H105 A Particular Case of Adipositas Cordis in a Girl Suffering From Lymphocytic Myocarditis and Hashimoto's Thyroiditis

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Learning Overview: After attending this presentation, attendees will have learned about a very unusual case of Adipositas Cordis (AC) in a young 14-year-old girl suffering from myocarditis and Hashimoto thyroiditis.

Impact on the Forensic Science Community: This presentation will impact the forensic science community by detailing how the forensic investigations were conducted for the diagnosis of AC in this rare case.

AC is a rare and poorly understood heart disease. This is proved by the fact that there are no guidelines on the subject. AC is characterized by structurally normal myocytes and diffuse adipose cell infiltration in the myocardium of both ventricles, especially in the right. In some cases, it could be a diagnostic challenge as it can closely mimic Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC). Preliminarily, there is no standard method for diagnosing AC before sudden death.[1] Secondly, in the scientific community, it is not known if AC is the first stage of ARVC. In fact, they could be two completely different diseases.

The primary differentiating characteristics between AC and ARVC are myocyte degeneration and interstitial fibrosis that are present in ARVC and responsible for the higher risk of Sudden Cardiac Death (SCD).[2] A case of AC is evidenced by prominent fatty infiltration associated with lymphocytic myocarditis and Hashimoto's thyroiditis is presented.

The young girl was apparently healthy and had a normal life, regularly playing sports. She had no symptoms. The girl was found dead at home with no apparent cause. During the autopsy, the organs were removed and fixed in formalin. Small portions of ventricular myocardium and 300ml of peripheral blood were frozen at -80° for further investigation. Formalin-fixed samples were stained with hematoxylin-eosin. Screening for drugs was carried out on blood and urine, after extraction with trichloroacetic acid and acetonitrile with immunoenzymatic method, gave negative results. The search for genetic material for the most common viral agents of myocarditis, with the Polymerase Chain Reaction (PCR) technique, on the heart and blood was also negative. Since myocarditis is associated with ARVC in some cases, the genome for known mutations for ARVC was examined with negative results.[3]

Histological investigation showed acute lymphocytic myocarditis on the anterior wall of the left ventricle, most likely on an autoimmune basis, and chronic lymphocytic thyroiditis. Adipose infiltration involved a large part of the right ventricle and extended for two-thirds of the wall. In some regions, the AC extended to the endocardium. There were no fibrous replacements. Myocytes did not show ARVC-typical changes.

With the results of the investigation, the SCD was attributed to AC. It is unlikely that the arrhythmic event was triggered by myocarditis, because it was too small and unique, and the central necrosis was not very extensive. Most likely in AC, the conduction of the electrical impulse is compromised by the dissociation of cardiomyocytes.

It is now known that AC is frequently associated with acute coronary syndrome. There have been a number of sudden AC deaths in recent years. The scientific community around the world must pay attention to AC, especially as affected patients have no symptoms and no related Electrocardiogram (ECG) abnormalities.[4,5] Unfortunately, AC can only be confirmed after autopsy; the real cause of AC remains unknown.[6,7] AC needs to be further explored, and the diagnostic criteria must be determined. Until the cause of AC is discovered, forensic pathologists should look for known ARVC mutations in these cases. In fact, many authors consider AC to be an early form of ARVC.

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