CASE REPORT

A case of congenital partial absence of the left pericardium presenting with atypical chest pain

Atipik göğüs ağrısı ile prezente olan konjenital parsiyel sol perikardın yokluğu olgusu

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Summary– Congenital absence of the pericardium (CAP) is a rare cardiac malformation and can be defined as the partial or total absence of the fibroelastic sac that surrounds the heart and great vessels. As the patients are often asymptomatic or have nonspecific symptoms, the diagnosis of this rare congenital anomaly is difficult. Therefore, it is usually diagnosed incidentally during imaging, intraoperatively, or during postmortem examinations. In this regard, it is important to keep specific images in mind during the examination and to suspect CAP to make an accurate diagnosis. In this report, we present a case of a 42-year-old male who presented with a complaint of atypical chest pain and was diagnosed with CAP using multimodality imaging.

Özet– Perikardın konjenital yokluğu nadir görülen bir kardiyak malformasyondur ve kalbi ve büyük damarları çevreleyen fibroelastik kesenin kısmen veya tamamen yokluğu olarak tanımlanabilir. Hastalar genellikle asemptomatik ya da non-spesifik semptomlara sahip oldukları için bu nadir konjenital anomalinin teşhisi zordur. Bu nedenle genellikle görüntüleme sırasında, intraoperatif olarak veya postmortem incelemeler sırasında tesadüfen teşhis edilir. Bu bakımdan, muayene sırasında spesifik görüntülerin akılda tutulması ve doğru bir teşhis için perikardın konjenital yokluğundan şüphelenilmesi önemlidir. Bu olgu sunumunda, atipik göğüs ağrısı şikayeti ile başvuran ve çoklu görüntüleme yöntemleri ile perikardın konjenital yokluğu tanısı alan 42 yaşında bir erkek hasta sunuyoruz.

Congenital absence of the pericardium (CAP) is a rare anomaly characterized by partial or total absence of fibroelastic pericardial membrane. As it is rare and usually asymptomatic, clinicians may have difficulties in the diagnosis. Here, we present a case of a 42-year-old male who presented with a complaint of atypical chest pain and was diagnosed with CAP.

CASE REPORT

A 42-year-old male patient was admitted to our cardiology outpatient clinic because of an atypical chest pain. His past medical history was unremarkable for any chronic illness. His vital signs were a heart rate of 80 bpm and blood pressure of 120/70 mmHg. Electrocardiogram showed normal sinus rhythm and non-specific inverted T-wave in anterior derivations. A chest x-ray was ordered during the diagnostic approach and showed levorotation of the heart and elongation of the left ventricular contour (Figure 1A). Transthoracic echocardiography was performed and revealed that the left ventricular apex was displaced toward the armpit and mid-axillary line (Figure 1B). With these findings, we suspected CAP and ordered chest computed tomography (CT). The chest CT scan indicated an extreme left-sided displaced heart with the apex pointing toward the mid-axillary line (Figure 1C) and interposition of lung parenchyma between the pulmonary artery and aorta (Figure 1D). With the addition of these findings, the diagnosis of CAP was confirmed. The patient was followed up medically.

DISCUSSION

Congenital absence of the pericardium is a rare cardiac malformation and can be defined as the partial or total absence of the fibroelastic sac that surrounds the heart and great vessels.[1] Partial defect of the pericardium is more common, and approximately 70%

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of it manifests as a left-sided defect.[1] More rarely, it may present as right-sided or diaphragmatic defects.[1] As patients are often asymptomatic or have nonspecific symptoms, the diagnosis of this rare congenital anomaly is difficult. Therefore, it is usually diagnosed incidentally during imaging, intraoperatively, or during postmortem examination.[2-4] In this regard, it is important to keep specific images in mind during the examination and to suspect CAP to make an accurate diagnosis.

For diagnosis, chest radiography, echocardiography, CT, and magnetic resonance imaging (MRI) are usually used as imaging modalities. On chest x-ray, flattening and elongation of the left ventricular contour are usually observed, at times accompanied by radiolucency between the main pulmonary artery and aortic knob.[3,5] Getting an imaging acquaintance and considering the finding of elongation in the left ventricular contour on chest x-ray, which we frequently use in our daily practice in emergency services and outpatient clinics, may help to diagnose CAP. During echocardiographic examination, optimal images are often obtained through modified windows owing to the abnormal heart position.[3,5,6] As in our patient, the LV apex is often imaged on the mid-axillary line. In addition, elongated atria may be seen together with bulbous ventricles on the modified apical view.[6,7] Another important point to consider during echocardiography is the fact that the right ventricle (RV), which appears to be enlarged owing to its anterior location, may cause patients to be mistakenly suspected for the diagnoses of arrhythmogenic right ventricular cardiomyopathy or atrial septal defect.[8,9] Therefore, when investigating the etiology of such RV enlargement, it is essential to remember the characteristic imaging findings of CAP in the differential diagnosis. When there are x-ray and echocardiographic findings suggesting CAP, CT or MRI is often ordered to confirm the diagnosis and to identify other associated lesions as further investigation. Detecting interposition of the lung parenchyma in areas where the pericardium is absent on CT and MRI is an almost specific finding for CAP.[6] Another advantage of CT and MRI is that these methods enable the detection of potentially fatal complications such as compression of coronary arteries that may occur because of herniation in partial type defect.[7]

Abbreviations:

CAP	*Congenital absence of the pericardium*
CT	*Computed tomography*
MRI	*Magnetic resonance imaging*
RV	*Right ventricle*

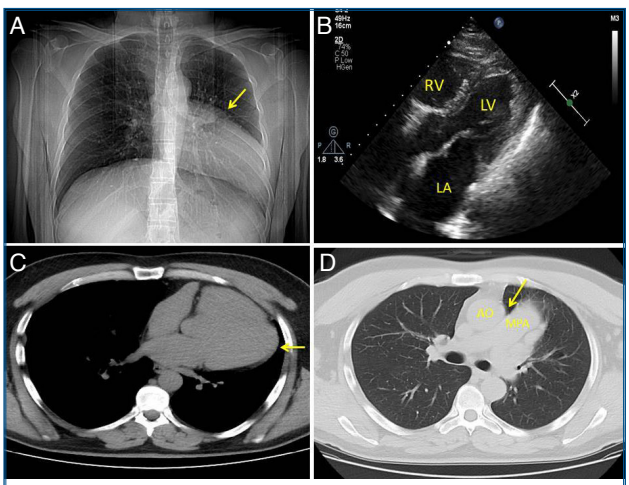

Figure 1. (A) Chest x-ray demonstrating levorotation of the heart and elongation of the left ventricular contour (arrow). **(B)** Transthoracic echocardiography apical 5-chamber view obtained through the modified window because of the abnormal left ventricular apical position. **(C)** Chest computed tomography scan in the axial plane demonstrating an extreme left-sided displaced heart with the apex pointing mid-axillary line (arrow). **(D)** Chest computed tomography scan in the axial plane demonstrating interposition of lung parenchyma between the pulmonary artery and aorta (arrow).

After clarifying the diagnosis with imaging modalities, patients are generally followed up conservatively. It is important to know the potential complications that may occur, especially in partial defects during the follow-up. Life-threatening complications such as left atrial appendage incarceration, ventricular herniation, torsion of the great vessels, and impingement of the coronary arteries may occur, which results in sudden cardiac death.[3] In such high-risk features, surgical intervention may be required.[3]

In conclusion, the diagnosis of this rare congenital anomaly requires evaluating the imaging findings and choosing appropriate imaging methods. In this case report, we have highlighted the essential imaging findings for clinicians to make an accurate diagnosis.

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Anahtar Kelimeler: Koroner fistül; perkütan kapama; vasküler tıkaç