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CASE IMAGE

Congenital absence of the pericardium

Konjenital perikard yokluğu

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A 15-year-old girl was referred to our outpatient clinic for further examination when heart sounds were heard in the left mid-axillary line from a hospital where she was previously admitted with the complaint of epistaxis. She did not have chest pain, shortness of breath, or any other symptoms. Her medical history showed no family history of sudden cardiac death or inheritable cardiac diseases.

On cardiac examination, a regular heart rhythm was heard without a heart murmur. Her apex beat, however, was grossly displaced to the left and more so on lying supine. Her blood pressure was 112/68 mmHg. A chest X-ray showed situs solitus, levocardia, with a left aortic arch and marked leftward displacement of the cardiac silhouette without tracheal deviation and a flattened and elongated left ventricular border. In addition, abnormal interposition of the lung tissue caused a lucent area between the aorta and the pulmonary artery and between the inferior myocardial wall and the left hemi-diaphragm. A 12-lead electrocardiogram showed coronary sinus rhythm with a heart rate of 65 beats per minute and a slightly right deviation of the heart axis (Figure 1). The transthoracic echocardiography showed a left laterally displaced left ventricle (LV) apex with a swinging motion, a globe-shaped heart, bulbous ventricle; and in the parasternal long axis view, paradoxical septal motion consistent with cardiac hypermobility

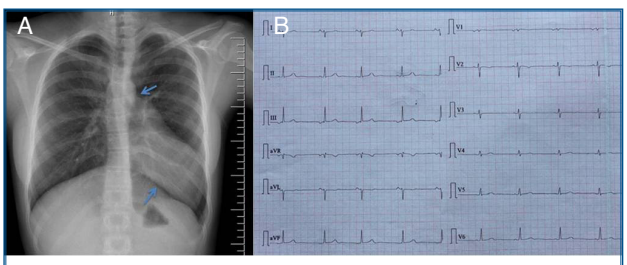

Figure 1. (A) Posterior-anterior chest x-ray demonstrating levoposition of the cardiac silhouette, obscured right heart border, and prominent main pulmonary artery along with midline trachea. There is a radiolucency between both the aortic knob and pulmonary conus and the inferior cardiac border and the left hemi-diaphragm; **(B)** electrocardiogram demonstrating rightward axis, low voltage with coronary sinus rhythm.

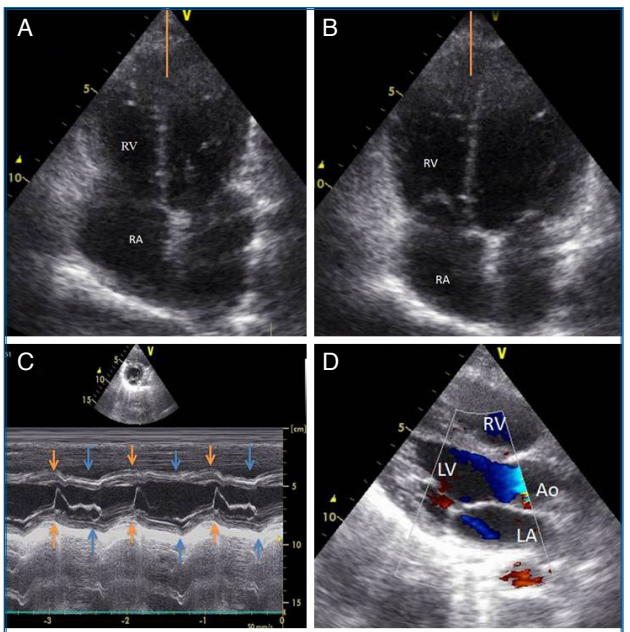

Figure 2. Transthoracic echocardiography demonstrating apical four-chamber view: left ventricular apex showed a swinging motion in **(A)** diastole and **(B)** systole; **(C)** M-mode imaging through the interventricular septum and mitral valve showing paradoxical septal motion consistent with cardiac hypermobility; **(D)** parasternal long-axis view demonstrating mild mitral valve regurgitation.

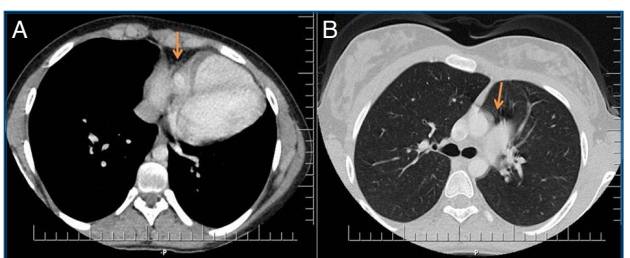

Figure 3. (A) Computed tomography (CT) of the chest confirms the levoposition of the heart and shows almost complete pericardial agenesis, with subtle pericardial remains overlying the right atrium; **(B)** CT demonstrating the interposition of the lung tissue between the aorta and pulmonary artery.

(Videos 1* and 2*). M-mode imaging through the interventricular septum also showed the paradoxical septal motion. A mild mitral valve regurgitation with normal valvular morphology was observed (Figure 2). The estimated LV ejection fraction was 72%. A chest computed tomography revealed an almost complete absence of pericardium on the left side with a small layer of the pericardium on the right atrium. The heart was grossly shifted leftward and posteriorly owing to enhanced mobility. The interposition of the lung tissue was seen between the ascending aorta and the pulmonary trunk (Figure 3). The constellation of findings was consistent with the diagnosis of a congenital absence of the left pericardium. As the patient did not have any additional cardiac or extracardiac pathology, chest pain, and exertional dyspnea or syncope, she was advised to have yearly follow-up without any further intervention.

Informed consent was obtained from the patient and her parents for the publication of the case image and the accompanying images.

*Supplementary video files associated with this article can be found in the online version of the journal.