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Right Atrial Myxoma with Pulmonary Tumor Embolus: Mid Term Follow up and Review of the Literature

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ABSTRACT
Cardiac myxomas are the most common primary cardiac tumor and usually develop in the left atrium. Right atrial myxomas are relatively rare and can be accompanied by pulmonary tumor embolism. The standard treatment for a right atrial myxoma with pulmonary tumor emboli is surgical removal of the cardiac tumor and pulmonary embolus. Although many successful surgical results have been reported, the mid- or long-term outcomes are rarely mentioned. This report is the 4-year follow-up of a case of a right atrial myxoma with a large tumor embolus in the left pulmonary artery. The patient underwent surgery for the removal of both the right atrial tumor and pulmonary embolus. Histopathological examination revealed both to be myxomas. Four years postoperatively, the patient was doing well. Although recurrence of the tumor was not observed, a few small pulmonary tumor emboli remained. Cardiac myxomas are benign neoplasms and proliferate slowly. However, careful follow-up observation for remote metastasis is crucial.

Keywords: myxoma, heart neoplasms, pulmonary embolism, thoracic surgery

1. Introduction

Primary cardiac tumors are rare entities, with an incidence of 0.0017%–0.19% in unselected patients at autopsy (Reynen, 1995). Metastasis to the heart from other primary malignant tumors is 30 times more common than primary cardiac tumors (Leja et al., 2011). Only 25% of primary cardiac tumors are malignant and 75% are benign. Approximately 50% of benign primary cardiac tumors are myxomas (Silverman, 1980).

Cardiac myxomas are neoplasms of endocardial origin. The tumor usually projects from the endocardium into the cardiac chamber. Approximately 75% of cardiac myxomas originate in the left atrium and 15%–20% in the right atrium. Cardiac myxomas originating in the left or right ventricles are rare (Demir et al., 2005).

Cardiac myxomas can be accompanied by three types of clinical presentations, including constitutional symptoms, intracardiac obstruction, and embolism. Constitutional symptoms include fever, general fatigue, weight loss, and anemia. These symptoms may be associated with the production of the cytokine interleukin 6 by the tumor itself (Seino et al., 1993). Intracardiac obstruction mimics mitral or tricuspid valve stenosis. The obstruction of filling of the left or right ventricles causes dyspnea, pulmonary edema, and right heart failure (Tok et al., 2007). Embolism can occur in 30%–40% patients with cardiac myxomas (St John Sutton et al., 1980). In cases of myxoma arising from the left cardiac chambers, systemic embolism, which

includes occlusion of cerebral, peripheral, and visceral arteries, is frequent (Braun et al., 2005). Meanwhile, right atrial and ventricular myxomas can be accompanied by pulmonary embolism (Keenan et al., 1982). Although many successful surgical results in cases of right atrial myxomas with pulmonary tumor emboli have been previously reported, follow-up reports are rare. This report is the 4-year follow-up of a case after the surgical removal of a right atrial myxoma with a large tumor embolus in the left pulmonary artery.

2. Case Report

This case was previously reported in 2014 (Ikeda et al., 2014). In summary, a 74-year-old male patient who had experienced a sudden onset of dyspnea was referred to our hospital for treatment of a right atrial tumor. Transthoracic echocardiography revealed a right atrial tumor that adhered to the interatrial septum and had a tail-like surface projection. The estimated right ventricle pressure was 63 mmHg, which suggested the presence of moderate pulmonary hypertension. Computed tomography (CT) showed that the left pulmonary artery was occluded by a large embolus. In addition, several small defects existed in the distal bilateral pulmonary arteries. The patient underwent an emergency surgery for the removal of both the right atrial tumor and embolus in the left pulmonary artery. Histopathological examination showed that both the right atrial tumor and pulmonary embolus were myxomas.

Follow-up observations of the patient were continued for 4 years after his operation. The patient was doing well. Just after the operation, transthoracic echocardiography showed mild pulmonary hypertension with an estimated right ventricle pressure of approximately 40 mmHg, which had not changed in 4 years. Although recurrence of the tumor was not observed 4 years postoperatively, CT revealed that a few defects remained in the distal bilateral pulmonary arteries (Figure 1). These defects had not changed in the last 4 years

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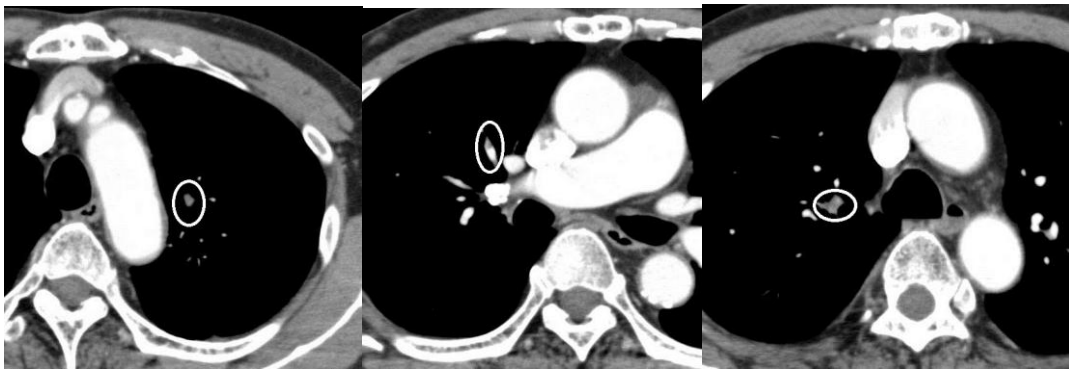

Figure 1. Follow-up computed tomography 4 years postoperatively shows that a few defects remained in the distal pulmonary arteries. These defects had not changed in the last 4 years.

3. Discussion

In cases of right atrial myxoma, pulmonary embolism is a serious complication (St John Sutton et al., 1980). The standard treatment for right atrial myxoma with pulmonary tumor embolism is surgical removal of both

the cardiac tumor and pulmonary embolus. Although successful surgical results have been previously reported, the mid- or long-term prognosis has remained unclear.

In the present case, small tumor emboli remained in the distal pulmonary arteries 4 years postoperatively. Although these small pulmonary emboli existed before surgery and did not grow, it is important to continue careful observation for remote metastasis. The malignant potential of cardiac myxomas remains doubtful (Reynen, 1995).

However, several remote metastases of cardiac myxomas have been reported (Kaynak et al., 2001; Rodrigues et al., 2006; Suzuki et al., 2008). Cardiac myxomas proliferate slowly, and the mechanism of the remote growth of the tumor is unknown.

In conclusion, surgical treatment for a right atrial myxoma with pulmonary tumor emboli produces excellent results. However, small tumor emboli can remain in the distal pulmonary arteries. Although remote metastasis of cardiac myxomas is rare, long-term follow-up should be performed.

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