G11 Sudden Death From Arteritis Involving a Surgically Repaired Coronary Artery - Right Atrium Fistula

Marc A. Krouse, MD, Tarrant County Medical Examiner, 200 Feliks Gwozdz Place, Fort Worth, TX 76104-4919*

After attending this presentation, attendees will be informed of the possibility of late complication of a repaired congenital coronary artery anomaly by an independent pathologic process.

This presentation will impact the forensic science community by revealing an unusual complication of surgically treated congenital cardiac malformation, specifically coronary artery - right atrium fistula.

After attending this presentation attendees will appreciate that an anomalous coronary artery (coronary artery – right atrial fistula), successfully repaired many years prior, may be involved by independently occurring disease processes such as pan-arteritis and may prove a cause of morbidity or mortality despite successful earlier treatment.

The subject of this presentation was an 11-year-old male who had a diagnosis of coronary artery – right atrium fistula some eight years prior. The anomalous vessel was ligated at the distal (right atrium) end and he was followed, without complication, for a period of some two years. He was well and active until the day prior to his collapse and demise with no complaints that could be related to cardiac disease. In early morning hours his family members responded to sounds of distress, he collapsed and began dry vomiting before becoming unresponsive. Resuscitation efforts, including ACLS protocol and emergency department treatment, were unsuccessful and he was declared dead less than two hours after onset. His history of previous surgery was initially reported (incorrectly) as repair of an abnormal right coronary artery.

At necropsy examination the body was normally developed. There were diffuse pericardial adhesions over the anterior and left side of the heart. Serial sectioning of the left coronary artery circulation revealed a slightly large (4-5 mm) left main coronary artery with a similar size anomalous branch passing posterior to the aortic root between the atria. In this area the vessel was markedly dilated (up to 2 cm) and filled with layered, clotted blood. The firmer clot had propagated retrograde and gelatinous, acute clot was found throughout the proximal part of the anomalous artery, into and occluding the left main coronary artery. The left coronary artery ostium was also large, some 1 cm. Microscopic sections of the coronary arteries and coronary artery fistula were notable for active pan-arteritis and healed arteritis in the dilated area of the fistula as well as layered blood clot without notable organization. There was no gross or microscopic evidence of ischemic myocardial injury.

The gross appearance of the artery fistula was reminiscent of Kawasaki disease and the pan-arteritis points to a similar pathogenesis of the vascular injury, aneurismal dilation and eventual thrombosis of the injured vessel. His recent medical history included only an episode of acute sinusitis with a four day course of an unknown prescribed medication, but in interviews with family a previous episode of a viral illness some four months prior was elicited.

This case study is presented to inform forensic and/or pediatric pathologists of the possibility of a late complication of a successfully repaired anomalous coronary artery, presumably by an immune- mediated vascular injury indistinguishable from typical Kawasaki disease.

Coronary Artery Fistula, Arteritis, Sudden Death

 ** Presenting Author*