

Case Records of the Massachusetts General Hospital

*Weekly Clinicopathological Exercises*

FOUNDED BY RICHARD C. CABOT

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Case 19-1997**PRESENTATION OF CASE**

A 57-year-old man was admitted to the hospital because of progressive edema and exertional dyspnea.

The patient had been well until eight weeks earlier, when he was admitted to this hospital because of recurrent chest pain. An electrocardiogram showed a normal rhythm at a rate of 69, with poor R-wave progression in the right precordial leads, and elevated ST segments, with a tendency toward biphasic T waves, in leads V_3 through V_5 . A radiograph of the chest revealed patchy air-space disease in the right lower lobe consistent with pneumonia; the cardiac silhouette was normal. The patient received alteplase intravenously. A coronary angiographic examination performed 90 minutes later showed 70 percent stenosis in the midportion of the left anterior descending artery, with normal flow and 30 percent ostial and distal stenoses in the left main coronary artery. There was no enzymatic evidence of a myocardial infarct. A low-level thallium stress test showed no additional ST-segment changes, and regional left ventricular perfusion was normal except for nonspecific apical thinning. A cardiac ultrasonographic examination revealed a left ventricular ejection fraction of 60 percent. Repeated electrocardiograms were unchanged. The patient was discharged on the seventh hospital day and remained free of chest pain during the next two weeks.

One month before admission, he was given ciprofloxacin and analgesic agents for prostatitis and Peyronie's disease. During the week before admission, edema gradually developed in both legs, accompanied by dyspnea on walking uphill and lightheadedness on standing. He returned to this hospital.

The patient was retired because of degenerative

joint disease of the spine. He had a history of hypertension and hyperlipidemia. He had drunk excessive amounts of alcohol until 10 months before admission and had a history of more than 100 pack-years of cigarette smoking. Evidence of chronic obstructive pulmonary disease was discovered 14 months before admission. Four months before admission, he underwent resection of a squamous-cell carcinoma of the tongue (tumor-node-metastasis [TNM] classification, T1N0M0), followed by radiation therapy, with the thorax shielded. There was no history of recent chest pain, dyspnea at rest, cough, fever, chills, syphilis, or exposure to tuberculosis, or of documented coronary disease in his family.

His temperature was 35.8°C, his pulse was 88, and his respirations were 20. His blood pressure was 95/70 mm Hg in the supine position, with a pulsus paradoxus of 10 to 20 mm Hg, and 90/70 mm Hg in the standing position.

On examination, the tongue was well healed; no lymphadenopathy was found. There was dryness of the oral mucous membranes. The jugular venous pressure was 12 cm of water, without inspiratory distention of the external jugular vein. Dullness and diminished breath sounds were present at both lung bases. A three-component pericardial friction rub was present. There was ++ pitting edema distal to the knees.

The urine was normal. The calcium, phosphorus, bilirubin, and alanine aminotransferase levels were normal. The results of other laboratory tests are shown in Tables 1 and 2. Tests for antinuclear antibodies, anti-native-DNA antibodies, and rheumatoid factor were negative. An electrocardiogram showed sinus tachycardia at a rate of 101, with no change in the elevated ST segments in leads V_3 through V_5 . Radiographs of the chest showed a band-like opacity that projected in an inferolateral direction from the right infrahilar region, blunting of both costophrenic sulci, and a soft-tissue opacity along the right major fissure. The heart was enlarged and globular, suggesting a possible pericardial effusion. A cardiac ultrasonographic examination showed Doppler evidence of minimal mitral and tricuspid regurgitation. Both ventricles appeared normal. The estimated left ventricular ejection fraction was 66 percent. A moderate circumferential pericardial effusion was present, without evidence of tamponade; echodensities along the visceral pericardium were considered to be consistent with the deposition of fibrin or other material. A computed tomographic (CT) scan of the chest (Fig. 1) showed a central mass-like consolidation in the

TABLE 1. HEMATOLOGIC LABORATORY VALUES.

VARIABLE	ON ADMISSION	ON THIRD HOSPITAL DAY
Hematocrit (%)	35.3	30.6
Mean corpuscular volume (μm^3)	82	—
Erythrocyte sedimentation rate (mm/hr)	35	—
White-cell count (per mm^3)	8,700	6,300
Differential count (%)		
Neutrophils	86	—
Lymphocytes	2	—
Monocytes	11	—
Eosinophils	1	—
Prothrombin time (sec)*	15	15.2
Partial-thromboplastin time (sec)	36	36.5
Platelet count (per mm^3)	233,000	212,000

*Control value, 12.2.

TABLE 2. BLOOD CHEMICAL VALUES.*

VARIABLE	ON ADMISSION	ON THIRD HOSPITAL DAY
Urea nitrogen (mg/dl)	27	32
Creatinine (mg/dl)	1.8	2
Total protein (g/dl)	—	6.3
Albumin	—	2.9
Globulin	—	3.4
Sodium (mmol/liter)	132	141
Potassium (mmol/liter)	3.6	4.1
Chloride (mmol/liter)	90	101
Carbon dioxide (mmol/liter)	29	32.5
Magnesium (mmol/liter)	0.65	Normal
Glucose (mg/dl)	47	—

*To convert the values for urea nitrogen to millimoles per liter, multiply by 0.357. To convert the values for creatinine to micromoles per liter, multiply by 88.4. To convert the value for magnesium to milliequivalents per liter, divide by 0.5. To convert the value for glucose to millimoles per liter, multiply by 0.05551.

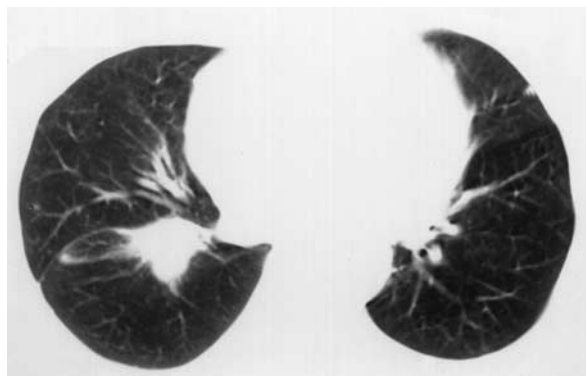


Figure 1. CT Scan of the Thorax.

A central, triangular, mass-like consolidation is present in the right lower lobe, extending to the major fissure, which is displaced posteriorly.

TABLE 3. FINDINGS ON EXAMINATION OF PERICARDIAL FLUID.

VARIABLE	VALUE
Quantity of fluid (ml)	392
Appearance of fluid	Grossly bloody
Initial pressure (mm)	16
Hematocrit (%)	6
Non-red-cell count (per mm^3)	980
Differential count (%)	
Neutrophils	63
Lymphocytes	12
Monocytes	8
Eosinophils	1
Nonhematic cells	16
Glucose (mg/dl)*	84
Total protein (g/dl)	4.9
Lactate dehydrogenase (U/liter)	1607
Staining for acid-fast and other microorganisms	Negative
Cytologic examination	Red cells only

*To convert the value for glucose to millimoles per liter, multiply by 0.05551.

TABLE 4. FINDINGS ON RIGHT-SIDED CARDIAC CATHETERIZATION.

VARIABLE	VALUE (mm Hg)
Right atrial mean pressure	14
Pulmonary arterial pressure	34.20
Mean pulmonary arterial pressure	27
Right ventricular pressure	34.16
Pulmonary-capillary wedge pressure	20

right lower lobe that displaced the major fissure slightly posteriorly, a finding consistent with volume loss in the lower lobe. A tiny, calcified, subpleural pulmonary nodule in the lingula, a tiny, noncalcified nodule in the right middle lobe, and a left internal mammary node, 1 cm in along the short axis, were present, as well as a pericardial effusion and a small left pleural effusion. A 24-hour specimen of urine was 1450 ml in volume; the sodium level was 11 mmol per liter, the potassium level was 30 mmol per liter, the chloride level was 1.5 mmol per liter, and the creatinine level was 0.75 mg per milliliter.

The patient's medications (metoprolol in a reduced dose, ciprofloxacin, isosorbide mononitrate, guaifenesin, and transdermal fentanyl) were continued. The temperature did not exceed 37.2°C, and the patient's hemodynamic status remained stable. On the evening of his admission, he expectorated bloody liquid on several occasions. Examination revealed a small ulcer in the buccal mucosa. On the second hospital day, an electrocardiogram showed

a normal rhythm at a rate of 64, without any other change. On the third day, the calcium, phosphorus, aspartate aminotransferase, alanine aminotransferase, and thyroid-stimulating hormone levels were normal. The results of other laboratory tests are shown in Tables 1 and 2. On the fourth hospital day, after the intravenous administration of fresh-frozen plasma, a pericardiocentesis was performed (Table 3), followed by right-sided cardiac catheterization (Table 4).

A diagnostic procedure was performed.

DIFFERENTIAL DIAGNOSIS

DR. FERDINAND J. VENDITTI, JR.*: May we review the radiographs?

DR. ELIZABETH A. DRUCKER: The chest film from the initial admission shows patchy consolidation in the right lower lobe. A radiograph of the chest obtained during the current admission shows enlargement of the cardiac silhouette, raising the question of pericardial effusion. The CT scan (Fig. 1) shows a perihilar mass-like consolidation in the right lower lobe that extends to the major fissure and reflects volume loss. A circumferential pericardial effusion, a small left pleural effusion, and an enlarged left internal mammary node are also visible.

DR. VENDITTI: This patient had signs and symptoms of heart failure, a hemorrhagic pericardial effusion, a process in the right lower lobe, and mediastinal lymphadenopathy. After the removal of 392 ml of pericardial fluid, the right-sided diastolic and pulmonary-capillary wedge pressures remained elevated. I believe that the initial chest pain and ST-segment and T-wave abnormalities did not indicate the presence of coronary artery disease but were instead the first manifestations of a pericardial process. Therefore, I shall discuss pericardial disease first.

The normal pericardium is a relatively inelastic envelope that helps maintain normal ventricular compliance, limits excessive ventriculoatrial regurgitation and acute dilatation, and helps maintain a functionally optimal cardiac shape. Membranous properties reduce friction due to cardiac contractions and prevent the spread of infection and inflammation from contiguous structures. Finally, the ligamentous function of the pericardium limits unnecessary cardiac displacement within the chest.¹ Diseases of the parietal pericardium, the visceral pericardium, or both can interfere with these important functions.

The pericardium normally contains 50 to 100 ml of fluid. Larger amounts of fluid can accumulate as a result of inflammation or other processes in the absence of evidence of cardiac compression or can compromise the hemodynamic function of the heart,

depending on the volume of fluid and the rapidity of its accumulation. Relatively small effusions that accumulate rapidly, such as those resulting from bleeding, can cause hemodynamic compromise or cardiac tamponade.

Cardiac tamponade is frequently manifested by arterial hypotension, pulsus paradoxus, and elevated systemic venous pressure. When the normally low intrapericardial pressure equals the pressures in the right atrium and right ventricle during diastole, the transmural pressure distending these chambers drops to zero. Further accumulation of fluid increases the intrapericardial pressure, which is transmitted to the right side of the heart, raising the right ventricular diastolic pressure, which can reach the level of the left ventricular diastolic pressure. Ultimately, equalization of the intrapericardial and ventricular filling pressures results in a decrease in diastolic and stroke volumes, with a subsequent decline in arterial pressure.²

Elevated systemic venous pressures during cardiac tamponade result from altered right-sided filling. During ventricular systole, blood usually surges from the venous system and enters the right atrium, resulting in an x descent on venous pressure tracings, and a second surge during ventricular diastole, when the tricuspid valve opens and ventricular filling commences, is represented by the y descent. When the heart is compressed by pericardial fluid, there is still considerable venous inflow into the right atrium during ventricular systole and a prominent x descent on the venous tracing, because the total volume of the heart declines during ventricular ejection. However, elevated intrapericardial pressures during ventricular diastole attenuate the usually rapid ventricular filling and blunt the y descent. In fact, the right atrial pressure may be lower than the intrapericardial pressure, resulting in diastolic collapse of the right atrium.

The negative intrathoracic pressure generated during inspiration further alters the pattern of left and right ventricular filling. Inspiration normally results in an increase in venous return to the right side of the heart and a corresponding small decrease in left ventricular diastolic volume. During cardiac tamponade, there is an exaggerated increase in right ventricular volume, accompanied by a marked decrease in left ventricular volume during inspiration. The decrease in left ventricular diastolic volume results in decreased aortic flow, a drop in systolic arterial pressure, and pulsus paradoxus. Removal of fluid from the pericardial space relieves the compression and restores the hemodynamic variables to normal values. Frequently, removing as little as 50 ml of fluid results in a return to normal arterial pressure.

Many systemic diseases can result in pericarditis with effusions (Table 5).

When the pericardium becomes thickened, fibrot-

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TABLE 5. CAUSES OF PERICARDIAL EFFUSION.

Infections
Viral
Bacterial
Tuberculous
Fungal
Inflammatory diseases with vasculitis
Systemic lupus erythematosus
Wegener's granulomatosis
Polyarteritis
Neoplasms
Primary cardiac tumor
Metastatic neoplasm
Metabolic disorders
Renal failure
Myxedema
Other causes
Sarcoidosis
Löffler's endomyocarditis
Syndromes due to myocardial or pericardial injury

ic, and adherent, signs and symptoms of constrictive pericarditis appear. Constriction follows an inflammatory process, with or without effusion. In most cases, the visceral pericardium and parietal pericardium fuse and stiffen, restricting diastolic filling. Symmetric constriction elevates and equalizes diastolic pressures in all four cardiac chambers, but early diastolic filling is unimpaired, because the total cardiac volume is lower. However, early, rapid filling is halted when the intracardiac volume reaches the limit of the noncompliant pericardium, resulting in a characteristic dip-and-plateau pattern in the pressure wave form recorded in both ventricles during diastole and a prominent y descent on the right atrial tracing. These findings are generally absent in patients with cardiac tamponade.

Whereas pulsus paradoxus is characteristic of cardiac tamponade, it is not a prominent feature of constrictive pericarditis. The negative intrathoracic pressure generated during inspiration is not transmitted to the pericardial space; therefore, there is no inspiratory augmentation of filling in the right heart and no decrease in left ventricular diastolic filling. As a result, systemic venous pressures do not fall with inspiration; indeed, they may rise as venous return to the thorax increases because of negative intrathoracic pressures. This finding on physical examination is called Kussmaul's sign, which is usually absent in cases of pure tamponade.

The causes of constrictive pericarditis are many. The most common is tuberculosis.³ Other infections can also result in constriction.

A review of the hemodynamic data in this case reveals elements of both effusive and constrictive disease. A moderate-sized effusion was present on echocardiographic examination, and the initial pressure during pericardiocentesis was markedly elevated. Af-

ter fluid had been drained from the pericardial cavity, however, the right atrial, right ventricular, and pulmonary-artery diastolic pressures, as well as the pulmonary-capillary wedge pressure, remained elevated, suggesting that ventricular filling was still impaired, perhaps by a noncompliant myocardium, as seen in cases of restrictive cardiomyopathy, or by a noncompliant pericardium, as in effusive-constrictive pericarditis. Restrictive cardiomyopathy is usually accompanied by right ventricular systolic pressures that exceed 60 mm Hg and left ventricular diastolic pressures that exceed the right ventricular diastolic pressures by more than 5 mm Hg. Since such pressures were not reported in this case, I believe that the patient's hemodynamic profile is most consistent with a diagnosis of effusive-constrictive pericarditis, causing both hepatic congestion and azotemia.

Subacute effusive-constrictive pericarditis was first characterized hemodynamically in 1971 by Hancock.⁴ The clinical and hemodynamic characteristics of 13 patients with a distinct pathophysiologic profile that differed from that associated with either pure tamponade or constriction were reported. The hallmark of effusive-constrictive pericarditis is persistent elevation of diastolic pressures after the removal of pericardial fluid has returned the intrapericardial pressures to normal values.⁴ The atrial and ventricular wave forms change from a pattern that is consistent with cardiac tamponade to one that is more consistent with constriction, a prominent y descent typical of constriction often becomes apparent on the right atrial pressure tracing, and the prominent diastolic dip and plateau of constriction becomes evident in the right ventricle. In this syndrome, involvement of the visceral pericardium produces constriction. In some cases, the parietal pericardium may be relatively spared. Effusive-constrictive pericarditis has been reported in association with many disorders, including tuberculous, viral, and neoplastic disorders, radiation damage, and hemopericardium. In most series, mediastinal irradiation, usually for a malignant tumor, is the most common cause of this syndrome. Effusive-constrictive pericarditis may progress to chronic constriction.

Patients with tumors who undergo mediastinal irradiation may have asymptomatic effusions, cardiac tamponade, effusive-constrictive pericarditis, or constrictive pericarditis. Pericardial effusions usually occur within two years after irradiation and are most often characterized by serous, protein-rich fluid, but the fluid can be hemorrhagic.⁵ Although this patient had received radiation for his tongue cancer, the use of thoracic shielding to prevent direct exposure of the chest during treatment makes it unlikely that the pericarditis was due to irradiation.

Effusive-constrictive pericarditis can result from hemopericardium.^{6,7} Although this patient received thrombolytic medication several weeks before pres-

entation, his illness would have been much more acute if hemorrhage into the pericardial cavity had occurred at the time of his initial presentation.

Tuberculous pericarditis is a potentially lethal manifestation of extrapulmonary tuberculosis.⁸⁻¹⁸ Usually the onset is insidious, developing over a period of weeks to months.¹⁴ The symptoms include cough, dyspnea, chest pain, night sweats, orthopnea, pedal edema, weight loss, and hemoptysis.^{8,9} Clinical findings include fever, cardiomegaly, a pericardial friction rub, tachycardia, distant heart sounds, pulsus paradoxus, hepatomegaly, edema, and neck-vein distention. Pericardial involvement can occur in the absence of pulmonary infiltrates, and in one series, only 32 percent of the patients had radiographic evidence of lung disease.⁹ Pericarditis can result from the rupture of adjacent involved lymph nodes or hematologic spread. Typically, the process begins as effusive-constrictive disease and progresses to constrictive pericarditis without effusion. In the later stages, acid-fast bacilli are usually not detected, but caseating granulomas involving the pericardium and epicardium may be present.

Making a definitive diagnosis of tuberculosis can be difficult. Microscopical examination of pericardial fluid seldom reveals acid-fast bacilli, but cultures frequently grow mycobacteria.⁸⁻¹¹ The pericardial fluid is usually serous and protein-rich, with a predominance of lymphocytes and monocytes, but hemorrhagic effusions can be seen.¹² Echocardiographic features of tuberculous pericarditis include visceral pericardial thickening, with a shaggy layer of material that is presumably fibrinous exudate,¹⁸ although this finding is not specific.

The pericardial process in this patient is consistent with a diagnosis of tuberculosis, but the pulmonary features are not typical of the disorder. The calcified pulmonary nodule in the lingula suggests previous tuberculosis, but the location of the infiltrate in the right lower lobe is not characteristic of secondary tuberculosis, in which upper-lobe infiltrates predominate.^{19,20} When there is lower-lobe involvement, a mass-like consolidation can be mistaken for carcinoma.²¹ Although the pulmonary involvement in this case is not typical of tuberculosis, it is consistent with that diagnosis. A malignant tumor, however, provides a better explanation of the findings in this case.

Primary cardiac tumors are rare, but autopsy studies have demonstrated an 8 to 18 percent frequency of metastatic disease involving the heart in patients who died from cancer.^{20,22} The most common primary cancers causing metastases are lung cancer, breast cancer, lymphoma, and leukemia, but melanoma, sarcomas, and many carcinomas, including those of the head and neck, have also been reported to spread to the heart.^{20,22,23} Cardiac involvement can occur by hematologic or lymphatic spread or by direct extension of the tumor.

Pericardial involvement occurs in most cases of cancer with cardiac involvement but is often not recognized clinically.²⁴ Not all pericardial effusions in patients with malignant tumors are due to metastatic spread. Up to half of patients with cancer and symptomatic pericarditis have nonmalignant effusions.²⁵ Although the pericardial fluid may be serous, it is more commonly serosanguineous or hemorrhagic.^{26,27} Cytologic studies of the fluid may have false negative results in some patients, especially those with lymphoma or mesothelioma.²⁵ Over half the patients with pericardial metastases in one series had cardiac tamponade,²⁶ but a subacute effusive-constrictive hemodynamic pattern may also be seen.

In one series of eight patients with malignant effusions, the typical pattern of effusive-constrictive pericarditis was seen at the time of pericardiocentesis.²⁷ Drainage of 200 to 2500 ml of fluid resulted in a decrease in the intrapericardiac pressure to 0, without a return of intracardiac pressures to normal values. In addition, there was a dip-and-plateau pattern of the ventricular diastolic wave form, a finding consistent with constriction. Five of six patients had grossly hemorrhagic effusions, with a fluid hematocrit of 5 to 20 percent. The visceral pericardium was involved in the malignant process, frequently with myocardial invasion, in the patients who were examined at autopsy.

The distribution of lymphoma depends largely on the cell type, but patients with non-Hodgkin's lymphoma frequently have disseminated disease.²⁸ Lymphadenopathy is detectable in 80 percent of patients, and systemic symptoms are common. In addition to other extranodal organs, lymphoma commonly involves the heart, particularly the pericardium. In one series,²⁹ almost 30 percent of the patients with non-Hodgkin's lymphoma had cardiac involvement at autopsy. In most cases, cardiac involvement is the result of direct spread from mediastinal masses or lymphatic spread from involved lymph nodes.³⁰ In view of the minimal mediastinal involvement, absence of lymphadenopathy on examination, and lack of systemic symptoms in this case, lymphoma is an unlikely diagnosis.

The extensive history of smoking and previous squamous-cell carcinoma make primary lung cancer a likely process in this patient. Pericardial involvement can be caused by direct invasion from an intrapulmonary or mediastinal mass but is more commonly caused by retrograde lymphatic spread from involved lymph nodes.³¹ Effusions can be large and are usually hemorrhagic. Occasionally, signs and symptoms of cardiac compression by the malignant effusion are the first manifestation of carcinoma.^{31,32} Cytologic examination of pericardial fluid usually leads to the diagnosis.²

Head and neck cancer, including cancer of the tongue, does not commonly metastasize to the lungs

and pericardium. The spread is usually local along the lymphatic vessels of the neck. In one series, distant metastatic disease was reported in only 2 percent of the patients with stage 1 oral squamous-cell carcinoma treated by surgical resection or irradiation.³³ When combination therapy was used, no distant metastases were seen during a minimal follow-up period of two years. There is one report, however, of a patient with a localized, well-differentiated squamous-cell carcinoma of the tongue that disseminated widely, causing death within nine months after presentation³⁴; that patient had cardiac involvement. Despite this report, since the disease in the patient under discussion was limited and combination therapy was used, I believe that widely metastatic squamous-cell carcinoma of the tongue is an unlikely diagnosis.

Primary cardiac tumors are 20 to 30 times less common than metastatic tumors, and only about 25 percent of primary cardiac tumors are malignant. The three most common forms are lymphoma, sarcoma, and mesothelioma.³⁵ Fewer than 20 cases of primary cardiac lymphoma have been reported in nonimmunosuppressed patients.³⁶ One diagnostic criterion is the absence of evidence of extracardiac involvement.³⁶ On the basis of the other findings in this case, I shall rule out a diagnosis of primary lymphoma.

The most common primary malignant tumor of the heart is sarcoma, and angiosarcoma is the most frequent histologic type.³⁷⁻⁴³ Typically, sarcomas are aggressive, and patients present late in their course. The diagnosis is often not made before death. Eighty percent of patients have systemic metastases at presentation.⁴⁰ The primary tumor typically arises on the right side of the heart, usually in the right atrium or atrioventricular groove. All layers of the heart can be involved, but the tumor usually starts in the epicardium or endocardium. Intracardiac growth can result in obstruction of valvular structures and the great veins. Pericardial involvement is common, with hemorrhagic effusions and obliteration of the pericardial cavity by the tumor.⁴⁴ Constriction has been reported in some cases.

Pericardial mesothelioma is rare, with the highest incidence in middle-aged adults. Nodular or diffuse involvement can occur. The tumor can present with constrictive pericarditis, hemorrhagic pericardial effusion with or without tamponade, acute myocardial infarction, obstruction of the superior vena cava, or congestive heart failure. The diffuse form typically causes marked pericardial thickening with obliteration of the pericardial cavity and constriction. The nodular variety may be restricted to one or two sites, with an associated hemorrhagic pericardial effusion. Like sarcomas, mesotheliomas usually present late in the course of the disease, with less than 30 percent of cases diagnosed before death.

In this case, I believe that the echocardiogram showed a mass-like lesion in the apical right ventricular wall, probably extending into the distal septum. Although there are no detailed views in this area, I suspect that this lesion is consistent with a sizable tumor invading the myocardium, such as a primary cardiac tumor, a tumor that metastasized from either the lung or the tongue, a primary angiosarcoma, or a nodular mesothelioma.

The diagnostic procedure was probably a bronchoscopic examination, with a biopsy of the abnormality in the right lower lobe or an echocardiographically guided biopsy of the right ventricular mass.

DR. JONATHAN G. ZAROFF: I saw the patient in consultation. His initial thrombolytic treatment seemed appropriate, even retrospectively. When he presented again with persistent ST-segment elevations without Q waves, I thought the original diagnosis was incorrect. I suspected a cardiac tumor, because the hemodynamic data from the pericardio-centesis were consistent with the presence of effusive-constrictive disease.

CLINICAL DIAGNOSIS

Malignant cardiac tumor.

DR. FERDINAND J. VENDITTI, JR.'S DIAGNOSIS

Malignant cardiac tumor, possibly primary, with effusive-constrictive pericarditis.

PATHOLOGICAL DISCUSSION

DR. GEOFFREY A. ROSE: A subsequent two-dimensional echocardiogram (Fig. 2) showed segmental dysfunction of the right ventricular apex associated with a mass apparently infiltrating the apical myocardium of both ventricles and the interventricular septum. Review of the previous echocardiogram at that time revealed the presence of the abnormality. A biopsy of the mass was performed under echocardiographic guidance.

DR. MICHAEL R. PINS: Microscopical examination of the specimen showed a grade 2 squamous-cell carcinoma infiltrating between myocytes and focally involving the epicardium (Fig. 3). After the diagnosis was established, the patient received a six-day course of cisplatin and fluorouracil. His hospital course was complicated by biventricular heart failure, and he died one month after the biopsy.

An autopsy revealed extensive squamous-cell carcinoma that replaced the pericardial space around both ventricles and infiltrated the apical myocardium of both ventricles and the septum (Fig. 4); the atria were minimally involved. There was no residual tumor in the tongue. The lung, however, contained an ill-defined squamous-cell carcinoma, 3 cm in diameter, in the hilar area that had spread around the adjacent vessels and metastasized to several lymph

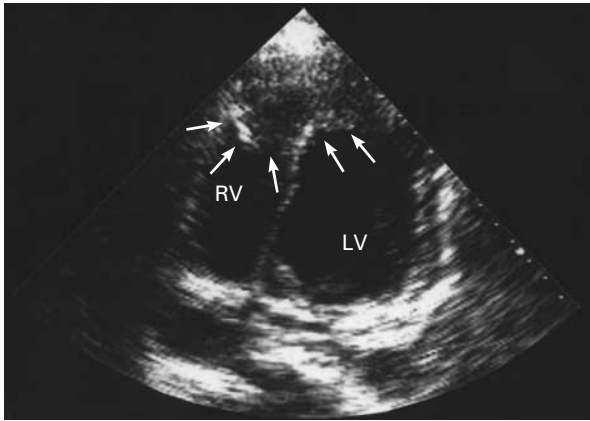


Figure 2. Two-Dimensional Echocardiogram Obtained after the Pericardiocentesis.

A large mass involves both the right ventricular (RV) and left ventricular (LV) apices (arrows).

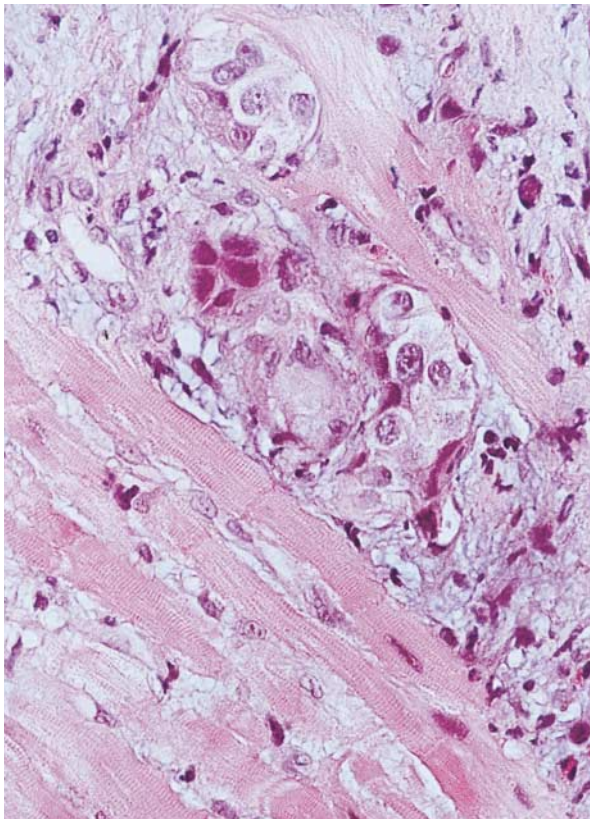


Figure 3. Specimen Obtained from a Biopsy of the Right Ventricular Endomyocardium, Showing Nests of Squamous-Cell Carcinoma Infiltrating between Pink Cardiac-Muscle Fibers (Hematoxylin and Eosin, $\times 480$).

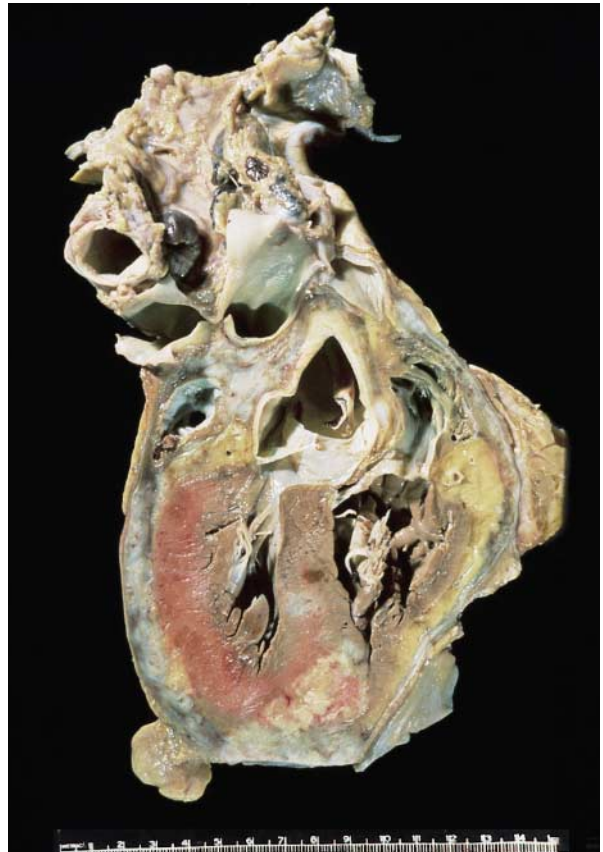


Figure 4. Heart and Great Vessels with Gray-White Tumor in the Pericardial Space and Invasion of the Interventricular Septum.

nodes. There was no evidence of direct extension of the tumor to the pericardium.

We are uncertain whether the lingual or pulmonary tumor had metastasized to the heart, but we favored the tongue as the primary site. Takagi et al.⁴⁴ reported that in cases of squamous-cell carcinoma of the tongue in which treatment had failed, the heart was the third most common site of hematogenous metastasis, with a frequency of 13.4 percent. Cases of carcinoma of the tongue with metastasis to the heart that were clinically similar to the current case have also been reported.^{34,45} Finally, in one autopsy study of patients with metastatic carcinoma, the heart and pericardium were involved in 8 percent of the cases.⁴⁶ In 24 percent of the cases with cardiac and pericardial involvement, the primary site of the carcinoma was the tongue; in only 7 percent of the cases with cardiac and pericardial involvement was the lung the primary site.

Metastatic squamous-cell carcinoma was also present in the liver and the penis. Additional findings included bilateral focal necrotizing pneumonia, pulmonary edema, and minimal coronary artery disease.

ANATOMICAL DIAGNOSIS

Squamous-cell carcinoma involving the tongue, lung, heart and pericardium, liver, and penis, probably with the primary tumor in the tongue.

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