Survival Following Treatment of a Cardiac Malignant Fibrous Histiocytoma*

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The authors report the successful surgical and chemotherapeutic management of a malignant fibrous histiocytoma with localization in the heart. This rare malignancy has specific morphologic characteristics and is diagnosed in adults when it is already locally aggressive or metastasized. The average survival time for patients with malignant fibrous histiocytoma is < 12 months. Neither clinical nor instrumental signs of recurrence have been detected in this patient after an aggressive surgical resection and a specific therapeutic approach.

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Key words: chemotherapy; heart; long-term survival; malignant fibrous histiocytoma

Abbreviations: MFH = malignant fibrous histiocytoma; RM = rhabdomyosarcoma

Malignant fibrous histiocytoma (MFH), formerly known as pleomorphic rhabdomyosarcoma (RM), is considered to be a quite rare malignancy that might grow within several localized areas, occasionally in the heart. It is usually diagnosed when it is locally aggressive or has already metastasized. At that stage, a relatively satisfactory therapeutic regimen seems to be complete resection followed by chemotherapy. Nevertheless, the average survival time of patients with this condition always has been described in the literature as being < 12 months.

We report the successful surgical management, with long-lasting chemotherapeutic survival, of a patient with localization in the heart and describe the histologic characteristics.

CASE REPORT

A 71-year-old woman was admitted to the hospital with a 2-month history of progressive dyspnea, chest discomfort, abdominal tension, asthenia, and 9-kg weight loss. Physical examination revealed hepatomegaly, ascites, and lower limb edema. Neither ECG signs of arrhythmias nor specific serologic marker elevation were reported. Abdominal ultrasonography excluded the presence of liver or vascular diseases. Bidimensional echocardiography displayed an enlarged right atrium that was partially filled with an echogenic thrombus-like mass. The mass was consistently protruding through the right atrioventricular orifice at diastole and was almost completely occluding the right ventricular inflow. A thoracoabdominal MRI (Fig 1) showed a large, nonhomogenous, solid mass in the right atrium that extended into the inferior vena cava and the suprahepatic veins. No metastases were detected elsewhere after a thorough survey, which included CT scans of the head, the abdomen, and the total body.

During surgery, a cardiopulmonary bypass was performed by cannulation of the ascending aorta, superior vena cava, and right common femoral vein to allow a more extensive inspection of the inferior vena cava. After aortic cross-clamping, cardiac arrest was induced by the infusion of a cold crystalloid cardioplegic solution into the aortic root. The right atrial wall was longitudinally incised to reveal the presence of a smooth and brownish-gray tissue firmly originating from the fossa ovalis but also strongly adherent to the posterior atrial wall itself (Fig 2). No involvement of the tricuspid valve was evident. The mass was entirely excised from the atrial wall, together with a rounded, 2-cm diameter isle of the interatrial septum, from which it originated. Circulatory arrest was not necessary during this phase. The inferior vena cava wall appeared irregularly hard to the touch, with several bumps all over its thickness, which is suggestive of consistent infiltration. Yet, its lumen appeared to be completely patent. The interatrial septal defect created to remove the original site of the mass was repaired with a patch of autologous pericardial tissue, and the right atrium was closed easily with a 5–0 polypropylene (Prolene) continuous suture. Steady hemodynamic parameters allowed a safe weaning of the patient from the cardiopulmonary bypass.

The patient had an uneventful recovery and was discharged from the hospital on postoperative day 7. One month later, she underwent three consecutive cycles of combined chemotherapy with doxorubicin (25 mg/m²), vincristine (2 mg/m²), and cyclophosphamide (37 mg/m²) at 3-week intervals. She showed neither instrumental signs nor clinical symptoms of recurrence during both her periodic follow-up and at 36 months after resection, which gave her an excellent long-term outcome.

At a macroscopic examination, the resected neoplasm had an oval shape, weighed 167 g, and had an overall dimension of 7.5 × 5 × 3.5 cm, with most of the tissue being necrotic. A lobulate architecture with a grayish color was evident at the cut surface. Hematoxylin-
eosin-stained sections (Fig 3) showed a pleomorphic, highly cellular tumor that did not extend beyond the fibrous capsule and that had a sarcomatous pattern consisting both of immature areas of oval cells arranged in sheets and areas of large spindle and giant multinucleated bizarre cells in strands (Fig 4). The presence of a larger cell population that reacted positively to immunohistochemical stains for actin, desmin, and myoglobin confirmed the final diagnosis of malignant fibrous histiocytoma. A positive response to vimentin and α-1 actin was also evident. Neoplastic cells otherwise appeared to react negatively to smooth-muscle actin, S-100, and creatine kinase-pool proteins.

**COMMENTS**

RM s are tumors found mainly in young patients. Although embryonal, botryoid, and alveolar RM s have been described chiefly in children and adolescents, pleomorphic RM has been considered the main type of RM in adults. In the 1960s, most of the tumors previously known as pleomorphic RM were reclassified as MFH. Nevertheless, MFH is currently defined as a rare malignanc y, and its diagnostic entity is continuously a subject of debate among pathologists. Despite several localizations having been described in the past, only exceptional cases primarily involve the heart.

An appropriate identification can be obtained by clinical presentation and light microscopy, but a definite diagnosis still requires either immunohistochemical or ultrastructural confirmation. These diagnostic procedures rely on several criteria, which already have been defined in literature, including the presence of typical spindle and polygonal (strap-like) cells that are filled with an abundant eosinophilic cytoplasm, cells with cross-striations, and, particularly, desmin- and myoglobin-positive immunoreactivity. However, some of these criteria remain controversial. Particularly, the presence of cytoplasmic filaments is not always found in MFH and makes it harder to establish a reliable differential diagnosis.

These tumors cause no clinical manifestations early in their course. Furthermore, there is no association with a serologic increase of specific neoplastic cellular markers. With localization to the heart, the impairment of the cardiac output leads to symptoms (e.g., dyspnea, chest pain, congestive heart failure, or arrhythmias) that inevitably require surgical resection. At this stage, instrumental diagnosis usually reveals a large mass or a diffuse metastasization with an extensive myocardial infiltration, which is the main cause of therapeutic failure. Nevertheless, an accurate resection of the tumor is required to attenuate symptoms and to improve postoperative survival. Because of such high aggressiveness, the average survival time is usually considered to be < 12 months. Despite the availability of valid chemotherapeutic regimens, the prognosis remains poor.

We have reported a case of malignant sarcoma of the heart in which the coupling of an extensive surgical resection with a mild chemotherapy regimen has resulted in an unprecedented survival time compared to cases previously described. The most recent reviews of the management of cardiac tumors highlight the predominance of sarcomas among all primary malignant neformations of the heart. They involve mainly adults (mean
age, 38.6 years) with an overall actuarial survival of 14% at 24 months. Although the effect of each therapeutic option is mainly palliative, the timing and extent of the approach are still a matter of debate. The future improvement in both surgical techniques and myocardial protection together with a thorough diagnostic survey might further improve the prognosis in these patients.

Our report demonstrates that, even in the presence of a reasonably bulky primary cardiac malignancy, and despite severe hemodynamic impairment, an aggressive surgical management with effective adjuvant therapy makes a complete remission still achievable, even in patients of advanced age.

REFERENCES
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