Congenital Partial Pericardial Defect and Herniated Right Atrial Appendage: A Rare Anomaly

Cemşit Karakurt, M.D., Deniz Oğuz, M.D., Selmin Karademir, M.D., Metin Sungur, M.D., and Burhan Öcal, M.D.

Pediatric Cardiology Unit, Dr. Sami Ulus Children's Hospital Ankara, Turkey

A congenital partial defect of the right-sided pericardium is a rare cardiac anomaly and it represents defective formation of the pleuropericardial membrane. Patients can be asymptomatic, but they may experience chest pain, myocardial ischemia, emboli, arrhythmia, and sudden death. In this report, we present an 8-month-old boy with pericardial defect and right atrial appendage herniation. It was diagnosed by echocardiography and cardiac magnetic resonance imaging. (ECHOCARDIOGRAPHY, Volume 23, October 2006)

congenital partial pericardial defect, *right atrial appendage*

A congenital defect of the pericardium occurs due to defective formation of the pleuropericardial membrane and may involve the entire pericardium. Defects of the left pericardium are commonly seen than right-sided defect.[1]

In this report, we present an 8-month-old boy with pericardial defect and right atrial appendage herniation. The diagnosis was performed by echocardiography and cardiac magnetic resonance imaging (MRI).

Case Report

An 8-month-old boy admitted to our hospital due to acute bronchiolitis. He was referred to the pediatric cardiology department for evaluation of cardiomegalia seen on chest x-ray examination. He had no known history of cardiac trauma. Cardiac examination of the heart did not reveal any abnormality. Electrocardiogram interpretation was normal. Chest x-ray examination revealed cardiomegalia with prominent right heart border. On transthoracic echocardiogram a large cystic mass adjacent and related to the right atrium was determined in the apical four-chamber view. This cystic mass reached the apex of the heart and compressed the right ventricle (Fig. 1). Left ventricular systolic functions were normal and there was no thrombus in the cardiac chambers. Contrast echocardiography performed through a peripheral venous line showed microbubbles filling the right atrium and large cystic mass simultaneously. Contrast echocardiography also showed slow and turbulated blood flow in this cystic mass (Fig. 2). Cardiac MRI showed right atrial appendage herniation due to partial congenital pericardial defect. MRI demonstrated leftward displacement of the heart and dark lines separating the high-intensity epicardial fat or the medium intensity pericardial fat. These low

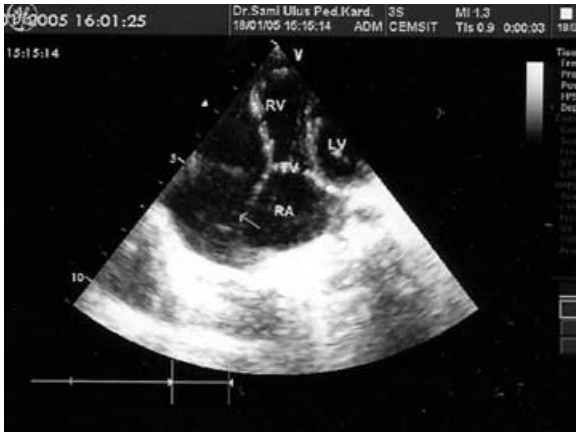

Figure 1. *Transthoracic echocardiogram in apical four-chamber view shows an enlarged right atrial appendage that looks like mass.*

Address for correspondence and reprint requests: Cemşit Karakurt, M.D., Dr. Sami Ulus Children's Hospital, Örnek mah. Babür cad. Altindağ, Ankara, Turkey. Fax: 0090-312 317 03 53; E-mail: ckarakurt@yahoo.com

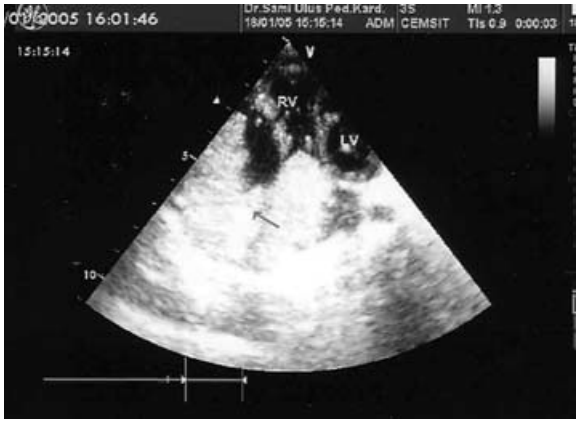

Figure 2. *Contrast echocardiography in apical four-chamber view shows microbubbles filling simultaneously the whole right atrium and herniated right atrial appendage.*

intensity lines were identified along the right atrial appendage and were thought to be partial absence of the right atrium (Fig. 3A and 3B).

After the treatment of acute bronchiolitis the patient was given aspirin 3 mg/kg per day because of slowed blood flow in the herniated right atrial appendage.

Discussion

The congenital absence of the pericardium is a rare congenital anomaly and can vary from partial to complete absence occurring more often on the left than the right side.[2,3]

The cause of defect is the persistence of pleuropericardial membrane during embryologic development. This may be caused by a deficient blood supply, and it has been hypothesized that the predominance of left-sided defects results from premature atrophy of the duct of Cuvier. Because the right Cuvier's duct remains as the superior vena cava, right-sided lesions and bilateral complete absence of the pericardium are extremely rare.[4]

Although, most cases of pericardial defect are identified incidentally, they may be associated with chest pain, syncope, arrhythmias, embolization from mural thrombus, pericarditis, and sudden death due to herniation or incarceration of the left atrial appendage, which have been reported.[2,5]

Diagnosis of pericardial defects may be difficult. Differential diagnosis includes pericardial defects resulting from surgery or trauma, pericardial cysts, idiopathic enlargement of right

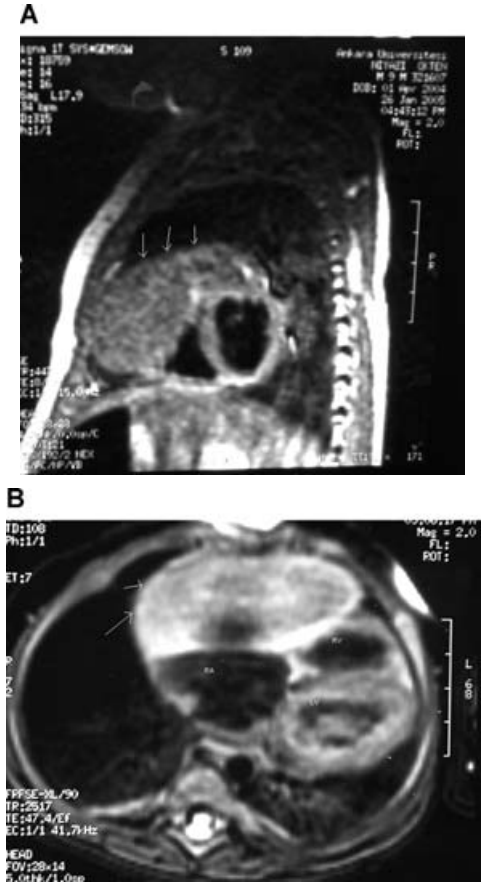

Figure 3. *T1* ***(A)*** *and T2* ***(B)*** *magnetic resonance images show enlarged right atrial appendage and pericardial defect on the right side.*

atrium. Chest radiography often raises suspicion; echocardiography, computed tomography, or MRI is used to establish the diagnosis.

MRI provides excellent images of the entire pericardium. MRI is a useful diagnostic tool for establishing the pericardial defect. High-resolution images synchronized with the cardiac cycle can be obtained in any plane. Additional anatomical and functional information can be obtained from adjacent structures such as aorta, lungs, pleura, and mediastinum.[7,8]

The present case referred to our clinic for evaluation of cardiomegalia. There was a cystic mass related to right atrium on echocardiography. We thought right atrial appendage herniation due to partial defect of the right pericardium and the appearance in the herniated appendage on MRI showed stasis. There was no thrombosis in the right atrial appendage.

Patients with partial absence of the left-sided pericardium carry high risk of the left atrial appendage or left ventricle, leading to fatal myocardial strangulation. Treatment of the right-sided defects is controversial but whenever a patient develops symptoms, surgical treatment is recommended.[6]

Our patient was asymptomatic. We did not detect any compression of great arteries, coronary arteries, and incarceration of cardiac structures but our patient carried thromboembolic risk due to slowed blood flow inside the herniated atrial appendage. Therefore, we gave aspirin treatment. He did not have any cardiac event and symptoms for 6 months.

We are not considering surgical treatment at the present but we will follow closely in frequent intervals due to thromboembolic events.

In conclusion, partial pericardial defects are a rare cardiac anomaly. Contrast echocardiography and cardiac MRI are useful diagnostic tools for the diagnosis.

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