Heart Vessels (2000) 15:100–102

CASE REPORT

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Primary leiomyosarcoma of the heart subsequent to double carcinomas of the thyroid and lung

Received: April 22, 2000 / Accepted: September 1, 2000

Abstract A 63-year-old woman underwent surgical operations for left lower lung cancer and for thyroid cancer. Nine months later, a third cancer developed in her heart and this tumor was removed by open heart surgery. A pathologic study revealed that the tumor was primary leiomyosarcoma of the heart and thus independent from the previous lung and thyroid carcinomas. This case was regarded as a triple carcinoma including a primary leiomyosarcoma arising from the left atrium. Reports in the literature on primary malignant tumors of the heart are reviewed briefly.

Key words Primary cardiac malignant tumor · Leiomyosarcoma · Triple carcinoma

Introduction

Malignant tumors derived from smooth muscle are extremely rare in the heart. Primary cardiac neoplasms are found in only about 0.002% of autopsies.[1] Approximately 25% of primary cardiac neoplasms are malignant.[2] Although a recent increase in multiple cancers has been noticeable, cases of double cancer concurrent with primary leiomyosarcoma of the heart are rare. This report describes a patient with leiomyosarcoma of the heart who had undergone surgical operations for lung and thyroid cancer 9 months previously.

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Case report

A 63-year-old housewife with diabetes mellitus that had been medically well controlled for 8 years was admitted to Muroran City General Hospital, on August 18, 1998, because of the sudden onset of dyspnea. In the previous year, on October 31, she had been treated for left lung cancer by endoscopic partial resection. The clinical stage classification of the lung cancer was differentiated papillary adenocarcinoma without lymph node metastasis or vessel invasion (T1N0M0; pT1N0M0, stage 1). She also underwent right hemithyroidectomy because of a cancer in the right lobe of the thyroid gland on November 10, 1997. The histopathological diagnosis of the resected tissue was papillary thyroid carcinoma. The postoperative course was smooth.

Until this admission the patient had not experienced exertional dyspnea, palpitation, wheezing, or ankle edema and cardiac murmur had not been pointed out. At the time of admission the patient was well nourished, but was suffering from moderate respiratory distress. Her lips and fingertips appeared slightly cyanotic. No significant cardiac murmur was audible. Moist rales were heard throughout both lungs and the liver was palpable 3 cm below the costal margin, its edge dull with slight tenderness. The abdomen was otherwise normal. Moderate pretibial edema was present. No neurological findings were observed. Her blood pressure was 95/60 mmHg, and pulse rate was 80 beats/min and regular. A chest X-ray revealed enlargement of the cardiac shadow with a cardiothoracic ratio of 54%, bilateral pleural effusion, and pulmonary congestion in both lung fields. A 12-lead electrocardiogram showed a regular sinus rhythm with a heart rate of 78 beats/min, slight depression of the ST-segment at leads I and aVL, and T-wave inversion at leads V1 and V2.

Laboratory findings were as follows: red blood cell count, $3.86 \times 10^6/\mu l$; hemoglobin, 11.2 g/dl; white blood cell count, 18400/μl; C-reactive protein, 22.9 mg/dl; lactate dehydrogenase, 469 mU/ml; fasting blood sugar, 177 mg/dl; and normal urine. Two-dimensional echocardiography revealed a large, mobile mass in the left atrium.

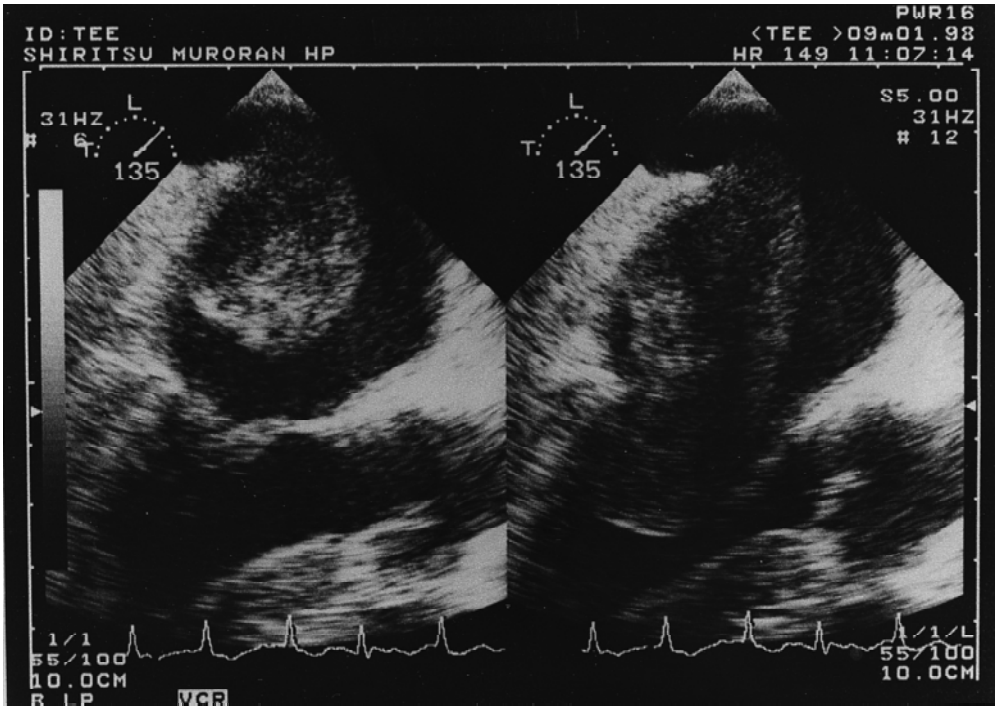

Fig. 1. Transesophageal echocardiography (systolic phase on the left and diastolic phase on the right)

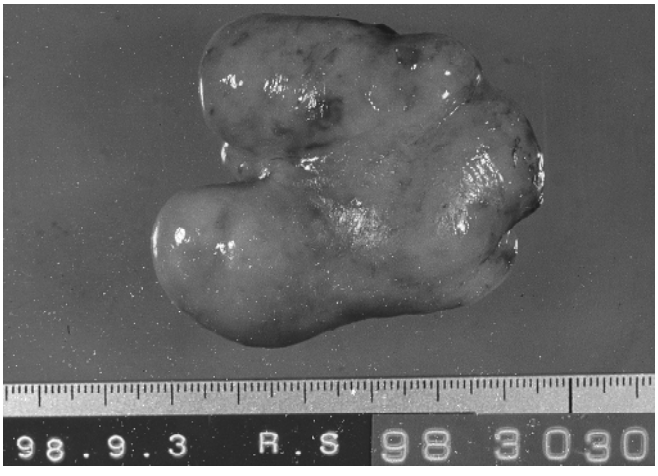

Fig. 2. Gross pathologic specimen of the removed larger tumor mass

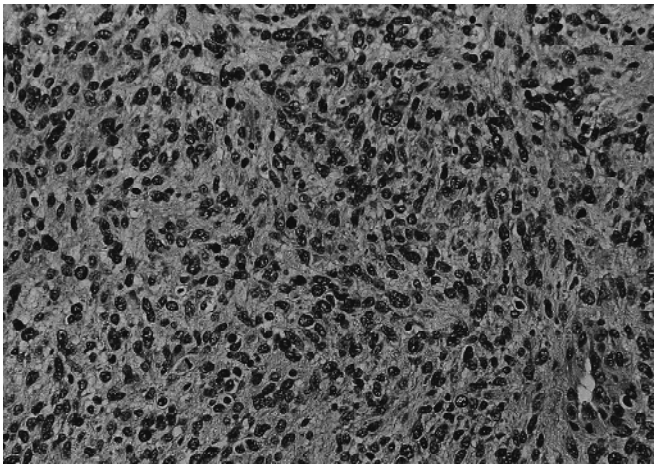

Fig. 3. Microscopic picture of the removed tumor mass (H&E, ×400)

Transesophageal echocardiography revealed a non-homogeneous mass which protruded into the left ventricular inflow tract (Fig. 1). Cardiac catheterization showed a pulmonary artery pressure of 37/24 mmHg (mean 30 mmHg) and a pulmonary wedge pressure of 19 mmHg. A pulmonary angiogram revealed a filling defect in the left atrium, suggesting stenosis of the mitral valve secondary to myxoma or sarcoma. Left ventriculography showed normal left ventricular dimensions and segmental wall motion.

On September 3, 1998, an emergency operation was performed because of recurring acute heart failure. At left atriotomy, two tumor masses were found. The larger one was a bilobed, pedunculated mass located on the posterior wall near the mitral valve, one head of which had penetrated into the left ventricle (Fig. 2). The other one was a sessile polypoid tumor located on the orifice of the left pulmonary vein. These two tumors were marginally resected along with a small part of the atrial wall.

Gross examination of the resected specimens revealed the two tumors to consist of a multinodular mass of elastic firm consistency. They were completely covered by smooth endocardium, and were 5.5 and 1.5 cm in size, respectively. The cut surface revealed solid, white tissue with bloody stains, and necrotic foci with a yellow tan.

Microscopically, both tumors showed similar histological findings. The tumor cells had elongated, rounded, or occasionally pleomorphic nuclei with coarse, dense chromatin, and eosinophilic cytoplasm with indistinct borders. Mitotic frequency was as high as 40/10 HPF (Fig. 3). The stroma contained a great deal of fine reticulin fibers and a small

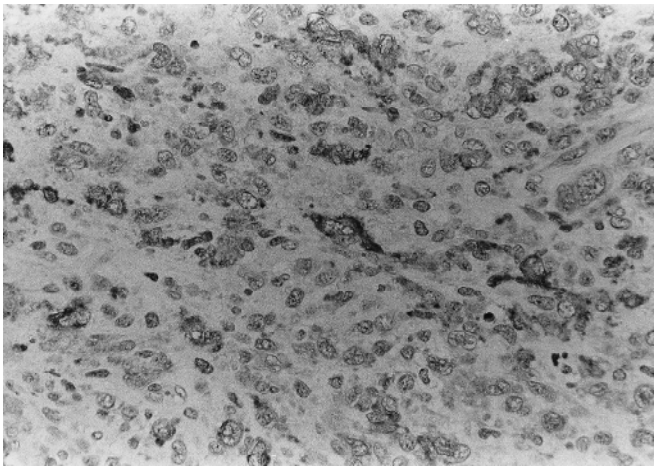

Fig. 4. Immunohistochemical examination of the removed tumor mass was positive for smooth muscle-specific actin (×400)

amount of acid mucopolysaccharide. Immunohistochemical examination was positive for vimentin, desmin, smooth muscle-specific actin (Fig. 4), and HHF35, and negative for S100 protein, factor VIII, and myoglobin. Altogether the morphological and immunohistochemical findings were considered consistent with a diagnosis of leiomyosarcoma.

Postoperative recovery was uneventful. However, the patient died of cerebral infarction on April 2, 1999.

Discussion

Primary cardiac leiomyosarcoma is a rare disease first described by Weir and Jones in 1941.[3] There have been few reports in the literature and the total number of primary cardiac leiomyosarcomas approaches only 37.[4–11] The clinical diagnosis of cardiac sarcoma is difficult because the symptoms are insidious and nonspecific. For most patients symptoms start with dyspnea, followed by cough, chest pain, palpitation, general malaise, and weight loss. In the present case, the patient had undergone surgical operations for lung cancer and thyroid cancer. Nine months later, she experienced the sudden onset of dyspnea. Physical signs, a chest X-ray, and an electrocardiogram showed nonspecific findings. The most useful diagnostic method in this case was transesophageal echocardiography since it clearly revealed protrusion of the tumor into the left ventricular inflow tract. Secondary invasion of the heart by metastatic tumors occurs much more frequently than primary cardiac neoplasm.[12,13] Autopsy studies indicate that 1.5%–21% of patients with primary malignancy in other body organs develop metastatic cardiac involvement.[14–17] It is necessary to exclude all other possible primary sites before deciding that a malignant cardiac neoplasm has in fact arisen in the heart. There are three reports of uterine leiomyosarcoma metastasizing to the heart.[18–20] The patient had never suffered menstrual irregularity and her preceding history ruled out the possibility that the myocardial tumor was a secondary deposit from a primary uterine lesion. Moreover, there was no local recurrence after the left lung cancer operation so in light of this and the above-mentioned facts a diagnosis of primary cardiac leiomyosarcoma was made.

While a recent increase in multiple cancers has been noticed, cases of double cancer concurrent with primary leiomyosarcoma of the hart are extremely rare. Patients with primary cardiac leiomyosarcoma have a poor prognosis, most of them dying within 1 year after the diagnosis. The effect of surgical excision and adjuvant therapy for cardiac leiomyosarcoma should now be evaluated by reference to the many well-described cases in the medical literature.

Acknowledgment. We are very grateful to Miss Youko Tanaka for her secretarial assistantance.

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