Sick Sinus Node Syndrome as the Presenting Manifestation of Reticulum Cell Sarcoma*

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Presented is an unusual case of reticulum cell sarcoma whose initial manifestation was a syndrome of cardiac arrhythmias which fit the description of the “sick sinus node syndrome.” In addition, the clinical presentation included pericarditis and refractory congestive heart failure. At autopsy, the sinus node and cardiac conduction system were found to be extensively involved with reticulum cell sarcoma.

The sick sinus node syndrome (sluggish sinus node syndrome) is characterized by periods of bradycardia and tachyarrhythmia. More precisely, a sequence of severe bradycardia, or episodes of sinoatrial block, or frank sinus arrest alternates with paroxysmal supraventricular tachycardia, often terminating in asystole or a slow subsidiary escape rhythm. Although described as early as 1916,† the syndrome has only recently received clinical attention and recognition.2-4 Little is known about its etiology or natural history, except it is generally confined to the older age groups and may present with syncope. Presented here is an unusual case of infiltration of the heart with reticulum cell sarcoma, presenting with features of the “sick sinus node syndrome,” atrioventricular and intraventricular conduction disturbances, pericarditis, and refractory congestive heart failure.

CASE REPORT

A 71-year-old white woman was admitted to Billings Hospital in March, 1959 with a two-day history of non-exertional chest pain. Laboratory and physical examination were essentially unremarkable, except for blood pressure of 200/100 mm Hg. After discharge, she had no cardiovascular symptoms for approximately nine years; thereafter, in December, 1968, she developed chest pains. Pertinent physical findings were low grade fever, blood pressure 190/90 mm Hg, regular pulse of 90, a prominent systolic uniphase friction rub, and an atrial sound. No lymphadenopathy, hepatosplenomegaly, or skin lesions were noted. The electrocardiogram (ECG) showed premature atrial beats and short runs of multifocal atrial tachycardia. Chest x-ray examination and fluoroscopy, blood count, urinalysis and other studies, including a percutaneous liver biopsy, were unremarkable.

After six days of hospitalization, her fever, pleuritic chest pain, and pericardial rub abated.

After more than a year without symptoms, congestive heart failure developed and digitalis was prescribed with only initial symptomatic improvement. Upon hospitalization, in April, 1970, pertinent physical findings included irregular pulse rate 78/min, blood pressure 170/80 mm Hg (without pulsus paradoxus), orthopnea with a respiratory rate of 34/min, jugular venous distention to 5 cm at 45° elevation (without Kussmaul sign), bilateral decreased breath sounds and dullness to percussion, moist râles, and two plus pitting edema of the ankles. Cardiac examination revealed a quiet precordium with the point of maximum impulse in the fifth intercostal space, 5 cm to the left of the midclavicular line, a prominent third heart sound, grade I/VI early systolic ejection murmur at the apex, and at the base, normal splitting of the second heart sound with a grade II/VI systolic ejection murmur. No pericardial friction rub was heard. Multiple, firm, nontender, 3 x 4 cm lymph nodes were palpable in both axillae, and abdominal examination showed a firm, tender spleen, extending 8 cm below the left costal margin. The liver was normal to palpation and percussion. Chest x-ray film revealed a widened mediastinum, gross cardiomegaly, bilateral pleural effusions, and marked pulmonary vascular congestion. The ECG demonstrated atrial fibrillation with a slow ventricular response, rare junctional escape beats, and incomplete right bundle branch block (Fig 1). Complete blood count, urinalysis, electrolytes, BUN, creatinine, liver function, calcium and phosphorus, serum protein electrophoresis, and immuno-electrophoresis tests were within normal limits. Furosemide was administered with an effective, moderate diuresis and symptomatic improvement. However, 18 hours later, the patient became acutely tachypneic, diaphoretic, and hypotensive with an irregular pulse of approximately 30 per minute. ECG showed a slow atrial rhythm with high-degree AV block, incomplete right bundle branch block, and an unstable subsidiary junctional pacemaker (Fig 2). Although a temporary transvenous demand pacemaker resulted in transient improvement, worsening heart failure, azotemia, confusion, and lethargy developed and she expired a week later.

![Figure 1. ECG in April, 1970 shows atrial fibrillation with a relatively slow ventricular response and incomplete right bundle branch block.](http://journal.publications.chestnet.org/pdftoasx?url=/data/journals/chest/21525/ on 04/28/2017)
SICK SINUS NODE SYNDROME

Figure 2. Rhythm strip (lead II) shows a regular, slow atrial mechanism (49 per minute) and an unsteady, slow junctional subsidiary pacemaker (P waves noted by arrows).

Necropsy

A large tumor mass in the anterior mediastinum, involving hilar lymph nodes, compressed the ascending aorta and extended into the pericardial cavity. The pericardium, infiltrated by tumor, was densely fibrotic and thickened; however, no evidence of frank constriction was noted. The heart weighed approximately 500 gm, and the spleen was massively enlarged (500 gm). Tumor nodules were noted in the spleen, as well as in the axillary and abdominal lymph nodes. Microscopically, sheets of malignant reticulum cells infiltrated the epicardium and myocardium along perivascular spaces and lymph channels. Although the right atrium was the most extensively involved part of the heart, lymphoma replaced the sinus node, both septae, AV node, bundle of His, and the right bundle branch (Fig 3). No significant coronary atherosclerosis, old or recent myocardial infarction, or valvular lesions were noted. In addition, tumor infiltration was seen in the spleen, lymph nodes, pancreas, small bowel, bone marrow, and skeletal muscles. Stains for amyloid were negative.

Discussion

The tumors that most frequently invade the heart include carcinoma of the lung and breast, malignant melanoma, leukemias and lymphomas. Cardiac involve-

ment, whether by direct extension (lung, breast, lymphoma) or by metastasis (melanoma, leukemia), is reported to occur in 10 to 20 percent of patients with malignant tumors. Roberts and other investigators have specifically noted that reticulum cell sarcoma involves the heart more frequently than other types of lymphoma (the incidence approaches 40 percent). However, signs and symptoms of cardiac dysfunction are quite unique and, because of the relative paucity of clinical significance, the antemortem diagnosis of cardiac involvement is rarely entertained. Unusually, systemic lymphoma has presented with cardiovascular derangements. However, to our knowledge there is no previous report of reticulum cell sarcoma presenting with the sick sinus node syndrome.

In the present case, although axillary lymphadenopathy, splenomegaly and a widened mediastinum suggested lymphoma, the antemortem diagnosis of cardiac involvement was not established because of the patient’s critical condition. In the appropriate clinical setting, the diagnosis of cardiac involvement must be entertained as irritation has been successful in treating lymphomatous extension to the pericardium, heart, and great vessels. More specifically, several authors stress that in patients with documented cardiac reticulum cell sarcoma, therapy may produce symptomatic improvement, tumor regression, or, occasionally, absolute cure. Therefore, early diagnosis is of paramount importance. Drug therapy of the changing arrhythmias in the sick sinus node syndrome is difficult and challenging. Atropine is usually ineffective in increasing the atrial rate, and antiarrhythmic drugs such as procainamide, quinidine, and propranolol can be used with safety only after the insertion of an artificial pacemaker.

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Recurrent Spontaneous Pneumothoraces in Systemic Lupus Erythematosus

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A 27-year-old Negro woman, with systemic lupus erythematosus had multiple episodes of spontaneous pneumothoraces. The pulmonary parenchymal cystic changes and rupture of the subpleural cysts which lead to recurrent spontaneous pneumothoraces were found to be secondary to systemic lupus erythematosus.

The pleural and parenchymal pulmonary changes in collagen diseases have been described extensively in the literature.1-6 Recurrent spontaneous pneumothoraces secondary to these changes have been described in histiocytosis X, scleroderma, chronic interstitial fibrosis, tuberous sclerosis, Marfan's syndrome, and a number of other related disorders.6-7 Extensive search of the literature, however, has not revealed a case of spontaneous pneumothorax complicating systemic lupus erythematosus (SLE). In the case reported here, spontaneous pneumothorax followed rupture of a pulmonary cyst in a patient with SLE.

CASE REPORT

A 27-year-old Negro housewife was referred to us by her family physician for treatment of spontaneous pneumothorax. Recently she had become markedly short of breath, and a chest roentgenogram (Fig 1) made at that time showed collapse of the lung and pneumothorax on the left side. She had had pleuritic chest pain and recurrent bilateral pneumonia for six months. Antibiotic therapy in the past did not relieve her symptoms. There was also a history of arthralgia and myalgia, without skin rashes during this period. One year ago the patient had congestive cardiac failure of unknown etiology.

Admission physical examination revealed a moderately wasted, dyspneic and acutely ill Negro woman. Oral temperature was 101°F; the pulse was 100/minute, and the respiratory rate 44/minute. Blood pressure was 150/90 mm Hg. The jugular neck veins were prominent at 45° angle. There was prethial pitting edema. Cardiac auscultation revealed a pro-

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FIGURE 1. Posteroanterior roentgenogram of the chest, showing collapse of the left lung with pneumothorax, enlarged right hilar lymph nodes, and cardiac enlargement. Pleural effusion and pneumonitis can be seen at the right base.

Closed thoracotomy was performed and a no. 24 rubber catheter was introduced in the second anterior left intercostal space under local anesthesia. When the rubber catheter was connected with the suction pump (through the underwateer-seal drainage) the lung expanded promptly. The catheter was removed on the fourth day and the lung remained expanded until the ninth day. Subsequently, the patient had recurrent episodes of spontaneous pneumothoraces on four occasions, on the same side, within a period of three months. Each time, closed thoracotomy was repeated with temporary relief. Finally, open thoracotomy was performed under general anesthesia and abrasion of the pleura was carried out. Small subpleural cysts were ruptured. Since then she did not have recurrence of pneumothorax.

The patient was investigated for collagen disease because of recurrent spontaneous pneumothoraces, history, and in-

FIGURE 2. Posteroanterior roentgenogram of the chest made after removal of the first catheter and immediately following thoracentesis on the right side. Cystic changes are visible at both bases.

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