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# Surgical Management of Recurrent Leiomyosarcoma in Heart

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Leiomyosarcoma may occur anywhere in the body but rarely occurs in the heart or great vessels. Leiomyosarcoma may be managed by surgical resection with or without chemotherapy or radiotherapy. Owing to the high rate of metastasis and poor prognosis, a definitive treatment modality for leiomyosarcoma has not yet been suggested. This case study reports the surgical management of the recurrent leiomyosarcoma of the heart and the great vessels in a 63-year-old woman.

Key words: 1. Neoplasm outcome
2. Surgery
3. Saphenous vein

## CASE REPORT

A 63-year-old woman with an impression of a recurrent tumor was admitted with a chief complaint of shortness of breath 10 months after surgery for cardiac leiomyosarcoma. In her past medical history, she had undergone surgery for the removal of leiomyosarcoma in the main pulmonary artery at a different hospital, where she had been admitted with a chief complaint of dyspnea (New York Heart Association Functional Classification II–III). In addition, she did not have hypertension or diabetes mellitus, other than the previous cardiac surgery.

Upon admission, her blood pressure was 130/90, and her heart rate was 90 beats/min showing a sinus rhythm. Her height, body weight, and body mass index were 160 cm, 60 kg, and 24.4 kg/m$^2$, respectively. The blood test taken upon her admission showed creatine kinase-MB 2.5 and troponin-I $<0.02$, which were within normal levels. Arterial blood gas analysis revealed pH 7.48, $PO_2$ 61.3 mmHg, $PCO_2$ 27.2 mmHg, and $O_2$ saturation 92.3%.

In her chest X-ray, no cardiomegaly was observed, but patchy haziness of the right middle lobe was seen. The cardiac computed tomography (CT) showed a massive amount of thromboembolism in the pulmonary trunk, right pulmonary artery, and left anteromedial basal segment artery (Fig. 1A). Lung cancer, pulmonary infarction, myocardial infarction, and coronary artery disease were not observed. Because of a suspicion of recurrent leiomyosarcoma, a surgical resection was decided upon (preoperative echocardiogram was not carried out). There was no evidence of lung metastasis in CT; therefore, lobectomy of the lung was not considered.

After carrying out median sternotomy, cardiopulmonary bypass was initiated. Transverse pulmonary arteriotomy was performed in the area straight above the pulmonary valve. Then, a mass with a volume of 4×4×3.5 cm hanging on the pulmonary valve with a stalk was observed. The mobile ge-

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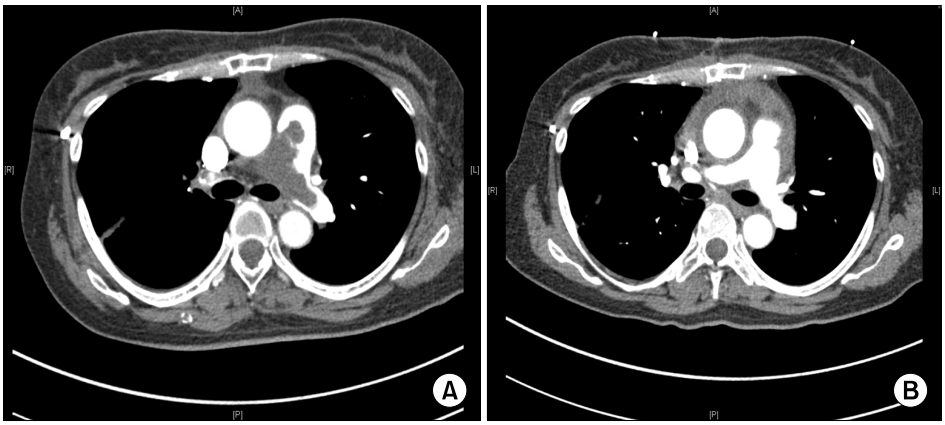

**Fig. 1.** (A) Preoperative computed tomography: Leiomyosarcoma in the main pulmonary artery and right pulmonary artery. (B) Postoperative computed tomography: no cardiac mass in the main pulmonary artery and right pulmonary artery.

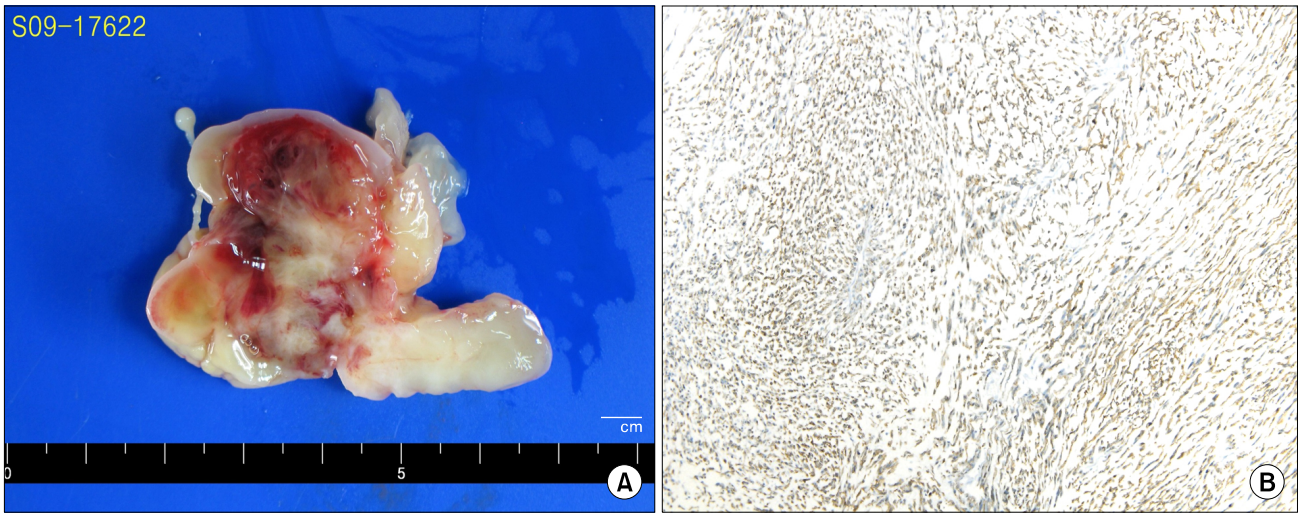


**Fig. 2.** (A) Gross sectional finding of resected leiomyosarcoma shows myxoid pinkish white soft tissue. (B) Microscopic finding of leiomyosarcoma (H&E, ×100).

latinous mass was hanging loosely in the right ventricular outflow track (RVOT), infundibulum, and main pulmonary artery. The incision of the right pulmonary artery was made up to the superior vena cava. An incision of the left pulmonary artery was extended to the first bifurcation area. Then, the myxoid mass was resected (Fig. 2). The sufficient back flow from the distal pulmonary artery was verified in both the right and the left pulmonary arteries. The harvested greater saphenous vein graft was used for the reconstruction and widening of the main pulmonary artery and left pulmonary artery. Because the myxoid mass had a well-defined capsule and stalk, we thought that there was no invasion to the nearby apparatus; therefore, we decided upon a pulmonary valve preservation operation.

The duration of extracorporeal circulation was 235 minutes, while that of aortic cross clamping was 85 minutes. Extubation was carried out 12 hours after surgery, and the patient was transferred to a general ward on the fifth postoperative day. Cardiac CT performed on the seventh postoperative day confirmed that there was no remnant mass in the pulmonary trunk, RVOT, or in either of the pulmonary arteries (Fig. 1B). The patient was discharged on the fourteenth postoperative day without any complications.

Three months after hospital discharge, she again developed dizziness and palpitation, and an echocardiogram was carried out and showed a generally hypoechoic and heterogeneous

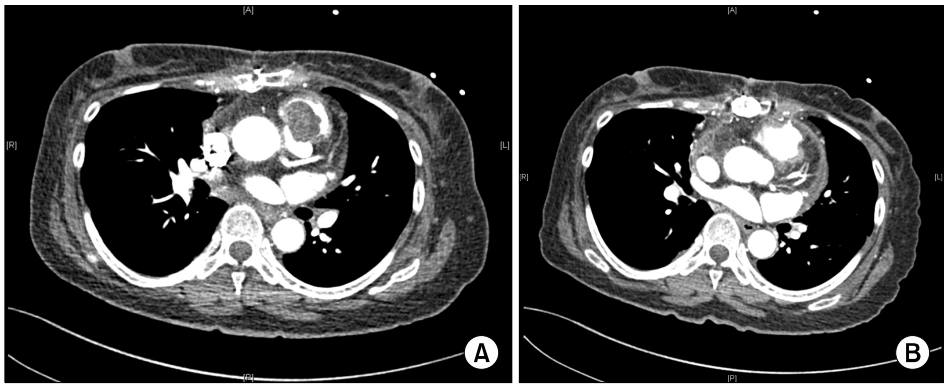

**Fig. 3.** (A) Preoperative computed tomography: 2.5×2.5 cm mass at the interventricular septal base of the right ventricular outflow track. (B) Postoperative computed tomography: complete mass resection of the pulmonary trunk and right ventricular outflow track.

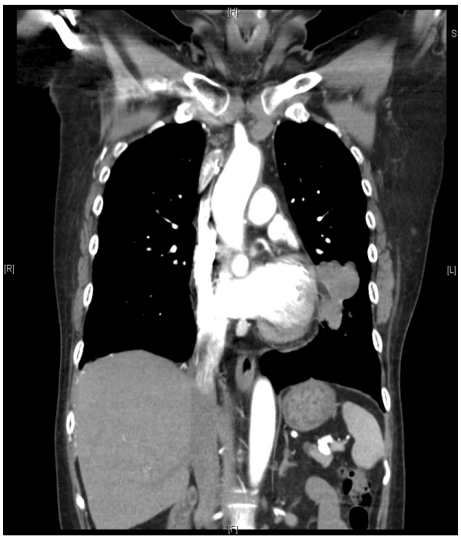

**Fig. 4.** Chest computed tomography shows lung metastasis of leiomyosarcoma to the left upper lobe and left lower lobe.

mass. The CT images showed a 2.5×2.8 cm mass at the base of the interventricular septum of the RVOT, as well as redundant movements in the direction of the pulmonary artery (Fig. 3A). As anticipated, the right ventricular systolic pressure increased to 70 to 75 mmHg. Upon the suspicion of recurrent cardiac leiomyosarcoma, we decided on a radical resection of the tumor.

Transverse pulmonary arteriotomy under cardiopulmonary bypass was performed in the area directly above the pulmonary valve. A gelatinous capsular mass causing RVOT obstruction was removed in the pulmonary valve and subvalvar infundibulum. Then, the transvalvar RVOT reconstruction was performed using a Prima plus 25 mm valve, and the vascular graft after the pulmonary valve was removed. The duration of extracorporeal circulation was 117 minutes. The postoperative course was uneventful, and an echocardiogram performed on the sixth postoperative day did not show a mass in the RVOT and revealed a well-functioning pulmonic valve. The images of cardiac CT carried out on the eighth postoperative day showed no residual mass of the pulmonary trunk, RVOT, or in either of the pulmonary arteries (Fig. 3B). The patient was discharged from the hospital on the tenth postoperative day.

During the follow-up six months after hospital discharge, a chest CT disclosed findings that leiomyosarcoma metastasized to left upper lobe (lingular segment) and left lower lobe (Fig. 4). Lingular segmentectomy and lower lobectomy were decided upon and performed under thoracotomy. A tumor 3×3×3 cm in size, dwelling apart from the pulmonary artery, was observed in the lingular segment of the left upper lobe. Another tumor mass with a size of approximately 3.5×3×3 cm was found in the anterior basal segment of the left lower lobe, for which lingular segmentectomy and lower lobectomy were carried out. The patient was discharged from the hospital 10 days after surgery. She has been asymptomatic and free of recurrence for 36 months after the last operation.

## DISCUSSION

Leiomyosarcoma is a rare tumor that develops in the smooth muscle cells and may occur systemically anywhere. However, this tumor, which comprises 8% of all sarcomas, largely occurs in the uterus, retroperitoneum, and intra-abdominal region [1-5]. Davidsohn was the first to report a case of leiomyosarcoma in 1908, and Cardes et al. reviewed 127

cases that had been reported up to 1999 [1].

The clinical symptoms of leiomyosarcoma in the heart or lung include chest pain and shortness of breath, which are similar to those of pulmonary embolism. Chest X-ray and CT findings are very similar to those of these two disorders, making it difficult to decide upon a diagnosis [4]. Transthoracic and transesophageal cardiac echography, as well as CT of the heart, have been used as the standard methods for the diagnosis of leiomyosarcoma that infiltrates the great vessels of the heart and pulmonary artery. However, recent utilization of a diagnostic approach with electrocardiogram-guided magnetic resonance imaging has increased diagnostic sensitivity [1,5].

Leiomyosarcoma that invades the heart is often discovered in the left atrium, but a tumor may also be found in the right heart structures, such as the pulmonary valve and RVOT on relatively rare occasions [2,4]. Treatment approaches to leiomyosarcoma are only surgical resection and adjuvant therapy after surgery [2,3]. With respect to surgical treatment, radial excision and aggressive excision are possible. A lower recurrence rate of leiomyosarcoma has been reported with the latter approach, but the necessity of further operations to a damaged heart, such as valve replacement, pulmonary trunk, and RVOT reconstructions, increase the rates of postoperative complications [6]. Either surgical resection only or adjuvant therapy after surgical resection shows an incredibly poor prognosis with a mean survival rate of 12 to 16 months after surgery [7]. An adjuvant therapy with doxorubicin, a chemotherapeutic agent, may extend the mean survival rate to 24 months, but will not change the natural history of leiomyosarcoma [7].

Leiomyosarcoma recurs frequently and has quite a poor prognosis. The result of pulmonary arterial repair with a greater saphenous vein graft and Prima plus-utilized pulmonary valve replacement showed no recurrences within the heart during 36 months of follow-up observations at the outpatient department. Regarding lung metastasis that supposedly occurred through the pulmonary vessels, a vigorous treatment attempt such as lobectomy was carried out with no current evidence of recurrence during the outpatient follow-up. In this case, the patients underwent heart surgery three times. After the third heart operation, the patient has remained alive for 36 months without recurrence of leiomyosarcoma. If a radical operation were done in the second heart operation, the third heart operation might not be required.

Pulmonary leiomyosarcoma recurs frequently and has poor prognosis. However, if a surgeon decides that surgical resection is possible, resection of leiomyosarcoma seems to be helpful in improving the prognosis. Thus, we report surgical experiences of multiple recurrences of leiomyosarcoma in cardiovascular and respiratory organs in this 63-year-old patient.

## CONFLICT OF INTEREST

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

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