



G54 Giant Cell Myocarditis as a Cause of Sudden or Unexpected Death: A Report of Two Cases and a Review of the Literature

Steven M. White, PhD, Mitra B. Kalelkar, MD, and James A. Filkins, JD, PhD, Office of the Medical Examiner, County of Cook, 2121 West Harrison Street, Chicago, IL 60612*

After attending this presentation, attendees will have an awareness of giant cell myocarditis as a cause of sudden or unexpected death and its diagnosis.

This presentation will impact the forensic science community by increasing knowledge and awareness of an uncommon cause of sudden or unexpected death with two case presentations.

Giant cell myocarditis (GCM), formerly known as Fiedler's myocarditis, is an inflammatory process of unknown etiology restricted to the heart, typically occurring in young and middle-aged adults. There is no clear gender predilection, but the prevalence of GCM is higher in caucasians than in other races. Because of its isolation to the heart and fulminant clinical course resulting in sudden or unexpected death, GCM is usually diagnosed at autopsy, and therefore may be encountered in a forensic setting. Gross findings at autopsy are variable. Microscopically, there is myocyte necrosis associated with an inflammatory infiltrate composed of histiocytic giant cells, lymphocytes, and scattered eosinophils. The differential diagnoses of GCM include other forms of granulomatous myocarditis, such as sarcoidosis and infectious etiologies. In contrast with sarcoidosis, GCM is typically localized to the heart and has a fulminant clinical course. Infectious etiologies can be excluded with the use of special stains. GCM is a rare cause of sudden or unexpected death with a very low prevalence as reported by other studies.

A search of the records of the Cook County Medical Examiner's Office identified 72 cases in which myocarditis was the principle or contributing factor to death in adults aged 18 and older. The search covered the period from January 1, 2000 through July 15, 2010. Of these 72 cases, only two were cases of giant cell myocarditis. The remaining 70 cases consisted of neutrophilic, lymphocytic, or mixed inflammatory infiltrates.

The first case is a 39-year-old African-American female with a each aforementioned disease entity, including a mixed immune-TTP history of hypertension and obesity, who presented to the emergency

room with shortness of breath and sinus tachycardia. Two days prior to this event, she was seen in the emergency room with fever, malaise, and an elevated white blood cell count, and was discharged. Soon after presentation to the emergency room, she developed pulseless ventricular tachycardia despite pharmacologic therapy. Following synchronized cardioversion and a brief period of asystole, she developed sinus bradycardia. Transcutaneous pacing was attempted, but she progressed to ventricular fibrillation, then asystole. She died within two hours of admission. At autopsy, the heart was enlarged, weighing 487 grams. Grossly, there was concentric left ventricular hypertrophy and the myocardium was uniformly red/brown with the exception of the papillary muscles of the left ventricle, which were pale yellow/gray. Microscopic examination of the heart revealed foci of myocyte loss, fibrosis, and chronic inflammation with scattered giant cells predominantly in the papillary muscle. Other findings at autopsy included cerebral edema, splenomegaly, and chronic passive congestion of the liver. Toxicologic studies were negative for ethanol, opiates, or cocaine.

The second case is a 33-year-old African-American female with no prior medical history, who collapsed suddenly at a nightclub. At autopsy, her heart was enlarged, weighing 426 grams. Grossly, there were geographic areas of pallor from base to apex involving the myocardium of the anterior, lateral, and septal walls of the left ventricle. Microscopically, there was extensive fibrosis and inflammation with numerous giant cells and only small islands of preserved myocardium. Other findings at autopsy included pulmonary congestion and an incidental ovarian teratoma. Toxicologic studies were negative for opiates or cocaine.

These cases are reported to demonstrate the variation in clinical presentation and autopsy findings of GCM, as well as to illustrate that GCM remains a rare cause of sudden or unexpected death even in a busy, urban medical examiner's office.

Myocarditis, Sudden or Unexpected Death, Heart Disease