CASE REPORT

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Sudden Death Due to Sarcoid Heart Disease

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ABSTRACT: A case of sudden death due to massive myocardial sarcoidosis is presented. Cardiac sarcoidosis is discussed. Since the deceased was a New York City police officer with death benefit entitlements under the Heart Bill, the implications of the medicolegal autopsy are emphasized.

KEYWORDS: forensic science, forensic pathology, sarcoidosis, myocardial sarcoidosis, chronic granulomatous disease

Sarcoidosis is a multisystem granulomatous disease of unknown etiology. It can be insidious and go unrecognized until it is discovered at autopsy. Only about 5% of patients with sarcoid have clinical evidence of myocardial involvement (1). Although myocardial involvement is difficult to diagnose, it may lead inexorably to death, and responds poorly to treatment. Myocardial involvement is found in up to one-third of patients with previously diagnosed sarcoidosis at autopsy (2). The case of a police officer who died suddenly from cardiac sarcoidosis follows.

Case Report

A 38-year-old, obese, white New York City police officer collapsed after descending a ski slope while on vacation. He had complained of chest pain shortly before he descended the slope. Despite resuscitative efforts, he was pronounced dead two hours later. A few days before death, he began taking amoxicillin clavulanate (Augmentin) for cold symptoms and an earache. Two weeks earlier, he had complained of chest pain. He had a history of syncope several years earlier, and a diagnosis of ''bradycardia'' was made. He had a remote history of cigarette smoking. A medicolegal autopsy was performed.

Gross Autopsy Findings

The gross examination revealed a diffuse granulomatous process involving the heart, spleen, liver, lung, and lymph nodes. The heart weighed 625 g. The myocardium was homogeneously pale yellow and shiny and massively infiltrated by the granulomatous process. Areas of the epicardium were white and firm. Several areas of the left side of the heart were firm, white, and fibrotic. The cardiac chambers were dilated, especially those of the right side of the heart. The coronary arteries were patent. The granulomatous process also involved the subpleural areas of the lungs. The liver was enlarged and weighed 3130 g. Cut surface of the liver was studded with multiple 1 to 3 mm gray-white lesions with irregular borders. The spleen weighed 710 g and cut surfaces of the spleen revealed innumerable firm, gray-white, 1-cm-wide nodules with a gritty consistency. Generalized lymphadenopathy was present with effacement of the nodal architecture. The brain revealed multiple acute infarcts in the temporal cortex, basal ganglia, and hippocampal gyrus indicative of a global ischemic episode.

Microscopic Examination of Tissues

Microscopic examination of the tissues revealed an extensive non-caseating granulomatous process with numerous giant cells and extensive fibrosis. A battery of histochemical stains, including Ziehl-Neelsen and Fite stains for acid-fast organisms, Brown & Brenn Gram stain for bacteria, as well as silver methenamine and Warthin Starry and Steiner & Steiner silver stains for fungi and spirochetes, respectively, were all negative. No birefringent material was identified on polarization. The post-mortem serum angiotensin-converting enzyme study was 70 mcg/mL (normal 8 to 52 mcg/mL). The post-mortem toxicology results were negative for drugs and alcohol.

Based on the pathological findings, the absence of any microorganisms, and the elevated serum angiotensin-converting enzyme level, a diagnosis of sarcoidosis was made. The cause of death was attributed to acute cardiac failure due to diffuse sarcoidosis with massive cardiac involvement. The manner of death was ruled natural.

Discussion

Sarcoidosis is a multisystem granulomatous disorder of unknown cause that affects approximately 2 in 10,000 adults, usually during the third and fourth decades of life (3). Familial occurrence has been reported, perhaps associated with HLA types B7, B8 and B27 (4). The basic lesion is a noncaseating granuloma composed of epitheliod cells, multinucleate giant cells occasionally containing eosinophilic asteroid bodies, and a scanty rim of lymphocytes. Granulomas may be multiple and discrete or confluent.

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Its clinical course varies according to the site and extent of organ involvement by the disease (3).

Sarcoidosis was initially described by Hutchinson in 1869 (5), but the pathologic pattern of cardiac involvement was not recognized until 1929 (6). Cardiac involvement is reported in 5 to 25% of cases and can occur at any time during its course (5,7–9). The predominant sites of myocardial involvement, in decreasing order of frequency, are: the left ventricular free wall, basal aspect of the ventricular septum, right ventricular free wall, and the atrial walls (5). When the left ventricle is involved, the granulomas are frequently located in the papillary muscles and adjacent free wall which may produce mitral incompetence. The clinical spectrum of disease includes: (1) conduction disturbances due to obliterative lesions of the conduction system, scarring or inflamation of the myocardium, or the development of a ventricular aneurysm; (2) disorders of impulse formation (ventricular arrhythmias account for approximately 55% and ventricular tachycardia is the most common of these); (3) chest pain with occasional electrocardiographic (ECG) changes mimicking transmural myocardial infarction; (4) progressive myocardial failure which occurs in approximately one-third of cases with primary cardiac sarcoidosis; (5) pericardial abnormalities, including effusion, constrictive pericarditis, and tamponade; (6) involvement of the great vessels, coronary arteries, and valvular and subvalvular structures; and (7) sudden cardiac death due to ventricular tachycardia or conduction block which accounts for 30 to 65% of sarcoid deaths (3,5,7,10–14).

Transvenous endomyocardial biopsy, introduced in 1962 and refined in 1972, is diagnostic of cardiac sarcoid when positive. However, it does not exclude the diagnosis when negative because of the focal nature of the lesions (5). The diagnosis of sarcoidosis may be supported by elevated serum levels of angiotensin-converting enzyme (ACE) which is thought to reflect the granulomatous load in the body. ACE is elevated in about 80% of patients with sarcoidosis (15). A determination of the serum level of ACE may be useful in establishing the diagnosis of sarcoidosis although the authors are unaware of any study that has analyzed the level of this enzyme in patients with sarcoidosis limited to the heart. Because of the grave prognosis in patients with cardiac sarcoidosis, it is recommended that patients receive 24-h holter monitoring during the initial workup. It is imperative that any holter or ECG abnormality be investigated further with two-dimensional echocardiography which may reveal abnormalities due to fibrosis and granulomatous reaction. Thallium 201 imaging may also detect sarcoid granulomas which appear as nonspecific segmental areas of decreased thallium 201 uptake (1).

Corticosteroid therapy has been shown to promote replacement of granulomas by fibrous tissue and may improve ventricular function, reduce the risk of sudden death, and prevent further progression of cardiac disease. However, its use has been associated with the development of ventricular aneurysms (5). The dose of prednisone should be at least 80 mg daily and be continued as long as necessary (1). In intractable cases, chloroquine and methotrexate may be prescribed and heart transplantation may be an option for younger patients. Diuretic, inotropic, and antiarrhythmic drugs, as well as pacemaker implantation may also be necessary.

In summary, a 38-year-old policeman died a sudden cardiac death from massive cardiac involvement by sarcoidosis. The disease was clinically quiescent for years until the patient presented suddenly with a fulminating clinical course that was first diagnosed at autopsy. Because the deceased was a member of the police force, his widow sought death benefits from the Heart Bill, which states that ''any condition of impairment of health caused by diseases of the heart, resulting in total or partial disability or death . . . which examination failed to reveal any evidence of such condition, shall be presumptive evidence that it was incurred in the performance and discharge of duty, unless the contrary be proved by competent evidence'' (16). The Heart Bill typically applies to police officers who die from coronary artery disease. In this case, although the heart was found to be massively infiltrated by the sarcoid process at autopsy, the absence of atherosclerotic coronary artery disease led to the denial of death benefits to the policeman's widow.

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