CASE REPORT
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Autotransplantation of the Heart for Recurrent Inflammatory Myofibroblastic Tumor

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We report a rare case of dyspnea caused by a cardiac tumor in a 53-year-old woman. The patient had undergone a cardiac tumor (inflammatory myofibroblastic tumor, 6.2 × 4.2 × 3.3 cm) resection at our institute 13 months earlier. We performed preoperative evaluations which revealed a cardiac tumor originating from the posterior wall of the left atrium. Cardiac autotransplantation surgery (cardiac explantation, ex vivo tumor resection, cardiac reconstruction, and cardiac reimplantation) was successfully performed for the complete resection of the recurrent tumor without major postoperative complications. The patient showed good physical conditions for 21 months after the surgery. Cardiac autotransplantation is a safe and feasible technique for the complete resection of complex left atrial tumors.

Keywords: Heart; Transplantation; Autologus; Myofibroblastic Tumor

INTRODUCTION

According to surgery and autopsy reports, primary cardiac tumors occur in only 0.3%–0.7% of all cardiac tumor cases (1). Only 25% of primary cardiac tumors are malignant, and 75% of all malignant tumors in the heart are sarcomas. Complete resection is the optimal treatment for cardiac tumors. Chemotherapy and radiotherapy have been employed with little success to prolong survival compared with surgery (2). However, complex cardiac tumors located in the left atrium (LA), which invade vital cardiac structures, are considerably difficult to access and completely resect with the standard open heart surgery technique. Cardiac autotransplantation (cardiac explantation, ex vivo tumor resection, cardiac reconstruction, and cardiac reimplantation) can be a useful tool in such cases for the complete resection of the cardiac tumor.

CASE DESCRIPTION

A 53-year-old woman visited our institute in July 2007 complaining of exertional dyspnea which had started 7 days earlier. She had undergone a heart surgery 13 months earlier (June 14, 2006) for the resection of a tumor in the LA at our institute. Intraoperative frozen biopsy of the mass revealed myxoma but the postoperative histological diagnosis was inflammatory myofibroblastic tumor (IMT). A crackle in both lung fields with grade I diastolic murmur was noticed on auscultation. Electrocardiography findings were normal but a simple chest roentgenography revealed pulmonary edema. Transthoracic echocardiography (Fig. 1A) and computed tomography (CT) (Fig. 1B) showed a 6.2 × 4.2 × 3.3 cm pedunculated mass originating from the posterior wall of the LA to the left ventricle obstructing mitral valve inflow.

Cardiac autotransplantation was planned and performed on July 13, 2007 for the complete resection of the tumor. Arterial cannulation for the right common femoral artery and venous cannulation for the superior vena cava (SVC) and common femoral vein were employed for a cardiopulmonary bypass (CPB). Moderate systemic hypothermia was obtained with the CPB. The ascending aorta was cross-clamped and a cardioplegic histidine-tryptophan-ketoglutarate (HTK) solution (Dr. Franz Kohler Chemie GmbH, Bensheim, Germany) was administered via a root cannula in the ascending aorta. The heart was explanted and preserved in a solution of iced saline & HTK solution

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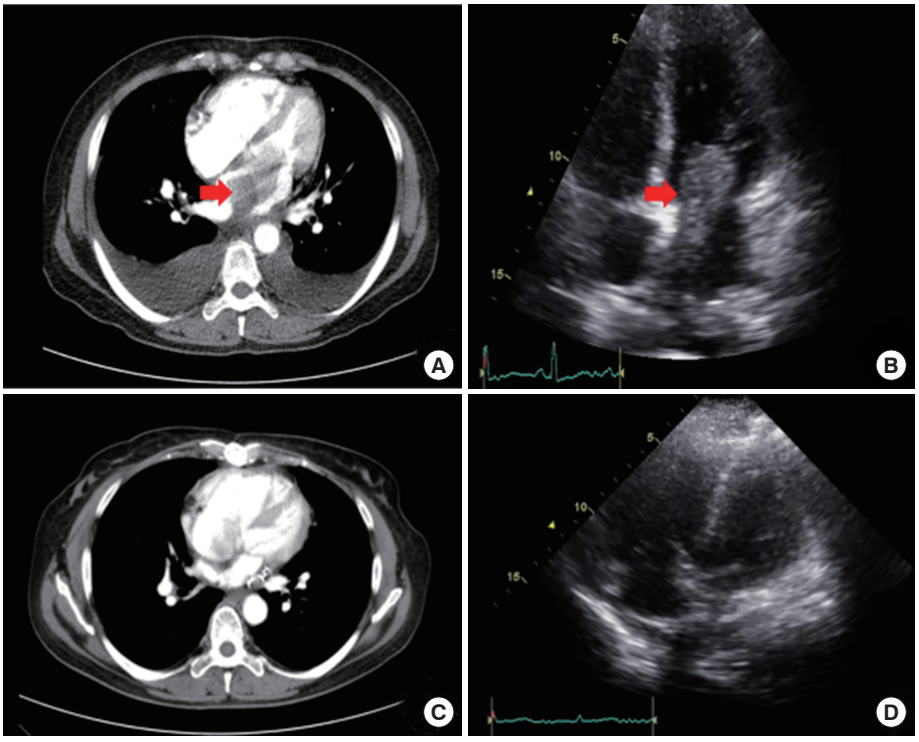

Fig. 1. Preoperative and postoperative images of CT and TTE. (**A**) Preoperative CT scan showing the stalk of the tumor arising from the posterior wall of the LA (arrow). (**B**) Preoperative echocardiography showing a huge mobile mass in the LA obstructing mitral valve inflow (arrow). (**C**, **D**). Postoperative CT and TTE showed no remnant mass.
CT = computed tomography, TTE = transthoracic echocardiography, LA = left atrium.

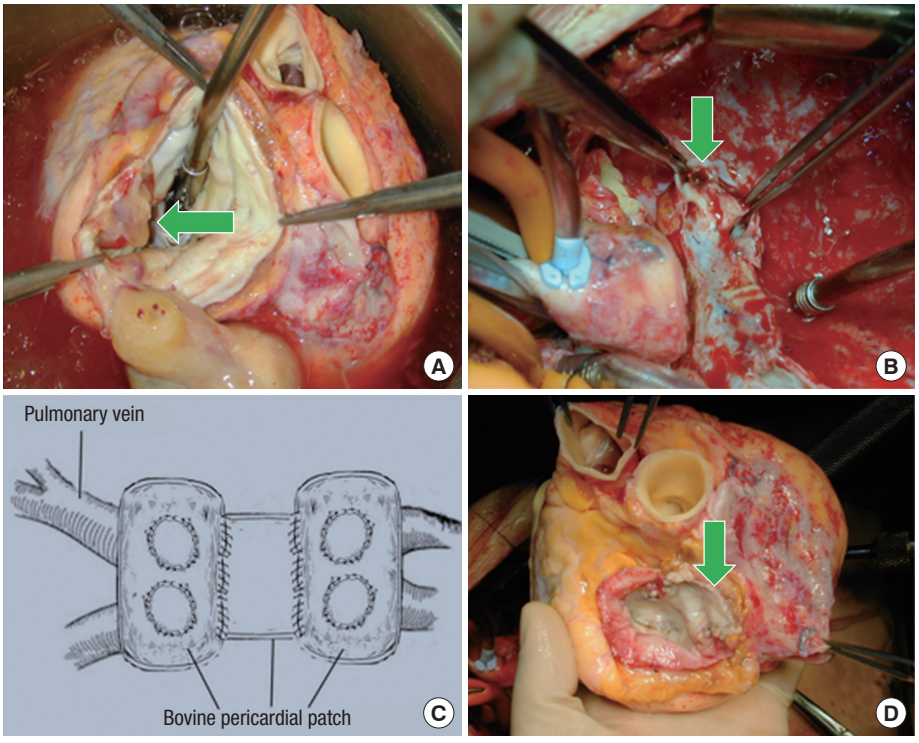

Fig. 2. Intraoperative findings. (**A**) Explanted heart was preserved in a solution composed of iced saline and HTK solution. The tumor was on the posterior portion of the left atrial wall and invaded the LA appendage (arrow). (**B**) After explantation of the heart. Pulmonary veins were invaded by the tumor (arrow). (**C**) The posterior wall of the LA was reconstructed using bovine pericardium. (**D**) Ex vivo reconstruction of the left atrial wall using autologous pericardium (arrow).
HTK = histidine-tryptophan-ketoglutarate, LA = left atrium.

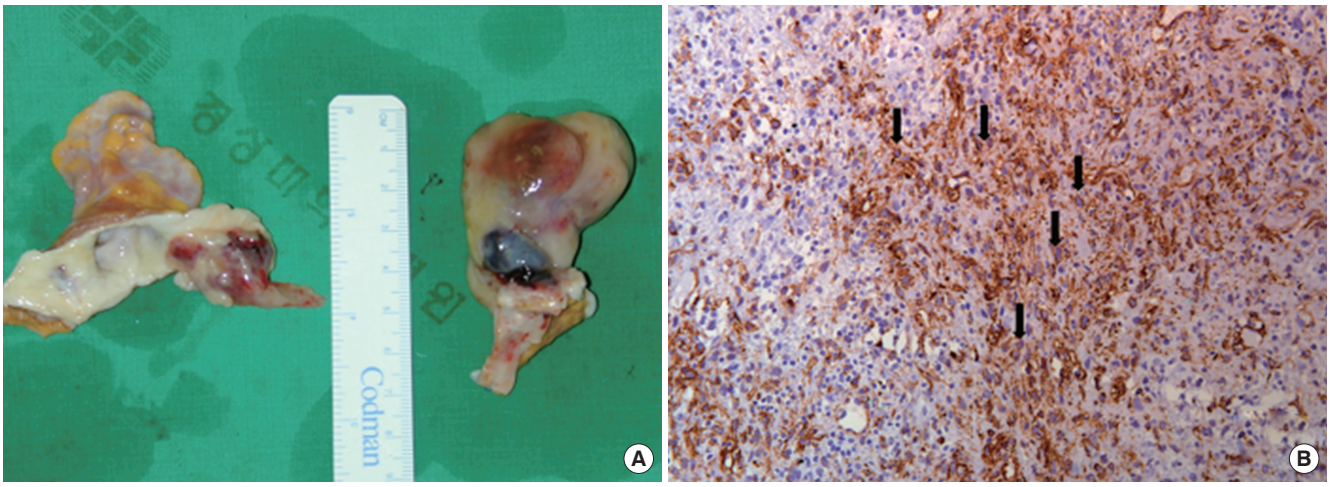

Fig. 3. Macroscopic and microscopic findings of inflammatory fibroblastic tumor. (**A**) Macroscopic view of the inflammatory fibroblastic tumor after resection. (**B**) Immunohistochemistry for smooth muscle actin. The tumor cells showing cytoplasmic staining for smooth muscle actin in the focal area (arrows) (×200).
HTK = histidine-tryptophan-ketoglutarate.

(Fig. 2A). Both inferior pulmonary veins had been invaded by the tumor (Fig. 2B). Consequently, the posterior wall of the LA and the tumor-invaded portion of the pulmonary veins were completely resected. The posterior wall of the LA was reconstructed with bovine pericardium (Fig. 2C). The LA of the explanted heart during ex vivo tumor resection was reconstructed with an autologous pericardium (Fig. 2D). The heart was reimplanted and the aortic cross-clamp released, and spontaneous cardiac activity resumed. The patient was weaned off CPB without difficulty. The total operation time was 13 hours, CPB time was 539 minutes and aorta cross-clamping time was 420 minutes.

Postoperative echocardiography and CT scan revealed no remnant mass (Fig. 1C and D). The presence of mild pulmonary hypertension (right ventricular systolic pressure was 32 mmHg) due to the small-sized LA and right inferior pulmonary vein stenosis was considered clinically insignificant. Gross examination revealed a 5 × 7 × 2 cm ivory piece of soft tissue mass with a smooth pericardium on one side (Fig. 3A). Tumor pathology revealed an IMT that had recurred locally (Fig. 3B). The patient's recovery was rapid but she was not discharged until the 25th day after the surgery because of a wound in the femoral vessel cannulation site. Periodic follow-up studies showed no local recurrence until 21 months after the operation.

DISCUSSION

The best treatment option for cardiac tumors has not been clearly established. Chemotherapy and radiotherapy can be used to treat cardiac tumors, but complete surgical resection of the tumor is considered the definitive treatment and has shown satisfactory outcomes. However, anatomic constraints due to inadequate visualization, along with incomplete resection of left atrial tumors of the heart, have often led to recurrence (1). In our patient, the cardiac tumor reoccurred at a similar site near the posterior wall of the LA 13 months after the previous operation. We performed autotransplantation to achieve a complete resection because the previous tumor may have recurred because of an incomplete removal, which was previously misdiagnosed as a myxoma using intraoperative frozen biopsy.

Complete resection is the optimal treatment for cardiac tumors. However, complete resection of complex tumors located in the LA is technically challenging. Access to this posterior location is difficult. For large complex tumors invading vital structures, pulmonary veins in this case, complete resection is almost impossible with a standard open heart surgery technique. The first option of the treatment for an unresectable non-metastatic cardiac tumor may be to consider an orthotopic heart transplantation (OHT). Gowdamarajan et al. (3) showed that 28 cases of cardiac transplantation published in the literature were for the treatment of a primary cardiac tumor. This study included 7 patients with benign histology and 21 patients with malignant histology. The mean survival rate for patients transplanted for a benign tumor is reported to be 46 months (range 8 to 105 months), while this number reduced to 12 months for malignant tumors (range 1 to 36 months).

Cardiac autotransplantation can achieve excellent exposure of the LA and its surrounding structures. The first attempted cardiac autotransplantation for the resection of a cardiac tumor was by Cooley et al. (4) in 1985. They reported a case where paragangliomas of the heart were located superiorly to the pulmonary veins. Ramlawi et al. (5) reported a large study of cases, 35 cardiac autotransplantations in 34 patients for malignant and complex benign tumors of the left heart. They suggested that standard surgical approaches could lead to incomplete re-

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section of tumors on the left side of the heart and that cardiac autotransplantation is a useful technique for the resection of complex left-sided tumors.

Cases of cardiac autotransplantation for primary cardiac tumors are limited in the literature. Despite this, we believe that cardiac autotransplantation is a safe and feasible technique for the complete resection of complex left atrial tumors not easily treated with the standard technique. It provides the surgeon with excellent visualization for complete resection of the tumor and accurate reconstruction.

DISCLOSURE

The authors have no potential conflicts of interest to disclose.

AUTHOR CONTRIBUTION

Conceptualization: Park HO, Yang JH, Choi JY, Kim JW. Data curation: Kim SH, Moon SH, Byun JH, Lee CE, Yang JW. Writing - original draft: Park HO, Yang JH, Kim JW.

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