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Case Report

A case of congenital total pericardial agenesis mimicking pneumothorax findings

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ABSTRACT

Congenital pericardial agenesis has been reported as usually asymptomatic cases, operated on for other reasons and found incidentally or in autopsy reports. Herein, we present a case of congenital total pericardial agenesis mimicking the findings of pneumothorax and incidentally detected in a uniportal video-assisted thoracic surgery (VATS). Total agenesis of the pericardium, which is very rare and usually asymptomatic, has a very good prognosis and does not require additional treatment unless it causes complications. It may mimic pneumothorax findings.

Keywords: pericardial agenesis, uniportal VATS, pneumothorax, congenital

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Introduction

Congenital agenesis of the pericardium is a very rare cardiac pathology [1,2]. It develops as a defect in the form of partial or total absence of the pericardium as a result of the defective separation of the pleura and pericardium joining in the midline, with an incidence of <1/10,000 [3,4]. Two cases and one case were reported in two large autopsy series of 13,000 and 14,000 patients [4,5]. The diagnosis of this anomaly, which is mostly asymptomatic, is usually made during autopsy or surgical intervention. Therefore, the true incidence is not known. The male gender is dominant, and the male–female ratio is 3:1. Familial transmission is rare [6,7].

This congenital defect manifests with complete or partial pericardial agenesis. A left-side pericardial defect is detected 70% more frequently, a right-side defect is detected in 14% of cases, and a complete bilateral pericardial defect is detected in 7-9% of cases [5,8,9]. The prognosis of total pericardial agenesis is better without mechanical complications [10]. A case of incidentally detected complete pericardial agenesis mimicking the radiological findings of pneumothorax is presented in this study.

Case Report

A 27-year-old man presented to our clinic with complaints of shortness of breath and chest pain. A chest X-ray showed a pneumothorax line on the left. The air-fluid level in the costadiaphragmatic sinus was pushed to the left diaphragm, with enlarged intercostal distances on the left and left retraction of the heart. Figures 1a,b shows the preoperative and postoperative chest X-ray. Left posterior lower lobe and lingula bulla, pneumothorax, and minimal pleural effusion were observed on computed tomography of the thorax. Cardiac herniation with a complete deviation of the heart to the left was considered (Figure 2). The patient's smoking history was 10 packs/year. No pathology was detected in blood biochemistry and laboratory tests. The pulse was 70/min, with normal sinus rhythm on ECG. The arterial blood pressure was measured as 120/70 mmHg. No additional disease was determined in the patient's past or family history. Surgery was recommended for the patient, who underwent tube thoracostomy and was found to have a giant bulla with no lung expansion. Wedge resection of the posterior lower lobe and lingula bullae (bulla resection) was performed with left uniportal video-assisted thoracic surgery (VATS). The exploration of the intraoperative thoracic organs showed an absence of pericardium (Figure 3). The opinion of the perioperative cardiovascular surgery team was received, and additional intervention was not considered. Congenital cardiopulmonary pathology was not detected in this case, which was detected incidentally, except for congenital bullae. The patient, who did not develop postoperative complications, was discharged on the 5th day. The retrospective findings included EF: 65%, PAP: 28 mmHg, and the absence of pericardium in the right and left heart. Other cardiac evaluations and parameters were normal on the echocardiogram performed at the postoperative 3rd-month follow-up. The patient has been followed up without problems in the 6th postoperative year. Written informed consent was obtained from the patient for the publication of his data.

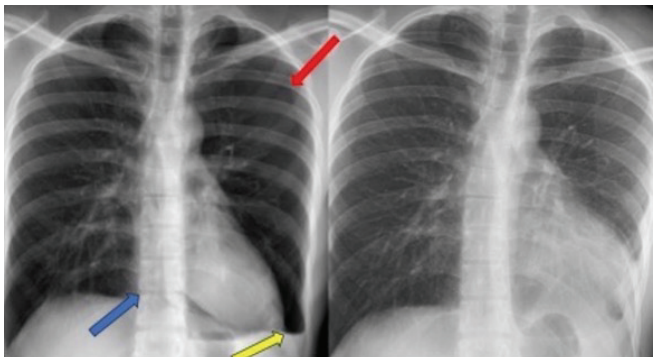

Figure 1. Preoperative chest X-ray showing enlarged intercostal spaces (red arrow), mediastinal shift (blue arrow), air-fluid level (yellow arrow) **(a)**, postoperative chest X-ray showing mediastinal shift **(b)**.

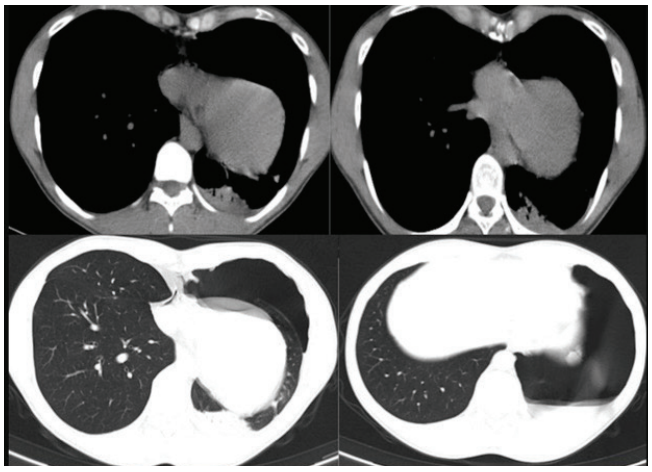

Figure 2. Thorax CT images showing the complete deviation of the heart.

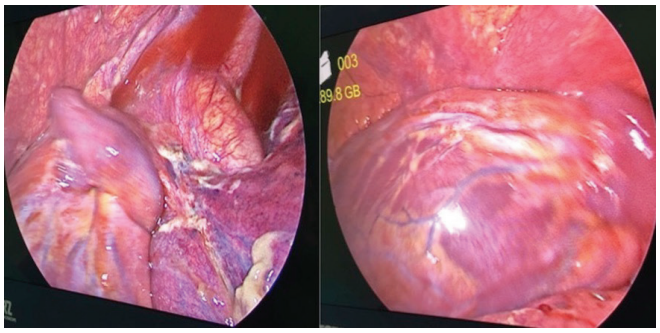

Figure 3. Operative images showing the absence of pericardium.

Discussion

The congenital absence of the pericardium is a rare malformation, characterized by the absence of the fibroserous membrane surrounding the heart. Inadequate blood supply to the pleuropericardium causes defective development of the pleuropericardial membranes because of early atrophy of the left main cardiac vein, causing pericardial insufficiency [8]. The absence of pericardium on the left side is the most common (70%), and the complete bilateral absence of pericardium (7-9%) is rare [5,8]. Although congenital pericardial agenesis is reported as an isolated pathology, it can be detected together with other congenital cardiac and pulmonary anomalies in 30% of cases [10]. No congenital cardiopulmonary pathologies other than congenital bullae were observed in our case, and isolated complete absence of the pericardium was detected.

Pericardial agenesis is usually asymptomatic and can be detected incidentally in autopsies or cardiothoracic operations. Rarely, clinical manifestations such as stabbing chest pain, shortness of breath, palpitations, and dizziness have been reported [8]. We think that the complete pericardial agenesis detected in this case was asymptomatic and the complaints of dyspnea and chest pain were related to acute pneumothorax. Only the left side was evaluated with VATS after the absence of pericardium on the left side and the absence of pericardium on both sides was detected on the postoperative echocardiogram. The retrospective re-evaluation with computed tomography of the thorax supported the findings. Therefore, the case was evaluated as complete pericardial agenesis. Right bundle branch block with sinus bradycardia, prominent P waves, and weak R waves have been reported on ECG in patients with pericardial agenesis [11]. A case with T-wave changes has also been reported [12]. The ECG was in normal sinus rhythm, and no pathological changes were detected, in our case.

Atelectasis may develop in pneumothorax because of the collapse of the lung, with possible retraction of the heart to the same side. There is an indication for emergency intervention because it presents a serious life-threatening situation. Ipsilateral retraction of the heart because of pulmonary atelectasis was interpreted as a sign of pneumothorax, and pericardial pathology was not considered, in our case. Although the lung was expanded with tube thoracostomy, no improvement in the cardiac shift was observed. In such pleural pathologies, if no improvement occurs in cardiac shift despite the expansion of the lung with the necessary intervention, the rare condition of pericardial agenesis should be considered.

The prognosis of complete pericardial agenesis without mechanical complications is very good. However, tricuspid regurgitation because of cardiac herniation, stretching of the chordae, myocardial strangulation, ischemia, atrial septal defect, mitral valve prolapse, and sudden death have been reported in the partial absence of pericardium [1,13-15]. The prognosis was good in our case, and the patient has been followed for 6 years without any problems.

In conclusion, no pathognomonic finding exists on clinical, radiological, ECG, and echocardiogram examination for the diagnosis of total agenesis of the pericardium, which is rare and usually asymptomatic. As long as complete pericardial agenesis does not cause any complications, it should be followed without the need for additional surgical intervention. Notably, it may mimic pneumothorax findings.

Declaration of conflicting interests

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Authors' contributions

HU; Data curation, formal analysis, project administration, resources, writing, review and editing, MA, NE,MRÇ; resources, co-writing, review, editing.

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