CASE REPORTS

Resection of metastatic pulmonary lesion of ossifying fibromyxoid tumor extending into the left atrium and ventricle via pulmonary vein

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Abstract
The ossifying fibromyxoid tumor (OFMT) of soft parts is a rare soft tissue neoplasm of uncertain lineage. The most common metastases are found in the lung. Herein, we present the first case report of pulmonary metastasis of ossifying fibromyxoid tumor with intracardiac extension, which was resected carefully using cardiopulmonary bypass and cardiac arrest. Subsequently pulmonary left lower lobectomy was performed. The patient recovered uneventfully and was discharged in about a few days. Recently, OFMT has been considered as a tumor of intermediate malignancy. We recommend wide surgical excision of primary tumor and radiotherapy. Early follow-up is mandatory.

Introduction
The ossifying fibromyxoid tumor of soft parts is a rare soft tissue neoplasm of uncertain lineage. It occurs primarily in the trunk and proximal extremities. The most common metastases are found in the lung, soft tissue and adrenal gland. Herein, we present the first case report of pulmonary metastases of ossifying fibromyxoid tumor with intracardiac extension, which was resected using cardiopulmonary bypass and cardiac arrest; subsequently, pulmonary left lower lobectomy was performed.

Case report
A 71-year-old man underwent a resection of an ossifying fibromyxoid tumor (OFMT) in the left leg in 1996. It was considered a low-grade sarcoma, and no follow-up was done. He has been admitted by emergency room because of 1-week progressive dyspnea, orthopnea and more physical signs of heart failure. Paroxysmal atrial tachycardia was
documented. Electrocardiogram showed sinus rhythm and complete right bundle branch block. Blood routine analysis was essentially normal. Chest X-rays showed a mass in the pulmonary left lower lobe. Transthoracic echocardiography was conclusive and revealed a large mass in the posterior region of the left atrium protruding into the left ventricle in diastole and coming back to occupy almost all the left atrium in systole (Fig. 1a,b). CT scan showed a large mass in the pulmonary left lower lobe, extending through the left inferior pulmonary vein and protruding into the left atrium and left ventricle. Fine-needle aspiration guided by CT scan pointed to OFMT. Bone gammagraphy showed a focal osteogenic lesion in the proximal fibula.

The tumor was resected carefully through left atriotomy using cardiopulmonary bypass and cardiac arrest. It partially adhered to the left inferior pulmonary vein and was completely free inside the heart. The tumor was large (17 × 3.5 cm), white and elastic with smooth surface and several cystic cavities (Fig. 2). After weaning off cardiopulmonary bypass, pulmonary left lower lobectomy was done.

The pathological study confirmed the diagnosis of pulmonary metastasis of ossifying fibromyxoid tumor. The postoperative course was unremarkable and the patient was discharged 2 weeks later. He is being followed-up by the Department of Oncology and currently he is healthy with no tumoral recurrence documented.

Discussion

Cardiac tumors, primary or secondary, are rare. Their potentially lethal course and the possibility of cure with adequate excision make early diagnosis and optimal therapeutic attainment crucial. Therefore, physicians should be knowledgeable about cardiac tumor pathology as well as their frequently atypical clinical presentations.

Metastatic heart tumors are far more common. The most common mean of spread is via the blood stream through coronary arteries and subdiaphragmatic vena cava. They can also reach the heart through lymphatic channels, direct extension from adjacent lung, breast, esophageal, and thymic tumors. The vast majority affects the right chambers.

The ossifying fibromyxoid tumor (OFMT) of soft parts is an uncommon soft tissue neoplasm of
uncertain lineage. It was described by Enzinger et al. in 1989. In most instances, OFMT is characterized by small size, subcutaneous location or deeply seated masses in the trunk, proximal extremities and in peripheral shell of bone. It has small and bland cells arranged in cords and nests within a fibromyxoid stroma. More than 120 cases have now been reported as case reports and in small series. Recently, Folpe and colleagues studied 70 cases of OFMT. Local recurrences were found in 18% and metastasis in 16%. The metastatic sites included the lung, soft tissue and adrenal gland. They elaborated pathological criteria for malignancy in these tumors and supported their reclassification as tumors of intermediate malignancy.

After perusing the medical literature, we did not find any other case of pulmonary metastasis of ossifying fibromyxoid tumor with intracardiac extension. The metastatic tumor extended through the left inferior pulmonary vein to the left cardiac cavities causing arrhythmias, obstruction of left ventricle inflow and mitral valve function interference. The intracardiac mass did not invade the cardiac tissue. Therefore, we recommend a careful excision of the tumor, particularly in the pulmonary vein, using cardiopulmonary bypass and cardiac arrest, and subsequent total pulmonary lobectomy. Because the OFMT was recently considered a tumor of intermediate malignancy, wide surgical excision of the primary tumor is essential and radiotherapy is strongly recommended. Early follow-up to discover early local recurrences or metastasis is mandatory.

References

Carcinoid crisis and reversible right ventricular dysfunction after embolization in untreated carcinoid syndrome

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