G16 Right Ventricular Lipomatosis and Fibrous Tissue in Cases of Non-Cardiac Deaths and Arrhythmogenic Right Ventricular Cardiomyopathy

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Upon attending this presentation, the audience will have a better appreciation for the fat and fibrous tissue content of the normal right ventricular myocardium, as compared to cases of arrhythmogenic right ventricular cardiomyopathy, and also appreciate the importance of selective sampling of the right ventricle to properly assess fat and fibrous tissue content.

This presentation will impact the forensic community by highlighting the regional differences normally present in the heart, which significantly impact upon the diagnosis of arrhythmogenic right ventricular cardiomy- opathy, particularly in the posterior basal right ventricular myocardium.

Arrhythmogenic right ventricular cardiomyopathy (ARVC), defined as variable replacement of the myocardium of the right ventricular (RV) free wall by fatty or fibrofatty tissue with degenerative changes in entrapped myocytes, is a form of cardiomyopathy that is often familial and typically presents as sudden death in young healthy individuals. Currently, a definitive pathologic diagnosis of ARVC diagnosis is often difficult because a certain amount of fat is always present within the RV myocardium that tends to increase with age, particularly in the anterior and lateral apical regions. We aim to quantitatively establish a normal range for regional RV lipomatosis and fibrous tissue to provide a basis for the pathologic diagnosis of ARVC. Anterior, lateral, and posterior regions of apical and basal RV myocardium were sampled from autopsy cases where deaths were due to non-cardiac causes (control; n=10; age = 21-84 years) and from individuals who had documented ARVC (ARVC; n=10; age = 15-60 years). Area fractions of RV myocardium (%) occupied by myocytes, fat, fibrous tissue, or blood vessels were measured on trichrome-stained slides by computer-assisted point counting, excluding epicardial adipose tissue and subendocardial trabeculations.

The results highlight significant regional differences in lipomatosis nor- mally present in the heart, with apical regions having significantly more lipo- matosis than corresponding basal regions of Control cases ($p<0.05$). The anterior apex showed the most lipomatosis (29.9 ± 4.2%), whereas the pos- terior base had the least lipomatosis (3.8 ± 1.1%). Comparatively, ARVC cases had a significantly greater amount of fat than Control cases ($p<0.05$), which was most apparent between corresponding posterior regions, particu- larly in the basal RV myocardium, which showed a seven-fold increase in lipomatosis (26.4 ± 8.5%; $p<0.05$). The large content of fat normally pres- ent in anterior and lateral apical RV myocardium indicates that diagnosis of ARVC may be difficult if based solely upon lipomatosis in these areas. Significant regional differences in fibrous tissue were not seen in Control hearts, but the amount of fibrous tissue within the posterior base of ARVC hearts was significantly higher than that of Control hearts (20.9 ± 3.7% and 12.2 ± 1.6%, respectively; $p<0.05$). Thus, the substantial amount and regional variation of lipomatosis that exists in normal RV myocardium indicate that changes in lipomatosis in posterior RV myocardium, particularly at the base, are the most reliable means of making a definitive diagnosis of ARVC. This interpretation also has relevance to cardiac imaging as it relates to diagnosis of ARVC.

Arrhythmogenic Right Ventricular Cardiomyopathy, Lipomatosis, Right Ventricle

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