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Pictorial Essay

Congenital absence of the pericardium and its mimics

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ABSTRACT

Congenital absence of the pericardium is a rare entity, with less than 400 cases reported in the literature. Pericardial absence is typically left sided, which results in herniation of the great vessels or portions of the heart. Patients may be asymptomatic, typical for complete defects, or can present with various degrees of chest pain in the setting of partial absence and strangulation. The finding may be isolated or associated with complex heart disease. We present a number of cases of pericardial absence that show isolated right- and left-sided defects, as well as entities in the differential diagnosis. Early recognition may decrease morbidity and mortality, resulting from delay in proper treatment.

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1. Introduction

Congenital absence of the pericardium is a rare condition that is most often incidentally discovered at cardiothoracic imaging. Dramatic presentations, such as atypical nonexertional chest pain or cardiac strangulation that requires prompt surgical management, are fortunately uncommon. The entity can be diagnosed by chest radiography, computed tomography (CT), magnetic resonance imaging (MRI), and ultrasound scanning, underscoring the importance of recognizing the imaging findings.

1.1. *Embryogenesis*

The pericardium is formed by an outer fibrous layer and an inner serous layer of mesothelial cells. During embryogenesis, the heart grows into and invaginates the inner serous pericardium, resulting in visceral and parietal layers that are in continuity. The parietal layer lines the outer fibrous layer, whereas the visceral layer lines the epicardial surface of the heart. A fluid-filled potential space between the visceral and parietal layers creates the pericardial cavity. The visceral layer can be up to 1 mm thick, thickening slightly over the right

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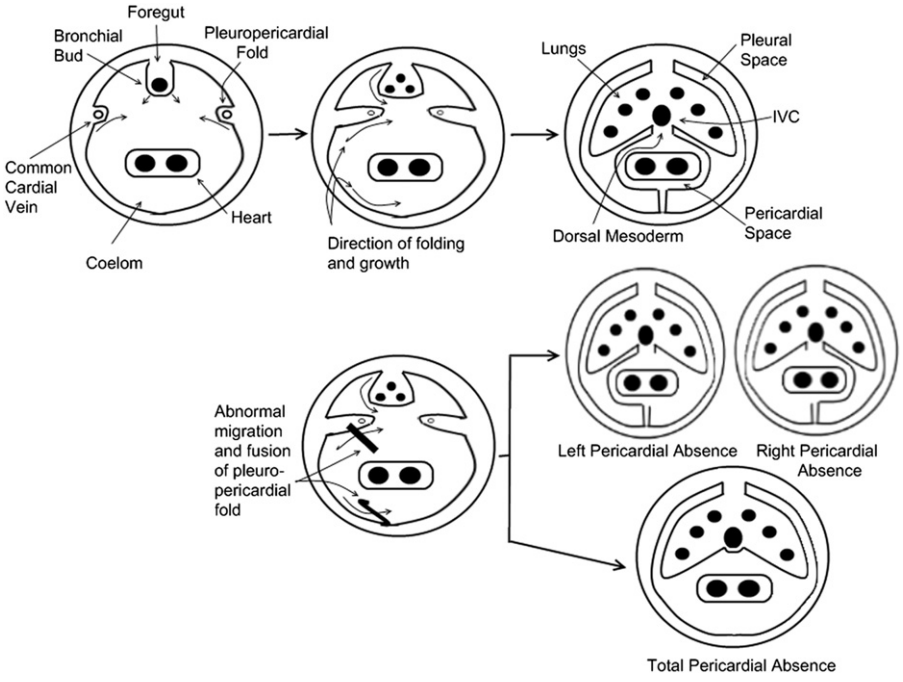

Figure 1 – Simplified depiction of the embryology of the pericardium and formation of defects. At the beginning of 5 weeks, 2 pleuropericardial folds develop from the lateral body wall mesoderm. Folds grow and extend toward the midline, which then fuse with the dorsal mesoderm. In addition, primary bronchial buds form from the foregut to develop into the lungs. Folding and fusion of the folds partition the coelom into the pleural and pericardial spaces. Failure of fusion from one or both folds, leads to pericardial defects.

ventricle and thinning over the left ventricle. It encloses the heart, proximal ascending aorta, pulmonary arterial trunk, and portions of left pulmonary veins. It serves to fix the heart in position, reduce friction by pericardial fluid, and isolate the heart from adjacent pathologic processes.[1]

Embryologically, the pericardial sac is formed from the embryonic coelom. Within the coelom, the pericardial sac is separated from the peritoneal cavity by the transverse septum and from the pleural by the right and left pleuropericardial folds. Pericardial absence is thought to be the result of malformation of the transverse septum or pleuropericardial folds, possibly because of a compromise of vascular supply (Fig. 1).[2]

1.2. *Prevalence*

Pericardial absence was first suggested by Italian anatomist and surgeon, M. Realdus Columbus, in 1559[3] with a definitive example in 1793 by Baille.[4] Ellis et al[2] in 1959 detailed 99 cases, which substantially improved awareness of this rare anomaly with a reported prevalence of 0.002% to 0.004%.[5] Up to 400 cases have been recorded in the literature to date.[6]

1.3. *Variations in pericardial absence*

Ellis et al[2] identified 6 categories of pericardial defects: right-sided (partial or complete), left-sided (partial or complete), total absence, and diaphragmatic defects. Thirty percent to 50% of defects are associated with other cardiopulmonary anomalies such as patent ductus arteriosum, Tetralogy of Fallot, atrial septal defect, sequestration, and bronchogenic cysts.[6,7]

1.3.1. *Absence of the left pericardium*

Left-sided defects are the most commonly reported, involving up to 70%–80% of cases of pericardial defects.[8,9] Complete left pericardial absence is 3:1 times more common than partial left-sided absence.[8] The congenital cause is thought to be premature atrophy of the left common cardinal vein (duct of Cuvier) which supplies the left pleuropericardial membrane, resulting in pericardial or diaphragmatic defects (Fig. 1). Patients with complete absence of the left pericardium are typically asymptomatic. Partial absence can cause no symptoms or can result in chest pain, dyspnea, and syncope related to herniation and strangulation of cardiac structures through the defect. Rarely, fatal incarceration of the left ventricle can occur.[6] Herniation of the left atrial appendage has been frequently reported.[10] Torsion of great vessels, coronary compression on the pericardial rim, and ventricular herniation are other potential complications.[7] Distortion of ventricular geometry will elongate the chordae of the anterior leaflet of tricuspid, contributing to regurgitation, a common finding in large partial or total left-sided absence.[7] Electrocardiogram findings usually show right-axis deviation, right bundle branch block, and poor R wave progression.[11]

CT or MRI is the mainstay for diagnosis, which usually can show the pericardial defect with cardiac herniation. Cardiac MRI may miss absence in up to 10% of patients because of