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Case Report

Cardiac Autotransplantation with Concurrent Pneumonectomy for Complete Resection of Primary Cardiac Intimal Sarcoma

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Primary cardiac sarcoma is rare, and intimal sarcoma is an extremely rare and highly lethal disease. We report a case of a 62-year-old woman who was incidentally diagnosed with a primary cardiac sarcoma originating from the left atrial appendage and extending to the left superior pulmonary vein. The location of the tumor was very complicated, posing a major challenge for complete resection. We successfully performed complete resection of the cardiac sarcoma via cardiac autotransplantation with left pneumonectomy. The patient recovered uneventfully, without any adjuvant therapy as of 6 months postoperatively. Autotransplantation of the heart may be suggested as a reasonable surgical option for extensive left atrial tumors.

Keywords: Autologous transplantation, Heart neoplasms, Sarcoma

Case report

A 62-year old woman who achieved complete remission after breast-conserving surgery for breast cancer in 2003 was referred to Jeju National University Hospital. She presented with a 1-month history of dry cough of unknown etiology. There were no abnormal findings on physical examinations, a chest X-ray examination, bronchoscopy, or echocardiography. However, chest computed tomography revealed a 3.5×2.5-cm bulging mass with low attenuation originating from the left atrial (LA) appendage and extending to the left superior pulmonary vein (LSPV) without lymph node enlargement. The mass had invaded a posterior wall of the LA (Fig. 1). To confirm the diagnosis, we performed video-assisted thoracoscopic surgery to obtain a tissue sample. We partially excised the mass between the LA appendage and LSPV after pericardiotomy and confirmed that it was a malignant neoplasm. 18F-fluorodeoxy-

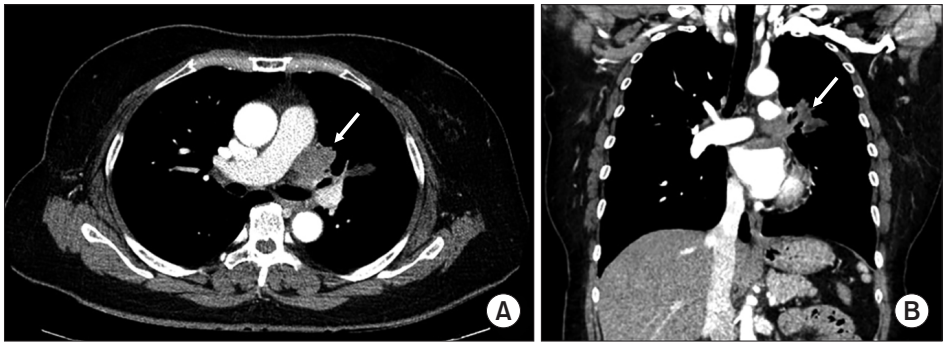

Fig. 1. (A) Preoperative computed tomography shows a 3.5×2.5-cm bulging mass with low attenuation located in the posterior wall of the left atrium (arrow). (B) It resulted in a tubular filling defect in the left superior pulmonary vein (arrow).

glucose positron emission tomography-computed tomography showed multiple hypermetabolic lesions, raising the suspicion of malignancy, without any metastatic lymph nodes in the mediastinum, pleural cavity, or abdomen. A conventional technique such as left atriotomy through the Waterston groove or a trans-septal approach could not provide adequate visualization of the tumor and a sufficient resection margin, because of the complicated location of the tumor on the posterior LA wall with invasion of the LSPV and central pulmonary parenchyma. We decided to perform cardiac autotransplantation with left pneumonectomy to achieve complete resection of the cardiac tumor.

Conventional cardiopulmonary bypass was established with bicaval venous cannulation via median sternotomy. After aortic cross-clamping, the aorta was divided in a transverse fashion, approximately 1 cm distal to the sinotubular junction. Both venae cavae and the pulmonary artery just proximal to the bifurcation were transected. The left atrium was then opened through the interatrial groove. These procedures showed that the tumor occupied the LA and had invaded the posterior wall of the LA and the LSPV. The incision was extended immediately behind the left pulmonary veins and above the mitral valve. The tumor had invaded the central portion of the lung parenchyma, with dense adhesions. After cardiectomy, we performed a left pneumonectomy with mediastinal lymph node dissection and extended pericardiectomy including the vagus and phrenic nerves. The posterior wall of the LA was reconstructed with a bovine pericardial patch, and the heart was re-implanted in reverse order. The aortic cross-clamping time was 246 minutes and the bypass time was 295 minutes. The resected pericardium was reconstructed using a 1-mm-thin Gore-Tex membrane (W.L. Gore & Associates Inc., Flagstaff, AZ, USA).

Fig. 2 shows the gross findings of the tumor, comprising atypical and spindle-shaped cells displaying low-grade pleomorphism (degree 1 out of 3 based on the *Federation Nationale des Centres de Lutte Contre le Cancer* system) (Fig. 3A). The final histopathology revealed a cardiac intimal sarcoma, which tested positive for smooth muscle actin and mouse double minute-2 homolog (MDM2) (Fig. 3B).

On postoperative day 3, the patient suffered from hoarseness and mild dyspnea after extubation. Cardiac and respiratory rehabilitation programs ameliorated the patient's symptoms, and she was transferred to the general ward on postoperative day 7. Follow-up echocardiography demonstrated a normal-sized LA and mild left ventricular diastolic dysfunction with an ejection fraction of 65%, almost identical to the preoperative findings. The patient continued to do well for 6 months postoperatively, with no need for additional adjuvant therapy.

The study was approved by the Institutional Review Board of Jeju National University Hospital (IRB approval no., 2019-07-003). Informed consent was waived.

Discussion

Primary cardiac sarcomas are rare, but represent the majority of primary malignant cardiac tumors. They can involve any area of the heart and great vessels, including the cardiac chambers, valves, and pulmonary artery. The his-

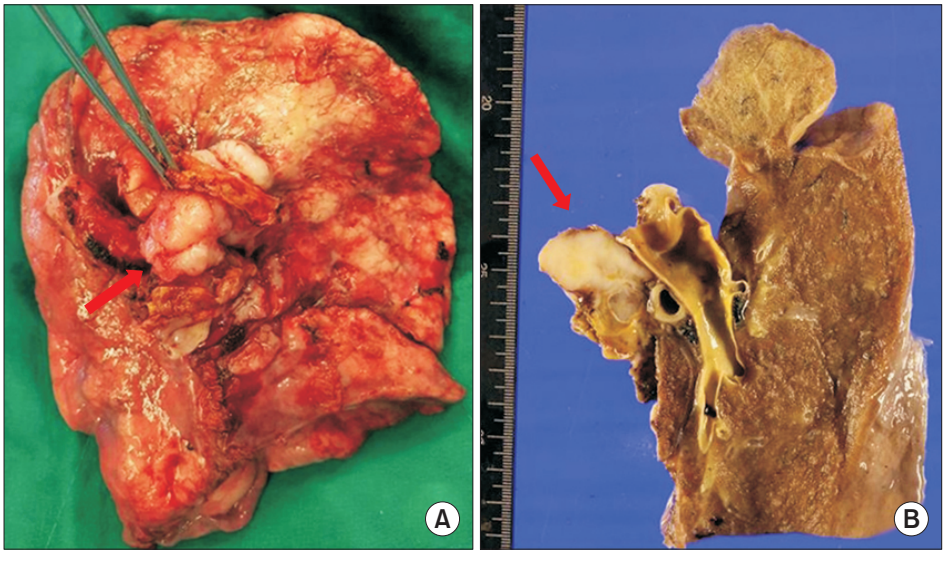

Fig. 2. (A) Grossly, a 3.5×2.5-cm and relatively well-circumscribed mass was found to have invaded the left atrium, pericardium, and lung parenchyma (arrow). (B) The cut surface of the resected specimen showed a diffuse fish-fleshy solid appearance without necrosis (arrow).

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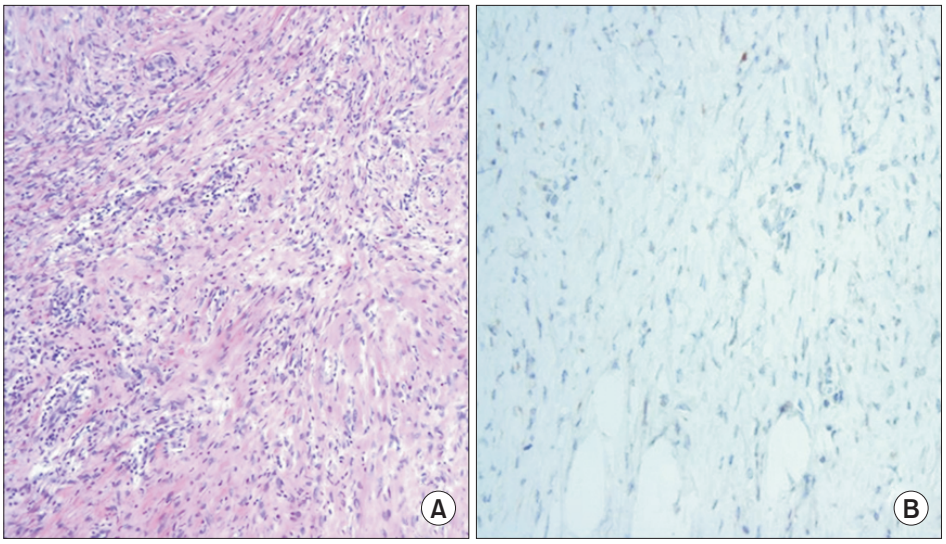

Fig. 3. (A) A pathological examination revealed a massive proliferation of pleomorphic spindle cells (H&E, ×100). (B) The human homologue of the mouse double minute 2 oncogene was overexpressed in these tumor cell nuclei (immunohistochemistry, ×200).

tological subtypes of cardiac sarcomas include angiosarcoma, leiomyosarcoma, fibrosarcoma, rhabdomyosarcoma, synovial sarcoma, osteosarcoma, undifferentiated pleomorphic sarcoma, and intimal sarcoma [1]. Intimal sarcoma is a mesenchymal tumor that arises from the tunica intima of large blood vessels including the pulmonary veins; however, it rarely involves the heart. It is a poorly differentiated malignant tumor characterized by the formation of spindle-shaped cells with a fascicular growth pattern. In a recent study, intimal sarcoma has been reported to be the most frequently occurring type of primary cardiac sarcoma (42%), with its characteristic pathological feature being *MDM2* gene overexpression and amplification [2].

Complete surgical resection is the most successful treatment for enhancing the survival rate of patients with cardiac sarcoma. All cases of local recurrence have been found in patients with incomplete resections [3]. Complete surgical excision, however, may not be possible in many cases because of the tumor location and involvement of the vital structures [4]. Tumor invasion of the resection margin is often unpredictable, and local recurrence and metastasis occur frequently and early, usually within 1 year [5]. Cardiac autotransplantation was first attempted by Cooley et al. [6] in 1985 for a LA paraganglioma. Park et al. [7] reported a large recurrent complicated tumor invading the pulmonary veins. The investigators also used cardiac autotransplantation to achieve complete resection. In our case, we employed autotransplantation and left pneumonectomy because of the direct invasion of the tumor of the posterior wall of the LA, LSPV, and central lung parenchyma.

Cardiac sarcomas are highly aggressive, with a mean survival of 3 months to 1 year [8]. Patients who undergo complete tumor resection are known to live twice as long as those who do not receive surgical resection [9]. Hamidi et al. [10] reported that the median survival was only 1 month in patients who did not receive any treatment, while the median survival was 12 months in patients who were treated with surgical resection. The use of adjuvant radiation therapy was associated with a median survival of 11 months, compared to 4 months in patients who did not receive adjuvant radiation therapy, although the association of radiation therapy use with patients' outcomes was not statistically significant [10]. However, adjuvant radiation therapy and chemotherapy do not appear to enhance survival in patients who have undergone incomplete resection [9].

In conclusion, complete resection of primary cardiac sarcomas is mandatory to improve survival. Autotransplantation of the heart seems to be a reasonable surgical option for left-sided cardiac tumors. Although adjuvant radiation therapy is not essential for the management of intimal sarcoma, it may facilitate survival in patients at risk of local recurrence because of positive resection margins and/or extensive invasion into the surrounding organs.

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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