Isolated Unilateral Absence of Pulmonary Artery Associated with Contralateral Lung Cancer

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Unilateral absence of a pulmonary artery (UAPA) is a rare congenital anomaly that may present with various symptoms, depending on the nature and severity of other cardiovascular anomalies. Furthermore, contralateral lung surgery in patients with UAPA is extremely rare, and clinical experience is limited. This report describes a case of surgical treatment of contralateral primary lung cancer in a patient with isolated UAPA. A 56-year-old man was diagnosed with primary lung cancer accompanied by isolated UAPA on the contralateral side. He underwent meticulous cardiorespiratory function tests preoperatively. We performed a right lower lobectomy. Although in the immediate postoperative period, the patient suffered from a mild decline in his respiratory function, he recovered uneventfully. The present case shows that preoperative awareness of UAPA and meticulous perioperative management enable contralateral lung surgery to be performed safely.

Key words: 1. Unilateral absence of pulmonary artery 2. Lung neoplasms 3. Surgery 4. Perioperative care

Case report

A 56-year-old man presented with a mass shadow in the right lung field on a chest X-ray. Chest computed tomography (CT) revealed a mass measuring roughly 4 cm in the right lower lobe (Fig. 1A), and a 3-dimensional reconstructed CT image confirmed left unilateral absence of a pulmonary artery (UAPA) with hypertrophic bronchial arteries reflecting collateral circulation (Fig. 1B). After percutaneous needle biopsy, the tumor was identified as adenocarcinoma. In room air, arterial blood gas analysis (ABGA) yielded results of 92.5 mm Hg for PO2, 34.7 mm Hg for PCO2, and 97.4% for oxygen saturation (SaO2). Pulmonary function testing revealed a forced vital capacity (FVC) of 3.1 L (70% of predicted), a forced expiratory volume in 1 second (FEV1) of 2.2 L (64% of predicted), and a FEV1/FVC ratio of 0.71. Echocardiography revealed no cardiac anomaly and the right ventricular systolic pressure (RVSP) was 23.7 mm Hg. Based on the results of these tests, we planned thoracoscopic lobectomy. During the operation, immediately after applying single-lung ventilation, SaO2 decreased rapidly. Because this event repeated, conversion to open thoracotomy was required, and the planned lobectomy was completed uneventfully under double-lung ventilation. Postoperatively, in the resting state, SaO2 was 97%–99% with an oxygen supply of 3 L/min via a nasal prong and 88%–92% in room air. Even during simple daily activities, he...
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Fig. 1. (A) Chest CT shows a mass larger than 4 cm in the right lower lobe (arrow). (B) Three-dimensional reconstructed CT image shows a normal RPA and absence of the left pulmonary artery, as well as compensatory hypertrophy of the collateral systemic arteries (arrows). Lung cancer is noted in the right lower lobe (asterisk). CT, computed tomography; RPA, right pulmonary artery.

Table 1. Reports of unilateral absence of a pulmonary artery associated with contralateral lung cancer

| Author (year) | Gender/age (yr) | UAPA | Lung cancer | Preoperative diagnosis of UAPA | Diagnostic modality | Treatment | Pathologic diagnosis | Outcome |
|---------------|-----------------|------|-------------|-------------------------------|--------------------|-----------|---------------------|---------|
| Ito (2010) [7] | M/57            | Left | Right middle lobe | Missed | Perfusion scintigraphy | Surgery (lobectomy) | Adenocarcinoma | Survived |
| Zhang (2016) [2] | F/60            | Right | Left lower lobe | Missed | CT | Surgery (lobectomy) | Adenocarcinoma | Expired |
| Kim (2018) [a] | M/56            | Left | Right lower lobe | Confirmed | CT | Surgery (lobectomy) | Adenocarcinoma | Survived |

UAPA, unilateral absence of a pulmonary artery; M, male; F, female; CT, computed tomography.

[a]Current study.

Table 1. Reports of unilateral absence of a pulmonary artery associated with contralateral lung cancer

Complained of dyspnea, and his SaO₂ fell to 80%. However, the dyspnea gradually improved through supportive management alone, and he was discharged on postoperative day 22. At a 10-month follow-up visit, the dyspnea had largely resolved. Follow-up echocardiography and ABGA revealed mild pulmonary hypertension (RVSP of 46 mm Hg) and an SaO₂ of 97.6% in room air.

Discussion

UAPA is a rare congenital vascular deformity in which the pulmonary artery is completely absent [1]. UAPA may occur as an isolated anomaly or be accompanied by other congenital cardiovascular malformations [2]. The prevalence of isolated UAPA ranges from 1 in 200,000 to 1 in 300,000 adults [1]. In 1868, Fraenzel [3] described the first case of UAPA, and recently, Bockeria et al. [4] in 2010 reported a total of 419 cases. UAPA is often accompanied by cardiovascular anomalies, especially in pediatric patients [5,6]. Adult patients may develop symptoms such as hemoptysis, recurrent lung infections, and exertional dyspnea. However, 13%–15% of patients with UAPA are asymptomatic [1], and in such patients UAPA generally remains undiagnosed until incidentally detected by chest imaging studies, as occurred in the present case.

Surgical treatment of UAPA is considered when associated cardiovascular anomalies are present or the condition is symptomatic. In particular, contralateral lung lesions that require surgical treatment should be given special consideration. The occurrence of primary lung cancer in patients with UAPA is extremely rare, and experience with contralateral lung surgery in such patients is very limited. Seven cases of the surgical treatment of lung cancer in patients with UAPA have been reported in the English-language literature [2], but only 2 involved lung cancer on the contralateral side [2,7]. To the best of our knowledge, the present case is the third in which surgery was performed for contralateral lung cancer (Table 1). In cases of lung cancer accompanied by contralateral UAPA, treatment options should be selected...
carefully according to the patient’s cardiopulmonary status. First, not only spirometric data, but also pulmonary artery pressure and right heart function should be evaluated carefully. In cases of marginal cardiopulmonary function, a preoperative pulmonary perfusion scan or preoperative pulmonary artery occlusion test might be useful to determine the extent of lung resection. Second, if the patients’ performance status and the results of all tests are within normal ranges, even lobectomy may be feasible.

Intraoperatively, repeated decreases in SaO$_2$ will inevitably occur during single-lung ventilation. Although in the present case, it was possible to perform lobectomy using double-lung ventilation, we would suggest that an extracorporeal membranous oxygenator be prepared preoperatively to help cope with potential troublesome situations, such as dense adhesions or anatomical complexity.

Postoperatively, it is inevitable for some degree of pulmonary hypertension to develop due to hyperperfusion of the remnant lobes. In a previous case [2], the patient suffered rapidly progressing lung failure and died of right heart failure on day 2. Therefore, it is extremely important to prepare for these potential critical situations in the immediate postoperative period. All possible options to control pulmonary hypertension, such as close monitoring, strict input/output control, vasodilators, and nitric oxide should be available. In the long term, follow-up with echocardiography to monitor whether cor pulmonale develops is mandatory.

We performed curative anatomical resection for primary lung cancer and achieved a good outcome despite contralateral UAPA. This outcome may have been due to the patient’s relatively good cardiorespiratory function, preoperative awareness of contralateral UAPA, and meticulous postoperative management.

### Conflict of interest

No potential conflict of interest relevant to this article was reported.

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