

Right Ventricle Mimics Right Atrium at First Glance: A Rare Case of Congenital Right Sided Partial Pericardial Defect

Meitzu Wang,¹ Tien-Yu Chang,² Wei-Hsian Yin³ and Yung-Nien Yang³

Among heart irregularities, congenital pericardial defect is an unusual anomaly, and is typically left dominant. However, cases of right pericardial defect with heart herniation are extremely rare. This is a case of congenital right pericardial defect with herniation of the right ventricle free wall and right ventricular outflow tract. The patient is asymptomatic and refused further intervention but even indolent discomfort underscores the risks of iatrogenic injuries to the heart and sudden death caused by mechanical pathogenesis due to changes in anatomical positions of the cardiac structures.

Key Words: Congenital right pericardial defect • Heart herniation

INTRODUCTION

It is generally believed that embryologic theory of the premature atrophy of the left duct of Cuvier, and arrested closure of the pleuropericardial membrane explain the left dominant pericardial defect. Fewer than 30 cases of isolated congenital right pericardial absence have been reported,¹ a number which is further reduced if concomitant with heart herniation. Such a circumstance presents variable clinical courses, ranging from asymptomatic to sudden death. Unexplored gene differences that may be associated with race make this case unusual and quite rarely observed in Asia.

CASE REPORT

A 37-year-old male was admitted to our hospital due to fracture of the distal end of the right clavicle, and was scheduled for a routine internal implantation. Upon pre-operative evaluation, the patient's chest X-ray (CXR) showed an increased prominence of the right heart border (Figure 1A). Additionally, early QRS transition with counterclockwise rotation appeared in lead V1 on electrocardiography. He was asymptomatic without cardiac risk or contraindication to the surgery, hence he opted for the surgery and was referred to our cardiology outpatient department after discharge. Post-surgical electrocardiography (ECG) showed early QRS transition with counterclockwise rotation (Figure 1C), and echocardiographic imaging displayed a retrocardiac echo-free space (Figure 1B). Therefore, right pericardial defect with herniation of right ventricle free wall (RVFW) and right ventricular outflow tract (RVOT) was suspected by chest computed tomography (CT) in comparison with the images of coronal versus axial planes as localizing sequences (Figure 2A); the diagnosis was further confirmed by cardiac magnetic resonance imaging (MRI) (Figure 2B, 2C, 2D). The patient was asymptomatic, thus he refused further intervention and was followed-up at our outpatient department.

Received: September 6, 2015 Accepted: February 1, 2016

¹Department of Internal Medicine; ²Department of Radiology; ³Division of Cardiology, Cheng-Hsin Medical Center, Taipei, Taiwan.

Address correspondence and reprint requests to: Dr. Yung-Nien Yang, Division of Cardiology, Cheng-Hsin Medical Center, No. 45, Cheng Hsin St., Paitou, Taipei, Taiwan. Tel: 886-2-2826-4400; E-mail: yangcvman@yahoo.com.tw

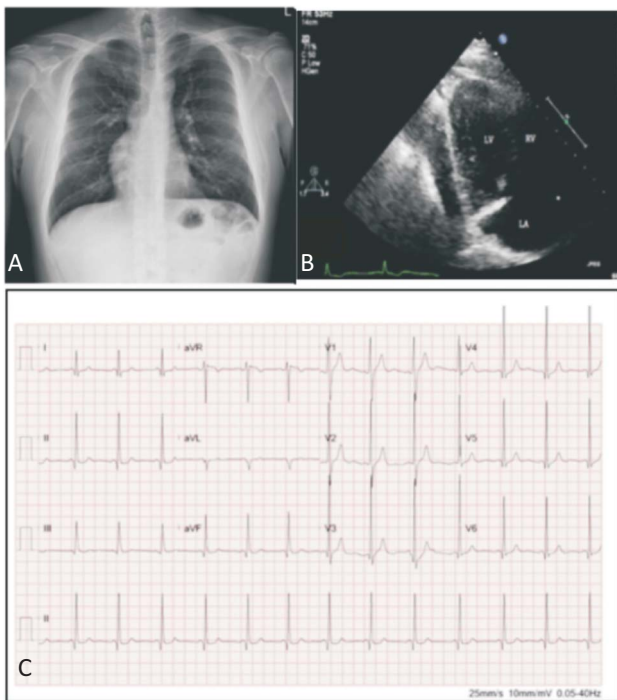


Figure 1. (A) Preoperative chest radiography shows bulging of right heart border. (B) Transthoracic echocardiogram in apical long-axis view shows retrocardiac echo-free space (asterisk). (C) ECG showed early QRS transition with counter-clockwise rotation. ECG, electrocardiography; LA, left atrium; LV, left ventricle; QRS, QRS complex; RV, right ventricle.

DISCUSSION

Congenital pericardial absence was first described in 1559 by Realdus Columbus, and is largely recognized as left dominant with a male preponderance.²

Cases of right pericardial defect with heart herniation are extremely rare. In Asia, only two cases were reported in Japan, in 2002 by Ikeda et al., and in 2015 by Ono and associates. Partial right anterior pericardial defect with reserved pleural membrane and concomitant herniation of RVFW and RVOT as found in our patient is a type of case rarely reported in Asia. In our case, antero-upward herniation rather than rightward protruding, herniated right ventricle free wall superimposed with right atrium lies behind it, which generally leads to increased right prominence on chest radiography (Figure 2A). Patients with partial defect of pericardium have an increased risk of morbidity than those with complete defects.² They manifest with paroxysmal non-exertional lancinating chest pain, dyspnea, syncope, arrhythmia, embolization from mural thrombus, pericarditis and

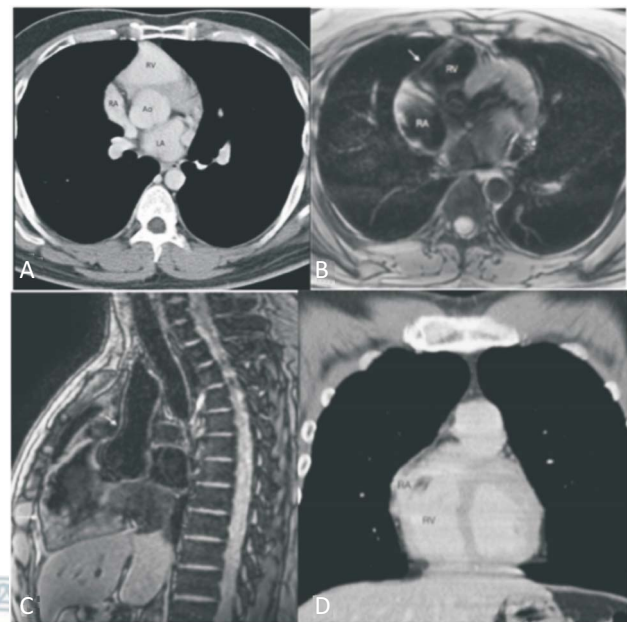


Figure 2. (A) Enhanced CT image shows loss of anterior mediastinal fat, anteroupward herniation of right ventricle. (B) T1 weighted MRI shows right pericardial defect (curved arrow) with right ventricle free wall. The other normal pericardium with linear dark signal intensity (arrow) surrounds aorta and left side right ventricle. (B) Normal pericardium with linear dark signal intensity surrounds non-protruding portion of right ventricle free wall (arrow). (C) T1 weighted MRI shows right anterior pericardial absence between the two ends of normal pericardium with linear dark signal intensity (arrow). (D) Coronal view of chest CT shows the increased right prominence consist of right ventricle free wall and partial right atrium lies behind it. Ao, aortic root; CT, computed tomography; LA, left atrium; MRI, magnetic resonance image; RA, right atrium; RV, right ventricle.

sudden death. This may due to the herniation of cardiac structures through a partial defect, resulting in strangulation of the heart, kinking of the great vessels, traction of pleuropericardial adhesions, postural distention of vessels, mechanical infringement and constriction of the coronary vessels by the fibrous thickened bands on the lower edge absent pericardium.³

Up to 33% of patients with complete absence of pericardium have vague chest pain, and dyspnea, dizziness and syncope have also been reported.² A slight rotation of the heart transforms 50% of these cases into cardiomegaly on CXR, and displacement of point of maximal impulse into the mid-axillary line without actual enlargement of the heart.² Nonspecific ejection systolic murmur is heard most frequently in the left second intercostal space next to the sternum. ECG changes contain right axis deviation, incomplete right bundle branch

block pattern, and clockwise rotation of the heart.² Conversely, physical examination and ECG are usually normal in these patients with partial defect.²

MRI provides excellent contrast of the entire pericardium compared with CT, with the added benefit of absence of ionizing radiation.² However, the retrocardiac echo-free space of our patient was not sufficiently clear to show the lesion of pericardial defect, but the absence of normal right atrium which lies behind the right ventricle provided us an indication of the abnormal anatomy or orientation of the heart structures. Further image workup showed evidence of the herniation and displacement of cardiac structures, while partial right pericardial defect was proved by MRI.

Why was the patient in our case asymptomatic? It may be related to his relatively young age and the pressure of fibrous band of absent pericardium had not constricted the coronary vessels, obviously leading to sufficient stenosis and ischemia. Thereafter, only when involvement is proximal to a major acute marginal branch of the right coronary artery does the RVFW longitudinal strain show significant attenuation.³ In another case, a symptomatic 73-year-old man presented by Ikeda et al., and an asymptomatic 12-year-old boy presented by Ono et al. may indirectly enhance the rationality of this hypothesis.

For a symptomatic patient, surgical intervention may lead to reduced symptoms, with acceptable associated morbidity. For patients diagnosed with moderate-sized pericardial defect, whether symptomatic or not, it may be necessary to undergo prophylactic operation such as left atrial appendectomy, pericardiectomy, defect extension and adhesion division with surgical liberation of the pericardial fibrous rim to reduce the risk of sudden death caused by cardiac structure herniation

and incarceration. Pericardioplasty with pericardial reconstruction were employed to immobilize the heart, and subsequently lead to symptomatic improvement. For asymptomatic patients, prophylactic surgical repair remains controversial. However, extremely small defects or complete absence of pericardium lack any coronary risk of morbidity and do not require surgical intervention.⁵ Additionally, antithrombotic therapy with aspirin treatment was reported in an asymptomatic patient.⁴ An elevated risk of injury to the heart should be noted if the patient undertakes thoracotomy due to changes of the anatomical position of cardiac structures.

TBX 18 and Wt1 are crucial regulators directing the process of the pleuropericardial membrane, to create a separation of pericardial and pleural cavities.⁶ Unexplored gene differences of gender and race may explain the male preponderance in congenital pericardial defect, and in those rare cases found in Asia.

REFERENCES

1. Koo CW, Newburg A. Congenital absence of the right pericardium: embryology and imaging. *J Clin Imaging Sci* 2015;5:12.
2. Faridah Y, Julsrud PR. Congenital absence of pericardium revisited. *Int J Cardiovasc Imaging* 2002;18:67-73.
3. Chang WT, Tsai WC, Liu YW, et al. Changes in right ventricular free wall strain in patients with coronary artery disease involving the right coronary artery. *J Am Soc Echocardiogr* 2014;27:230-8.
4. Gatzoulis MA, Munk MD, Merchant N, et al. Isolated congenital absence of the pericardium: clinical presentation, diagnosis, and management. *Ann Thorac Surg* 2000;69:1209-15.
5. Nasser WK. Congenital absence of the left pericardium. *Am J Cardiol* 1970;26:466-70.
6. Norden J, Grieskamp T, Christoffels VM, et al. Partial absence of pleuropericardial membranes in Tbx18- and Wt1-deficient mice. *PLoS One* 2012;7:e45100.