H37 A Sudden Unexpected Infant Death (SUID) Associated With Coronary Arterial Fibromuscular Dysplasia

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After attending this presentation, attendees will appreciate some causes of sudden cardiac death in infancy, have an understanding of the pathophysiology, incidence and epidemiology of fibromuscular dysplasia, and be able to identify coronary artery abnormalities (or coronary artery hyperplasia) grossly and microscopically.

This presentation will impact the forensic science community by illustrating the aspects of a thorough and complete postmortem cardiac examination, stressing the importance of including sections of the coronary arteries for histologic examination in cases of SUID.

SUID is a traumatic burden for families and a significant diagnostic challenge for forensic pathologists. There is increasing evidence that certain cardiac abnormalities presenting as SUID may be underemphasized and may have implications for surviving family members.

Reported here is the case of a 4-month-old male infant who was laid down by his mother on the date of death and was found unresponsive approximately ten minutes later. Resuscitative efforts were unsuccessful. The infant had a history of prematurity, born at 29 weeks, 4 days, and weighing 1, 600 grams. At that time, he was hospitalized in the neonatal intensive care unit for 52 days with respiratory distress syndrome, apnea of prematurity, a transient flow murmur which resolved, and retinopathy of prematurity. His prenatal history was significant for maternal smoking (one-half pack per day) and a positive maternal drug screen for amphetamines and marijuana. According to his mother, the infant had upper respiratory symptoms prior to death.

Autopsy revealed a well-nourished, 7,400-gram infant measuring 59.5cm from crown to heel. The major autopsy finding was limited to the heart, which exhibited a 1.2cm x 0.8cm area of epicardial fibrosis on the left ventricular apex with scattered petechiae on the anterior visceral epicardium. Grossly, the epicardial coronary arteries were each diffusely thickened and fibrotic, although patent. Histologically, all of the coronary arteries were diffusely, mildly stenosed by a concentric musculoelastic intimal thickening. The subendothelial connective tissue contained an infiltrate of spindled mesenchymal cells set in an abundant fibrocollagenous tissue matrix. The proliferating cells showed smooth muscle actin and vimentin positivity and desmin negativity, confirming myofibroblastic origin. By elastic/trichrome stain, the internal elastic membrane and elastic fibers of the media were fragmented, especially at sites of greater intimal proliferation. An Alcian blue/Periodic acid Schiff stain revealed increased intimal mucinous ground substance. There was no evidence of inflammation, foam cells, cholesterol clefts, or other evidence of atheroma. Other anatomic regions of the heart, including the cardiac valves and the atrioventricular nodal region, were free of abnormalities. Other vessels, including the aorta, internal carotid arteries, and renal arteries, showed no vascular stenosis, aneurysmal changes or dissections. Cultures of the lungs, cerebrospinal fluid, and blood showed postmortem overgrowth, and a nasopharyngeal swab viral panel was negative. Postmortem toxicological analysis was negative. No other natural disease processes or injuries were identified that caused or contributed to the infant's death. Based on the lack of clinically significant coronary artery stenosis or chronic or acute ischemic cardiac lesions, the cause of death was listed as SUID, and the coronary arterial fibromuscular dysplasia was listed as a contributing condition.

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A non-inflammatory, non-atherosclerotic disorder, fibromuscular dysplasia is characterized by abnormal fibroblastic proliferation in the walls of small- to medium-sized arteries leading to stenosis, aneurysm, dissection, or occlusion. The etiology is unknown but is believed to be related to hormonal, environmental/mechanical, and genetic factors. It is familial in 7%-11% of cases and commonly affects the renal (60%-80% of cases), extra-cranial cerebrovascular (25%-30% of cases), and vertebral arteries. Isolated involvement of the coronary arteries is rare and can present with unstable angina, myocardial infarction, left ventricular dysfunction, or even sudden cardiac death.

Sudden cardiac death in infancy can be attributed to cardiomyopathies, channelopathies, congenital cardiac anomalies such as valvular abnormalities, endocardial fibrosis, and coronary artery disease. Anatomic findings of these entities can be subtle or sometimes obscured by prolonged survival time leading to secondary myocardial ischemia; however, a thorough cardiac examination can elucidate certain structural pathologies. This case highlights the importance of a thorough cardiac examination including microscopic sections of the coronary arteries in cases of SUID.

Sudden Unexpected Infant Death, Coronary Artery, Fibromuscular Dysplasia

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