G45 Sudden Death Due to Segmental Arterial Mediolysis Involving the Right Coronary Artery

Harold Sanchez, MD, Department of Pathology, Hospital of Central Connecticut, New Britain, CT 06050; and Rebecca Irvine, MD, Dept of Forensic Medicine, 50 Parramatta Road, Glebe, NSW 2037, AUSTRALIA*

After attending this presentation, attendees will learn to recognize segmental arterial mediolysis as a potential cause of sudden cardiac death and to distinguish it from other causes of coronary artery dissection.

This presentation will impact the forensic scientific community by focusing attention on nonatherosclerotic causes of lethal coronary artery disease in general and on one potentially overlooked cause, segmental arterial mediolysis, specifically.

A 65-year-old woman with a past medical history significant for hypercholesterolemia and hypertension presented approximately three months prior to death with a chief complaint of rib pain. Radiographic and laboratory studies and a bone marrow biopsy were consistent with multiple myeloma. She was treated with bortezimib, dexamethasone, zoledronic acid, and lenalidomide and tolerated it well. On the day of her death, she complained of back pain, and late that night she suddenly became pale and unresponsive. Emergency medical personnel found her in ventricular fibrillation, initiated resuscitative efforts, and transported her to the emergency department. Physical examination revealed a pulseless, unresponsive, afebrile, slightly overweight (BMI of 26) woman with an unremarkable body habitus. Laboratory results were significant for metabolic acidosis and a troponin I value that was elevated but below the range considered consistent with acute myocardial infarction. The clinical cause of death was given as hypoxia due to metabolic acidosis due to multiple myeloma.

Autopsy revealed cardiomegaly and left ventricular hypertrophy. The left-sided coronary arteries showed minimal to mild coronary artery disease. The posterior interventricular branch of the right coronary artery appeared thrombosed, and the gross impression was of probable coronary artery disease with superimposed thrombosis. Histology revealed instead a right coronary artery dissection with medial hemorrhage and luminal occlusion. The surrounding myocardium showed histologic evidence of recent infarction. Changes similar to those seen in the right coronary artery were incidentally found in a medium-sized branch of the splenic artery. Neither of the affected vessels showed atherosclerotic changes, vasculitis, or giant cells. Both showed disruption of the elastic lamina highlighted by elastic stains. An incidental small pulmonary embolus was found in the right middle lobe. The aorta and its great branches showed trace atherosclerotic changes. No histologic evidence of myeloma was found in the marrow.

As a group, nonatherosclerotic coronary artery lesions are a rare cause of sudden death. The list includes congenital coronary artery anomalies (e.g., hypoplasia and anomalous origin), coronary arteritis (e.g., polyarteritis nodosa, giant cell arteritis, Kawasaki disease, and luetic arteritis), coronary artery spasm, and mechanical obstruction resulting from coronary artery dissection. Spontaneous Coronary Artery Dissection (SCAD) is most often seen in women younger than 45, often in the peripartum period, and typically in the left-sided coronary arteries. Dissection has also been associated with arteritis, connective tissue disorders such as Marfan syndrome and Ehlers-Danlos syndrome, cocaine use, and strenuous exercise. The findings in the present case are consistent with Segmental Arterial Mediolysis (SAM), another rare and potentially under-recognized cause of coronary artery dissection and sudden cardiac death.

First described in 1976, SAM is characterized by destruction of the media of small- to medium-sized arteries with loss of the elastic lamina and separation of the media from the adventitia. There is no evidence of significant inflammation or atherosclerotic change. The resulting structural compromise can lead to aneurysm formation, dissection, or rupture of involved vessels with infarction of supplied tissue. While SAM most often involves abdominal arteries and presents as abdominal pain and ischemia, cases have been reported involving arteries throughout the body, including coronary arteries. Compared to SCAD, SAM is typically seen in older patients. There can be considerable clinical and radiographic overlap between SAM and other nonatherosclerotic causes of coronary artery dissection. Histology remains the gold standard for definitively distinguishing between these conditions, a distinction which can have important therapeutic implications.

Sudden Cardiac Death, Coronary Artery Dissection, Segmental Arterial Mediolysis

 ** Presenting Author*