H93 A Rare Case of Sudden Cardiac Death Associated With Isolated Congenital Coronary Artery Anomalies (CAA): Autopic and Histopathological Results

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Learning Overview: The goal of this presentation is to demonstrate the importance of an early diagnosis of anomalies of origin of the coronary arteries.

Impact on the Forensic Science Community: This presentation will impact the forensic science community by illustrating a case of sudden death due to CAA in a young girl and the analysis of the autoptic and histopathological results.

CAAs are the second cause of Acute Coronary Syndrome (ACS) under 35 years of age, after hypertrophic cardiomyopathy.[1-8] ACS usually occurs during or shortly after strenuous exercise. In most cases, people with CAA are asymptomatic. The anomaly is recognized only during the autopsy. The prevalence of CAA in the population is between 1%–2%.[9] CAAs are divided into: anomalies of origin, course anomalies, and termination anomalies.[10]

A case of a young student who complained of dyspnea after physical exertion for several years is presented. The girl was found dead in bed. At autopsy, examination of the coronary artery circulation showed that the Right Coronary Artery (RCA) ostium was born at the union between the right and left cusp. The right ostium also had a higher origin, specifically, the ostium was 0.4cm higher than the left ostium. The anomalous ostium had a "flute beak" shape. The first section of the RCA passed between the pulmonary trunk wall and the aorta. The inter-aorto-pulmonary extension was 0.4cm. The coronary artery circulation was left dominant. The left coronary artery showed no anomalies. The atria and ventricles were normal. The mitral valve showed myxoid degeneration with a "parachute" appearance. The other valves were normal. The histology of the heart showed wavy myocardial fibers; interstitial and perivascular sclerosis; dilation of small intramyocardial and subepicardial vessels.

The cause of death was an arrhythmia secondary to an anomaly of the right coronary artery origin. The abnormal origin of the right coronary artery from the left Valsalva sinus is documented in 0.03%–0.17% of patients who undergo traditional angiography. This anomaly is associated with ACS because the first section of the coronary artery has an inter-arterial course. In fact, the first section is compressed during systolic expansion causing ischemia in the downstream areas. This compression typically occurs during physical exercise. On the other hand, if the course is retro-aortic, pre-pulmonary, or septal, there would be no risk of ACS because the walls are not compressed. When a young patient experiences symptoms during exercise, evaluating the coronary circulation is recommended. In young people, Computed Tomography (CT) or Magnetic Resonance Imaging (MRI) angiography as an initial screening is recommended. In this way, candidates can be selected for life-saving surgery.

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