H65 Sudden Unexpected Deaths Due to Sarcoidosis: A Forensic Autopsy Study

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After attending this presentation, attendees will better understand sudden death due to sarcoidosis, particularly the pathological distribution and pattern of sarcoidosis. The clinicopathological correlation of sarcoidosis deaths will also be addressed.

This presentation will impact the forensic science community by providing attendees with a deeper understanding of the role forensic pathologists/medical examiners play in the determination of the cause of sudden death. This presentation will also demonstrate the inter-relationship between forensic pathologists and clinicians in counseling family members about sarcoidosis.

Sarcoidosis is a multisystem disease of uncertain etiology characterized by multifocal areas of discrete and confluent granulomatous inflammation that may be responsible for sudden and unexpected death.[1,2] Sarcoidosis affects young and middle-aged adults without sex predilection.[1] Environmental, occupational, and infectious causes may act as immunologic triggers in genetically predisposed individuals.[2] Sarcoidosis commonly involves the lymph nodes, lungs, cardiovascular system, liver, spleen, central nervous system, and kidney. Involvement can be widespread or limited to involvement of only a single system at a time. The failure to diagnose sarcoidosis clinically is partly attributable to the relative rarity of clinically apparent forms of the disease.[3,4] In a significant proportion of patients with sarcoidosis, the initial presentation is sudden death.[3] There have been few reported autopsy series of patterns of multisystem involvement by sarcoidosis.

A retrospective search of deaths caused by sarcoidosis was performed from the Office of the Chief Medical Examiner, State of Maryland, over a seven-year period from 2005 to 2011. In all cases, medical history, circumstance of death, and autopsy findings including toxicological testing results were reviewed. Distribution of disease was determined both by gross and microscopic examination. Gross evaluation included heart weight and measurements of the ventricular septum and left ventricular free wall thicknesses. Histologic sections were taken in a standardized way and included the Lateral Left Ventricular (LLV) wall, Posterior Left Ventricle (PLV), Anterior Left Ventricle (ALV), right ventricular free wall, interventricular septum, and interatrial septum. Cardiomegaly was defined based on established criteria depending on body weight.4.

A total of 6,442 natural deaths were identified and sarcoidosis was listed as cause of death in 29 cases (0.62%). Of 29 sarcoidosis cases, cause of death was certified as cardiac arrhythmia due to sarcoidosis in 25 cases and pulmonary sarcoidosis in four cases. Of the 25 cardiac sarcoidosis deaths, sarcoid lesions involved left ventricle in 24 cases, followed by interventricular septum (N=20), right ventricle (N=11), mitral and tricuspid valve (N=7), and interatrial septum (N=2). Other organ system involvement included lung in 20 cases, followed by lymph nodes (N=17), liver (N=7), spleen (N=6), and brain (N=1). Of the four pulmonary sarcoidosis cases, two had extensive granulomatous change with diffuse pulmonary fibrosis, one complicated by bronchopneumonia, and one complicated by hypothermia and cocaine use. Other organ system involvement included lymph node (N=4), spleen (N=2), liver (N=1), kidney (N=1), and brain (N=1). Of the 29 cases, 11 patients were witnessed sudden collapse and 18 were unwitnessed. Twenty-one (21/29) cases were clinically undiagnosed. The majority (76%, 19/25) of the cardiac sarcoidosis cases were either overweight (N=4) or obese (N=15), whereas, the body mass index was normal in all four pulmonary sarcoidosis cases.

In conclusion, presented here are characteristics of a series of autopsy cases of sarcoidosis from a state-wide single medical examiner's office with review of literature and clinicopathological correlation.

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Sarcoidosis, Non-Caseating Granulomas, Sudden Death