review of cardiac tamponade as the presenting sign of extracardiac malignancy revealed only 29 cases, of which four were sarcomas and 25 were carcinomas. In this series, the most common primary type of extracardiac malignancy presenting with cardiac tamponade was lung cancer, though other varieties included pancreas, stomach, ovary, kidney, and lymphoma. Primary neoplasms of the heart and pericardium are quite rare, with a reported incidence rate of 0.001 to 0.28 percent of autopsy series. However, metastatic disease of the heart is surprisingly common with an incidence of about 10 percent of cancer-related deaths in autopsy series. Although cardiac symptoms during life are uncommon, clinical manifestations in such patients fall into recognizable patterns: 1) pericardial involvement, with findings such as effusion, tamponade, and constriction; 2) myocardial involvement, with findings such as rhythm disturbances, pump failure, cardiac rupture, and coronary artery occlusion; 3) endocardial involvement, with findings mimicking valvular stenosis and bacterial endocarditis; and 4) tumor thrombosis of cardiac chambers, with findings of chamber inflow and/or outflow obstruction.

The lymphatic circulation of the heart and pericardium has been described in detail both in human and animal subjects. An extensive subendocardial plexus exists which empties through myocardial channels into a subepicardial plexus. Branches from this plexus then drain into larger trunks which generally parallel the coronary vasculature, ultimately exiting the heart into mediastinal nodal structures. Spread of tumor emboli through these lymphatics and direct invasion of the pericardium appear to be the major mechanisms for development of malignant pericardial disease.

We emphasize the rapid and lethal progression of neoplastic cardiac tamponade which the current case exemplifies and which has been noted by others. However, Fraser et al. noted that the median survival time of those patients with neoplastic cardiac tamponade receiving pericardiectomy was 5.5 months as compared to those who received either supportive care with pericardiocentesis (seven days) or radiation therapy or chemotherapy (five weeks). These considerations support an aggressive policy in dealing with this problem. Our experience with this case and others suggests that CT can be decisive in the establishment of this diagnosis and subsequent treatment planning.

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Benign Mesenchymal Tumor of the Heart

Spontaneous Regression and Disappearance of Pulmonary Artery Stenosis

Yu-Chen Lee, M.D.; Robert T. Singleton, M.D.; and Chik-Kwun Tang, M.D.†

We describe a patient who was found to have a large benign mesenchymal tumor of the right ventricular wall and right pulmonary artery stenosis at the age of 20 months. Following biopsy of the tumor, the patient's respiratory distress improved gradually and cardiac catheterization at the age of ten years showed normal intracardiac pressures and disappearance of right pulmonary artery stenosis. He remained asymptomatic at age 20. Possible factors for the spontaneous regression of the tumor are discussed.

Partial or complete disappearance of a tumor in the absence of treatment is a rare but well known phenomenon. However, to our knowledge, spontaneous regression of cardiac tumors has not been reported. The purpose of this article is to describe a patient who had a large benign mesenchymal tumor of the heart which has regressed; pulmonary artery stenosis disappeared following biopsy of the tumor.

CASE REPORT

A 20-month-old baby was admitted to the University of Maryland Hospital on December 14, 1959. The prenatal history was unremarkable and the delivery was uneventful. Since the age of 14 days, he had been admitted to a local hospital on numerous occasions due to dyspnea and wheezing. He was found to have marked cardiomegaly, and treatment with digitalis failed to improve the symptoms. Physical examination on admission revealed a well-developed, well-nourished boy without cyanosis or clubbing. Blood pressure was 100/60 mm Hg, and pulse rate was 150/min with frequent premature beats. The chest was symmetric and coarse rhonchi were heard throughout. The heart was markedly enlarged; a grade 3/6 ejection systolic murmur was heard over the base and the second heart sound at the pulmonic area was widely split. The liver was palpable 2 cm below the right costal margin. Hemoglobin was 10 g/dl, white blood cell count was 11,500/ml with 65 percent polymorphs, 22 percent lymphocytes, 12 percent monocytes,

*From the Division of Cardiology, Department of Medicine, and Department of Pathology, University of Maryland Hospital, Baltimore. Reprint requests: Dr. Lee, Division of Cardiology/Medicine, 22 Greene Street, Baltimore 21201
Right heart catheterization was performed on December 21, 1959. There was no evidence of intracardiac shunt by blood oxygen analysis. The pressures (mm Hg) at the various sites were as follows: right atrium mean, 9; right ventricle, 60/5; main pulmonary artery, 60/18; left pulmonary artery, 60/18; right pulmonary artery, 25/16; and the pulmonary artery wedge mean, 12. Angiocardiogram showed a large, soft, tissue mass arising anteriorly on the right side of the heart and compression of the right pulmonary artery. There was no filling defect of the cardiac chambers.

Exploratory thoracotomy was performed on March 10, 1960. There was a large multinodular, whitish, rubbery mass arising from the right ventricular wall extending to the pulmonary artery which appeared inoperable. A small section of the tumor was obtained for pathologic study and the chest was closed. The postoperative course was uneventful.

The biopsy specimen consisted of fragments measuring 0.8 x 1.2 cm in aggregate. The tissue was whitish in color, soft and homogeneous, partly covered by a greyish glistening membrane. Microscopically, some areas were more cellular than others. The tumor was composed almost exclusively of spindle cells which possessed elongated nuclei with inconspicuous nucleoli. The cytoplasm was relatively scanty and eosinophilic. Long cytoplasmic processes were frequently observed. In the more cellular areas, the tumor cells formed bundles or fascicles, whereas in the less cellular areas, the tumor cells were arranged in a haphazard fashion and were separated by clear spaces. Masson trichrome stain demonstrated abundant extracellular collagen fibers. Cross striations were not identified by PTAH stain. No acceptable elastic fibers were found with Verhoff-von Gieson stain. There was a rich vasculature, mainly capillaries, within the tumor tissue. This tumor is unusual as it did not conform exactly to any of the recognized soft tissue lesions. It was believed to be a benign mesenchymal tumor and most likely fibroblastic.

During the ensuing years, dyspnea and wheezing improved gradually, although digitalis was discontinued, and he did not receive any form of treatment. He was able to attend school and participate in sports activities. He was readmitted to the University Hospital on April 14, 1969 for cardiac catheterization at which time he was asymptomatic. Physical examination revealed a well-developed, well-nourished boy. The blood pressure was 120/78 mm Hg, and pulse rate, 62/min with occasional premature beats. The heart was not enlarged, no murmur was heard and the pulmonic second sound was physiologically split. The chest x-ray film showed no evidence of cardiomegaly and there was pleuropericardial scarring on the right mediastinal area (Fig 3). The electrocardiogram showed sinus rhythm with occasional unifocal ventricular premature beats and incomplete right bundle branch block. Cardiac catheterization revealed no evidence of intracardiac shunts; intracardiac pressures (mm Hg) were as follows: right atrial mean, 7; right ventricle, 30/4; main pulmonary artery, 28/14; right pulmonary artery, 28/14; left pulmonary artery, 26/16.

Following discharge from the hospital, he has been examined periodically at the cardiology clinic. His exercise tolerance remains excellent. Chest x-ray examination revealed no cardiomegaly; ECG was unchanged and echocardiogram was normal.

Benign Mesenchymal Tumor of the Heart (Lee, Singleton, Tang)
DISCUSSION

Benign primary cardiac tumors are rather rare entities.\textsuperscript{4,5} Correct clinical diagnosis is usually not made before surgical exploration or autopsy. Clinically, it may mimic various cardiac lesions such as endocardial fibroelastosis,\textsuperscript{6} subaortic stenosis,\textsuperscript{7} or pulmonic stenosis.\textsuperscript{8} Although classified as benign, such tumors frequently cause sudden death by encroachment upon the cardiac cavities or compression of the conducting systems.\textsuperscript{9,10} More recently, successful removal of the intramural ventricular cardiac fibroma have been reported in approximately 16 cases.\textsuperscript{3,4,5,9,10} Symptomatic improvement may occur as the patient becomes older. Hoen and associates\textsuperscript{4} reported a patient who had cardiac failure in childhood in whom symptoms improved as the patient became older. At the age of 11 years, the patient underwent exploratory thoracotomy and a $6 \times 4$ cm tumor was found in the left ventricular wall. Biopsy of the tumor demonstrated benign fibrous tumor. One and one-half years after the diagnosis of fibroma of the left ventricle was established, the patient’s physical examination, chest x-ray film, and ECG revealed no change and exercise tolerance was unlimited. It was assumed that the tumor was present during the first year of life and there was no progressive enlargement of the tumor out of proportion to the growth of the normal myocardium. Our patient likewise had a large tumor producing recurrent episodes of dyspnea and wheezing, as well as right ventricular hypertension and right pulmonary artery stenosis up to age 20 months. Following exploratory thoracotomy and biopsy of the tumor, his symptoms gradually improved. In addition to the symptomatic improvement, chest x-ray examination revealed normalization of the cardiac silhouette, and right heart catheterization revealed disappearance of right ventricular hypertension and right pulmonary artery stenosis. These changes occurred without any form of treatment. Therefore, it is reasonable to assume that spontaneous regression of the tumor took place.

Several possible factors have been speculated upon for the spontaneous regression of tumors. These include: immunologic factors; elimination of a carcinogen; body response to operative trauma; hormone factors; role of irradiation; infection and/or fever; drugs.\textsuperscript{8} Our patient had gradual improvement of his symptoms following biopsy for his tumor and he did not receive any medication or radiation. Therefore, it is speculated that the most likely factor responsible for the spontaneous regression of the cardiac tumor in this case is due to operative trauma. It has been stated that operative trauma invokes a reaction that increases the patient’s immunologic resistance to the tumor.\textsuperscript{8}

Of additional interest in this patient is the presence of pulmonary hypertension in the face of unilateral pulmonary artery obstruction and subsequent disappearance of both conditions. Clinically, in unilateral pulmonary artery stenosis, there is no major change in cardiac dynamics.\textsuperscript{11} However, in animal studies, Falkenbach et al\textsuperscript{12} observed progressive pulmonary hypertension after partial occlusion of the left pulmonary artery, but none when the left pulmonary artery was ligated. It has been speculated that hypoxia or ischemia of the lung may stimulate the right ventricle to increase pressure proportionately to the pulmonary artery coarctation.\textsuperscript{12}

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