H130 Is It Proper to Cite Mitral Valve Prolapse (MVP) as the Cause of Sudden Cardiac Death (SCD)?

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After attending this presentation, attendees will be armed with the required information when dealing with skeptical clinicians who do not accept the opinion that SCD due to MVP is a proper cause of death.

This presentation will impact the forensic science community by raising awareness about the appropriateness of the diagnosis of SCD due to MVP. This presentation will also correct the impression among clinical colleagues in whose opinion patients with MVP only succumb to the disease after developing multiple secondary complications of Congestive Heart Failure (CHF). An index case and the response from the decedent's treating cardiologist are described. This response provoked a ten-year review of SCD cases that were determined to be due to MVP.

The deceased was a 43-year-old man with a past medical history of myocarditis, diagnosed approximately a year prior to the terminal event. He was evaluated by a cardiologist who also diagnosed him with mild mitral valve regurgitation and mitral valve thickening. Terminally, he complained of headache, rapidly developed chest pain, and anoxic-type seizures. He was transported to a hospital in asystole and was pronounced dead after failure of resuscitation.

The autopsy revealed a well-nourished and well-developed man with a body mass index of 24.7kg/m^2. The external examination revealed no significant physical trauma and was otherwise essentially unremarkable. Internally, there was concentric biventricular myocardial hypertrophy; the heart weight was 549 grams (average heart weight for body weight 371 grams, with a range between 281 and 489 grams). There was generalized mild (less than 25%) atherosclerotic stenosis of the proximal aspect of all major epicardial arteries. There were no changes of acute, subacute, or remote myocardial infarct or significant myocarditis. The mitral valve revealed marked redundancy and, when viewed from the left atrium, looked like what has been classically described as a deployed parachute. The circumferential measurement of the mitral valve was 13.5cm. The anterior leaflet of the mitral valve measured 4cm in length with a thickness of 0.3cm. The mitral valve annulus revealed focal calcification. The chordae tendinae of the mitral valve were thickened. The other cardiac valves were unremarkable. There was pulmonary edema (combined lung weight 1,095 grams) and changes of passive venous congestion. The remainder of the autopsy was essentially unremarkable. The toxicology screen was negative. The evaluation of the conduction system was non-contributory.

The cause of death of this decedent was listed as sudden cardiac arrhythmia secondary to mitral valve prolapse. This opinion was challenged by the decedent's treating consultant cardiologist, in whose opinion, "No one dies suddenly of MVP."

A ten-year look back of cases certified as related to mitral valve prolapse was performed. Fifteen cases, aged between 25 and 62 years (mean 46 years) which included 8 males and 7 females, predominantly Caucasians (ten), few (four) of African descent, and a rare (one) Asian descent were identified. The decedents had varied clinical histories with five cases having no history of medication use or illicit drug abuse. Among the rest, there was one case each with histories of alcohol abuse, warfarin and loratadine use, cocaine abuse, fluconazole and valaciclovir use, escitalopram and metformin use, acetaminophen/oxycodone use/abuse, and the last decedent had a history of the use of tadalafil. Adequate information was unavailable in two cases. None of the decedents had a prior history of cardiac dysrhythmia.

Anatomically, the mitral valve consists of the mitral valve annulus, the anterior and posterior leaflets, chordae tendinae, and the papillary muscles. Histologically, the mitral valve consists of the fibrosa, spongiosa, and atrialis layers. The spongiosa is made up of connective tissue, proteoglycans, and some elastic fibers, all being most prominent at the free edge of the valve. The spongiosa is the major load-bearing layer of the valve and is where one sees the myxomatous degeneration due to accumulation of glycosaminoglycan (dermatan sulfate). This accumulation secondarily attenuates the fibrosa and atrialis layers leading to the "parachuting" that is classically seen in MVP.

Chesler, et al, found that MVP induced endocardial friction lesions with thrombotic lesions at the angle formed by posterior leaflet of the valve and left atrial wall could lead to fatal cardiac dysrhythmias.[1]

Reference:

3. The myxomatous mitral valve and sudden death. E. Chesler, R. A. King, J. E. Edwards. *Circulation*. 1983 March; 67(3): 632–639

Sudden Cardiac Death, Mitral Valve Prolapse, Myxomatous Degeneration