



Pathology Biology Section – 2007

G113 A Chest Full of Blood: Hemothorax as a 26 Year Delayed Consequence of Repair of Transposition of the Great Vessels

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After attending this presentation, attendees will learn about an uncommon consequence they may see long after surgical correction of transposition of the great vessels.

This presentation will impact the forensic community and/or humanity by providing useful information about a potentially lethal delayed complication of a common surgical procedure to correct a congenital heart problem; describing newer procedures that do not have this kind of problem; and illustrating signs and symptoms to look for in a similar autopsy case.

A 27-year-old woman had been born with transposition of the great vessels (TGV), a congenital heart disorder that was always fatal before corrective surgery was developed. In TGV, blood oxygenated by the lungs never reaches the rest of the body, and blood returning from the body never reaches the lungs. This occurs because, in TGV, the pulmonary artery is attached to the left side of the heart and the aorta is attached to the right side of the heart.

Twenty seven years ago, a standard treatment for this disorder was the Mustard procedure, in which the pulmonary veins are detached from the left atrium and surgically reattached to the right atrium, and the superior and inferior vena cava are detached from the right atrium and reattached across an "atrial baffle" to the left atrium. This procedure allows oxygenated blood to flow to the body, and deoxygenated blood from the body to return to the lungs. It also allows the coronary arteries to remain in their native position in the aorta. It has the disadvantage that the right ventricle is responsible for pumping blood to the body, which causes it to hypertrophy, and the left ventricle pumps blood to the lungs, under too much pressure.

The patient's Mustard procedure was performed at 11 months of life. She recovered well and lived for twenty-six more years. Approximately three months prior to her death, she was diagnosed with pneumonia, and was treated with antibiotics. The day before her death, she complained of chest pain. The night of her death, while talking on the phone, she had a fit of coughing. The expectorant contained blood. She went to the emergency room, and during the initial work-up, collapsed and died.

At autopsy, external examination revealed a slender, gracile and nearly cachectic female who had pallor of the face and body. There was a midline chest scar as evidence of her previous cardiac surgery, but no recent injuries or surgical procedures. Blood for toxicology was hard to obtain by inguinal or subclavian puncture.

Internal examination revealed a right hemothorax with 1700 cc of blood and clots, visceral pallor, and depletion of the vascular tree. The heart showed evidence of the Mustard procedure, with all anastomoses intact. However, there were bilateral pulmonary artery aneurysms. The right ventricle showed evidence of extreme hypertrophy, with the wall measuring up to 1.5 cm thick. The right pulmonary artery showed a 10 cm aneurysm at the hilum that extended into the lung. The left pulmonary artery revealed a 3cm aneurysm. A fresh rupture was found in the right pulmonary aneurysm near the hilum, creating the hemothorax and causing the young woman's death.

Extensive recent and older thrombosis of pulmonary artery branches leading from the aneurysm into the right lower lobe was noted, suggesting this might have prompted the diagnosis of pneumonia. Thrombosis of these vessels might have been responsible for raising the pressure in the aneurysm, ultimately leading to rupture.

The Mustard procedure has been known to extend victims' lives for ten years or more. However, pulmonary artery aneurysms, although rare, have been reported in the literature as a long term sequela. Commoner problems with the Mustard procedure include cardiac arrhythmias and baffle leaks. Transposition of the great vessels is now treated by a different procedure, the arterial switch.

This case report will compare the long term complications and advantages of the arterial switch to the Mustard procedure in light of this catastrophic delayed result. This will aid forensic personnel in the future when they have a case for forensic autopsy with a history of surgical repair of transposition of the great vessels. They will know what to expect on heart dissection from these surgeries, and to anticipate the possible long term problems that can accompany these types of techniques.

Transposition of Great Vessels, Aneurysm, Sudden Death