






Incidentally Detected Pericardial Defect in a Patient with Pneumothorax as Confirmed on Video-Assisted Thoracoscopic Surgery

흉강경 수술로 확인한 우연히 발견된 기흉을 동반한 심막결손

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Congenital defects of the pericardium, which are generally asymptomatic, are rare disorders characterized by complete or partial absence of the pericardium. Here, we report a rare case of a 19-year-old male who was incidentally diagnosed with congenital absence of the left pericardium during examination for symptoms of pneumothorax. Chest radiography and CT revealed a collapsed left lung without any evidence of trauma, no unusual findings of free air spaces along the right side of the ascending aorta, heart shifted toward the left side of the thorax, and a shallow chest. Subsequent thoracoscopy confirmed the absence of the left pericardium and displacement of the heart toward the left thoracic cavity. We further discuss the correlation between radiologic images and surgical findings of a congenital pericardial defect associated with spontaneous pneumothorax.

Index terms Pericardium; Pneumothorax; Thorax; Thoracoscopy; Computed Tomography, X-Ray

INTRODUCTION

Pericardial defects are rare disorders and are classified as either acquired or congenital (1). Congenital defects of the pericardium can be partial or complete, and complete



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absence of the pericardium is more common. Most cases of congenital pericardial defects (CPDs) are asymptomatic and diagnosed incidentally (1, 2). Chest radiography, electrocardiography, and CT can provide valuable evidence for diagnosis (3). Herein, we presented the case of a 19-year-old male who showed the symptoms of pneumothorax and was diagnosed with CPD through chest radiography and CT. Furthermore, shallow chest was observed to occur concomitantly with CPD. Incidental diagnosis of CPD following the symptoms of pneumothorax is rare; moreover, this diagnosis was surgically confirmed by thoracoscopy. This case demonstrated radiologic findings of CPD associated with pneumothorax and was confirmed by subsequent surgical findings.

CASE REPORT

A 19-year-old male was referred to our emergency room for left-sided chest pain. He had a history of left-sided spontaneous pneumothorax 1 year prior, for which he was treated by chest tube drainage, and his symptoms were relieved. Except that, other previous medical history and family history were unremarkable. Initial chest radiograph showed a radiolucent area with the absence of vascular marking and sharp visceral pleura in the left upper lung zone, which indicated pneumothorax (Fig. 1A). There was no definite abnormality in the bony thorax and no history of trauma; thus, the physician regarded the pneumothorax to have possibly developed spontaneously. However, unlike the usual spontaneous pneumothorax, the cardiac silhouette in this case was displaced to the ipsilateral side (left) and the right heart border was obscured by the spine. Moreover, there were radiolucent bands on the right side of the ascending aorta and between the left hemidiaphragm and the base of the heart, which was suggestive of pneumomediastinum or pneumopericardium. After chest tube insertion, he underwent chest CT examination. The CT scan was obtained using a 64-row detector CT scanner (SOMATOM Definition AS, Siemens, Erlangen, Germany). The scan parameters were as follows: 100 kVp tube voltages, 100 mA tube current. Chest CT revealed several blebs in both upper lobe apical segments (Fig. 1B). There was no evidence of bony fracture in both the ribs and the sternum. The lung tissue was interposed between the aortic arch and the prominent pulmonary trunk. The heart was displaced to the left side and the thoracic cage appeared flat. The Haller index was 4.89, obtained by dividing the internal transverse distance of the thorax by the antero-posterior height at the most depressed segment of the thorax. The coronal reconstruction scan images revealed free air spaces located in the superior margin of the pericardial space along the ascending aorta, superior vena cava (SVC), and right atrium (Fig. 1C). Three-dimensional reconstruction with volume-rendering images showed abnormally decreased intercostal spaces of the anterolateral arc of the ribs (Fig. 1D) compared with the intercostal spaces of the posterolateral arc of the ribs. Unlike a typical case of pectus excavatum, the front chest wall appeared flat in this case. The patient was scheduled for video-assisted thoracoscopic wedge resection of the left upper lobe apical segment. During the surgery, we observed that the heart was displaced into the left thoracic cavity, and the epicardial fat and coronary arteries were exposed due to a lack of fibrous pericardium (Fig. 1E); thus, the patient was diagnosed with a pericardial defect.

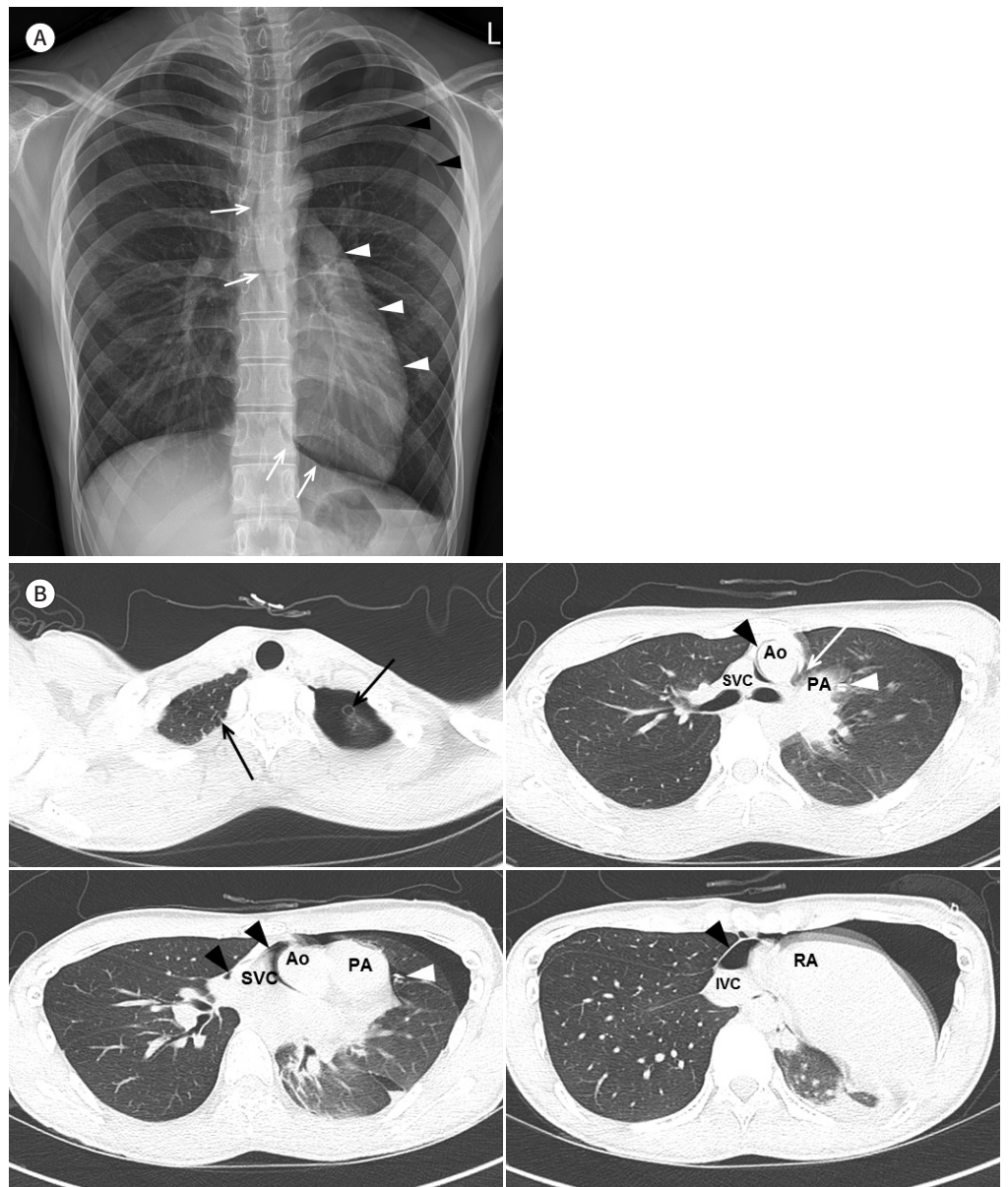
After surgery, he received conservative treatment for postoperative chest pain. Follow-up

Fig. 1. A 19-year-old male presented with pneumothorax and a congenital pericardial defect.

A. Posteroanterior chest radiograph shows a radiolucent area without vascular marking and a sharp white line (black arrowheads) in the left upper zone, suggesting pneumothorax. The left heart border appears straightened (white arrowheads), and free air spaces (arrows) are seen between the heart and the left hemidiaphragm and on the right side of the ascending Ao.

B. Chest CT scans with lung window settings show several blebs (black arrows) in both the upper lobe apical segments and left pneumothorax. Free air spaces (black arrowheads) are seen between the SVC and the ascending Ao and adjacent to the IVC and the RA. Abnormal interposition of the lung tissue (white arrow) is seen between the ascending Ao and the PA. It appears to be rotated clockwise away from the mediastinum. The Haller index is 4.89, obtained by dividing the internal transverse distance (235 mm) of the thorax by the anteroposterior height (48 mm) at the most depressed segment of the thorax. The heart appears to be shifted toward the left thorax. Chest tube was inserted (white arrowheads).

Ao = aorta, IVC = inferior vena cava, PA = pulmonary artery, RA = right atrium, SVC = superior vena cava



chest images showed no more pneumothorax or other complications, and the patient was discharged. Chest radiography was performed 1 month later, and it did not reveal any abnormally located free air spaces in the mediastinum or thoracic cavity because the air in the pleural space was absorbed through treatment (Fig. 1F). However, it still showed a straightened left

Fig. 1. A 19-year-old male presented with pneumothorax and a congenital pericardial defect.

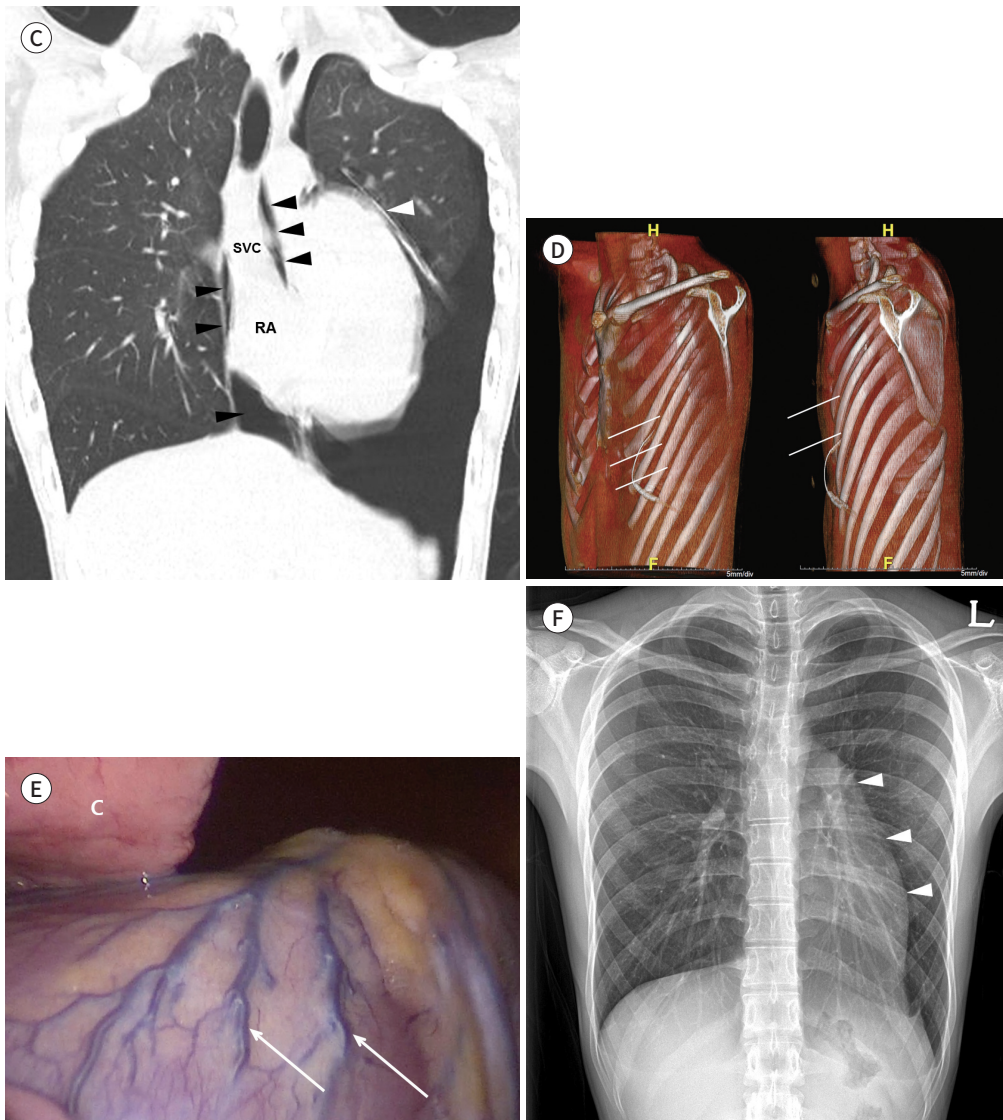
C. Coronal reformatted CT scan shows free air spaces (black arrowheads) located at the superior margin of the pericardial space along the ascending aorta, SVC, and RA. The chest tube (white arrowhead) is located in the left upper lung and pleural space.

D. Three-dimensional volume-rendering images show abnormally decreased intercostal spaces of the anterolateral arc of the ribs (white lines) relative to the intercostal spaces of the posterolateral arc of the ribs.

E. During video-assisted thoracoscopic surgery wedge resection, thoracoscopy shows the heart positioned toward the left thoracic cavity. Exposed epicardial fat and coronary arteries (arrows) are seen without the fibrous pericardium. Note collapsed lung in left side (C).

F. Follow up chest radiography obtained 1 month later shows improved abnormal air-collections. The left heart border still appears straightened (arrowheads).

RA = right atrium, SVC = superior vena cava



heart border and the obscured right heart border by the spine.

DISCUSSION

Here, we present a unique case of congenital absence of pericardium with unilateral spontaneous pneumothorax. Some cases of congenital absence of pericardium have been reported (1). However, our case shows incidental findings of CPD which was diagnosed following the symptoms of spontaneous pneumothorax. Additionally, pectus excavatum is a possible deformity associated with CPDs (1). In this case, we could find several evidences of CPD on chest radiography and chest CT. Some radiologic features were characteristic because of the associated pectus excavatum and accompanied by spontaneous pneumothorax. The chest radiography, CT, and surgical findings were well correlated. Additionally, subsequent surgical findings of CPD through thoracoscopy are very rare.

Pericardial defects can be classified as either acquired or congenital. Acquired defects of the pericardium are found after pericardiectomy due to constrictive and recurrent pericarditis (1). CPD is a rare clinical entity with an incidence of < 1 in 10000 (1). The most common type of pericardial defect is complete absence of the left pericardium (70%), followed by complete bilateral absence of the pericardium (9%). Right pericardial defect and partial absence of the pericardium are very rare (1, 4). Pericardial defects have a variable pathogenesis. In the 5th and 7th weeks of embryonic life, premature atrophy of the left common cardinal vein (left duct of Cuvier) may compromise the blood supply to the left pleuropericardial fold. This may lead to a failure to close the embryonic pleuropericardial foramen and cause pericardial defect (1, 3).

Thirty to fifty percent of patients with CPDs have associated anomalies, including atrial septal defect, bicuspid aortic valve, patent ductus arteriosus, and tetralogy of Fallot (1, 3). Other mesodermal defects such as chest and abdominal anterior wall abnormalities can be associated with CPDs. Nasser et al. (5) noticed the pectus excavatum deformity in 5 of 6 patients with congenital defects of the pericardium. Pectus excavatum is generally believed to have developed because of the overgrowth of sternocostal cartilage or ribs (6). In our case, the Haller index on chest CT was 4.81 and it corresponded to severe degree of pectus excavatum; A Haller index 3.5 or greater is considered a severe deformity (7). However, the case in our study showed a flat anterior chest wall, unlike the typical case of pectus excavatum which presents as a sunken appearance of the anterior chest wall. According to Chen et al. (8), imbalanced development of anterior and posterior thorax can lead to shallow chest and it can be causative factor of scoliosis. We speculated that the imbalanced development of anterior and posterior thorax might be related to the absence of pericardium which lead to shifting of mediastinum, and produced abnormally decreased intercostal spaces of the anterolateral arc of the ribs. This mechanism may be different from the pathogenesis of classical pectus excavatum.

Clinical manifestations of pericardial defects depend on the size of these defects. Patients with complete pericardial defects or small pericardial defects usually do not have symptoms. Occasionally, a complete pericardial defect leads to increased heart mobility and can be a source of positional symptoms (feeling of heart beating against the chest wall and pain dyspnea). Patients with large pericardial defects may have atypical chest pain in up to 33% of the patients.

Life-threatening complications, such as incarceration of the left arterial appendage, ventricular herniation, and torsion of the great vessels, require surgical correction for immobilization of the heart (1, 9, 10). Left-sided pneumothorax with consequent protrusion of the heart into the left thoracic cavity was also reported, and our case showed similar clinical features and surgical findings (9). Electrocardiograms are usually normal, and our case also showed a normal electrocardiogram. In some patients with complete absence of the left pericardium, poor R-wave progression can be seen on electrocardiogram because the precordial transitional zone is displaced leftward (1, 9).

The imaging findings of CPD on chest radiography were leftward-shifted cardiac silhouette, straightened left heart border, and elongated left heart border (Snoopy sign). Mostly, the right heart border is not visible as it is obscured by the spine, and radiolucent bands can be seen between the aortic knob and the main pulmonary artery (1). These signs were seen on our follow-up chest radiograph, and particularly, the finding of the concealed right heart border was suggestive of a CPD associated with pectus excavatum. Unusually located free air spaces were seen on our initial chest radiograph on the right side of the ascending aorta, which revealed the margin of the ascending aorta and between the left hemidiaphragm and the base of the heart. The presence of free air was an additional finding of associated pneumothorax.

Typical findings of CPD on chest CT include abnormally located lung tissue between the aorta and the pulmonary artery or between the diaphragm and the base of the heart. Leftward cardiac displacement is also seen usually. Although the pericardium is usually identified on CT, it is not typically visualized on the CT of a patient with pericardial defect (1). Our case showed abnormal lung tissue located between the aorta and the pulmonary artery, however it was difficult to evaluate the presence of left pericardium because we did not use the ECG-gated CT and the motion artifact was so severe. In addition, free air spaces were also seen along the SVC, inferior vena cava, and between the aorta and the SVC, which suggests abnormally located air displaced from the left pleural cavity associated with the underlying pericardial defect. Pneumothorax combined with CPD resulted in these CT findings, which correlated with the chest radiograph findings.

In summary, we reported a case of CPD that was incidentally found with pneumothorax and diagnosed using radiographs and CT. It was confirmed by thoracoscopy and the surgical findings correlated well with multimodality radiologic findings.

Author Contributions

Conceptualization, K.E.; investigation, all authors; supervision, K.E.; visualization, C.H., K.E., J.S.; writing—original draft, C.H., K.E.; and writing—review & editing, L.K., K.M.S.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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흉강경 수술로 확인한 우연히 발견된 기흉을 동반한 심막결손

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선천성 심막결손은 대부분의 환자가 무증상을 보이는 드문 질환으로 전체 혹은 부분 심막결손으로 나타난다. 본 논문에서는 기흉 증상으로 인해 우연히 좌측 선천성 심막결손을 진단받은 19세 남성 환자를 보고하고자 한다. 일반 흉부 X선 사진 및 컴퓨터단층촬영에서 외상의 흔적이 없는 무기폐, 상행대동맥의 우측에 비정상적으로 위치한 공기, 좌측 흉부로 전위된 심장, 그리고 납작한 흉곽이 보였다. 뒤이은 흉강경검사서 좌측 심막결손과 왼쪽 흉강으로의 심장 전위가 확인되었다. 이는 영상의학적 소견과 수술적 소견이 잘 일치하는 자발성 기흉이 동반된 선천성 심막결손에 관한 보고이다.

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