Partial Anomalous Pulmonary Venous Connection Coexisting with Lung Cancer: A Case Report and Review of Relevant Cases from the Literature

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A 45-year-old man had an abnormal shadow in the right lung field on an annual screening chest X-ray. He was diagnosed with Stage IA (cT1bN0M0) lung cancer. Initially, we did not notice an anomalous vein on non-contrast computed tomography. However, we found that the right upper lobe bronchus branched from the lateral wall of the right main bronchial orifice, above the level of the common right upper lobe bronchus. Therefore, the bronchus was thought to be a tracheal bronchus. We carefully reevaluated the patient using three-dimensional computed tomography angiography. This technique showed that the anomalous right superior pulmonary vein drained into the azygos vein along the superior vena cava. These findings confirmed a partial anomalous pulmonary venous connection of the right upper lobe. We performed video-assisted thoracoscopic right upper lobectomy and mediastinal lymph node dissection for definitive treatment for lung cancer and partial anomalous pulmonary venous connection. No hemodynamic problems occurred in the postoperative course.

Keywords: partial anomalous pulmonary venous connection, lung cancer, three-dimensional computed tomography angiography

Introduction

Partial anomalous pulmonary venous connection (PAPVC) is a comparatively uncommon congenital vascular anomaly. PAPVC is frequently associated with congenital heart disease, such as atrial septal defect. However, PAPVC that is incidentally found on computed tomography (CT) in adults without heart disease is uncommon, and generally clinically insignificant. We report here a case of lung cancer coexisting with asymptomatic PAPVC, which was diagnosed with preoperative contrast three-dimensional computed tomography angiography (3D-CTA).

Case Report

A 45-year-old man, who never smoked, was referred to our hospital for evaluation of an abnormal shadow in the right upper lung field on an annual screening chest X-ray taken in January 2015 (Fig. 1A). His past and family histories were unremarkable. He likes to run, and he had finished a full marathon several times. A physical examination showed no abnormalities and no lymphadenopathy. There was no evidence of vascular shunt sounds on the chest and neck. Routine laboratory tests were within the normal range. A chest X-ray showed a slightly high-intensity shadow in the right upper lung field. A chest non-contrast CT scan showed an irregular, circumscribed, ill-defined,
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partly solid nodule in the right upper lobe, measuring 2.5 cm in diameter. There was no evidence of enlargement of the lymph nodes. We could not detect any distant metastasis by systemic examinations, including brain magnetic resonance imaging and 18F-fluorodeoxy-glucose positron emission tomography/CT.

We planned bronchofiberscopy to confirm the histological diagnosis of lung tumor. On careful review of the patient’s non-contrast CT images before bronchofiberscopy, we observed that the right upper lobe bronchus (RULB) was branching from the lateral wall of the right main bronchial orifice, above the level of the common RULB. Therefore, the bronchus was thought to be a tracheal bronchus (Fig. 1B). The length of the RULB was approximately 3 cm, which is longer than the common variation, and segmental bronchi of the right upper lobe had two branches. We also observed that the right upper lobe pulmonary vein (PV1-3) did not connect to the left atrium, and the right middle lobe pulmonary vein drained into the left atrium as normal. We performed a contrast CT

Fig. 1  (A) Chest X-ray shows an abnormal shadow in the right upper lung field. (B) Chest CT shows the RULB branching from the lateral wall of the right main bronchial orifice, as a tracheal bronchus. CT: computed tomography; RULB: right upper lobe bronchus

Fig. 2  (A) Frontal view of 3D-CTA showing PV1-3 draining into the azygos vein along the superior vena cava. (B) Back view of 3D-CTA showing the RULB branching from the lateral wall of the right main bronchial orifice, as a tracheal bronchus. The length of the RULB is approximately 3 cm, which is longer than the common variation. 3D-CTA: three-dimensional computed tomography angiography; RULB: right upper lobe bronchus
scan and 3D-CTA, which showed that the PV1-3 drained into the azygos vein along the superior vena cava, and it confirmed PAPVC of the right upper lobe (Figs. 2A–2B and 3A). Pathological examination of right transbronchial lung tumor biopsies confirmed the diagnosis of adenocarcinoma. The patient was diagnosed with Stage IA (cT1bN0M0) lung cancer coexisting with of PAPVC in the same lobe. Spirometry showed that the vital capacity was 5010 mL and forced expiratory volume in 1.0 s was 4100 mL. Electrocardiography showed normal cardiovascular activity, and his preoperative Qp/Qs ratio by Doppler echocardiography was only 1.08. Lung cancer was located in the right upper lobe, in the same location as PAPVC. Therefore, we planned to perform right upper lobectomy and mediastinal lymph node dissection for definitive treatment of lung cancer and PAPVC. We considered that no hemodynamic problems would occur during and after the operation.

Under general anesthesia and double-lumen intubation, we performed video-assisted thoracoscopic approach using three ports. An anomalous vessel, PV1-3, was noted after dissecting the pleura, which drained into the azygos vein from the right upper lobe (Fig. 3B). The PV1-3 was divided using a stapler as the usual maneuver, and right upper lobe lobectomy and lymph node dissection were performed. The operative time was 2 h and 51 min and the amount of bleeding was 30 mL. The patient’s postoperative course was uneventful, and no hemodynamic problems occurred. The patient was followed up with no recurrence for 16 months.

**Discussion**

PAPVC is a comparatively uncommon congenital vascular anomaly that is found in only 0.1%–0.2% of the adult population using CT scans. In this vascular anomaly, one or more pulmonary veins drain into a systemic vein, resulting in a left-to-right shunt. Anomalous right-sided pulmonary veins drain into the superior vena cava, inferior vena cava, right atrium, azygos vein, portal vein, or hepatic vein. Additionally, anomalous left-sided pulmonary veins can drain into the left brachiocephalic vein, coronary sinus, or hemiazygos vein. The patient’s clinical severity is determined by the degree of the left-to-right shunt. Patients with a small amount of shunts are usually asymptomatic and these tend to not be found during their lifetime. These anomalous veins are sometimes incidentally detected during investigation of another disease, while patients with large shunts present with dyspnea, palpitations, and chest pain. Our patient had no symptoms, and he finished a full marathon. His lung cancer was located in the right upper lobe in the same location as PAPVC. Lobectomy was the definitive treatment for lung cancer and PAPVC, and no hemodynamic problems occurred during and after the operation. However, if PAPVC is located in a different lobe to be preserved, major lung resection for conditions, such...
as lung cancer, could increase the volume of left-to-right shunt flow and cause right-sided heart failure.\(^4\)

In the English and Japanese literature, there are 17 reported cases of PAPVC associated with lung cancer.\(^4\)–\(^20\) Including our case and these previous reports, preoperative diagnosis of PAPVC was obtained in nine (50%) cases. The other nine cases were not diagnosed as PAPVC before pulmonary resection. One patient developed heart failure after right pneumonectomy for lung cancer with coexisting left-sided PAPVC.\(^4\)

Fortunately, six cases of PAPVC were recognized in the same lobe as lung cancer, and two cases of PAPVC were recognized in ipsilateral different lobe during the operation, and the postoperative course of each cases were uneventful (Table 1).

Similar to these reported series, some cases of PAPVC could be overlooked or undetected at the initial radiological diagnosis because of the rare association, low incidence of symptoms, and lack of knowledge of physicians. In our case, the RULB had a tracheal bronchus, with branching from the lateral wall of the right main bronchial orifice, above the level of the common RULB, and this was a bronchial anomaly. The length of the RULB was approximately 3 cm, which is longer than the common variation, and segmental bronchi of the right upper lobe had two branches. We did not notice the coexisting PAPVC in the first non-contrast CT image. However, based on the anomalous findings of the bronchus, we reevaluated our patient using contrast 3D-CTA, and diagnosed him with coexisting PAPVC. Recently, the quality of CT has greatly advanced, and it is able to detect minor vessels or bronchial anomalies. Oshiro et al.\(^1\) reported that 42% (5/12) of cases of PAPVC were associated with major bronchial anomalies, such as tracheal bronchus and accessory cardiac bronchus. If we encounter a bronchial anomaly on preoperative CT, we need to consider the possibility of coexisting PAPVC.

<table>
<thead>
<tr>
<th>Preoperative diagnosis of PAPVC (N)</th>
<th>Positional relationship of PAPVC and LC (N)</th>
<th>Location of PAPVC (N)</th>
<th>Surgical procedures for PAPVC</th>
<th>Surgical procedures for LC (N)</th>
<th>Outcome after OP</th>
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<tr>
<td>Yes (9)</td>
<td>Same lobe (6)</td>
<td>RU (3), RL (1), LU (1)</td>
<td>Simultaneous resection</td>
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<td>Revascularization</td>
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<tr>
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<td>Contrarateral (1)</td>
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<td>LU (1)</td>
<td>None</td>
<td>RP (1)</td>
<td>Heart failure</td>
</tr>
</tbody>
</table>

PAPVC: partial anomalous pulmonary venous connection; N: number of patients; LC: lung cancer; OP: operation; RU: right upper lobe; RL: right lower lobe; LU: left upper lobe; LL: left lower lobe; LUS: left upper segmentectomy; RLL: right lower lobectomy; LLL: left lower lobectomy; LP: left pneumonectomy; RMLBL: right middle and lower bilobectomy; RP: right pneumonectomy

Conclusion

PAPVC is a rare congenital anomaly. Thoracic surgeons should be aware of the possibility of the presence of PAPVC if they plan to perform a major lung resection of not only the affected lobe in lung cancer, but also the preserved lobe before a major lung resection to prevent unexpected complications. The presence of bronchial anomalies could indicate coexisting PAPVC.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

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Disclosure Statement

The authors declare that they have no competing interests.

References


