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Coronary artery aneurysm in Kawasaki disease.

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Congenital Absence of Pericardium

Clinical summary:

A 16 year old girl presented to the emergency department with orthopnea and exercise intolerance for 1 week. Electrocardiogram (ECG) at that time showed rightward deviation of the cardiac axis, with nonspecific T wave inversion in lateral precordial leads and flattening in inferior leads. She was discharged from the emergency department with cardiology follow-up. Subsequent echocardiogram demonstrated that “the heart is in a funny position in the chest”, and CT of the chest was requested to evaluate for a mediastinal mass causing leftward shift of the heart.

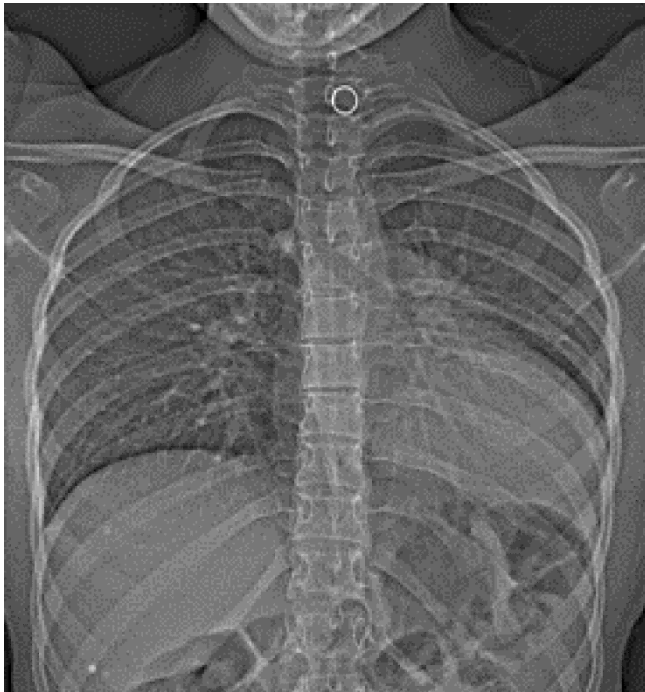


Figure 1: Scout image from CT chest demonstrates obscuration of right heart border by the spine due to levoposition of the heart.



Figure 2: Parasternal long-axis transthoracic echocardiogram image showing posteriorly directed apex and unusual imaging windows.

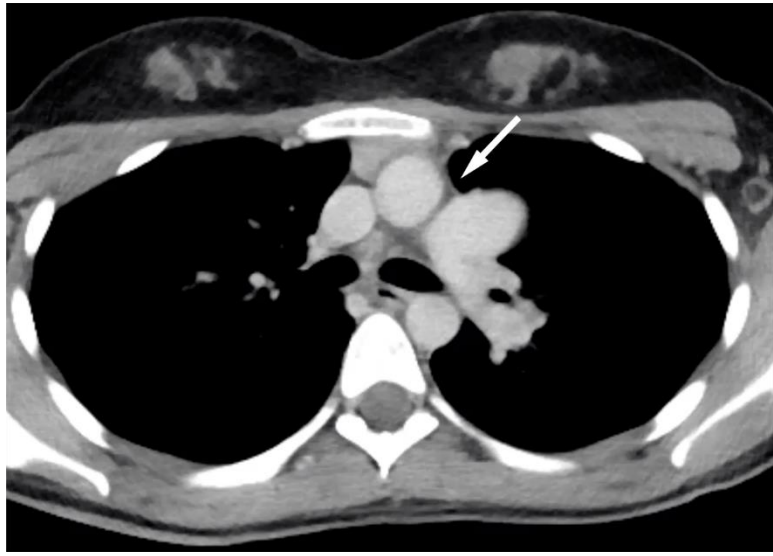


Figure 3a

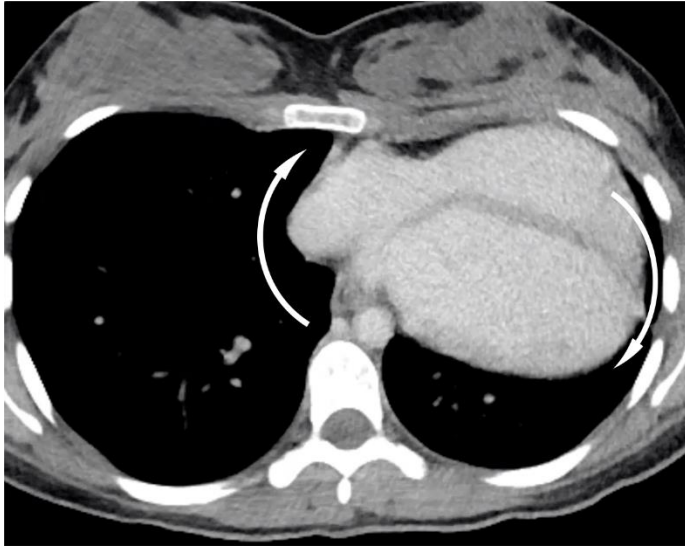


Figure 3b



Figure 3c

Figure 3a, 3b and 3c: Two axial and one coronal image from contrast enhanced chest CT demonstrates interposition of lung tissue in areas of absent pericardium between the aorta and PA (straight arrows) and marked levoposition of the cardiac silhouette (curved arrows)

Differential diagnosis:

Acute coronary syndromes, cardiac aneurysms, tumors of the lung or heart, mitral valve disease, atrial septal defects, pulmonic stenosis, idiopathic dilation of pulmonary artery, and hilar lymphadenopathy.

Discussion:

The normal pericardium is an avascular sac consisting of two layers: an outer fibrous layer and an inner serosal layer. Serous fluid normally occupies the space between the two layers.

Congenital absence of pericardium (CAP) is a rare condition that results from a failure of the pleuropericardial membranes to fuse completely on one or both sides. CAP has an incidence of less than 1 in 10,000 [1]. Left-sided defects are the most common with a prevalence of 70% of all pericardial defects [2]. 30–50% of patients with congenital absence of pericardium have other congenital abnormalities, including atrial septal defect, patent ductus arteriosus, and tetralogy of Fallot [3].

CAP is generally benign but can be confused with other pathologic conditions on imaging. The characteristic findings on chest imaging include a “snoopy dog” appearance of the cardiac silhouette, and a teardrop appearance with a bulbous ventricle on the chest radiograph (CXR). CXR commonly shows marked levoposition of the cardiac silhouette, loss of the right heart border, prominent pulmonary artery, and lung tissue between the diaphragm and inferior border of the heart [4]. Scout image from CT demonstrates loss of right heart border with levoposition of heart (Figure 1). TTE features of CAP include posterior orientation of the apex, cardioptosis in the parasternal long-axis view [5] (Figure 2). CT imaging demonstrates interposition of lung tissue between the heart and diaphragm and between the aorta and the main pulmonary artery (Figures 3a, 3b and 3c).

Patients with partial pericardial defects can be symptomatic with chest pain noted in one-third of patients [6]. They are also at risk of complications resulting from strangulation of any herniating cardiac structures. On the contrary, complete absence of pericardium does not commonly cause complications or symptoms.

Although CAP is a rare congenital abnormality, most cases are found incidentally and it is important for clinicians to recognize the constellation of abnormal clinical and imaging findings for accurate diagnosis.

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