Asymptomatic congenital pericardial defect: an aspect of diagnostic modalities and treatment

Asemptomatik konjenital perikardiyal defekt: Tedavi ve tanısal yönleri

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Introduction

Pericardium invests the heart and has two main functions: maintenance of the heart in mediastinum and prevention of sudden cardiac distension by cardiac volume overload. Most of the congenital abnormalities of pericardium are asymptomatic and discovered at routine chest imaging, cardiac surgery, or investigation of other diseases. They are rare disorders associated with cardiac, pulmonary and skeletal abnormalities in 30% of patients. Pericardial defects comprise of partial left sided (70%), right sided (17%) and complete defects (13%) (1). They occur in 1 out of 14,000 births and have a male predominance. Chest pain is the most common complaint in the evidence of pericardial defects. We present a case with partial absence of left sided pericardium without any symptoms. We also discuss diagnostic modalities and treatment strategies.

Case Report

A 34-year-old asymptomatic male in the army was seen in our outpatient clinic during his periodical examination. His past medical history was normal. Chest X-ray examination (Fig. 1) revealed an enlarged cardiac silhouette and he was referred for further cardiologic work-up. His physical examination was normal. Electrocardiography (ECG) showed normal sinus rhythm with negative T waves in DIII, aVF and V4-6.

On echocardiographic examination, left ventricle was imaged at a more lateral position. Left ventricle was elongated into the apical portion resembling a ventricular aneurysm. Wall motions were normal and no thrombus was observed in left ventricle. Mitral valve was prolapsing with a mild regurgitation (Fig. 2).

Pericardial defect was suspected and magnetic resonance (MR) images of the pericardium were obtained. The ECG-triggered MR imaging was performed with a 1.5T scanner in axial, coronal, and sagittal planes (TR: 2.6 ms; TE: 1.3; flip angle 80; slice thickness: 8 mm, field of view: 410; matrix: 183*256, number of signal acquired: 1; scan time: 9.9 s). Pericardium over the right atrium and ventricle was delineated. On MR scans, we found rotation and displacement of the heart into the left hemithorax, interposition of the lung tissue between the aorta and the main pulmonary artery and extension of the left atrial appendage and main segment of the pulmonary artery beyond the mediastinal margins that was consistent with partial absence of left sided pericardium (Fig. 3-5).

Exercise stress testing was performed to exclude asymptomatic coronary ischemia and to evaluate exercise capacity. There was no ischemic ST segment changes, but exercise was terminated at 85% of the target heart rate due to fatigue. Results of further examination including pulmonary function test in terms of fatigue were in normal limits and it was thought to be related to anxiety of a new disease in psychiatry consultation. Since the patient was asymptomatic initially despite rigorous exercise for years and no cardiac structure was at risk of incarceration in imaging modalities, conservative treatment was preferred.

Discussion

There are no specific ECG changes for pericardial defects. Chest X-ray raises only suspicion but has no definite diagnostic role in pericardial defects. Pericardial pathologies are initially assessed by echocardiography after abnormal findings on ECG or chest X-ray for differential diagnosis. Partial pericardial defects can be suspected by paradoxical septal motion, bulging of left ventricle through the defect and displacement of the heart towards more lateral position in thorax. In our case left ventricular apex was bulging through the defect and mitral valve was prolapsing with a mild regurgitation. Total pericardial defects may be suspected by bulging of the inferior left ventricular wall during diastole (2). Right-sided defects are difficult to image echocardiographically. Magnetic resonance study provides excellent images of the entire pericardium without contrast injection or ionizing radiation and seems to be the best diagnostic moda-

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lity in the diagnosis of pericardial defects (3). Computed tomography (CT) is useful especially in quantification of pericardial thickening but can also delineate the extension and defect of the pericardium and relation of the defect to great vessels and cardiac structures (4). In our case, MR did not demarcate the pericardium over the aorticopulmonic window, posterior and lateral aspects of the left ventricle while pericardium appeared lying over the right atrium and right ventricle. Pericardium over the right atrium and right ventricle can mostly be visualized with CT and MR imaging, however delineation of the pericardium over the lateral and posterior walls of the left ventricle may not be possible in all cases due to insufficient epicardial fat and motion artifact. Therefore, as in our case, indirect findings such as interposition of the lung tissue between the aorta and the main segment of the pulmonary artery and between the inferior aspect of the left heart and left diaphragm, bulging of the left atrial appendage and main segment of the pulmonary artery beyond the mediastinal margins, displacement of the heart into left hemithorax, rotation of the heart towards the left, and blurring of the heart-lung borders are used as diagnostic criteria in the recognition of congenital absence of pericardium in MR images. Although non-visualization of the ventricular pericardium is a usual finding on CT and MR imagings, combination of aforementioned indirect findings and visualization of the right atrial and ventricular pericardium suggested a partial absence of the left heart pericardium in the current case. Cardiac catheterization may reveal constriction of coronary arteries by pericardial edges and herniation of left ventricular structures or atria through the defect.

Figure 1. Chest X-ray showing enlargement of cardiac silhouette and apical bulging

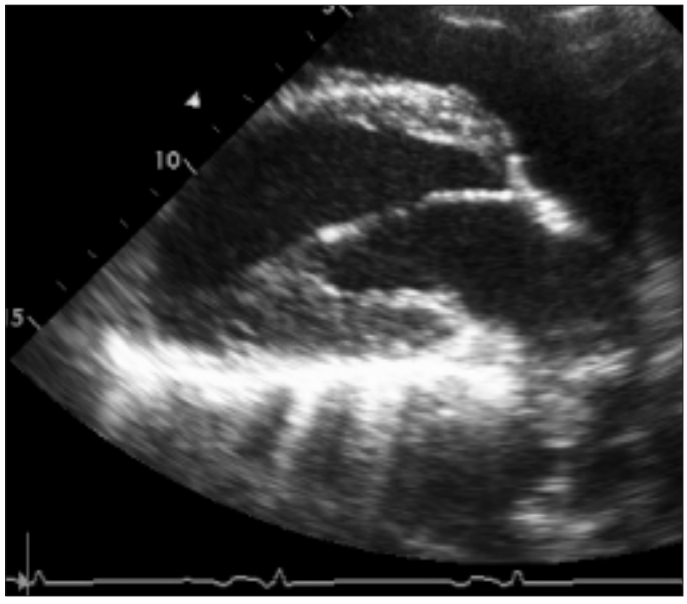

Figure 2. Parasternal long axis view showing apical bulging of the left ventricle through the defect

Figure 3. An axial balanced turbo field echo MR image through the main pulmonary artery reveals interposition of the lung tissue (arrow) between the main pulmonary artery and aorta due to absence of the pericardium. Extension of main pulmonary artery beyond the mediastinal borders was also noted (arrowhead)

MR - magnetic resonance

Figure 4. The axial balanced turbo field echo MR images at the ventricular level demonstrate significant displacement of the heart in to the left hemithorax. Heart apex rotates posteriorly

MR - magnetic resonance

Figure 5. Coronal balanced turbo field echo MR image shows significant displacement of the heart into the left hemithorax with enlargement of the left atrial appendage that is consistent with the absence of pericardium.

MR - magnetic resonance

Asymptomatic patients may be managed without undergoing surgery. Yamaguchi et al (5) have advocated pericardial repair if thoracotomy is performed for other reasons in asymptomatic patients. However, if patients are symptomatic or there is a risk of incarceration of left ventricle or atrial appendage, left atrial appendectomy, pericardioplasty or extension of pericardial defect may be beneficial (6). We suggested observation alone in our patient because of the absence of symptoms, objective evidence of ischemia as well as a risk of incarceration.

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