Marked levoposition of the heart – a clue to diagnosis of complete pericardial agenesis

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Introduction: Congenital pericardial agenesis (CPA) is a rare cardiac condition with an incidence of < 1 in 10000. It can be found in isolation or associated with other congenital cardiac abnormalities. The most common form is complete left-sided CPA and patients are usually asymptomatic. On the other hand, patients with partial CPA may be more symptomatic and at higher risk for complications due to cardiac herniation.[1,2]

Case report: A 54-year-old lady with history of hypertension was referred to our outpatient clinic because of dyspnea on exertion and palpitations. Physical examination revealed BP of 180/85mmHg and mild systolic murmur. An EKG showed normal sinus rhythm, with a heart rate of 65 bpm, vertical heart axis, Q waves from V1-V3 and slow R wave progression in precordial leads. A 24-hour Holter EKG and a treadmill exercise stress test were unremarkable. During the echocardiography study a marked leftward and posterior displacement of the apical 4-chamber window was noted. Impression of cardiac hypermobility, left ventricular bulbous appearance and elongated, compressed atria were observed. Parasternal LAX window was also shifted more laterally and from this view right ventricle appeared dilated. All these findings raised the suspicion of complete CAP. The CMR confirmed the diagnosis by showing typical indirect signs of complete left-sided CAP. Pericardial continuity was only seen on the right side, close to atria (**Figure 1**).

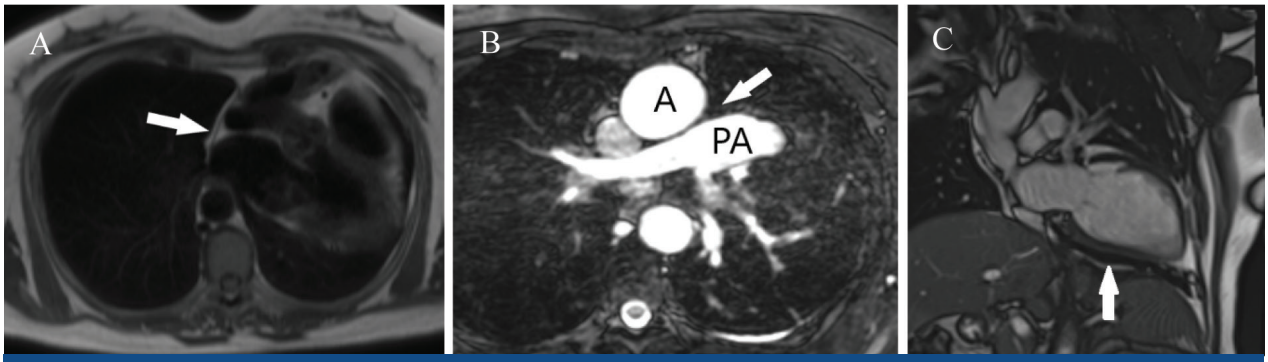

FIGURE 1. A) Axial thoracic half-Fourier acquisition single-shot turbo spin-echo (HASTE) image depicts levorotation of the heart with posterolateral shift of the left ventricular apex. Pericardial continuity on the right side close to atria is preserved (arrow); B) 3D non-contrast MR angiography. Interposition of lung tissue (arrow) between the ascending aorta (A) and the main pulmonary artery (PA) is typical for complete left-sided pericardial agenesis; C) Two-chamber steady-state free precession (SSFP) view of the heart. Interposition of the lung parenchyma (arrow) between the left hemidiaphragm and the inferior cardiac surface.

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Conclusion: Complete CPA is usually incidentally diagnosed on cardiac imaging, cardiac surgery or postmortem. Marked leftward and posterior shift of the heart, as the most prominent sign on chest X-ray or echocardiography, should raise suspicion of this entity and CMR or cardiac CT should be performed in order to confirm the diagnosis. Although majority of patients with complete CPA are asymptomatic with good long-term prognosis, some may experience debilitating symptoms and may require surgery.

LITERATURE

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