A case of congenital complete absence of the pericardium

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A 20-year-old male was evaluated for the complaint of atypical chest pain. His physical examination was normal except for the notification of left lateral displaced apical impulse at the left lateral position. Electrocardiogram demonstrated normal sinus rhythm at 78 bpm with incomplete right bundle branch block. The posteroanterior chest X-ray was found to be normal. Transthoracic echocardiography was performed to clarify the cause of the displaced apical impulse. During transthoracic echocardiographic examination, an apical four-chamber view could be obtained by means of lateral and slightly superior positioning of the probe, almost near the subaxillary region, that was well-matched with the physical examination. For further evaluation, a cardiac computed tomography (CT) scan was carried out. This revealed that the cardiac axis was rotated leftward and the left lung tissue herniated between the ascending aorta and the main pulmonary artery (Fig. 1) as a pathognomonic sign of the complete absence of the pericardium (Wang ZJ et al. CT and MR imaging of pericardial disease. Radiographics, 2003; 23: 167–180). We followed up this case without surgical prophylactic intervention, given its excellent prognosis with no risk of fatal myocardial herniation and strangulation, which can be seen in patients with partial absence of the pericardium.

Figure 1. A–D. Contrast-enhanced chest computed tomography images; asterisk — herniated left lung tissue between ascending aorta and main pulmonary artery

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