Mitral valve abnormalities in decedents of sudden cardiac death due to hypertrophic cardiomyopathy and idiopathic left ventricular hypertrophy

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Background: The sole identification of left ventricular hypertrophy (LVH) in a young individual that died suddenly may often lead to an erroneous diagnosis of hypertrophic cardiomyopathy (HCM). Emerging data suggests that idiopathic LVH (ILVH) and HCM may be separate entities.

Aim: We aimed to report on the prevalence and nature of mitral valve (MV) abnormalities, in a cohort of sudden cardiac death (SCD) victims with a post-mortem examination consistent with HCM and ILVH.

Methods: We reviewed 6860 consecutive cases of SCD referred to our specialist cardiac pathology centre between 1994 and 2020. SCD was defined as death from a cardiovascualar cause within 12 hours of apparent well-being. HCM was defined by the presence of LVH, in the absence of abnormal loading conditions and characterised by myocyte disarray at histology. ILVH was defined as unexplained LVH (heart weight >500 g in males and >400 g in females) and left ventricular (LV) wall thickness >15mm, in the absence of myocardial disarray or secondary causes of LVH. The MV was examined for patency, circumference, thickening, nodularity, ballooning, bulging between cords, perforation, and the presence of impact lesions in the LV outflow tract (LVOT) and aortic outlet.

Results: Of the total cases of SCD, 264 (4%) were due to HCM (mean age 41±18 years, 78% males, LV maximal wall thickness 19±6 mm) (Figure 1). Ante-mortem symptoms were reported in 44 (17%) cases and for the majority (n=217, 82%) HCM was established at post-mortem. Death was attributed to ILVH in 253 (3%) cases (mean age 43±16 years, 80% males, LV maximal wall thickness 18±3 mm). MV abnormalities were found in 58 (22%) decedents with HCM (mean age 38±17 years; 72% males) and in 13 (5%) decedents with ILVH (mean age 55±15 years; 77% male), p<0.001. Amongst the 58 (22%) cases with HCM and MV abnormalities, 15 (6%) cases had multiple MV abnormalities. These included impact lesions associated with thickening of the anterior leaflet of the MV (n=39) and degenerative changes such as bulging and ballooning; and thickening and nodularity. Decedents with HCM exhibiting MV abnormalities were younger than decedents with normal MV (38±17 versus 45±19 years; p=0.08). Among the 253 decedents with ILVH, 13 (5%) cases exhibited MV abnormalities, which largely included degenerative changes (n=12). Among decedents with HCM and ILVH exhibiting MV abnormalities, the former was significantly younger (38±17 versus 55±15; p<0.001). Myocardial fibrosis was observed in 162 (61%) cases of HCM and 99 (39%) cases of ILVH, p<0.001.

Conclusions: MV abnormalities are over four-fold more common in individuals with HCM than those with ILVH and may be considered as additional macroscopic features to differentiate between these two entities. Furthermore, the inherent descriptive terminologies used when assessing the MV, support a greater emphasis on the standardisation and quantification of MV abnormalities as part of the autopsy in victims of SCD.

Graphical abstract

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