Case Report

Congenital partial absence of the pericardium presenting with a rare concurrent abnormality of vascular ring diagnosed by cardiac magnetic resonance imaging

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Abstract

Congenital absence of the pericardium is a rare abnormality that can be diagnosed by cardiac imaging procedures. A 49-year-old male needed medical attention due to the appearance of palpitation with a systolic murmur, and a notable aortic arch deviation was seen in the chest X-ray. In the echocardiogram, a poor echo window was detected. A cardiac magnetic resonance imaging (MRI) showed a rare concomitant anomaly of partial absence of the pericardium including a rare defect of the right-sided aortic arch. Using cardiac MRI, the pericardium can be easily visualized, and thus, its absence more easily detected, aiding appropriate clinical decision-making.

Key Words: Absence of pericardium, cardiac magnetic resonance imaging, vascular ring

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INTRODUCTION

Congenital absence of the pericardium is a rare abnormality characterized by the absence of part or all of the pericardium frequently due to the failure of pericardial development secondary to premature atrophy of the left duct of Cuvier, which then fails to nourish the left pleuropericardial membrane.[1,2] This defect is three times more common in men than in women, and though it can be diagnosed at any age, it is seen most often in those under 20 years old.[3] Regarding its location, complete absence of the left side is seen in 35% of cases, and other defects include diaphragmatic surface in 17%, total bilateral absence in 9%, and right-sided defect in only 4% of cases.[3] Clinically, this defect is discovered incidentally; its manifestations may include tachycardia, palpitation, right bundle-branch block, and chest pain that may even be overlapped with ischemic events.[4] Congenital absence of the pericardium is technically diagnosed by cardiac imaging procedures, including echocardiography and axial computed tomography scans; however, it may remain hidden in some cases requiring the use of supplemental imaging techniques such as magnetic resonance imaging (MRI).[5] Moreover, this abnormality may be accompanied by

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other simultaneous defects that may only be diagnosed by MRI and not by initial imaging assessments. Here, we report a case of congenital absence of the pericardium that was finally discovered by MRI and which was bundled with a rare concomitant defect of vascular ring created by a right-sided aortic arch with aberrant origin of the left subclavian artery (LSCA) and Kommerell's diverticulum.

CASE REPORT

A 49-year-old male sought medical attention for palpitations and was referred to our echocardiography laboratory for structural assessment of the heart. The patient suffered from an occasional palpitation in the prior weeks without a history of dyspnea, chest discomfort, or any notable extracardiac symptom. He had no coronary artery disease-related risk factors. On physical examination, normal pulses with normal heart sounds were revealed; however, a systolic murmur grade of 2/6 to 3/6 could be heard in the left sternal border. The point of maximal impulse was palpated in the axillary area and was hyperdynamic. The initial electrocardiogram indicated normal sinus rhythm with normal axis without any evidence of arrhythmias, blocking, or ischemia. The chest X-ray showed normal heart size; however, it was noted that the aortic arch deviated to the right side. Transthoracic echocardiography was technically challenging due to poor echo-windowing. The best site for image taking was in the second to third intercostal of the mid-axillary line. The pericardium seems normal in long and short axis views; however, there was a hyperdynamic heart and dilated right ventricular outflow tract (RVOT) in the short axis view [Figure 1]. The view also showed hyper-exaggerated motion of all cardiac chambers and floating right cardiac chambers.

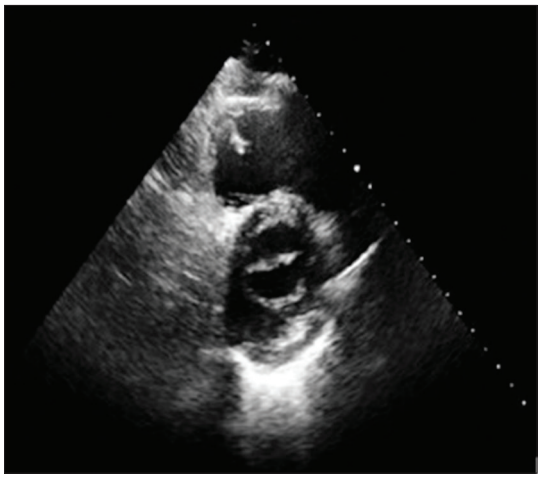

Figure 1: Parasternal short axis from an uncommon site showing a dilated right ventricular obstructive tract

A suprasternal view was not possible in the standard site. The aortic arch was visualized whenever the probe moved to the right side which was suggestive of the right-sided aortic arch. The regional left ventricular ejection fraction was normal. For further assessment, a transesophageal echocardiography revealed no evidence of intracardiac shunt and normal cardiac valves with mild tricuspid regurgitation, normal systolic pulmonary artery pressure, normal drainage of pulmonary veins to left atrium (LA), and normal-sized and intact coronary sinus. However, in this echocardiography viewing, the apex of the right ventricular seemed aneurysmal. The patient was referred for MRI imaging to rule out or confirm arrhythmogenic right ventricular dysplasia. The cardiac MRI showed upward rotation in addition to a well-distinguished and extreme levoposition of the heart and the partial absence of the pericardium, especially around the LA and left ventricle (LV), with obvious remnants of the pericardium around the anterior right atrium and basal part of the right ventricle (RV) free wall, followed by a discontinuity of the parietal pericardium around the mid-anterior RV free wall [Figure 2a and b]. The cardiac MRI showed a projection of lung tissue interposed between the aorta and mean pulmonary artery (MPA) [Figure 3]. Furthermore, a focal bulge and dilation of RVOT

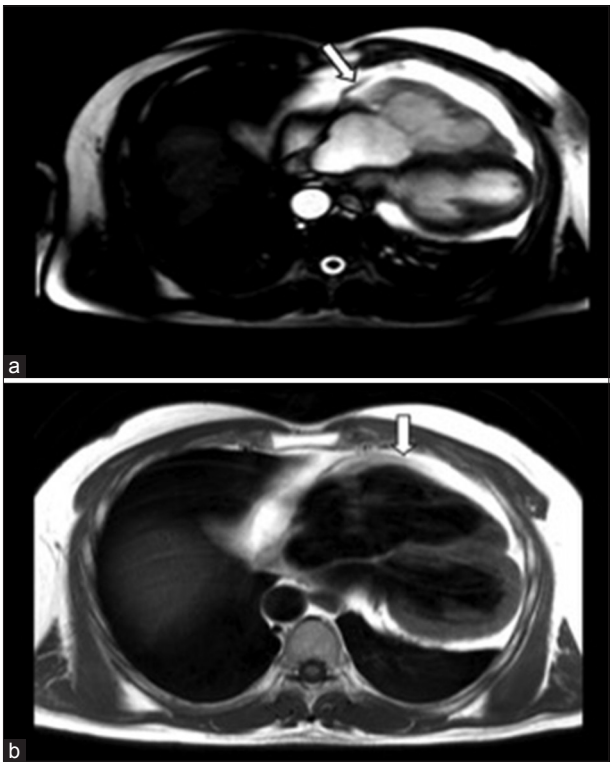

Figure 2: (a) Axial view in true FISP sequence showing an extreme cardiac levoposition with obvious discontinuity of the parietal pericardium. (b) Half Fourier acquisition single-shot turbo spin echo sequence in true FISP sequence showing an extreme cardiac levoposition with obvious discontinuity of the parietal pericardium

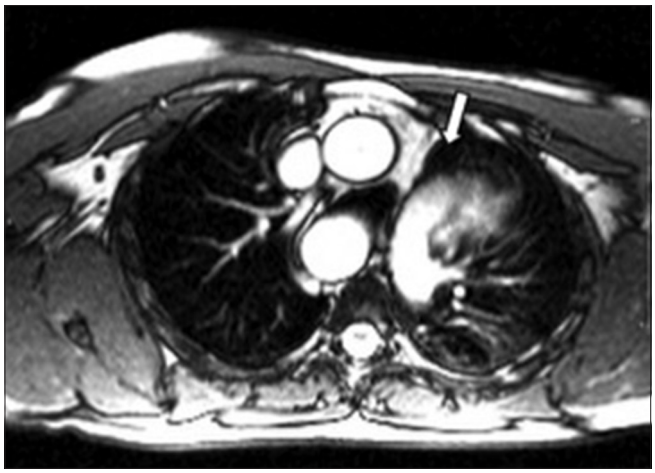

Figure 3: True FISP sequence on axial image showing a long projection between the pulmonary artery and the ascending aorta

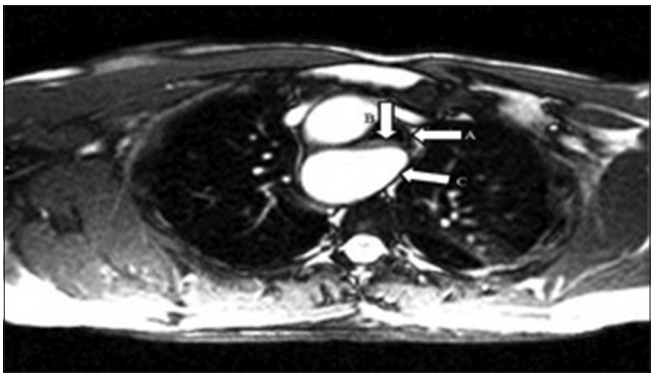

Figure 4: True FISP sequence of axial image showing how Kommerell's diverticulum creates a compressive effect on the posterior aspect of the esophagus and trachea

(about 39 mm) with normal size MPA and its branches were seen. The aortic arch was right-sided with aberrant origin of the LSCA from the upper descending aorta in addition to a saccular aneurysmal dilatation of the LSCA's origin (Kommerell's diverticulum) resulted compressive effect on posterior wall aspect of esophagus and trachea (vascular ring) [Figure 4]. In summary, there was partial absence of the pericardium (especially around LA and LV) with vascular ring due to aberrant origin of the LSCA from the upper descending aorta of the right-sided aortic arch in addition to a Kommerell's diverticulum.

DISCUSSION

In cases with congenital absence of the pericardium, the common image findings include a focal bulge in the area of the main pulmonary artery or LA in partial defects, rotating-up of the heart, interposing lung between the heart and left hemidiaphragm, levopositioning of the heart, or even pneumopericardium due to pneumothorax.[6] Although in previous reported cases with absence of the pericardium, a single defect was revealed, other congenital abnormalities may also be also found, including bronchogenic cysts, atrial or ventricular septal defects, patent ductus arteriosus, mitral stenosis, Tetralogy of Fallot, pulmonary sequestration, or diaphragmatic hernia.[4] For the first time, we report a rare concomitant anomaly of partial absence of the pericardium including a rare defect of the right-sided aortic arch with aberrant LSCA's origin and Kommerell's diverticulum creating a vascular ring. In most cases, such abnormalities are frequently diagnosed by echocardiography. With nonspecific symptoms, a combination of diagnostic procedures, such as chest radiography, electrocardiography, transesophageal echocardiography, and even coronary angiography, should be done to confirm or rule out the abnormalities; however, as presented in our report, these defects may not be correctly diagnosed by these procedures which demands the use of advanced imaging techniques such as MRI.[7] This modality has been accepted as the gold standard in delineating the extent of defects, reducing complications, and detecting concomitant anomalies, which can be further managed by minimally invasive surgical techniques.[8-10] Using this procedure, the pericardium can be easily visualized and thus its absence more easily detected, aiding appropriate clinical decision-making.

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Conflicts of interest
There are no conflicts of interest.

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