Primary cardiac tumor is rare and most commonly benign. We report a case of right atrial tumor associated with metastatic pulmonary angiosarcoma. A 38-year-old man came to our hospital with exertional dyspnea and hemoptysis. Chest film showed diffuse lung nodules and computed tomography of chest discovered right atrial tumor. Wedge resection of one lung lesion provided histological diagnosis of angiosarcoma.

**Key Words:** Cardiac tumor • Angiosarcoma • Lung metastasis

**INTRODUCTION**

Primary cardiac tumors are rare and most commonly benign tumors. Angiosarcoma is the most common primary malignant heart tumor and frequently has lung metastasis when diagnosed. Herein we report such a case proved histologically by thoracoscopic wedge resection.

**CASE REPORT**

A 38-year-old man who had no major medical illness such as hypertension, diabetes mellitus or hyperlipidemia, reported general weakness 3 months before admission. Dry cough, especially in the morning, and exertional dyspnea followed. Body weight loss from 70 to 63 kg during recent 3 months was also noted. He then presented to clinics due to small amount of hemoptysis (less than 25 mL). The ENT fibroscopy and bronchoscopy could not find a bleeder in the airway.

A chest X-ray film showed diffuse multiple nodules of various size and normal heart contour (Figure 1A). The chest computed tomography showed one cardiac tumor in the right atrium with multiple nodular lesions over bilateral lung field (Figure 1B). A magnetic resonance image was also obtained (Figure 1C). Cardiac echo showed a tumor mass from the whole right atrial free wall toward the posterior leaflet of the tricuspid valve (size up to 3.2 x 3.6 cm) without inferior vena cava involvement, mild pulmonic and tricuspid regurgitation, and normal left ventricular size and systolic function.

Right-sided cardiac catheterization showed normal pulmonary artery, right ventricular, right atrial and central venous pressures. Right atrial endomyocardial biopsy was performed, and the pathology showed fragments of fibrotic tissue with increasing elastic fiber, confirmed by orcein stain. No definite evidence of tumor was seen. Parts of the arterial wall or fibroelastosis of the endocardium was favored. Pulmonary function test showed normal ventilatory function, within normal limits of gas exchange. The whole-body bone scan showed increased tracer activity at manubrium sterni and Louis angle of the sternum, which was suspected to be metastatic lesions. Wedge resection of a pulmonary lesion by video-assisted thoracic surgery was done. Pathology showed a small tumor measuring 0.6 cm in diameter grossly. Microscopically, it showed a poorly differenti-
ated tumor with epithelioid tumor cells having large vesicular nuclei, prominent nucleoli and pale to pink cytoplasm arranged in sheet-like pattern (Figure 2A). Numerous mitotic figures are noted. Tumor emboli are also seen (Figure 2B). Immunohistochemically, the tumor cells were positive for CD31 and vimentin but negative for cytokeratin. A metastatic angiosarcoma was considered (Figure 2C). After discussion with the patient, he wished to be discharged to receive other kinds of therapy such as herbal medicine. He was then discharged and lost to follow-up.

DISCUSSION

Primary tumors of the heart are rare, approximately
0.02% based upon 22 large autopsy series.1 Three-quarters of these tumors are benign; nearly half of the benign heart tumors are myxomas.2 About a quarter of cardiac tumors are malignant, and 95% of these are sarcomas; the other 5% are lymphomas.3 Among malignant primary cardiac tumors, the more common tumors are angiosarcomas (37%), undifferentiated sarcomas (24%), malignant fibrous histiocytomas (11-24%), leiomyosarcomas (8-9%), and osteosarcomas (3-9%).4

Angiosarcomas occur more commonly in men, and almost exclusively in the right atrium.3 Pulmonary metastases are frequent, and survival after diagnosis rarely exceeds six months.5 The symptoms and signs of angiosarcomas include dyspnea, cough, fever, weight loss, hemoptysis, right heart failure, pericardial involvement with pleuritic pain, and cardiac tamponade. But symptoms of left-sided heart failure rarely occur.3,6

Electrocardiographic abnormalities are nonspecific, including right axis deviation, nonspecific or pericarditic ST-T wave changes, low QRS voltage and supraventricular arrhythmia.6 Chest roentgenograms often show cardiomegaly.6 Other abnormalities include right-sided heart enlargement, widened mediastinum, hilar adenopathy, pulmonary congestion, or pleural effusion.7

Echocardiography usually shows a broad-based right atrial mass near the inferior vena cava. Epicardial, endocardial or intracavity extension is common, and local spread of the tumor to pleura or mediastinum is often found.3

Two major types of angiosarcoma have been morphologically described, well-defined protruding type and diffuse infiltrative type. In the well-defined type, tumor mass protrudes into a cardiac chamber, usually the right atrium, often sparing the atrial septum. Grossly, it shows hemorrhagic, necrotic, and sometimes adherent to the pericardium. CT frequently shows a low-attenuation irregular or nodular mass with central necrosis in communication with the chamber. Contrast enhancement is heterogeneous. At MRI, areas of increased signal intensity on T1-weighted image may be focal or peripheral and are thought to represent blood products. Local nodular areas of increased signal intensity are interspersed within areas of intermediate signal intensity on T1- and T2-weighted images so as to have “cauliflower” appearance.

In diffuse infiltrative type, tumor mass extends along the pericardium. The pericardial space may be obliterated with hemorrhagic, necrotic tumor debris, which may appear on CT as pericardial effusion or thickening. On MRI, linear contrast enhancement along vascular lakes has been described as “sunray appearance”.4 In our case, the cardiac tumor was confined to the right atrial wall and had no pericardial effusion or significant pericardial extension, thus well-defined protruding type was more favored.

Histologically, angiosarcoma is a tumor of endothelial and mesenchymal cells. They are usually hemorrhagic and often have poorly defined borders. They can both invade and encase the myocardium. Characteristic areas of vascular channels lined by pleomorphic and atypical cells are seen. The presence of irregular anastomosing sinusoidal structures with papillary intraluminal tufting is diagnostic of these tumors. Higher mitotic rates and necrosis are correlated with poorer outcome.7 Typically, these tumors’ stains are positive for a variety of immunohistochemical markers, including CD34, CD31, factor VIII, Von Willebrand factor, and vimentin.7,9 The hallmark of cardiac angiosarcoma at electron microscopy is Weibel-Palade bodies, which are characteristic for endothelial cells, but these structures are not often seen.7 In our case, pleomorphic, atypical cells with numerous mitotic figures were noted and stains for CD31 and vimentin were positive.

The mean survival time without surgical resection is approximately 4 months, which improves to 10 months following surgery.10 Due to the unfavorable location in our patient, conventional surgery could only resect the tumor partially instead of radical resection to attain tumor-free surgical margins. Uberfuhr et al. attempted radical tumor resection with heart transplantation but distant metastasis emerged later.11 Due to early metastasis, effective chemotherapy is needed to eradicate micrometastasis. However, angiosarcomas are thought to be unresponsive to these traditional treatments.12

REFERENCES

2. Centofanti P, Di Rosa E, Deorsola L, et al. Primary cardiac
7. Best AK, Dobson RL, Ahmad AR. Best cases from the AFIP: cardiac angiosarcoma. Radiographics 2003;23(suppl):141-5.
右心房腫瘤併轉移性肺部血管肉瘤 — 一病例報告

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原發性心臟腫瘤並不常見，而且大部分是良性的。我們報告一例右心房腫瘤併轉移性肺部
血管肉瘤。一個三十八歲男性，因為運動性喘氣及血痰求診，胸部 X 光檢查發現廣泛肺部
結節。胸部電腦斷層檢查發現心臟右心房腫瘤。最後用胸腔鏡切下部分肺部病灶，病理證實
是血管肉瘤。

關鍵詞：心臟腫瘤、血管肉瘤、肺轉移。