G115 Sudden Cardiac Death Due to Atrophy and Fibrous and/or Fatty Substitution of Right Ventricle: Pathologic Substrates and Postmortem High Resolution MRI

Massimo Grillo, Department of Biotechnology and Legal Medicine – Section of Legal Medicine, Via del Vespro, n. 129, Palermo, 90127, ITALY; Pierangela Fleres, MD, and Cettina Sortino, Via del Vespro, n. 129, Palermo, , ITALY; Antonino Bonifacio, MD, Institute of Legal Medicine, Via del Vespro, n. 129, Palermo, 90127, ITALY; Livio Milone, PhD, Via del Vespro, n. 129, Palermo, 90127, ITALY; Paolo Procaccianti, PhD, Palermo University, Via del Vespro, n. 129, Palermo, 90100, ITALY; and Emiliano Maresi, PhD, Via del Vespro, n. 129, Palermo, 90127, ITALY*

After attending this presentation, attendees will be introduced to some cases arrived to observation of Section of Legal Medicine of Palermo about sudden cardiac arrhythmogenic death, in order to show the possible disease that can cause death. This presentation emphasizes the difficulty of making diagnosis of Arrhythmogenic right ventricular cardiomyopathy (ARVC) without genetic analysis: myocardial atrophy and fatty/fibro-fatty substitution is diagnostic of ARVC at autopsy only in absence of any other cardiac and extracardiac remarkable injuries.

This presentation will impact the forensic community by demonstrating how data showed highly frequent association between ARVC and the fatty variant with cardiomyopatic pattern. MRI is more sensitive to detect the fatty variant with the caridomyopatic pattern rather than fibro-fatty and/or infiltrative substrate. With this presentation authors will show cases reached to their attention in the last years, characterized by atrophy and fibrous and/or fatty substitution of right ventricle's muscle, with consequent sudden cardiac arrhythmogenic death. Besides, authors will try to make differential diagnosis between different nosologic entities compatible with histological findings, in order to reach the most probable diagnostic hypothesis.

ARVC is a primitive disease of myocardium with not-well-known aetiology, characterized by structural and functional abnormalities of right ventricle, that consist in an hemodynamic, rhythmic and electric disarrayes, due to progressive myocardial atrophy with fatty or fibro- fatty substitution.

Sectorial or widespread atrophy of the myocardium of free wall of right ventricle is the typical morphologic case, with normal characteristics of left ventricle and interventricular sept. Loss of myocardium set the trans-lightning of the wall, that appears to be yellowish for the fatty substitution and/or whithish for the fibrous substitution. Another constant find is the right ventricular dilatation, from mild to severe, with hypokinesia or akinesia of the muscle.

Less frequent are noticed single or multiple right parietal aneurysms, more often at the back side, infundibulous or apex. Besides, frequently there's mural thrombosis of right ventricle and septal fibrosis. This can cause a consequent bilateral atrial dilatation with the high risk of auricular thrombosis with possible systemic and pulmonary embolism.

From 1990 to 2006 474 autopsies of sudden cardiac death (SCD) were performed. In this series, ARVC accounted for 27 cases (16.4%), including 21 males and 6 females (16 to 43 years old; median 26 years). Circumstances of death were during physical exercise in 20 cases, and at rest in seven cases. The family history of ARVC and/or SCD was negative.

The transmural loss of the myocardium in the right ventricular free wall was diffuse in 11 cases and segmental in 16 cases. External bulging of right ventricular free wall and left ventricular was present in four and six cases respectively. The myocardial atrophy was replaced by fatty tissue in 20 cases and by fibro-fatty tissue in seven cases. The interface between residual cardiomyocytes and tissue replacing the myocardium (fatty or fibro-fatty), was wave front (cardiomyopathic pattern) in 21 cases, and lacelike (infiltrative pattern) in six cases. Active myocarditis was detectable in the fibro-fatty variant only. MRI describe the fatty replacement on T1-weighted images with moderate inter-intra observer variability, thus sequende like "fat suppression" or "triple inversion recovery" to evaluate fatty replacement might provide a significant improvement in the diagnosis of ARVC.

The data showed highly frequent association between ARVC and the fatty variant with cardiomyopatic pattern. MRI is more sensitive to detect the fatty variant with the caridomyopathic pattern rather than fibro-fatty and/or infiltrative substrate.

With this presentation cases reached to their attention in the last years, characterized by atrophy and fibrous and/or fatty substitution of right ventricle's muscle, with consequent sudden cardiac arrhythmogenic death will be discussed. Besides, authors will try to make differential diagnosis between different nosologic entities compatible with histological findings, in order to reach the most probable diagnostic hypothesis.

Sudden Cardiac Death, Arrhythmogenic Right Ventricular Cardiomyopathy, Magnetic Resonance Imaging

 ** Presenting Author*