Partial absence of pericardium

Case 16725

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Section: Cardiovascular
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Imaging Technique: MR
Case Type: Clinical Cases
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Patient: 44 years, male

Clinical History:

We present a case of a 44-year-old Caucasian man who presented with acute chest pain and mildly raised troponin levels. Invasive angiography revealed patent and normal coronary arteries. Echocardiography, although challenging due to an unusual cardiac position, revealed trabeculated apical ventricles with dyskinetic inferoseptum. He had suffered an embolic stroke 8 years ago with residual right-sided weakness.

Imaging Findings:

Contrast-enhanced cardiac MRI (CMR) was performed using arrhythmogenic right ventricular cardiomyopathy viability protocol. The heart was markedly displaced into the left hemithorax (Fig. 1). There was interposition of lung tissue between normally configured thoracic aorta and pulmonary trunk (Fig. 2, 3). Both ventricles had an elongated appearance (Fig. 4), normal indexed volumes and normal biventricular function. There was indentation of the right ventricular (RV) free wall at mid-ventricular level with relative dilatation of the apex (Fig. 5). Dyssynchronous contraction was observed on cine images. The pericardium was predominantly absent with some pericardium seen adjacent to the RV (Fig. 6, 7).

Discussion:

The heart and origin of the great vessels are enveloped with a two-layered membrane - the pericardium, as most other organs are covered, i.e. lungs by the pleura and abdominal organs by the peritoneum. Parietal and visceral pericardial layers are separated by a thin layer of serous fluid (up to 50 ml) so as to facilitate cardiac contractions and to limit spread of infection and inflammation [1]. Complete or partial absence of pericardium is a rare condition with a reported incidence of 0.002 to 0.004% in a surgical and pathological series [2]. Premature atrophy of the common cardinal vein is a precursor to pericardial agenesis [3]. Patients with pericardial defects and without associated congenital abnormalities can be asymptomatic, otherwise they may present with chest pain and cardiac conduction defects.

Echocardiography, computed tomography (CT) and cardiac magnetic resonance (CMR) imaging have been used to evaluate the pericardium and its absence. Normal pericardium is 2 mm thick and appears dark on T2-weighted CMR images [4]. Pericardial agenesis may be complete or partial. It is mainly incomplete and left-sided resulting in the interposition of the lung between major vessels. Right-sided and complete absence is rarer. The other features include leftward rotation and elongation of the heart chambers.
Agenesis of the pericardium is associated with bicuspid aortic valve, atrial septal defect, patent ductus arteriosus, mitral valve stenosis, or tetralogy of Fallot, bronchogenic cyst and pulmonary sequestration [5]. Complications of congenital pericardial defect may include herniation and entrapment of a cardiac chamber, especially the left atrial appendage or compression of the left coronary artery [6]. Asymptomatic patients may not be diagnosed until scanned for other reasons and will not require any treatment. Patients with uncomplicated absence of pericardium may need symptomatic treatment. In complicated cases surgical closure or enlargement of the defect may be contemplated to alleviate herniation.

Conclusion: Congenital absence of pericardium is a rare anomaly and may only be detected incidentally. Patients usually present with chest pain and conduction abnormalities and may need symptomatic treatment. Surgical management should be reserved for complicated cases.

Written informed patient consent for publication has been obtained.

**Differential Diagnosis List:** Partial absence of pericardium, Complete absence of pericardium, Arrhythmogenic right ventricular cardiomyopathy, Coronary artery disease, Atypical chest pain

**Final Diagnosis:** Partial absence of pericardium

**References:**


Figure 1

Description: Half fourier single-shot turbo spin echo (HASTE) image depicts heart displaced in the left hemithorax. Origin: Dr. Sohail Iqbal, North West Heart Centre, Wythenshawe Hospital, MFT, Manchester.
Figure 2

Description: Steady state free precision (SSFP) image shows interposition of lung in between great vessels Origin: Dr. Sohail Iqbal, North West Heart Centre, Wythenshawe Hospital, MFT, Manchester
**Description:** Black-blood image shows interposition of lung in between great vessels

**Origin:**
Dr. Sohail Iqbal, North West Heart Centre, Wythenshawe Hospital, MFT, Manchester
Figure 4

**Description:** Elongated ventricles

**Origin:** Dr. Sohail Iqbal, North West Heart Centre, Wythenshawe Hospital, MFT, Manchester
Description: RV mid ventricular indentation
Origin: Dr. Sohail Iqbal, North West Heart Centre, Wythenshawe Hospital, MFT, Manchester
Description: Partially absent pericardium

Origin: Dr. Sohail Iqbal, North West Heart Centre, Wythenshawe Hospital, MFT, Manchester
Description: Partially visible pericardium

Origin: Dr. Sohail Iqbal, North West Heart Centre, Wythenshawe Hospital, MFT, Manchester