H123 Severe Pulmonary Artery Hypertension Persisting After a Late Atrial Septal Defect (ASD) Closure: Presentation of a Case

Serenella Serinelli, MD, State University of New York Upstate, Department of Pathology, Syracuse, NY 13210; Lorenzo Gitto, MD*, State University of New York Upstate, Department of Pathology, Syracuse, NY 13210*

Learning Overview: After attending this presentation, attendees will understand the pathophysiology and the autopsy findings of pulmonary artery hypertension after a late ASD closure.

Impact on the Forensic Science Community: This presentation will impact the forensic science community by demonstrating that pulmonary hypertension can persist after a late surgical repair of ASD.

ASD is the most prevalent congenital cardiac anomaly in adults, representing ~35% of all congenital heart defects. It consists of a heart defect in which blood flows between the atria of the heart: oxygen-rich blood can flow directly from the left side of the heart to mix with the oxygen-poor blood in the right side of the heart, or vice versa. This can lead to lower-than-normal oxygen levels in the arterial blood that supplies organs and tissues. Due to the high blood flow from left to right, an ASD can also cause right ventricular remodeling and failure, with enlargement of right cardiac chambers, severe arrhythmias, and pulmonary artery hypertension. The left ventricle can be smaller than the right one and a reduced coronary perfusion can occur.

Most adults with ASD are safely treated using available techniques of surgical or percutaneous repair and become free of complications over the long term. However, there is a small fraction of patients who are at risk for post-operative complications, particularly the persistence of elevated pulmonary vascular resistance and pulmonary artery hypertension.

This presentation describes the case of a 47-year-old Asian female (height: 152cm; weight: 36kg) who was found unresponsive by her husband in the bathroom of their house. The husband promptly called the emergency medical services. A cardiopulmonary resuscitation was performed with negative results and the woman was pronounced dead.

According to the medical records, the woman underwent a surgical repair of an atrial septal defect at the age of 37 and suffered from severe pulmonary hypertension, high blood pressure, paroxysmal atrial fibrillation, and multivalvular insufficiency. A few months prior to death, she had an episode of acute heart failure and underwent a chest-abdomen Computed Tomography (CT) and an echocardiogram that showed: persistence of a left superior vena cava probably draining in the coronary sinus, enlargement of the right heart with massive tricuspid regurgitation, enlargement of the pulmonary arteries with severe pulmonary regurgitation, bilateral pleural effusion, atelectasis of the left lung, chronic passive congestion of the liver, and ascites.

At the autopsy, the heart weighted 650 grams and showed: an enlargement of the coronary sinus, a correctly positioned interatrial patch, a notable right ventricular hypertrophy with a small left ventricle, enlarged pulmonary arteries with fatty streaks, and pulmonary and tricuspid valve insufficiency. It was not possible to identify the left superior vena cava detected during the imaging studies; this could be due to the relatively small size of the vessel or to the presence of massive adhesions from prior surgery.

At the microscopic examination, the heart showed multiple myocardiocites with enlarged nuclei and multiple areas of fibrosis. The lung showed hypertrophy of the media of the arterioles and plexiform lesions, findings consistent with a severe pulmonary artery hypertension.

In this case, since the woman did not have the defect closed until such an advanced age (37 years old), probably all the autoptic findings could be attributed to the ASD. The left superior vena cava persistence may have contributed. A prolonged flow from the left atrium across the ASD to the right atrium caused the right ventricular hypertrophy and pulmonary hypertension. Moreover, the left ventricle was reduced in size and a myocardial fibrosis (presumably due to low coronary perfusion) was observed. It was not possible to understand if, in this case, the delay in surgical treatment was due to a late presentation of the defect or due to different reasons. In any case, the late surgical treatment allowed the subject to become free from the interatrial communication, but not from the advanced right ventricular remodeling and pulmonary vasculopathy, which caused death.

Atrial Septal Defect, Pulmonary Hypertension, Heart Remodeling