

**Fig. 2.** Photomicrograph from operative biopsy specimen of right atrial mass shows tufts of endothelial-like cells forming and lining vascular channels. Nuclei are pleomorphic and atypical, but mitotic figures are not abundant. Immunocytochemical staining of endothelial-like cells with antihuman factor VIII was positive (not shown). Findings are consistent with diagnosis of angiosarcoma. (Hematoxylin & eosin stain; original magnification  $\times 100$ .)

Results of a follow-up NMR study in November, 1986 (Fig. 1, *E* and *F*), confirmed extension of the mass along the aorta and superior vena cava. In spite of this, she remained free of symptoms attributable to the tumor until March, 1987, when upper body swelling and dyspnea developed. Chest x-ray film demonstrated a mediastinal mass with tracheal compression. She was admitted to the hospital for terminal care and died shortly thereafter. Postmortem examination was not performed.

This case represents the longest reported observation of angiosarcoma of the heart, shown at an early stage by NMR imaging and echocardiography. Mean survival time after presentation has been reported to be 9 months, with 29 of 33 patients in the review by Glancy et al.<sup>1</sup> surviving less than 10 months. The exceptionally long survival time in this instance is most likely the result of early detection of the tumor during investigation of new-onset atrial fibrillation. NMR imaging, by virtue of its ability to obtain high-resolution images in an unrestricted field of view, was the most helpful technique in the preoperative

evaluation of this patient. Echocardiography provided less complete information than did NMR imaging; whereas repeated attempts at endomyocardial biopsy did not yield diagnostic tissue, NMR imaging correctly predicted the diagnosis of invasive tumor. Use of NMR imaging to identify tissue type has been reported for both cardiac and noncardiac lesions, for example, lipomatous hypertrophy of the interatrial septum<sup>5</sup> and cavernous hemangioma of the liver,<sup>6</sup> respectively. It is possible that, with the accumulation of data such as are presented here, NMR imaging may provide noninvasive determination not only of the extent of cardiac tumor but of tissue type as well, facilitating preoperative assessment of resectability.

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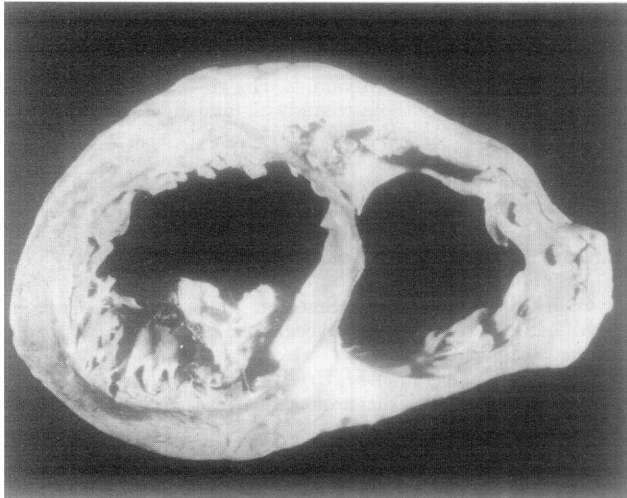
### Rapidly progressive heart failure resulting from cardiac sarcoidosis

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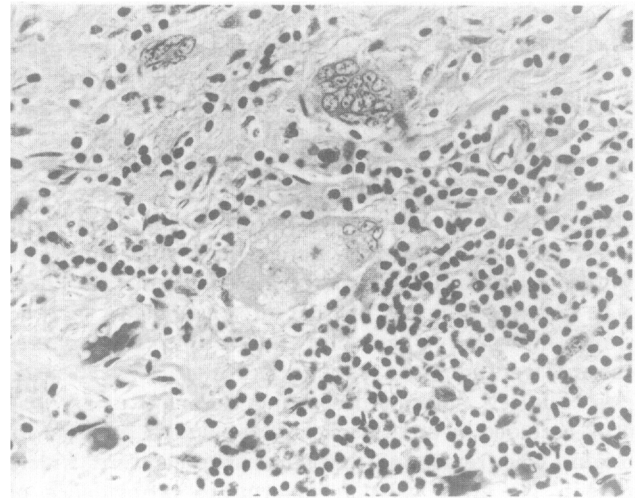
Progressive congestive cardiac failure has been shown to be the cause of death in 25% of patients with cardiac dysfunction resulting from sarcoid granulomatous infiltration.<sup>1</sup> Excluding sudden death, fatal arrhythmias, and valvular disease, a common clinical course for patients with worsening congestive heart failure (CHF) resulting from sarcoidosis is death within 9 to 12 months after the onset of symptoms.<sup>2,3</sup> We describe a patient with cardiac

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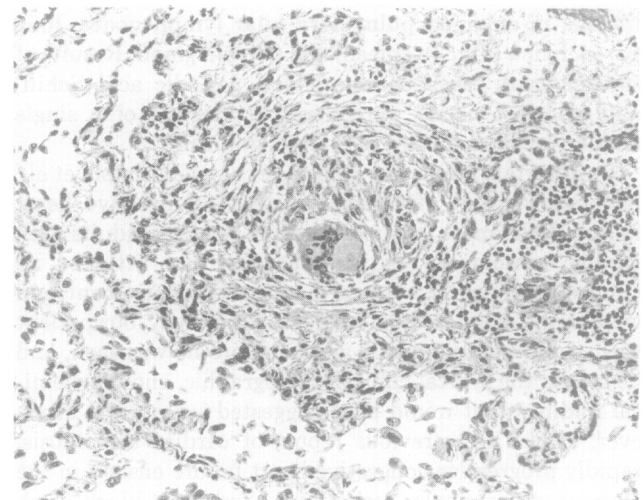
**Fig. 1.** Gross transverse slice of heart showing biventricular dilatation and marked thinning of interventricular septum. Gray-white lesions are interspersed throughout myocardium.



**Fig. 2.** Microscopic section of heart from one of the gray-white lesions. Multinucleated giant cell is present and noncaseating granulomatous inflammation is evident.

sarcoidosis who had rapidly progressive CHF that led to death within 30 days of presentation.

A 50-year-old athletic man was initially seen after 9 days of increasing weakness, fatigue, and nonproductive cough. Results of an outpatient chest x-ray examination showed bilateral infiltrates in the middle and lower lung fields interpreted as pneumonia. Oral antibiotic therapy was prescribed. The patient denied having ankle edema, orthopnea, or fevers before hospitalization. At the time of physical examination, blood pressure was 88/65 mm Hg, heart rate was 120 and regular, respirations were 24, and temperature was 36° C. Auscultation of the lungs showed bilateral inspiratory crackles to the scapular tips. There was marked jugular venous distention. The apical impulse was laterally displaced, and there was a quadruple rhythm (S4 and S3) without murmurs. The remainder of the examination results were notable for 1+ to 2+ pitting pedal edema. Laboratory assessment included a normal complete blood count and normal results of serum biochemical screening. The creatine kinase level was 172 IU/L (normal 40 to 180 IU/L) with an 8% MB fraction. Results of chest x-ray examination showed cardiomegaly with pulmonary infiltrates but no hilar adenopathy. An ECG showed sinus tachycardia and right bundle branch block with left anterior fascicular block. Right-sided cardiac catheterization showed that the pulmonary capillary wedge pressure was 31 mm Hg, pulmonary artery pressure was 46/24 mm Hg, and mean right atrial pressure was 15 mm Hg. The cardiac index, according to the thermodilution technique, was 1.54 L/min/m<sup>2</sup>, and the systemic vascular resistance index was 1569 dynes/sec/cm<sup>5</sup>/m<sup>2</sup>. Two-dimensional echocardiography confirmed global impairment of left ventricular (LV) contractility with moderate LV enlargement and normal wall thickness. A gallium-67 scan on the third hospital day showed no



**Fig. 3.** Microscopic section of lung showing noncaseating granulomatous changes and multinucleated giant cell.

increase in myocardial uptake to suggest myocarditis, and the resting LV ejection fraction was 13% by radionuclide ventriculography. Acute viral titers for coxsackie, echo, and cytomegalovirus were not elevated. Symptoms of congestive heart failure and prerenal azotemia became refractory to medical management despite aggressive treatment with diuretics, multiple inotropic agents, and vasodilators. Methylprednisolone, 1 mg/kg/24 hr, was prescribed on hospital day 19. Paroxysmal supraventricular arrhythmias were suppressed with procainamide. On hospital day 22 the patient was transferred to another facility for further assessment and possible insertion of an LV assist device, cardiac transplantation, or both.

Attempted coronary arteriography and a single right ventricular endomyocardial biopsy were poorly tolerated. After the procedure electromechanical dissociation developed and the patient could not be resuscitated.

At autopsy the heart weighed 420 gm. Both the ventricles and atria were dilated. The anterior and posterior walls of the left ventricle and interventricular septum were irregularly thinned and largely replaced by soft gray-white lesions (Fig. 1). Patchy gray-white infiltrates were also present in the walls of the right ventricle. There was marked thinning of the interventricular septum, which measured 4 mm near the cardiac base. Mural thrombi were adherent to the endocardium in both ventricles. The left anterior descending coronary artery was 60% occluded by an atherosclerotic plaque 5 cm from its origin. The remaining coronary arteries were widely patent. No gross valvular deformities or evidence of ventricular perforation were identified. A histologic section of the heart showed extensive myocardial destruction with fibrosis and noncaseating granulomatous inflammation (Fig. 2). The diagnosis of sarcoidosis was based on numerous multinucleated giant cells containing intracytoplasmic asteroid bodies in the myocardium and similar areas of noncaseating granulomatous inflammation and fibrosis in enlarged pulmonary hilar lymph nodes, lung parenchyma (Fig. 3), spleen, and thyroid gland. Results of staining with methenamine silver, periodic acid-Schiff, and acid-fast bacilli were negative. Results of a single endomyocardial biopsy were not significant.

Postmortem studies by Roberts et al.<sup>1</sup> and Matsui et al.<sup>2</sup> have shown that after sudden death, congestive heart failure is the most frequent mode of death in adults with cardiac sarcoidosis. Patients usually had little or no clinical evidence of extracardiac organ involvement and were rarely diagnosed before death.<sup>4</sup> Similarly, our patient was asymptomatic until 2 weeks before he was seen, and he lacked the typical clinical, radiographic, and biochemical findings that would have suggested sarcoidosis. However, unlike any previous report of cardiac sarcoidosis, rapidly progressive congestive heart failure ensued in the absence of ventricular arrhythmias and heart block. Such an unfortunate course, leading to death within weeks, would be more typical for viral, toxic, or giant cell myocarditis.<sup>5</sup> Extensive involvement of the heart and other organs by a noncaseating granulomatous reaction strongly suggests that the clinical events were the result of advanced cardiac sarcoidosis. Analysis of the present case suggests that cardiac sarcoidosis should be considered in patients with acute-onset, rapidly progressive, or unexplained congestive heart failure. Moreover, early endomyocardial biopsy is recommended<sup>6</sup> because it can frequently establish the diagnosis of cardiac sarcoidosis, which may alter initial patient management and outcome.

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### Dilated cardiomyopathy associated with natural killer cell deficiency

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An increasing number of reports suggest that the immune system may be involved in the pathogenesis of dilated cardiomyopathy (DCM).<sup>1,2</sup> Anderson et al.<sup>3</sup> reported a deficient activity of natural killer (NK) cells in a patient with DCM. However, the actual role of the immune mechanism has not been established in terms of the pathogenesis of DCM. We present a case of DCM with NK cell deficiency that was not responsive to treatment with recombinant interleukin-2 (rIL-2) and a small number of mature NK cells.

A 46-year-old man was admitted to the hospital because of cardiomegaly. He had a several-year history of exertional dyspnea. Pulse rate was 96 beats/min and regular, and blood pressure was 114/80 mm Hg. A third heart sound was heard. Results of a chest x-ray examination showed cardiomegaly. An ECG showed ST segment depression in the left precordial leads. Results of left-sided ventriculography showed diffuse hypokinesis, with a cardiac index of 1.53 L/min/m<sup>2</sup> and an ejection fraction of 15%. Results of coronary angiography were normal. Results of endomyocardial biopsy showed interstitial fibrosis and infiltration of a few lymphocytes. Immunologic studies of peripheral blood were performed. Serum immunoglobulin concentra-

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