Abstract: A missed and misdiagnosed case of pericardial agenesis

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Introduction: Pericardium is a fibroserous sac surrounding the heart and the roots of the great vessels. Pericardial agenesis which is a rare condition can be complete or partial and the most common form is absence of left hemipericardium. Pericardial agenesis is often an incidental finding, as it is usually asymptomatic. Moreover patients presenting with symptoms can be missed diagnosed, because it can be difficult to visualise the pericardium clearly with imaging modalities. Herein we present a misdiagnosed pericardial agenesis case.

Case Report: A 49 years old male was admit to our cardiology clinic for confirmation, and follow-up of a previous diagnosis of right ventricular cardiomyopathy (CMP). He had been previously diagnosed based on transthoracic and transesophageal echocardiography findings 13 years ago. Since then he had been attending routine follow-ups, he had also underwent several imagining modalities even cardiac magnetic resonance (CMR) to confirm the diagnosis of right ventricular CMP. No arrhythmia had been detected in Holter monitoring. He had no dyspnea, or other specific signs and symptoms. His medical and family histories were otherwise unremarkable. A 12-lead electrocardiogram (ECG) showed normal sinus rhythm without having specific abnormalities. The chest X-ray revealed laevorotation of the heart (Fig A). The transthoracic echocardiography showed a laterally displaced left ventricle (LV) apex, and an enlarged right ventricle (RV) with normal systolic function (Fig B). Coronary computed tomography angiography (CCTA) revealed normal coronary arteries, leftward displacement of the heart with mild RV dilatation. The pericardium was only visible around right atrium (Fig C). CMR demonstrated mild RV dilatation (end-diastolic volume 167ml) with reasonable systolic function (EF 50%), normal LV dimensions with normal systolic function (Fig D). The absence of pericardium at both left and right sides, except around right atrium, was confirmed, whereas there was no additional finding such as wall motion abnormality. Therefore the patient was diagnosed with partial pericardial agenesis and scheduled for follow-ups.

Conclusion: The pericardial agenesis which is a rare disorder is generally benign. Although MRI is believed to be the gold standard technique, it might not be able to delineate entire pericardium, because of various artefacts or paucity of surrounding fat. Therefore physicians should be aware of the indirect signs on various imaging modalities like excessive laevorotation of the heart, as direct recognition can be tricky, and the patient can be easily missed and/or misdiagnosed. Our patient was asymptomatic. His pericardial defect was almost total and this abnormality hadn’t been recognized in his previous CMR or echocardiography, leading to a misdiagnosis of CMP. Complete or unilateral absence of the pericardium is considered to be benign, surgical pericardioplasty may only be considered for highly symptomatic patients.
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