Case Report

# Complete Left-sided Pericardial Defect in a Lung Cancer Patient Undergoing Pneumonectomy without Closure of the Defect

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A 61-year-old asymptomatic man underwent a left pneumonectomy for Stage IIIA lung cancer. At thoracotomy, the pericardium was found to be completely absent; however, we did not close the defect. Although the heart was rotated toward the left pleural cavity in the postoperative chest computed tomography (CT), the postoperative course was uneventful, and the patient has remained asymptomatic for 7 months, since the resection. We reviewed the preoperative chest CT, which showed the heart extending unusually to the left, but the pericardial defect was not evident. Complete pericardial defects usually do not endanger the lives of patients, and if the patient is asymptomatic, surgical repair of the defect may be unnecessary even during a left pneumonectomy.

**Key words:** congenital pericardial defect, lung cancer, pneumonectomy

### Introduction

A congenital pericardial defect is a rare anomaly that is seldom diagnosed. Its incidence is reported in about 1 of 14,000 autopsies<sup>1)</sup> and in 15 of 34,000 cardiovascular surgical operations.<sup>2)</sup> Cases detected outside of a surgical intervention are even rarer, and only 400 cases have been reported in the literature.<sup>3, 4)</sup> To the best of our knowledge, this is the first report of a lung cancer patient with a complete, left-sided congenital pericardial defect, who underwent ipsilateral pneumonectomy. How to deal with this defect in such a patient has never been reported, so without experience or information on how to proceed, we left the defect as it was, and the patient recovered

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uneventfully. He has been well for 7 months since the surgery.

## **Case Report**

A 61-year-old man was referred to a community hospital for further examination for chest X-ray abnormality on a routine medical check-up (Fig. 1). Computed tomography (CT) of the chest demonstrated a 4.4 cm tumor in the left upper lobe that was in direct contact with the mediastinal fat adjacent to the aortic arch (Fig. 2). Bronchoscopic biopsy revealed non-small cell lung cancer. At thoracotomy, a complete left-sided pericardial defect was unexpectedly discovered (Fig. 3). The anterior free edge was fused to the diaphragm, and the posterior free edge was fused to the mediastinal pleura covering the aorta. The tumor extended to the adjacent mediastinal fat, and the peribronchial metastatic lymph nodes directly invaded the left main pulmonary artery. We performed a left pneumonectomy and systemic lymph node dissection and did not close the pericardial defect. The pathological diagnosis was primary large cell carcinoma of the lung, T3N1M0, stage IIIA.



Fig. 1 Chest X-ray of a mass shadow in the left upper lung field adjacent to the mediastinum.



Fig. 2 Computed tomographic scan of a tumor shadow in the left upper lobe in direct contact with the mediastinal fat adjacent to the aortic arch (arrow).

The postoperative course was uneventful, and the patient was discharged 9 days later. He underwent adjuvant chemotherapy and was well without any signs of recurrence at 7 months after resection.

We reviewed the preoperative electrocardiogram and chest CT. The electrocardiogram was normal. Preoperative chest CT showed the heart extending unusually to the left, but the pericardial defect was not evident. After the pneumonectomy, the heart rotated clockwise around the venae cavae (**Fig. 4**); however, the postoperative course was uneventful, and the patient has remained asymptomatic for seven months since the resection.

#### **Discussion**

A congenital pericardial defect is a rare entity and is believed to result from incomplete development of either the transverse septum or of the pleuropericardial folds.<sup>5)</sup> The defect is either complete or partial, occurring more often on the left than on the right side.<sup>4, 6–9)</sup> Ellis et al. summarized the majority of the defects reported in the literature, and 78%, were on the left side.<sup>6)</sup> Perna et al. suggested that congenital left-sided pericardial defects were due to premature atrophy of the left duct of Cuvier, which resulted in incomplete development of the left

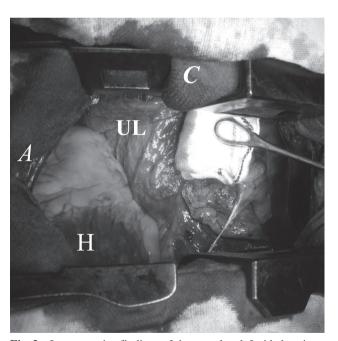


Fig. 3 Intraoperative findings of the complete left-sided pericardial defect immediately after thoracotomy No pericardium covering the heart (H) can be observed. UL, left upper lobe; A, anterior side; C, cephalad side

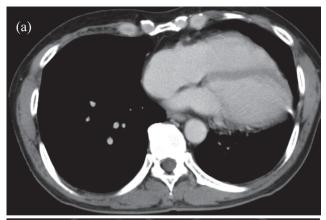




Fig. 4

- (a) Preoperative CT of the heart extending into the left pleural cavity, with no pericardial defect shown.
- (b) 3-month, post-operative CT of the heart rotated clockwise around the venae cavae.

pleuropericardial fold. The right duct of Cuvier normally develops into the superior vena cava, thereby ensuring adequate nutrition to the right plueropericardial membrane. Therefore, right-sided or bilateral complete defects are extremely rare.<sup>7,8)</sup>

Most patients with a congenital pericardial defect remain asymptomatic, and only a few defects are discovered by chance. Van Son et al. reported that 93.3% of patients with defects were asymptomatic.<sup>2)</sup> The symptoms attributable to the defect include nonspecific chest pain and awareness of heart beats, especially when patients are lying on the left side,<sup>6)</sup> and the beats are more frequent when the defect is partial. Although one-third of these defects is associated with congenital anomalies of the heart, lungs, chest wall, or diaphragm<sup>2, 10)</sup>; our patient had none of these.

Congenital pericardial defects are seldom diagnosed or encountered before autopsy. Small, left-sided defects do not usually show up as an abnormality on a chest X-ray unless there is hernation of the lung or heart. Ellis et al. reported that complete pericardial defects are suggested, when the chest X-ray reveals a leftward displacement of the heart, a long, prominent and distinct pulmonary artery segment, and a flattened left ventricular contour. Gassuner et al. reported that magnetic resonance imaging is useful in distinguishing complete and partial left-sided defects. In the present case, although the heart extended unusually to the left on the preoperative CT, we did not find it remarkable enough to make diagnosis, preoperatively.

Patients with a partial pericardial defect sometimes present with symptoms due to cardiac herniation and compression of the epicardial coronary arteries and are at risk of sudden death.<sup>1)</sup> When partial defects are diagnosed, it is necessary to resect a part of the pericardium and to leave it open, or to close it with a prosthetic membrane. In contrast, complete pericardial defects do not usually endanger the lives of patients, and if the patient is asymptomatic, surgical repair of the defect is not necessary.<sup>12)</sup> However, congenital complete left-sided pericardial defect patients who underwent ipsilateral pneumonectomy have never been reported in the literature.

We unexpectedly found a complete left-sided pericardial defect during ipsilateral pneumonectomy for lung cancer, and left the pericardial defect as it was. One reason for not repairing the defect was because it was complete and left-sided. Goldstraw et al. concluded that, in acquired pericardial defects from surgical pericardiectomy for lung cancer, the left-sided pericardial defect could be opened widely to allow free movement of the heart without strangulation, whereas, a right-sided defect required repair because the defect might cause cardiac herniation with consequent torsion of the great veins, resulting in cardiac arrest.<sup>13)</sup> Another reason for not repairing the defect was because it is too difficult to suture artificial or autologous material (such as the diaphragm or fascia lata) securely to the surrounding mediastinal surface and adventitia of the descending aorta. Yamaguchi et al. reported a congenital complete left-sided pericardial defect in one patient, who developed cardiac herniation after lobectomy despite patch closure of the defect, and emphasized the importance of secure closure.<sup>14)</sup> However, some researchers concluded that, for complete defects, nothing needed be done if the patient is asymptomatic.<sup>6, 12)</sup> In the present case, although the heart was rotated toward the left pleural cavity, the postoperative course was uneventful, and the patient has been asymptomatic for 7 months since surgery. Our experience and previous reports suggest that repair of a complete left-sided pericardial defect during ipsilateral pneumonectomy may be unnecessary, if the patient is asymptomatic.

## Acknowledgments

The authors thank Professor J. Patrick Barron of the International Medical Communications Center, Tokyo Medical University, for reviewing the manuscript. This work was supported in part by a Grant-in-Aid for Cancer Research from the Ministry of Health, Labour and Welfare, Japan.

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