The pericardium is a serous sac that surrounds and helps to maintain the heart within the thoracic cavity. It is composed of two layers: the outer parietal layer and the inner visceral layer. The pericardium serves several functions, including protection, absorption of shock, and a place for extracellular fluids. Pericardial effusions can be caused by various conditions, such as infection, trauma, or malignancy, and can lead to symptoms such as dyspnea, chest pain, and cardiac tamponade.

Case

An 80-year-old female with a history of diastolic heart failure, asthma and type II diabetes mellitus presented with shortness of breath and a systolic ejection murmur. She was subsequently found to be in septic shock secondary to a urinary tract infection, as well as new onset atrial fibrillation.

HR: 76
BP: 87/55
SpO2: 97% on 2L NC

Admission ECG revealed new onset atrial fibrillation with controlled ventricular rate, and no evidence of ischemia.

2D Echocardiography, Figures 1 and 2, noted an atypical exam with apparent absence of the pericardial sac and significantly deviated viewing windows.

The parasternal long axis view was significant for a prominent right ventricle with marked right ventricular dilatation, and a posteriorly displaced left ventricle. The apical 4 chamber window was severely displaced both posteriorly and laterally, and was obtained posterior to the mid-axillary line.

2D Echo noted an ejection fraction of 50-55%, severe right ventricle dilatation, moderate to severe tricuspid regurgitation, and estimated right ventricular systolic pressures of 40-50 mmHg, suggestive of moderate pulmonary hypertension.

Computed Topography of the Chest, Figures 3 and 4, showed bilateral pleural effusions with marked enlargement of the cardiac silhouette, aumorous appearance of the left ventricle, and severely enlarged main pulmonary arteries.

Errors in the embryologic development of the pericardium can result in atrophy/agenesis of either the entire pericardium, or solely the right or left pericardial sac. The majority of patients with congenital absence of the pericardium (CAP) are asymptomatic, therefore diagnosis is often made as an incidental finding.

Cardiac Computed Topography (CTT) and Cardiac Magnetic Resonance Imaging (CMR) provide better visualization of the thin pericardial membrane than surface echocardiography and can aid in diagnosis. Accurately identifying and describing the defect has both important diagnostic and prognostic implications, and will help guide long term management. CTT and CMR findings suggestive of pericardial agenesis include exaggerated levoposition of the heart, and interposition of lung tissue in the anterior space between aorta and pulmonary artery, or between the diaphragm and the base of the heart. In our case, the patient was found to have elevated right ventricle systolic pressure, as well as a severely enlarged main pulmonary artery suggestive of moderate pulmonary hypertension. While these findings only suggest the presence of pulmonary hypertension, it was suspected in our case that the significant deviation of the heart within the hemithorax resulted in compression atelectasis of the left lower lobe of the lung, physiological pulmonary shunting, and increased pulmonary artery pressures. While enlargement of the main pulmonary arteries and tricuspid regurgitation are described in association with congenitally absent pericardium, pulmonary hypertension is not explicitly described in the literature as a consequence of CAP.

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