Bronchial Carcinoid Tumor Arising from an Intralobar Bronchopulmonary Sequestration

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We report a rare case of a 38-year-old woman with a bronchial carcinoid tumor arising from an intralobar bronchopulmonary sequestration. The vascular supply to the sequestered left lower lobe originated from the descending thoracic aorta. A left lower lobe lobectomy was performed. The findings of the pathological examination revealed an atypical carcinoid tumor that was immunopositive for chromogranin and synaptophysin. At the 3-year follow-up examination, the patient was healthy.

Key words: 1. Lung neoplasms  
2. Intralobar pulmonary sequestration  
3. Carcinoid tumor

CASE REPORT

A 38-year-old woman with recurrent pneumonia, who had been repeatedly hospitalized every year since 1988, was admitted to our hospital. She was treated for pulmonary tuberculosis 9 years before admission to our hospital. She was a nonsmoker and did not have any other disease or a family history of any disease.

Chest radiography showed a calcified wall in the left lower lung field and multifocal nodular calcification in the adjacent lung parenchyma (Fig. 1A). Computed tomography (CT) angiography (Fig. 1B) showed a multiseptated cystic mass in the left lower lobe, systemic arterial supply from the descending thoracic aorta, and venous drainage to the left inferior pulmonary vein. Therefore, she was diagnosed with intralobar pulmonary sequestration. A posterolateral thoracotomy with single-lung ventilation was performed through the fifth intercostal space, sparing the serratus anterior muscle. This procedure was performed under general anesthesia, with the patient lying in the right lateral position. A necrotic mass 10 cm in diameter was located in the left lower lobe with severe pleural adhesion, and 3 feeding arteries from the descending aorta were observed. After ligation and division of the feeding arteries, a standard left lower lobectomy was performed.

The resected mass measured 5×5×2 cm and showed multiseptated cystic spaces interposed by consolidated or fibrotic lung parenchyma and bronchus-like structures (Fig. 2A). The mass was located at a distance of 2.5 cm from the bronchial resection margin. Microscopically, the mass consisted of congested alveoli, dilated bronchi and bronchioles, and accompanying thick walled vessels, consistent with pulmonary sequestration. A few atypical cells were identified in the cystic...
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Fig. 1. Preoperative chest radiograph (A) and chest computed tomography (CT) angiography (B). (A) Posteroanterior chest radiograph shows multifocal nodular calcification in the posterior left lower lobe (circle). (B) Axial CT scan shows a multiseptated cystic mass in the left lobe. Systemic arterial supply from the descending thoracic aorta (arrow).

wall, spanning 0.5 cm. They were arranged in nests and cords without stromal reaction. The cells showed salt-and-pepper type nuclear features without prominent nucleoli. Small amounts of cytoplasm were eosinophilic with an indistinct boundary (Fig. 2B). The nest and cord-like growth pattern and cellular features suggested a carcinoid tumor. A subsequent immunohistochemical study revealed tumor cell reactivity for synaptophysin (Fig. 2C) and chromogranin (Fig. 2D), which indicated neuroendocrine differentiation. Mitosis and necrosis were not identified. The diagnosis of a carcinoid tumor was rendered.

The patient was discharged on the sixth postoperative day without any complications. No tumor recurrence or distant metastasis was detected on positron emission tomography (PET/CT) performed 6 months after the operation. We followed up with the patient for 3 years and performed chest CT every 6 months; tumor recurrence or metastasis was not observed.

DISCUSSION

Pulmonary carcinoid tumors are neoplasms of neuroendocrine origin. They account for approximately 25~30% of all carcinoid tumors and approximately 3% of all bronchopulmonary neoplasms [1]. The 2004 World Health Organization (WHO) classification recognizes 4 major types of lung neuroendocrine tumors (NETs): typical carcinoid (TC), atypical carcinoid (AC), large-cell neuroendocrine carcinoma (LCNEC), and small-cell lung cancer (SCLC). The most common lung NET is SCLC (20%), followed by LCNEC (3%), TC (2%), and AC (0.2%).

Pulmonary sequestration is defined as a segment or lobe of the lung that has no bronchial communication with the normal tracheobronchial tree. There are 2 types of sequestration: intralobar sequestration (ILS) and extralobar sequestration (ELS). Patients with ILS frequently show signs of respiratory tract infection, whereas those with ELS are generally asymptomatic [2].

Intralobar pulmonary sequestration with a carcinoid tumor has rarely been reported in the literature. To the best of our knowledge, it has not yet been reported in Korea. Juettner et al. (1985) reported the occurrence of a carcinoid tumor associated with intralobar bronchopulmonary sequestration. There have been the 3 reports with similar findings. In the case studied by Juettner et al., the tumor arose from the sequestered segment, and this tumor was thought to be a complication rather than a cause of the sequestration [3-6]. However, in the case studied by Eustace et al. (1996), the carcinoid tumor occluded the airway proximal to the sequestered segment, resulting in distal stasis and thereby creating a potential site for chronic infection; these findings suggest that the carcinoid tumor caused the sequestration [7].

In our case, which we believe is the first such case reported in the country, the patient had a medical history of long-term inflammation in the form of recurrent pneumonia that persisted for several years. The carcinoid tumor in our
Fig. 2. Pathologic features (A) The mass contains cystic spaces interposed by consolidated lung parenchyma (hematoxylline-eosin stain, original magnification ×100). (B) The cells identified in the cystic wall are arranged in nests and cords, and have granular chromatin in a salt and pepper pattern, and a small amount of eosinophilic cytoplasm. Nucleoli are indistinct (hematoxylin-eosin stain, original magnification ×400). (C) The cells express synaptophysin (immunohistochemical stain, original magnification ×200) and (D) chromogranin (immunohistochemical stain, original magnification ×400).

patient did not occlude the airway, and therefore, the tumor probably did not cause the inflammation. In addition, the cyst was located at a distance of 0.5 cm from the bronchial resection margin, and a mass of atypical cells with a diameter of 0.5 cm was found around the cystic wall. Therefore, we assumed that the carcinoid tumor was a complication of the intralobar pulmonary sequestration. However, to date, the etiology of the intralobar sequestration in association with the carcinoid tumor remains disputed.

Patients with pulmonary sequestration are frequently treated for benign diseases such as the common cold and pneumonia. If the symptoms of patients with pulmonary sequestration are not serious, the condition may not warrant intervention and thus persist for several years. However, as previously mentioned, carcinoid tumors are low-grade malignancies. Without treatment, a carcinoid tumor with sequestration can worsen a patient’s condition and spread to other organs.

Patients with intralobar pulmonary sequestration and carcinoid tumors should receive regular follow-up examinations after treatment to detect recurrence or metastasis of carcinoid tumors. Further studies are required to investigate the pathogenesis of carcinoid tumors associated with pulmonary
REFERENCES