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CASE REPORT

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Complete congenital pericardial defect found during S^{1+2} segment excision of the left upper lung by single-aperture thoracoscopy: a case report

Xiao Wang[1*], Jian Liu[1], Bichen Zhan[1] and Mingbo Gu[1]

Abstract

Background Congenital pericardial defect (CPD) is a rare congenital heart malformation with atypical clinical symptoms.

Case presentation The 69-year-old woman was referred because a 23-mm mixed ground glass shadow was found in the apicoposterior segment (S^{1+2}) of the left lung. The S^{1+2} segment excision of the left upper lung was performed under uni-portal fluorescence thoracoscopy. Intraoperatively, an unintentional complete absence of the pericardium was discovered, with the phrenic nerve running behind the sternum. To reduce the intraoperative stimulation of the heart and prevent postoperative friction of the bronchial stump against the left atrial appendage, the pulmonary hilum was not separated, and the intersegmental vein ($V^{1+2}b+c$) was not cut off. Given the patient's lack of symptoms and the low risk of cardiac hernia, no specific pericardial surgery was performed. The patient was well at the 1-month postoperative follow-up.

Conclusions This paper reports a case of complete pericardial defect accidentally found during left upper lung S^{1+2} segmentectomy under single-hole thoracoscopy, summarizes the main points of diagnosis and surgical precautions, and provides experience reference for similar clinical cases.

Keywords Congenital pericardial defect, Pulmonary segmentectomy, Pulmonary nodules, Thoracoscope, Case report

Background

Congenital pericardial defect (CPD) is a rare anomaly due to the congenital absence of the pericardium, with a prevalence of 0.007-0.015% at autopsy and 0.044% at surgery [1–3]. Of the CPD cases, approximately 9% were congenital complete pericardial defects (CCPD), with 70% involving left-side defects, 4–6% involving right-side defects, and 17% being diaphragmatic pericardial defects [4]. CPD is rare in clinics, without typical symptoms, and most cases are found inadvertently during lung or heart surgery [5]. As a result, most physicians do not know enough about the disease to diagnose and manage it properly. This report presents a case of a CCPD incidentally discovered during a single-aperture thoracoscopic S^{1+2} segment resection of the left upper lung.

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Case presentation

A 69-year-old woman was referred to the Department of Cardiothoracic Surgery of Anqing Municipal Hospital on March 12, 2024, because a 23-mm mixed ground glass shadow was found in the apicoposterior segment (S^{1+2}) of the left lung on the chest computed tomography (CT). The patient had a history of hypertension, and reported no family history of heart disease or lung cancer. Physical examination was unremarkable. Preoperative chest CT showed a mixed ground glass shadow with a maximum tumor diameter of 23 mm, and the consolidation tumor ratio (CTR) was about 39% (Fig. 1. A). The cardiac shadow was tilted to the left side of the chest (Fig. 1. B). A prolonged Q-T interval was observed on the electrocardiogram (ECG). Echocardiography showed that the cardiac function was maintained, but the left atrium was slightly enlarged (39 mm). The S^{1+2} segment excision of the left upper lung was performed under uni-portal fluorescence thoracoscopy (Figure S1). The incision length was about 4 cm, located at the 5th intercostal line of the anterior axillary line. The complete absence of the pericardium was observed during the operation, and the phrenic nerve ran behind the sternum (Fig. 2). The S^{1+2} segment of the left upper lung was excised. Invasive adenocarcinoma was diagnosed by rapid intraoperative examination. Lymph node dissection was performed.

In order to reduce the intraoperative stimulation of the heart and prevent postoperative friction of the bronchial stump against the left atrial appendage, the pulmonary hilum was not separated, and the intersegmental vein ($V^{1+2}b+c$) was not cut off. Postoperative pathological examination revealed invasive adenocarcinoma (pT1bN0M0, Stage IA2). The chest drainage tube was removed on the 2nd postoperative day. The patient was discharged 4 days after surgery. At the one-month postoperative follow-up, the patient was found to be in good condition, with no reported complications or discomfort.

Discussion and conclusions

CPD is a rare congenital heart malformation with atypical clinical symptoms. This report presented a case of unintentional discovery of a complete pericardial defect during S^{1+2} segment excision of the left upper lung by single-aperture thoracoscopy.

The premature regression of the ducts of Cuvier (common cardinal vein) during the embryonic period delays the growth of the thoracic pericardial folds [6]. Iijima et al. [4] summarized the CPDs found during lung operations, mainly lobectomy and total lung resection. The case reported here was a patient who underwent left upper lung excision S^{1+2} segment by single-aperture fluorescence thoracoscopy, during which complete pericardial absence was found. Previous literature has reported on congenital pericardial defect during cardiopulmonary surgery, but there are few reports on resection of left upper lung with S^{1+2} segment, and the relevant surgical experience is lacked.

Most patients with CPD have no clinical symptoms, while some patients have atypical precardiac tingling or left-side chest pain, dyspnea, and other symptoms [7, 8]. CT and magnetic resonance imaging (MRI) are important imaging bases for diagnosing CPD. Identifying the pericardium by CT and MRI depends on the epicardium and the adipose layer of the pericardium. Two-dimensional cardiography of real-time MRI images can show right ventricular enlargement, excessive cardiac activity, "teardrop" heart, and abnormal atrioventricular angle [9–11]. Other clinical tests, such as ECG, can indicate right axis deviation, complete right bundle branch block, right ventricular lumen, and ventricular septum enlargement

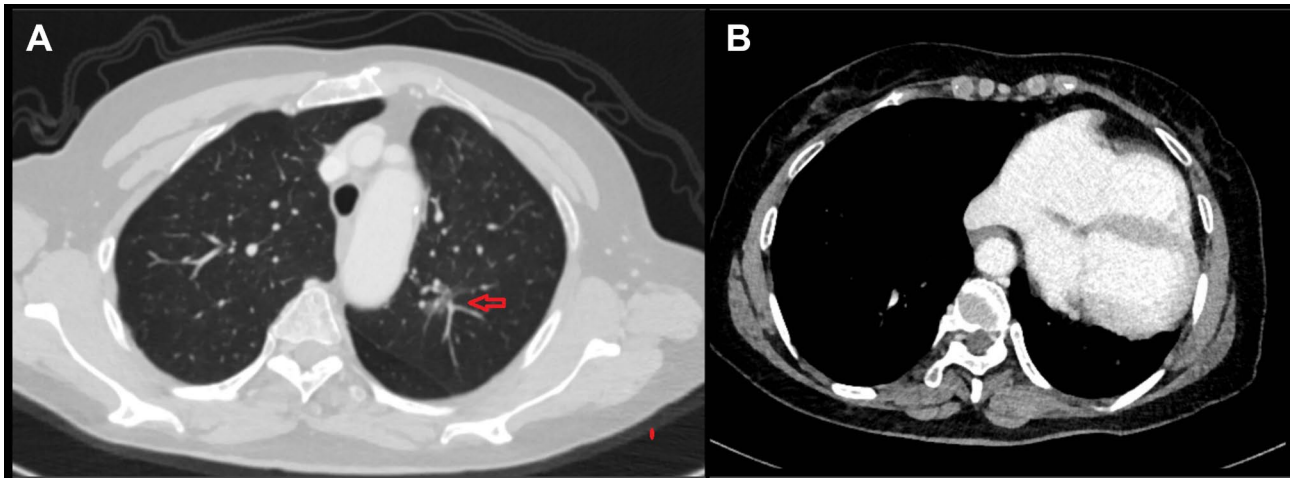

Fig. 1 Pulmonary CT images. (**A**) The pulmonary nodule (red arrows) was located in the S^{1+2} segment of the left upper lung. (**B**) The cardiac shadow was tilted to the left side of the chest

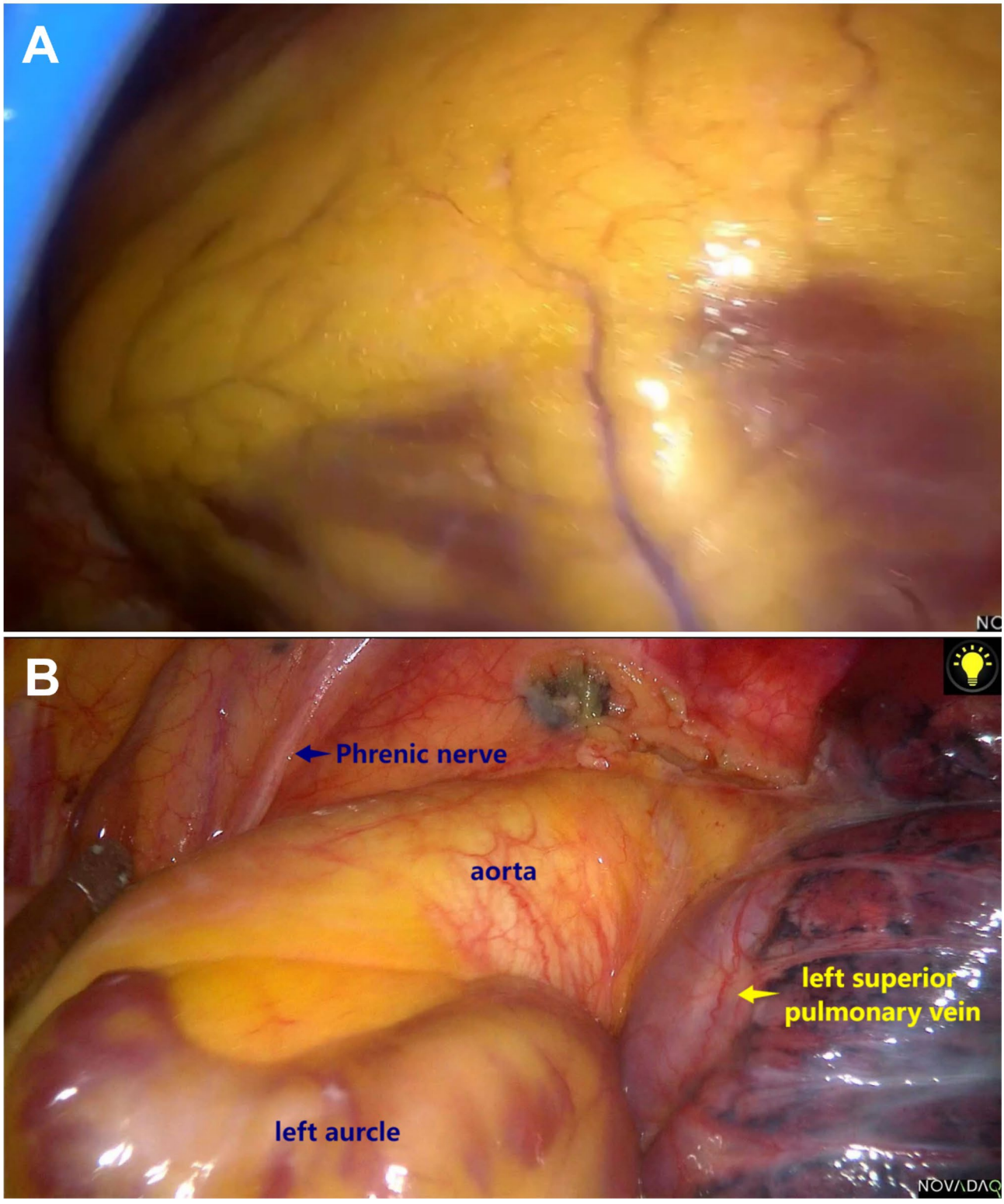

Fig. 2 **(A)** Intraoperative view showing a complete pericardial defect with the heart exposed; **(B)** Variation in the position of the phrenic nerve, located posterior to the sternum

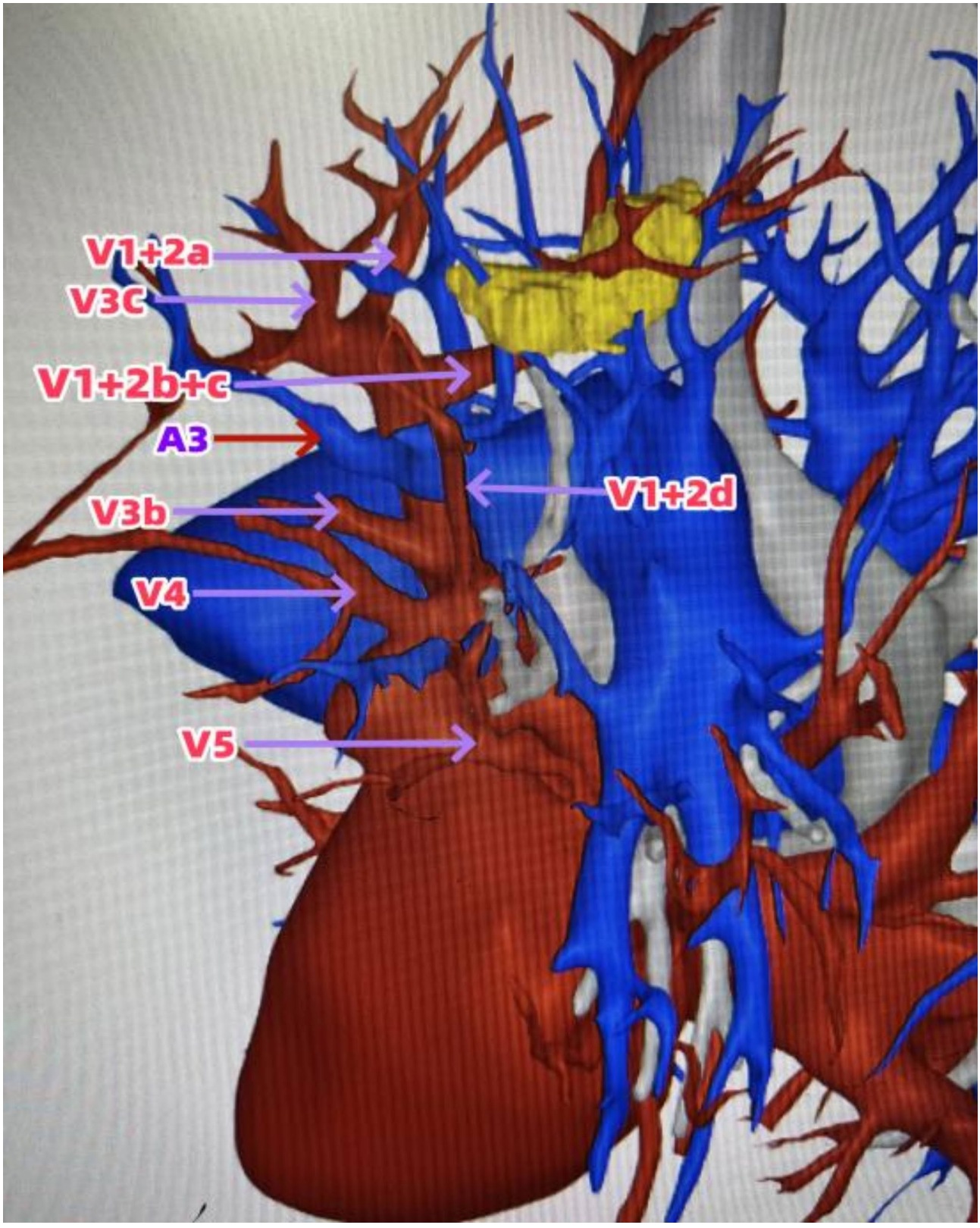

Fig. 3 3D reconstruction shows malformation of the left upper pulmonary vein, absence of V3a, V5 emanating directly from the left atrium

or have no clinical specificity [12]. In the case reported here, the patient had no symptoms before surgery.

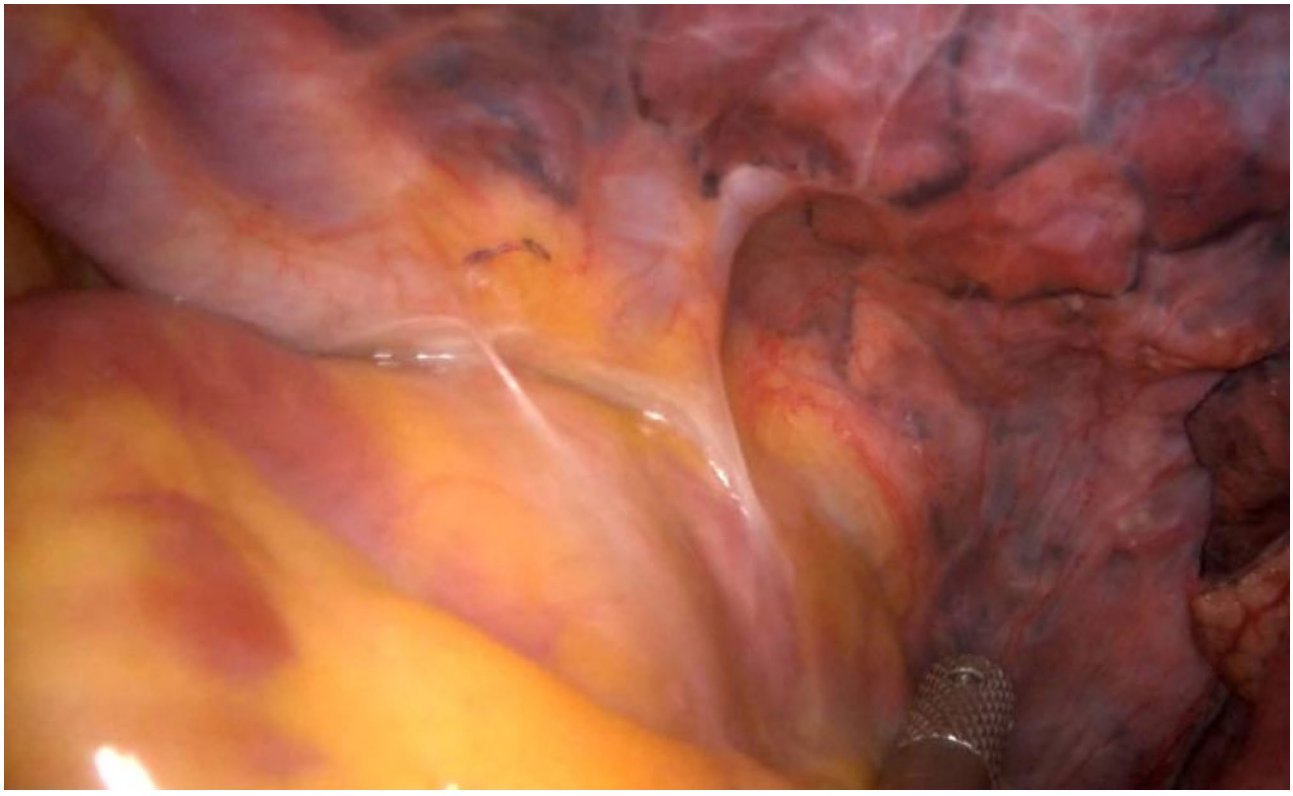

Fig. 4 The left upper pulmonary vein is thickened and dilated

Although the preoperative ECG in this patient indicated a prolonged Q-T interval, there was no diagnostic specificity.This patient's preoperative CT showed a deviation of the heart into the left thoracic cavity. However, the CT report did not mention the diagnosis of CPD. Although cardiac MRI is the gold standard for diagnosing pericardial defects [9, 13], it is not a routine test before lung surgery.

Most CPDs, especially CCPD, do not require surgical treatment. Only some cases of congenital partial pericardial defects (CPPD) have a risk of myocardial infarction and require surgical pericardium plasty [5, 14, 15]. In previous case reports, extreme left deviation of the heart was reported after excision of the upper left lung or of large lung cysts, resulting in angina pectoris and sudden death [16, 17]. Therefore, pulmonary surgery in patients with CPD needs to be performed with caution. According to the results of JCOG1211 in Japan, segmentectomy can effectively treat early lung cancer with diameter ≤ 3CM and CTR value less than or equal to 0.5. The patient had a mixed ground glass nodule with a diameter of 2.3CM and a CTR of 0.39, and was therefore suitable for segmental resection [18–20]. Still, the patient underwent resection of the S^{1+2} lung segment of the upper left lung by single-port thoracoscopy on the premise of ensuring complete resection of the nodule and sufficient surgical margin. During the operation, the left upper pulmonary vein in the anterior mediastinum was not dissected. Instead, the dissection started from the oblique fissure, and $A^{1+2}c$, $A^{1+2}a+b$, and B^{1+2} were successively dissected on the right dorsolateral ventral side (Figure S1). After dissection, the fluorescence was displayed, and S^{1+2} was resected along the segment plane from the dorsalateral ventral side with a closure device so that V^{1+2} and lung tissue were closed together (Figure S1). This method of non-separation of V^{1+2} has the following advantages. (1) It avoids the mechanical stimulation of the heart, which could lead to ventricular arrhythmia or even cardiac arrest. (2) The patient had left upper pulmonary vein malformation with multiple branches, and misjudgment of V^{1+2} was avoided(Fig. 3). (3) Reduces the risk of major bleeding due to venous injury. During the operation, the patient's left upper pulmonary vein was dilated and thickened, and the wall of the vein was thin. Meanwhile, frequent cardiac arrhythmias were seen when the heart was pressed to expose the pulmonary hilus, resulting in difficulty in exposing the left upper pulmonary vein(Fig. 4). There are advantages to left upper lung S^{1+2} resection in the presence of CCPD. (1) Dissociation and amputation of $A^{1+2}c$, $A^{1+2}a+b$, and B^{1+2} in oblique cleft results in less mechanical stimulation of the heart and less risk of intraoperative malignant ventricular arrhythmia. (2) The left upper pulmonary vein was retained during the operation, which separated the B^{1+2} stump from the left atrial

appendage to prevent perforation of the heart caused by mechanical wear of the bronchial stump after surgery. (3) Most of the volume of the left lung was preserved to prevent cardiac complications caused by excessive cardiac torsion after surgery. (4) The preserved upper left lung tissue was conducive to postoperative recovery. The difficulty lies in group 7 lymph node dissection, which requires compression of the heart and causes arrhythmia. Therefore, group 7 lymph node sampling was performed.

In conclusion, a case of CCPD was incidentally found during left upper lung S^{1+2} resection. The surgical experience was summarized, and it has certain reference significance for the intraoperative management.

This case only focuses on a single individual, with a small sample size and a serious lack of representativeness. As a result, the results of the study are difficult to be generalized to a wider population, and cannot reflect the manifestations and rules of diseases under different individual characteristics (such as differences in age, gender, region, and genetic background, etc.). There is a risk of overgeneralization, which greatly limits the universality of the research conclusions.

Abbreviations

CPD	Congenital pericardial defect
CCPD	Congenital complete pericardial defects
MRI	Magnetic resonance imaging

Supplementary Information

The online version contains supplementary material available at https://doi.org/10.1186/s13019-025-03390-1.

Supplementary Material 1

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None.

Author contributions
Jian Liu performed the surgery. Bichen Zhan and Xiao Wang participated in design. Mingbo Gu participated in the draft of the manuscript. All authors read and approved the final manuscript.

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Data availability
All data generated or analysed during this study are included in this published article.

Declarations

Ethics approval and consent to participate
This work has been carried out in accordance with the Declaration of Helsinki (2000) of the World Medical Association. This work was approved by the Medical Ethics Committee of Anqing Municipal Hospital (2024 – 134). Informed consent was obtained from the patient.

Consent for publication
Not applicable.

Competing interests
The authors declare no competing interests.

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