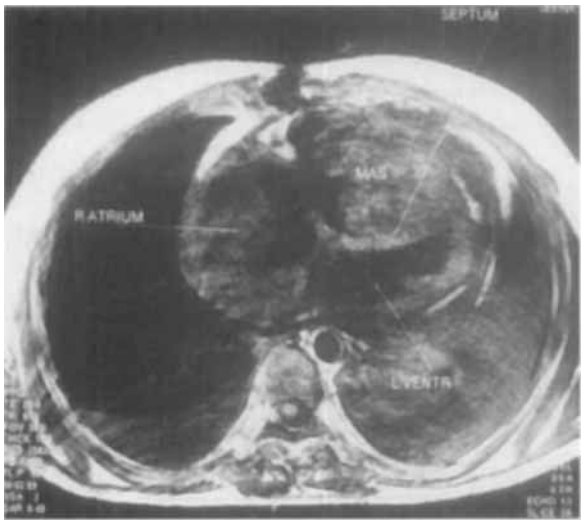

Figure 12. *Magnetic resonance imaging showing almost complete occlusion of the right ventricle with left ventricular compression.*

References

1. Dapper F, Görlah G, Hoffmann C: Primary cardiac tumors—Clinical experiences and late results in 48 patients. *Thorac Cardiovasc Surg* 1988;36:80-85.
2. Dein J, Frist W, Stinson E, et al: Primary cardiac neoplasm—Early and late results of surgical treatment in 42 patients. *J Thorac Cardiovasc Surg* 1987;93:502-511.
3. Verklala K, Kupari M, Maamies T, et al: Primary cardiac tumors—Operative treatment of 20 patients. *Thorac Cardiovasc Surg* 1989;37: 361-364.
4. Putnam J, Sweeney M, Colon R, et al: Primary cardiac sarcomas. *Ann Thorac Surg* 1991; 51:906-910.
5. Burke A, Cowan D, Virmani R: Primary sarcomas of the heart. *Cancer* 1992;69:387-395.
6. Bear PA, Moodie DS: Malignant primary cardiac tumors. *Chest* 1987;92:860-862.
7. Tazelaar HD, Locke TJ, McGregor CG: Pathol-

ogy of surgically excised primary cardiac tumors. *Mayo Clin Proc* 1992;67:957-965.
8. Moriyama Y, Saigenzi H, Shimokaça S: Surgical treatment of primary cardiac tumors. *J Jpn Assoc Thorac Surg* 1993;41:367-371.
9. Laya M, Milliard J, Bewtra C: Malignant fibrous histiocytoma of the heart. A case report and review of the literature. *Cancer* 1987;59: 1026-1031.
10. Ovcak Z, Masera A, Lamovec J: Malignant fibrous histiocytoma of the heart. *Arch Pathol Lab Med* 1992;116:872-874.
11. Sasaki K, Tamaka S, Ikashita M, et al: Malignant fibrous histiocytoma of the heart. A case report and review of the literature. *J Jpn Assoc Thorac Surg* 1991;39:2240-2245.
12. Korbmacher B, Doering C, Schulte HD: Malignant fibrous histiocytoma of the heart—A case report of a rare left atrial tumor. *Thorac Cardiovasc Surg* 1992;40:303-307.
13. Wahba A, Liebold A, Birnbaum DE: Recurrent malignant fibrous histiocytoma of the left atrium in a 27 year old male. *Eur J Cardiothorac Surg* 1993;7:387-389.
14. Suziki Y, Ikeda H, Kanemaki: An autopsy report of malignant fibrous histiocytoma of the left atrium and a review of the pertinent literature. *Jpn J Cancer Clin* 1990;36:169-175.
15. Kasugai T, Sakurai M, Yutani C: Sequential malignant transformation of cardiac myxoma. *Acta Pathol Jpn* 1990;40:687-692.
16. Zalosnik AJ, Pezzella AT: Management of cardiac metastasis from malignant fibrous histiocytoma. *Cancer* 1988;61:602-604.
17. Cutton C, Shulman H, Rusthoven J: Gated magnetic resonance imaging of metastatic intracardiac malignant fibrous histiocytoma. *Can Assoc Radiol J* 1990;41:96-97.
18. Freedberg R, Kronzon I, Rumancik W: The contribution of magnetic resonance imaging to evaluation of intracardiac tumors diagnosed by echocardiography. *Circulation* 1988;77:96-103.