

A Case of Surgically Resected Lung Cancer in a Patient with Kartagener's Syndrome

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Kartagener's syndrome is a rare inherited disorder with a triology of symptoms (bronchiectasis, sinusitis and situs inversus) and is also associated with abnormalities of the cilia of the respiratory epithelium. Lung cancer arising in Kartagener's syndrome is very rare and to date only 5 cases have been reported in the English and Japanese literature. We report on a case of a 65-year-old Japanese male Kartagener's syndrome patient with squamous cell carcinoma of the lung. A left pneumonectomy was performed and no recurrence was found within 2 years.

Key words: non small cell lung cancer, Kartagener's syndrome, surgery

INTRODUCTION

Kartagener's syndrome is a rare inherited disorder characterized with the triad of bronchiectasis, sinusitis and situs inversus, and is also combined with abnormalities of the cilia of the respiratory epithelium [1]. Lung cancer in patients with Kartagener's syndrome is very rare. In this paper, we report a case of a surgically resected lung cancer in a man with Kartagener's syndrome.

CASE REPORT

A 65-year-old man presented with reproductive cough and hemoptum. He exhibited the classical triad of Kartagener, that is, situs inversus, chronic sinusitis and bronchiectasis, and had been diagnosed as having Kartagener's syndrome in his childhood. He had a history of recurrent episodes of cough, expectoration and sinusitis and was followed up by his family doctor.

When he first consulted his family doctor with the symptoms of reproductive cough and hemoptum, he was diagnosed as having bronchitis and oral antibiotics were prescribed. However, his symptoms persisted, so he was referred to our institution for further investigation and treatment after a month from the appearance of the first clinical symptoms.

He had a 90-pack/year smoking history. A chest examination revealed crackles and rhonchi in the left upper region. His chest roentgenogram revealed dextrocardia and consolidation of the left upper lung field (Fig. 1). A chest CT scan revealed a mass measuring about 6.5 cm and bronchiectasis in the left upper lobe (Fig. 2A, B).

Results of routine blood examination were all

normal. His carcinoembryonic antigen (CEA) was 3.2 ng/ml and cytokeratin fragment 19 (Cyfra 21-1) < 1.0 ng/ml, and all findings were normal.

Positron emission tomography with 2-[fluorine-18]-fluoro-deoxy-d-glucose (FDG-PET) revealed a hot spot in the tumor (Fig. 2C). No apparent finding of hilum and mediastinal lymph node metastasis or tumor invasion into the left main pulmonary artery and upper pulmonary vein was detected. No apparent distant metastasis was shown in the routine workup.

Bronchoscopy showed bronchus transposition. The tumor was not visible endoscopically and the diagnosis of squamous cell carcinoma was confirmed through a transbronchial lung biopsy (TBLB) from the left B3 bronchus. His forced expiratory volume in one second (FEV 1.0) was 2.24 L (67% predicted). No apparent cardiac risk was found during the routine work up. Accordingly, with the diagnosis of squamous cell lung cancer (cT2bN0M0, stage IIA), resection of the tumor was planned.

Because the possibility of adhesion of the tumor and vessels was taken into consideration, the operation was performed via a median sternotomy. Intraoperatively, the tumor and left upper lobe were tightly adherent to the chest wall. Increased vascularity in the connective tissue surrounding the lesion was observed and was susceptible to bleeding during dissection. Although the superior vein could be divided extrapericardially, exposure of the superior trunk of the pulmonary artery was impossible because of the surrounding adherent solid connective tissue and pneumonectomy was therefore necessitated. The total operation time was 5 h 13 min and the total blood loss was 3539 ml. The tumor measured 65x55 mm. There was no apparent tumor invasion of the main pulmonary artery (Fig. 3). A

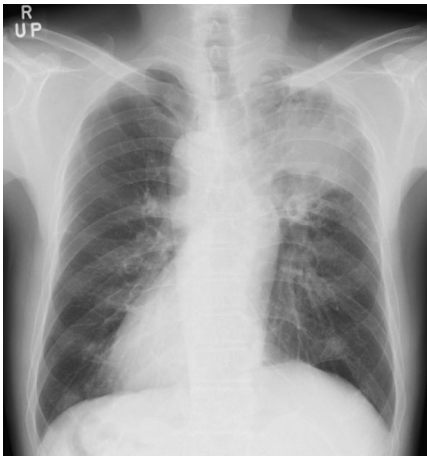


Fig. 1 Chest x-ray image showing dextrocardia and consolidation in the left upper lung field.

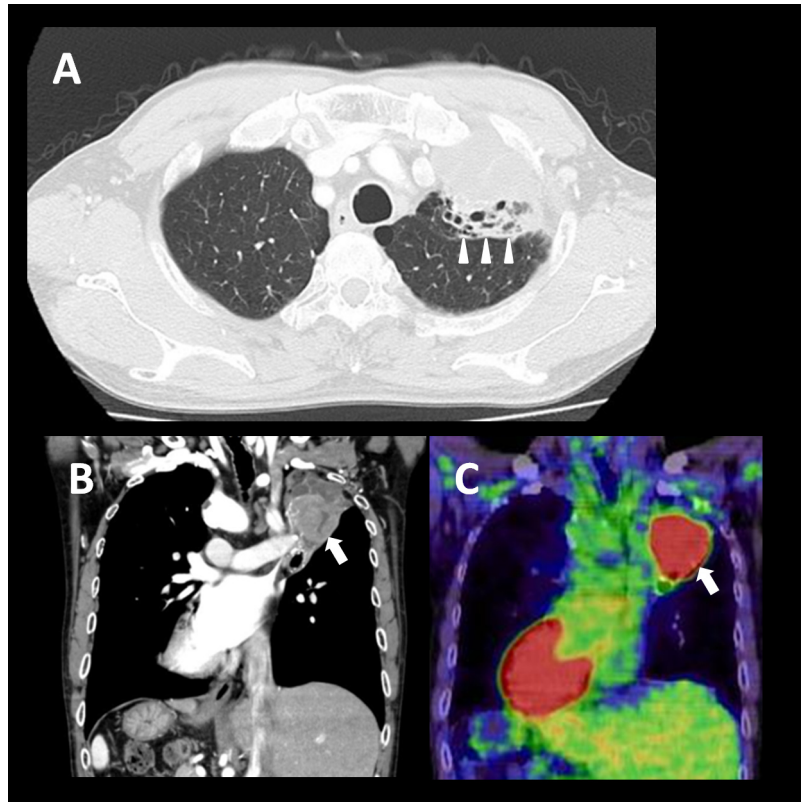


Fig. 2 Chest CT scan shows bronchiectasis (A, arrowhead) and a mass measuring about 6.5 cm (B, arrow) in the left upper lobe. Positron emission tomography with 2-[fluorine-18]-fluoro-deoxy-d-glucose (FDG-PET) revealed a hot spot in the tumor (C, arrow).

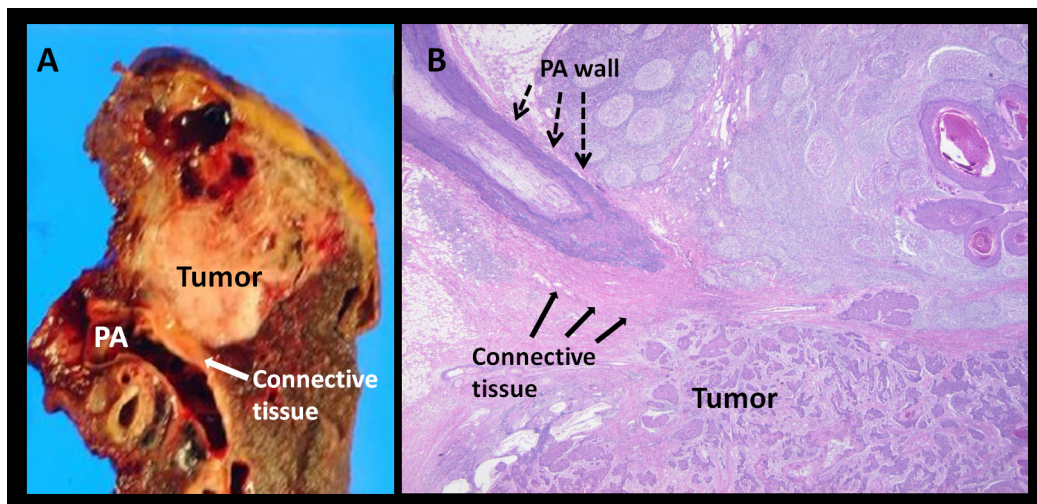


Fig. 3 Macroscopic and microscopic findings of the tumor (A and B, respectively). Connective tissue exists between the tumor and pulmonary artery, and there was no apparent tumor invasion of the main pulmonary artery (PA, pulmonary artery).

metastatic lesion was detected in the interlobar lymph node (station 11s). The final diagnosis was squamous cell lung cancer (pT2bN1M0, stage IIB).

The patient's postoperative course was uneventful. He was discharged on the 14th postoperative day. At present, 2 years after the surgery, the patient is under follow-up at the outpatient clinic and is doing well with no signs of recurrence of the tumor.

DISCUSSION

Kartagener's syndrome is an autosomal recessive disorder primarily manifesting as a **ciliary movement** disorder [1]. Initially, Sievert described the combination of situs inversus, chronic sinusitis and bronchiectasis in 1904 [2]. However, Kartagener first recognized this clinical triad as a distinct congenital syndrome in 1933 [3]. The frequency has been reported to be 1 case per 30000-60000 live births [4].

There is little knowledge concerning the correlation between Kartagener's syndrome and malignancy, including lung cancer. Only 5 cases of lung cancer arising in Kartagener's syndrome patients could be found in the English and Japanese literature, namely 1 bronchogenic carcinoma, 3 squamous cell carcinomas, and 1 small cell carcinoma [5-9] (Table 1). The review of the literature and our case are listed in Table 1. All patients were male, ranging in age from 52 to 77 years (mean, 65.5 years): in 4 cases the tumor arose from the right side (all from left upper lobe), and in 2 cases from the left side (1 from the middle lobe and 1 from the intermediate bronchus).

Kartagener's syndrome is associated with a motility defect in the cilia of the respiratory mucosa in the lungs and sinuses and broncho-pulmonary tree clearance. Theoretically, this can result not only in poor bacterial clearance but also chronic oncogenic substance exposure. In our review, there is no smoking information in 2 out of 6 cases and another 4 cases whose smoking history was described as considerable (60-90 pack year) which is a clear risk factor associated with lung cancer (Table 1). Because the information is not sufficient, we consider that it is difficult at present to discuss the definitive influence of Kartagener's syndrome on oncogenesis. Further study about the influence of Kartagener's syndrome in oncogenesis in the respiratory tract is necessary.

Intraoperatively, strong adhesion and increased vascularity in the connective tissue surrounding the lesion was observed, which was susceptible to bleeding. This made surgical resection very difficult and the patient underwent very high operative stress. We consider that the findings in our present case arose from chronic and recurrent infection of the broncho-pulmonary tree and are probably common in patients with Kartagener's syndrome. Surgeons should therefore take these possibilities into consideration and make appropriate preoperative preparation for such cases.

In our present case, the patient presented with a reproductive cough and hemospitum. He was initially diagnosed as having a respiratory infection, and was treated for a month based on this diagnosis which caused a delay in the accurate diagnosis of lung cancer. A similar clinical course has also been reported previously [6, 7]. Our experience and previous reports sug-

Table 1 Literature review: Characteristics of Kartagener's syndrome patients with lung cancer

No.	Author, Year of Publication	Age	Sex	Smoking history (pack-year)	Sites of tumor	Histology	Treatment	Outcome
1	Baruah, 1952	52	Male	NR	Right upper lobe	Bronchogenic ca	NR	Dead at 4 months, hemoptysis
2	Kawanishi, 1980	57	Male	76	Left bronchus intermedius	Squamous cell ca	Left pneumonectomy	Alive at 12 months without recurrence
3	Hachiya, 1983	71	Male	60	Right upper lobe	Squamous cell ca	Best supportive care	Dead at 1 month, pneumonia
4	Higashi, 1994	77	Male	NR	Left middle lobe	Small cell ca	Chemotherapy	Alive at 17 months with disease
5	Horie, 2010	71	Male	60	Right upper lobe	Squamous cell ca	Chemoradiation therapy	Dead at 12 months pneumonia
6	Inoue, 2011, current case	65	Male	90	Right upper lobe	Squamous cell ca	Left pneumonectomy	Alive at 24 months without recurrence

NR, not reported. ca, carcinoma.

gest that it can be difficult to distinguish Kartagener's syndrome-related symptoms and radiologic image findings from those caused by malignancy of the lung. We suggest that the possibility of lung malignancy should be taken into consideration when we medically examine a patient with Kartagener's syndrome whose respiratory infection-related symptoms are persistent and who are refractory to the usual treatment.

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